A Rare Complication After Transcanalicular Dacryocystorhinostomy: Tissue Necrosis and Nasal-Cutaneous Fistula

Baris Yeniad, M.D., Lale Kozer Bilgin, M.D.*, Atahan Cagatay, M.D.†, and Ismet Aslan, M.D.‡*

Abstract: **Transcanalicular dacryocystorhinostomy (TCDCR) with diode laser is a minimally invasive technique with good cosmetic results. The reported complication rate is low. In this brief report, the authors describe a patient with tissue necrosis and nasal-cutaneous fistula after TCDCR. A 65-year-old woman attended the authors' clinic 1 month after TCDCR. Examination revealed a large tissue defect and nasal-cutaneous fistula in the medial canthal region. *Aspergillus* growth was noted in culture specimens taken from the nasal cavity. The patient was treated with intravenous amphotericin B. The treatment resulted in granulation and closure of the defect.**

Transcanalicular dacryocystorhinostomy (TCDCR) with a diode laser is a minimally invasive approach that has been shown to restore nasolacrimal patency.[1,2] It operates at a wavelength of 810 nm or 980 nm, inducing excellent hemostasis due to its high absorption by melanin and hemoglobin.

This technique may decrease operating time and reduce morbidity with good cosmetic results. The reported complication rate is low and is usually related to problems with probing and intubation.[1] Thermal injury to adjacent tissues is very rare and was reported in only one of 118 patients in a study by Hong et al.[2]

We herein report a patient with tissue necrosis and nasal-cutaneous fistula after TCDCR.

Case Description. A 65-year-old woman attended our clinic 1 month after TCDCR and bicanalicular silicon intubation at another institution. Further evaluation revealed that she had developed redness and tenderness in the medial canthal region 3 days after the initial surgery. By 2 weeks, skin ulceration had developed, followed by a large area of adjacent tissue necrosis. Topical and systemic broad-spectrum antibiotics were instituted, but no improvement was noted. At this stage, an advancement flap was performed; however, the flap failed and the defect recurred in the same medial canthal area.

Our examination revealed a large tissue defect and nasal-cutaneous fistula in the medial canthal region (Fig. A). Assessment by the Ear Nose and Throat team showed inflammation and destruction of the associated nasal mucosa. Culture specimens taken from the nasal cavity demonstrated *Aspergillus* growth, consistent with fungal infection. The patient was started on a regimen of intravenous followed by oral amphotericin B and was observed closely for 2 months. With adequate treatment of her fungal infection, our conservative management resulted in granulation and closure of the tissue defect (Fig. B, C).

CONCLUSION

Tissue necrosis after external dacryocystorhinostomy has been reported in some studies.[4,5] In one of these studies, a destructive vasculitis was thought to be the cause.[4] In another study, the cause was attributed to poor circulation and extensive cauterization in a patient with diabetic vasculopathy. The authors recommended avoidance of overcauterization and cautious control of postoperative infections to prevent undesired wound problems.[5]

The diode laser system delivers laser energy via an optical fiber. The physical properties of the fiber, in particular its shape and coating, convey and confine the laser energy to the tip of the probe, where the energy is maximal. Wavelengths of 810 nm to 980 nm with a power setting of 3–12 watts are typically used in TCDCR surgery. These settings are critical to perforate only the desired area of bone and nasal mucosa while avoiding retrograde heating that may result in canalicular thermal damage.

In our patient, TCDCR was complicated by tissue necrosis and was associated with fungal infection. Contributing factors may have included high laser energy, fiber optic tip problems, or surgical technique. Laser energy greater than 15 watts has been shown to lead to heat lateralization and excessive tissue charring.[3] This may lead to inadvertent canalicular damage. The fiber optic tip and its properties outlined above are critical for regulating the appropriate amount of energy to achieve its desired effects. Surgeons need to be well versed in their technique for this procedure and have a thorough understanding of relevant anatomical structures to minimize complications.

In conclusion, TCDCR is a promising technique that can be performed in nasolacrimal duct obstruction. Understanding of relevant technical issues and possible complications is important to achieve the best possible outcomes.

REFERENCES

1. Plaza G, Beteré F, Nogueira A. Transcanalicular dacryocystorhinostomy with diode laser: long-term results. *Ophthal Plast Reconstr Surg* 2007;23:179–82.
2. Hong JE, Hatton MP, Leib ML, Fay AM. Endocanalicular laser dacryocystorhinostomy: analysis of 118 consecutive surgeries. *Ophthalmology* 2005;112:1629–33.
3. Henson RD, Henson RG Jr, Cruz HL Jr, Camara JG. Use of the diode laser with intraoperative mitomycin C in endocanalicular laser dacryocystorhinostomy. *Ophthal Plast Reconstr Surg* 2007; 23:134–7.
4. Jordan DR, Miller D, Anderson RL. Wound necrosis following dacryocystorhinostomy in patients with Wegener's granulomatosis. *Ophthal Surg* 1987;18:800–3.
5. Salour H, Montazerin N. Incision site tissue necrosis after dacryocystorhinostomy. *Ophthal Plast Reconstr Surg* 1998;14: 146–8.

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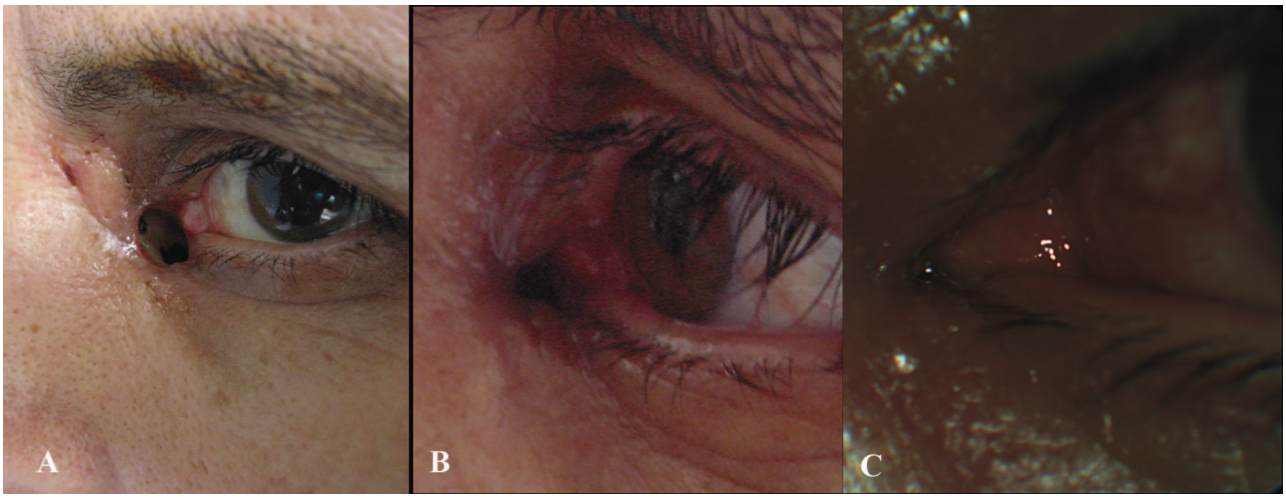

A, A large tissue defect and nasal-cutaneous fistula in the medial canthal region. Note the scar tissue due to unsuccessful advancement flap superior to the defect. **B,** The tissue defect got smaller after intravenous amphotericin B treatment. **C,** Granulation and closure of the tissue defect.

Cicatricial Entropion Following Docetaxel (Taxotere) Therapy

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Abstract: **We report eyelid margin changes in 3 patients after docetaxel treatment for breast cancer. These patients were referred to Cincinnati Eye Institute for eyelash abnormalities and/or epiphora and all had been treated with docetaxel in the near past. The ophthalmic complaints started soon after chemotherapy. All 3 cases showed varying degrees of palpebral mucosal inflammation and cicatricial changes leading to marginal entropion. The first patient exhibited very prominent eyelid margin inflammation and diffuse trichiasis without madarosis. The second patient had extensive madarosis and abnormally curved, thin eyelashes with punctal and canalicular obstruction. Her marginal entropion was most prominent at the medial lower eyelids. The last patient showed significant lash misdirection in a scattered distribution and patchy madarosis along with pseudomembranes blocking the puncta. All of these eyelid abnormalities occurred soon after docetaxel treatment for advanced breast cancer. Such changes in the absence of questioning for previous docetaxel use in history taking.**

Docetaxel (Taxotere, Aventis, Bridgewater, NJ) is a taxane-class antineoplastic agent widely used in the treatment of several solid tumors, most commonly advanced breast cancer.[1,2]

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Canalicular obstruction causing epiphora is the best known ocular side effect of docetaxel.[3] A few other reports have also associated docetaxel with erosive conjunctivitis and punctal stenosis,[4] erythematous skin reaction, madarosis and ectropion,[5] and meibomian gland inflammation causing chalazion.[6]

Severe epiphora was reported in up to 77% of the patients receiving weekly docetaxel therapy.[7] Epiphora and canalicular stenosis were more severe and occurred more frequently after weekly docetaxel regimen in a study that documented epiphora in 64% of patients on weekly docetaxel treatment versus in 39% of patients in a group receiving treatment every 3 weeks.[8] Initial reports claimed docetaxel-induced conjunctivitis as the cause of tearing[9]; however, Esmaeli et al.[3] proved that epiphora was in fact due to punctal/canalicular obstruction in such cases. The same group also demonstrated histopathologic alterations in the nasolacrimal system mucosa that resulted in anatomical narrowing in patients using docetaxel.[10]

We describe 3 patients showing cicatricial marginal entropion and structural eyelash abnormalities including trichiasis and lash ptosis after treatment with docetaxel for advanced breast cancer. Trichiasis is a universally observed phenomenon after any cytotoxic chemotherapy and not unique to docetaxel; however, to our knowledge, marginal structural changes causing eyelash abnormalities after docetaxel therapy have not been reported before.

METHODS

The medical records of 3 patients with a history of docetaxel use and a diagnosis of marginal cicatricial eyelid changes and various eyelash abnormalities were reviewed. Information regarding the chief complaint and history of the current problem, medical health history including systemic treatments, and ophthalmic plastic examination findings were extracted from the files. Informed consent was obtained from each patient, and the study adhered to the principles of the Declaration of Helsinki.

All patients were Americans referred from other centers for ophthalmic plastic evaluation. There was no history of trauma, exposure to chemicals, topical ophthalmic medications, recent eye infections, or travel history to a geographic location of endemicity where a possible infection such as trachoma could have resulted in a preexisting conjunctival scarring. All the patients had been treated with docetaxel in the near past, and none of them had any

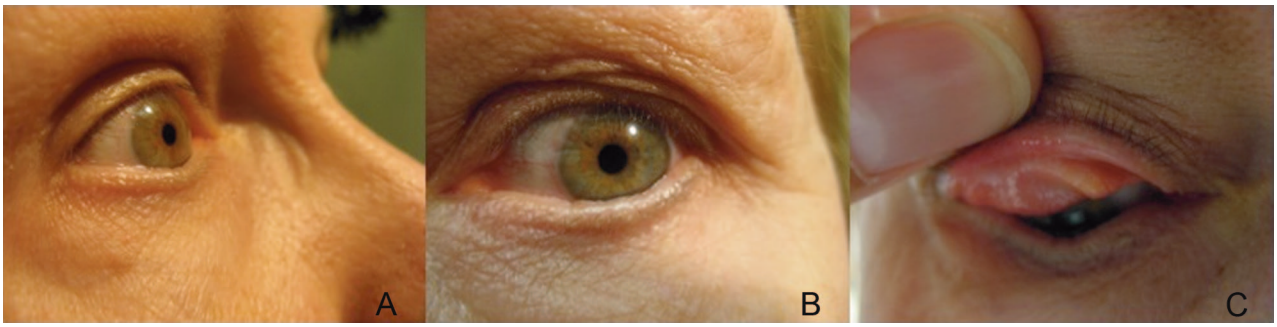

FIG. 1. **A,** Patient 1 right eye. Misdirected eyelashes involving the lateral half of the upper eyelid. **B,** Patient 1 left eye. Diffuse lash ptosis involving the entire length of the upper eyelid. **C,** Patient 1 right eye. Significant inflammatory and cicatricial changes within the temporal palpebral conjunctiva that corresponds to the area of eyelash abnormalities.

of the ophthalmic problems before chemotherapy. The first patient received an every-3-week regimen, whereas docetaxel was used weekly for 12 sessions in the other 2 patients.

Patient 1

A 54-year-old woman was referred for trichiatic upper eyelashes touching her cornea for more than 4 months. Her eyelashes were epilated twice during this time by her regular optometrist, who thought the problem was due to her recent chemotherapy. She denied any tearing and in fact reported dryness and foreign body sensation in both eyes. She stated that she had received chemotherapy for breast cancer 7 years ago and completed 6 sessions/4-month therapy with docetaxel and carboplatin 8 months ago for an unrelated uterine/ovarian carcinoma. Since then, she had experienced marked changes in her nailbeds, palms, toes, and eyelids. Slit-lamp examination showed diffuse lash misdirection in the lateral half of the right eyelid (Fig. 1A) and all through the left eyelid (Fig. 1B), along with moderate blepharitis and meibomitis on both upper eyelids. Eversion of the eyelids demonstrated marginal mucosal cicatrization and extensive inflammation (Fig. 1C). All 4 puncta were patent and apposed to globe. She could not recall any history of eyelash problems or history of blepharitis at any of her prior annual optometrist visits. Blepharitis was treated with medical treatment; however, no change was noted in the lash misdirection. She refused to have any kind of surgical treatment for her eyelashes and was advised to keep using contact lenses, since she described relief of her symptoms during contact lens use.

Patient 2

Patient 2 was a 43-year-old black woman who was referred for excessive tearing and mucous in her eyes for the previous 4 to 5 weeks. Tearing started 5 days after the last dose of her weekly docetaxel therapy for metastatic breast cancer. She denied any previous episodes of epiphora or ocular surface disease. Meniscus was elevated in both eyes, worse in the left. Patchy madarosis and structural eyelash changes were evident in all 4 eyelids. Cicatricial marginal changes in the medial one third of the lids and medial entropion with punctal inversion were evident in both lower eyelids, with no evidence of blepharitis (Fig. 2). All 4 puncta were stenotic, and only the right lower eyelid could be popped open with the dilator. The other 3 puncta underwent snip punctoplasty, and bicanalicular stents were inserted successfully after the canalicular obstruction was forced open using lacrimal trephine.

Patient 3

Patient 3 was a 50-year-old Caucasian female who was referred for tearing and excessive mucous formation in both eyes for the previous 4 months. She received 12 sessions of weekly docetaxel treatment for breast carcinoma. Tearing started by the end of the first month of treatment and improved to some extent after the cessation of treatment about 2 months before her visit. Meniscus was elevated equally in both eyes. All puncta appeared stenotic; however, probing revealed normal-sized puncta plugged with overlying inflammatory membrane-like structures. Irrigation was free bilaterally, revealing no canalicular or nasolacrimal duct obstruction. She exhibited near-total madarosis on the lower lids and patchy madarosis on the uppers. Misdirected lashes were evident predominantly on the temporal side of both upper eyelids (Fig. 3). The mucosal margins were diffusely thinned and inverted due to cicatricial changes. She was treated with short-term topical steroids, and tearing was resolved on follow-up examinations without major benefit on the entropion.

DISCUSSION

Docetaxel is a frequently prescribed chemotherapeutic agent that is highly effective against high-incidence cancers

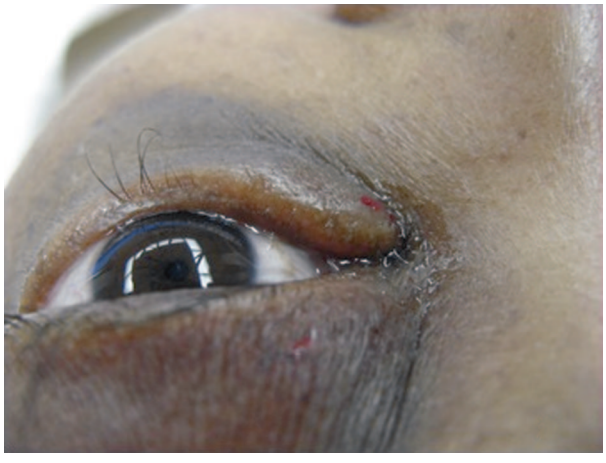

FIG. 2. Patient 2 left eye. Careful observation shows marginal entropion in the medial 1/3 of the lower eyelid. When the lid margin is inspected lateral to medial, meibomian gland orifices seem to disappear medially and the lash line seems to stand closer to the globe due to cicatricial entropion.

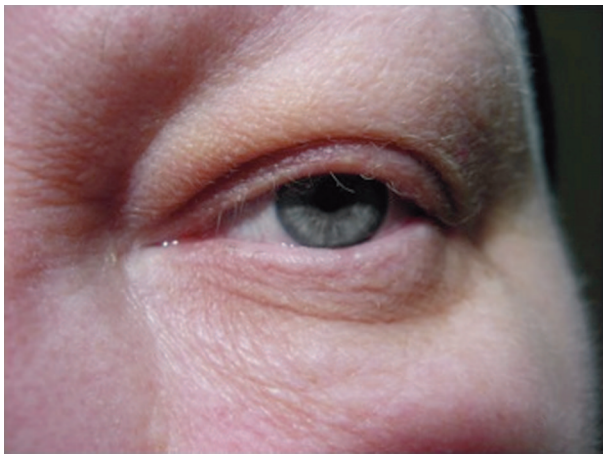

FIG. 3. Patient 3 left eye. The upper eyelid margin is thinned and inverted. There is patchy madarosis, but the more striking feature is the trichisis all along the eyelid.

involving the breast, prostate, and lung. Since its use has become widespread, specialists/subspecialists need to know about any of the possible side effects related to their practice. Commonly reported general side effects of docetaxel therapy include neutropenia, fluid retention, hypersensitivity reactions, asthenia, myalgia, peripheral neuropathy, alopecia, nail changes, mucositis, and acral erythema.[11,12]

The most prominent and extensively studied ocular adverse effect of docetaxel is epiphora. Esmaeli et al.[3,7,13] have brought up the relation and provided the literature with several informative studies. In 2001, they acknowledged their careful observation about docetaxel-induced canalicular stenosis in 3 patients and subsequently showed that canalicular stenosis and nasolacrimal duct blockage were the underlying mechanisms of epiphora secondary to docetaxel use. The same group also showed that docetaxel was secreted in the tears.[14] In a histopathologic study, they demonstrated significant inflammation and stromal fibrosis in the mucosal lining of the lacrimal drainage apparatus in 3 patients suffering from docetaxel-induced epiphora.[10] The authors speculated that the direct contact between the docetaxel in the tears and the epithelial lining of the lacrimal drainage apparatus caused changes in the mucosa, ultimately leading to anatomic narrowing. The authors also speculated that chronic changes were probably evident in the entire lacrimal drainage apparatus.

Docetaxel is a semisynthetic taxane that inhibits mitotic spindle apparatus, stabilizing tubulin polymers.[15] Cells with rapid turnover, including mucous membrane linings, are therefore more susceptible to the cytotoxic effects of chemotherapy. We believe that the chronic inflammatory and fibrotic changes from exposure to the drug, as evident in the nasal mucosa and lacrimal sac mucosa, could be evident in any mucosal tissue that docetaxel touches. Patient 2 had the most striking structural changes proximal to the punctum. The puncta were totally obstructed causing the tears (and possibly docetaxel) to accumulate and further worsen the inflammatory reaction around the ampulla, the neighboring palpebral conjunctiva, and the mucosal margin. Patient 3 also had inflammatory membranes blocking the puncta, causing prolonged tear contact with the eyelid margins, thus probably causing inflammation and fibrosis in the irritated mucosa.

Another cancer chemotherapeutic, 5-fluorouracil (5-FU), is also known to cause epiphora as a consequence of canalicular stenosis.[16] Lee at al.[17] showed that fibrosis of the punctum and the adjacent tissues may be seen after 5-FU use. Systemic 5-FU has also been associated with other ocular adnexal problems, including blepharitis, ankyloblepharon, eyelid dermatitis, and cicatricial ectropion, all attributed to the inflammation and fibrosis caused by the agent.[18–20] Since both agents are similarly documented to induce mucosal fibrosis and are secreted in the tears, it may well be expected that docetaxel-exposed eyelids could show similar reactions to 5-FU as well.

All our patients exhibited cicatricial entropion to some degree. Cicatricial entropion is usually known to occur as a result of trauma, chemical burns, Stevens-Johnson syndrome, ocular cicatricial pemphigoid, infections, or local response to topical medications. In our cases, we believe the etiology was inflammatory reaction and fibrosis incited by chemical trauma similar to the previously reported cases for 5-FU and docetaxel in the lacrimal sac and nasal mucosa. It may be speculated that blepharitis could possibly be the cause of marginal entropion and eyelash changes in one of our patients; however, we would argue otherwise for 2 reasons. First, the pattern of lash misdirection was not typical of the usual blepharitis cases. Trichiasis was temporal in only one eyelid and diffuse along the upper eyelid in the other, whereas blepharitis induced trichiasis is frequently limited to the central eyelid. More importantly, the patient could not recall any eyelash problems before the chemotherapy and she was not aware of the blepharitis problem despite routine annual ophthalmologic examinations. Therefore, we believe blepharitis in her case was actually part of the picture induced by the contact of docetaxel-loaded tears to the eye surface.

In summary, all the 3 cases we present showed varying degrees of inflammation and cicatricial changes leading to marginal entropion. The first patient exhibited the most prominent eyelid margin inflammation and diffuse trichiasis without madarosis. In the other 2 patients, entropion was more subtle and lacrimal changes were more prominent. This may be due to the difference in the regimen, as the first patient received thrice-weekly administration and the other 2 were treated with weekly sessions. The second patient had extensive madarosis and abnormally curved, thin eyelashes with punctal and canalicular obstruction. The last patient showed significant scattered lash misdirection and patchy madarosis along with membranes blocking the puncta.

Our findings in these 3 patients suggest that docetaxel may produce extensive surface inflammation that could result in profound alterations involving the eyelid margin epithelium and the lacrimal drainage system epithelium. It should not be unexpected for docetaxel to cause mucosal fibrosis in the palpebral conjunctiva or the mucosal margin of the eyelids as it does anywhere else in the body. Ophthalmologists may consider asking about docetaxel use in patients with marginal inflammation and cicatricial changes causing misdirected eyelashes in the absence of other suggestive etiology.

REFERENCES

1. Crown J, O'Leary M, Ooi WS. Docetaxel and paclitaxel in the treatment of breast cancer: a review of clinical experience. *Oncologist* 2004;9:24–32.
2. Burris HA III. Single-agent docetaxel (Taxotere) in randomized phase III trials. *Semin Oncol* 1999;26:1–6.
3. Esmaeli B, Valero V, Ahmadi MA, Booser D. Canalicular stenosis secondary to docetaxel (Taxotere). A newly recognized side effect *Ophthalmology* 2001;108:994–5.
4. Skolnick CA, Doughman DJ. Erosive conjunctivitis and punctal stenosis secondary to docetaxel (Taxotere). *Eye Contact Lens* 2003;29:134–5.
5. Kaya AO, Buyukberber S, Coskun U, et al. Acute erythema and edematous skin reaction and ectropion following docetaxel in a patient with non-small cell lung cancer. *Cutan Ocul Toxicol* 2008;27:327–31.
6. Gupta S, Silliman CG, Trump DL. Docetaxel-induced Meibomian

duct inflammation and blockage leading to chalazion formation. *Prostate Cancer Prostatic Dis* 2007;10:396–7.
7. Esmaeli B, Hortobagyi GN, Esteva FJ, et al. Canalicular stenosis secondary to weekly versus every-3-weeks docetaxel in patients with metastatic breast cancer. *Ophthalmology* 2002;109:1188–91.
8. Esmaeli B, Amin S, Valero V, et al. Prospective study of incidence and severity of epiphora and canalicular stenosis in patients with metastatic breast cancer receiving docetaxel. *J Clin Oncol* 2006; 22:3619–22.
9. Burstein HJ, Manola J, Younger J, et al. Docetaxel administered on a weekly basis for metastatic breast cancer. *J Clin Oncol* 2000;18:1212–9.
10. Esmaeli B, Burnstine MA, Ahmadi MA, Prieto VG. Docetaxel-induced histologic changes in the lacrimal sac and the nasal mucosa. *Ophthal Plast Reconstr Surg* 2003;19:305–8.
11. Gligorov J, Lotz JP. Preclinical pharmacology of the taxanes: implications of the differences. *Oncologist* 2004;9:3–8.
12. Ferraresi V, Milella M, Vaccaro A, et al. Toxicity and activity of docetaxel in anthracycline-pretreated breast cancer patients: a phase II study. *Am J Clin Oncol* 2000;23:132–9.
13. Esmaeli B, Hortobagyi G, Esteva F, et al. Canalicular stenosis secondary to weekly docetaxel: a potentially preventable side effect. *Ann Oncol* 2002;13:218–21.
14. Esmaeli B, Ahmadi MA, Rivera E, et al. Docetaxel secretion in tears: association with lacrimal drainage obstruction. *Arch Ophthalmol* 2002;120:1180–2.
15. Perry MC, Anderson CM, Donehower RC, Chemotherapeutic agents. In: Abeloff MD, Armitage JO, Niederhuber JE, et al., eds. *Clinical Oncology*. 3rd ed. Philadelphia, PA: Churchill Livingstone, 2004:483–535.
16. Loprinzi CL, Love RR, Garrity JA, et al. Cyclophosphamide, methotrexate, and 5-fluorouracil (CMF)-induced ocular toxicity. *Cancer Invest* 1990;8:459–65.
17. Lee V, Bentley CR, Olver JM. Sclerosing canaliculitis after 5-fluorouracil breast cancer chemotherapy. *Eye* 1998;12:343–9.
18. Eisman AS, Flanagan JC, Brooks AB, et al. Ocular surface, ocular adnexal, and lacrimal complications associated with the use of systemic 5-fluorouracil. *Ophthalmic Plast Recon Surg* 2003;19:216–24.
19. Straus DJ, Mausolf FA, Ellerby RA, et al. Cicatricial ectropion secondary to 5-fluorouracil therapy. *Med Pediatr Oncol* 1977;3:15–9.
20. Insler MS, Helm CJ. Ankyloblepharon associated with systemic 5-fluorouracil treatment. *Ann Ophthalmol* 1987;19:374–5.

Dacryocystitis Secondary to *Stenotrophomonas maltophilia* Infection

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Abstract: **An 88-year-old man developed recurrent bouts of dacryocystitis and underwent dacryocystorhinostomy. Cultures from the lacrimal sac demonstrated *Stenotrophomonas maltophilia*. This uncommon bacterium is highly antibiotic resistant, and clinicians should consider it in atypical cases of infection and should guide antibiotic therapy appropriately.**

An 88-year-old male patient with a medical history that was significant only for hypertension developed 3 prior episodes of right-sided dacryocystitis. Each of these episodes was notable for marked tenderness along his right medial canthus, mucopurulent discharge from his puncta, and considerable discomfort. These recurrent infections reportedly cleared with 10-day courses of oral amoxicillin clavulanate, and the patient generally experienced several weeks of relief between recurrences of his infection. Given these findings, he was ultimately referred to the oculoplastic and orbital surgery service for more definitive management of his epiphora and periodic infection.

On initial examination, the patient had a 7-mm elevated, firm nodule immediately medial to his right medial canthus. While this lesion was not erythematous or painful, copious mucopurulent discharge emanated from his right punctum with digital pressure. Probing and irrigation of his lacrimal system revealed a complete nasolacrimal duct obstruction.

In light of these findings, the patient underwent an external dacryocystorhinostomy with silicone stent intubation. Notably, copious mucopurulent discharge emanated from the lacrimal sac upon its incision. The sac was aggressively irrigated with antibiotic solution, and the pus was retrieved and sent for microbiologic culture. The remainder of the patient's surgery was uneventful, and his tearing resolved postoperatively. Furthermore, he remains infection free several months after surgery. A review of the microbiologic specimen revealed aerobic Gram-negative bacilli, and the official culture demonstrated *Stenotrophomonas maltophilia*.

Stenotrophomonas maltophilia is a rare cause of human infection and has been previously demonstrated in water, soil, plants, and animals.[1] While this bacterium has been previously implicated in endocarditis, mastoiditis, pneumonia, wound infection, systemic sepsis, and urinary tract infections, it is typically considered to be of low virulence[2] and is an extremely rare cause of ophthalmic disease.[1] Penland and Wilhelmus[1] documented 15 cases of *Stenotrophomonas maltophilia* ocular infections, and the majority (53%) of the patients in their series had bacterial keratitis. Nonetheless, they documented 2 cases of infantile dacryocystitis. In their review of the microbiology of lacrimal abscesses, Briscoe et al.[3] documented one case (2.5% of their series) of *Stenotrophomonas maltophilia* infection. In their review of the microbiology of chronic dacryocystitis in their center, Chaudhry et al.[4] discovered 4 additional cases of infection from this bacterium.

From a thorough literature search, this case highlights the infrequent nature of adult dacryocystitis secondary to colonization with this bacterium. Furthermore, it underscores several important features of this uncommon infection. While *Stenotrophomonas maltophilia* infection generally occurs in immunocompromised individuals, our patient did not have any risk factors for suppression of his immune system. However, lacrimal sac abscesses containing rarer Gram-negative bacteria appear to be increasingly common and may represent an emerging trend.[3] While the sac is not truly immunologically protected, the combination of stasis and moisture inherent to an obstructed lacrimal system may represent an optimal environment for such atypical bacteria to develop, as *Stenotrophomonas maltophilia* is known to colonize moist environments.

Furthermore, the management of *Stenotrophomonas maltophilia* infection is often difficult, as evidenced by the recurrent nature of the dacryocystitis in this case. *Stenotrophomonas maltophilia* is often resistant to multiple bacteria and expresses β-lactamases, efflux pumps for quinolone resistance, and aminoglycoside acetyl transferases for aminoglycoside resistance.[5]

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These resistance mechanisms likely explain the multiple repeat episodes of infection that this patient experienced.

The rate of dacryocystitis from uncommon Gram-negative bacteria appears to be on the rise.[3] This report illustrates an example of such a case with a unique bacterium and represents only the second known case of dacryocystitis from *Stenotrophomonas maltophilia* in the adult population. Recurrent episodes of dacryocystitis or an atypical course after antibiotic treatment warrant consideration of this bacterium and appropriate microbiologic culture and antibiotic selection.

REFERENCES

1. Penland RL, Wilhelmus KR. *Stenotrophomonas maltophilia* ocular infections. *Arch Ophthal* 1996;114:433–6.
2. Falagas ME, Kastoris AC, Voulamanou EK, et al. Community-acquired *Stenotrophomonas maltophilia* infections: a systematic review. *Eur J Clin Microbiol Infec Dis* 2009;28:719–30.
3. Briscoe D, Rubowitz A, Assia EI. Changing bacterial isolates and antibiotic sensitivities of purulent dacryocystitis. *Orbit* 2005;24: 95–8.
4. Chaudhry IA, Shamsi FA, Al-Rashed W. Bacteriology of chronic dacryocystitis in a tertiary eye care center. *Ophthal Plast Reconstr Surg* 2005;21:207–10.
5. Falagas ME, Valkimadi PE, Huang YT, et al. Therapeutic options for *Stenotrophomonas maltophilia* infections beyond co-trimoxazole: a systematic review. *J Antimicrob Chemother* 2008;62:889–94.

Hematic Cyst in a Barrier-Covered Porous Polyethylene/Titanium Mesh Orbital Floor Implant

Lisa D. Mihora, M.D., and David E. E. Holck, M.D.

Abstract: **Hematic cyst formation after placement of nonporous alloplastic orbital floor implants is well described. A benefit of porous polyethylene is fibrovascular ingrowth without capsular formation. This ingrowth inhibits hematic cyst formation. In the case presented here, a hematic cyst developed on a screw-fixated porous polyethylene/titanium mesh implant (Medpor Titan B-T-M, Porex Surgical, Newnan, GA, U.S.A.). The risk factor for this patient was the nonporous barrier surface of the implant and inadequate fracture reduction. Proper surgical technique, including implant sizing, placement, and fixation, are factors reported to prevent hematic cyst formation. This patient's symptoms resolved after removal of the implant and cyst and replacement of the orbital implant.**

We report a case of a hematic cyst developing over a screw-fixated porous polyethylene/titanium mesh orbital floor implant with a nonporous barrier surface.

CASE REPORT

A 29-year-old male presented with acute onset of binocular vertical diplopia upon awakening (Fig. 1A). He had undergone right orbital floor fracture repair by a different service 3 years earlier after a sports injury. His visual acuity was 20/20 OD and OS. Pupils were isocoric without an afferent pupillary defect. Intraocular pressures were 16 mm Hg OU. He demonstrated 2.5 mm of right-sided hyperglobus with 1.5 mm of inferior scleral show OD. Marco exophthalmometry was 17 mm OD and 17.5 mm OS as measured by Hertel exophthalmometer. Sursumduction was limited OD. Computed tomography demonstrated a cystic lesion above the right orbital floor implant, consistent with a hematic cyst (Fig. 2A). The patient did not recall recent trauma and denied oral anticoagulant use.

Intraoperatively, an encapsulated hematic cyst was found with blood breakdown products within. The cyst was drained, and the capsule was partially excised. The previously placed Medpor Titan B-T-M (Porex Surgical) implant was

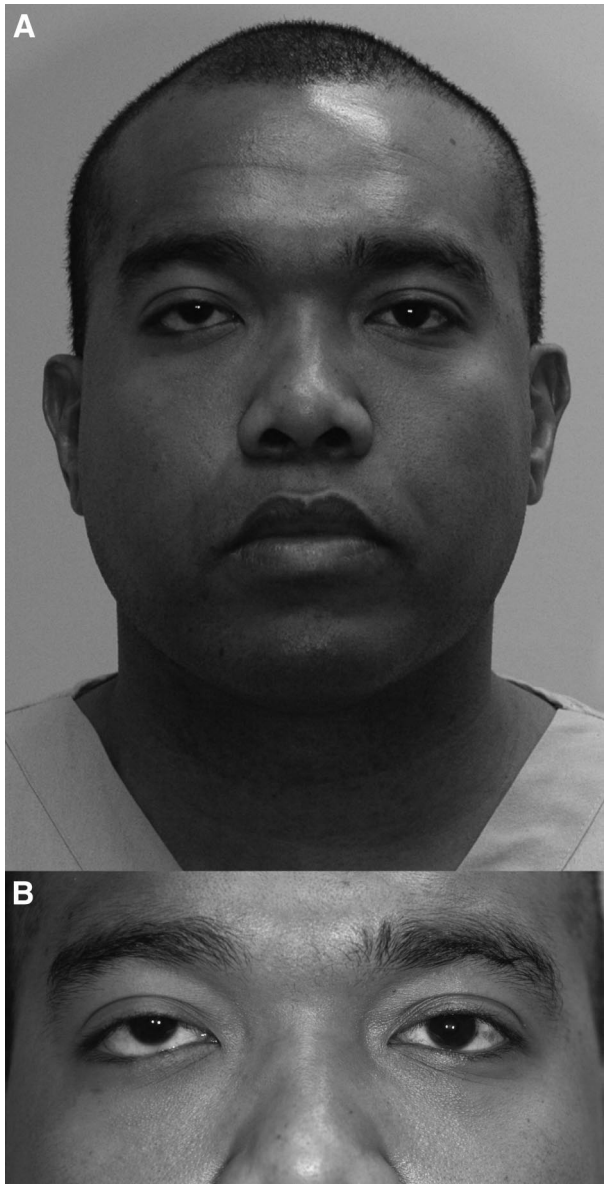

FIG. 1. **A,** Patient presenting with right-sided hyperglobus. **B,** 12 month postoperatively demonstrating resolution of hyperglobus.

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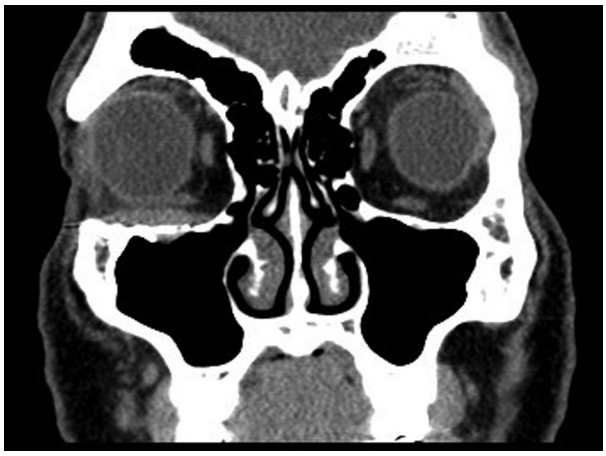

FIG. 2. Preoperative coronal computed tomogram view demonstrating cystic lesion superior to the orbital floor implant. Preoperative computed tomogram sagittal view demonstrating inadequate posterior orbital floor fracture reduction.

removed and replaced with a 0.6-mm-thick porous polyethylene/titanium mesh implant without a nonporous barrier surface (Fig. 3A, B). The original implant was fixated with a single screw to the anterior orbital rim. Additionally, the original implant inadequately reduced the posterior aspect of the fracture (Fig. 2B). With 12 months of follow-up, the patient's diplopia and hyperglobus had resolved (Fig. 1B).

DISCUSSION

Numerous postoperative complications of nonporous alloplastic orbital floor implants, including silicone (Silastic, Dow Corning, Midland, MI, U.S.A.), polytetrafluoroethylene (Teflon), and nylon (SupraFOIL, S. Jackson, Inc., Alexandria, VA, U.S.A.) have been reported. These include implant migration, extrusion, infection, fistula formation, acute hemorrhage, and hematic cyst formation.[1–3] To our knowledge, all reports of hematic cyst formation have been associated with nonporous alloplastic floor implants.[4] A capsule forms around the nonporous implant, which creates a potential space for fluid and hemorrhage accumulation. Hematic cysts may present with diplopia, pain, proptosis, vision loss, and globe displacement. A frequent presentation of a hematic cyst is diplopia upon awakening, possibly incited from rapid eye movement during sleep.

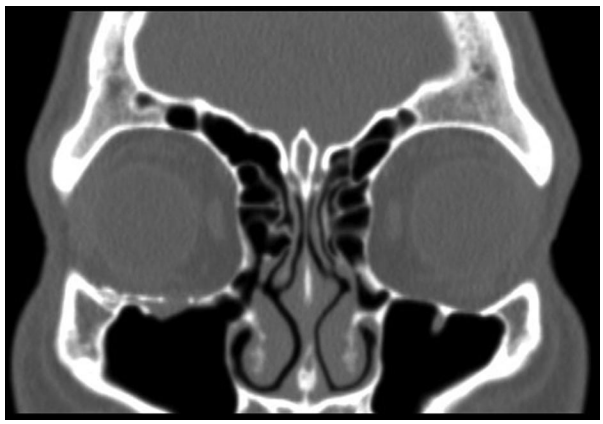

FIG. 3. Postoperative coronal computed tomograms demonstrating the reduced fracture with resolution of the hematic cyst.

Postulated risk factors for hematic cysts include subclinical trauma and implant migration causing erosion of the capsule and periosteum, erosion in the infraorbital vessels, and subclinical hemorrhage in the area of lymphatic drainage.[5] Incomplete resorption may lead to chronic granulomatous inflammation with formation of a pseudocapsule around the blood breakdown products. Adequate sizing and fixation at the time of initial fracture repair have been suggested to decrease the likelihood of hemorrhage.[1]

Porous implants such as porous polyethylene and fenestrated implants such as titanium mesh and plates provide stability through fibrovascular ingrowth in the porous channels and fenestrations, respectively. We know of no reports of hematic cyst formation in any of these implants. Recently, a combination porous polyethylene/titanium mesh has been introduced (Medpor Titan, Porex Surgical; and Synpor, Synthes Corp., Paoli, PA, U.S.A.) to provide increased implant strength with less "memory" after bending to conform to the orbital defect. Additional implant stability is promoted by screw fixation of the plate. This provides implant strength, malleability, and stability. Initial reports of this material have been positive, with no cases of delayed hematic cyst formation.[6] Available variations of this implant include impermeable barriers to one side (Medpor Titan B-T-M) or both sides (Medpor Titan B-T-B). These variations prevent orbital soft tissue adhesions to one or both sides of the plate.

In this patient, a hematic cyst developed after placement of a Titan B-T-M floor implant. This occurred despite screw fixation and fibrovascular ingrowth to the undersurface of the implant—2 stabilizing factors. We believe that contributing factors for this occurrence included chronic erosion and irritation of the periosteum and the capsule overlying the nonpermeable barrier from inadequate posterior fracture reduction. This resulted in bleeding in the capsule. We strongly agree with other authors that surgical technique is still the most important factor to prevent hematic cyst formation: proper sizing, placement, and fixation of the orbital implant greatly reduce the chance of postoperative complications.[1,4] Management of hematic cysts may include imaging with subsequent cyst drainage, removal of the fibrous wall and implant, and replacement of the implant if necessary. These steps were successful in our patient. Additionally, we are more inclined to use a porous polyethylene/titanium mesh implant without a barrier surface in those fracture patients requiring this type of implant.

REFERENCES

1. Custer PL, Lind A, Trinkaus KM. Complications of supramid orbital implants. *Ophthal Plastic Recon Surg* 2003;19:62–7.
2. DeBacker CM, Dutton JJ, Lipham W. Hemorrhagic cyst following remote alloplastic implantation for orbital floor fracture repair. *J Craniomaxillofac Trauma* 1998;4:16–8.
3. Glavas I, Lissauer B, Hornblass A. Chronic subperiosteal hematic cyst formation twelve years after orbital fracture repair with alloplastic orbital floor implant. *Orbit* 2005;24:47–9.
4. Park DJJ, Garibaldi DC, Iliff NT, et al. Smooth nylon foil (SupraFOIL) orbital implants in orbital fractures: a case series of 181 patients. *Ophthal Plastic Recon Surg* 2008;24:266–70.
5. Sutula FC, Palu RN. Delayed chocolate cyst after blow-out fracture. *Ophthal Plast Reconstr Surg* 1991;7:267–8.
6. Garibaldi DC, Iliff NT, Grant MP, Merbs SL. Use of porous polyethylene with embedded titanium in orbital reconstruction: a review of 106 patients. *Ophthal Plast Reconstr Surg* 2007;23:439–44.

Dedifferentiated Adenoid Cystic Carcinoma of the Lacrimal Gland

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Abstract: **A rare variant of adenoid cystic carcinoma is the dedifferentiated sarcomatoid form, which has previously been reported in the hard and soft palate, maxillary sinus, submandibular glands, and nasal cavity. The authors report the first case of a dedifferentiated sarcomatoid adenoid cystic carcinoma occurring in the lacrimal gland, that of a 52-year-old man. The patient presented with a 4-month history of diplopia, decreased vision, and right upper eyelid swelling. Radiographic imaging showed a soft tissue mass in the extraconal compartment superolateral to the right eye. The patient subsequently underwent surgical debulking. Histologic examination of the tissue revealed classic cribiform adenoid cystic carcinoma and a sarcomatous component consisting of malignant spindle cells and fusiform cells arranged in whorls. Dedifferentiation is a well-established phenomenon in salivary gland tumors that is associated with aggressive behavior and poor prognosis; however, the exact nature of such dedifferentiated neoplasms remains unclear.**

Orbital adenoid cystic carcinoma (ACC) typically presents with varying degrees of proptosis and globe dystopia. Pain usually accompanies these signs as perineural invasion occurs. Radiographic features of the tumor include bony erosion, irregular margins, and focal calcification within the lesion. The most common histologic pattern is cribiform, which is seen in up to 45% of these neoplasms, while the basaloid pattern is generally associated with the worst prognosis, having a 5-year survival rate of only 21%.[1,2] A rare variant of ACC is the dedifferentiated sarcomatoid ACC. Thus far, it has been reported 3 times in the submandibular glands, twice in the maxillary sinus, nasal cavity, and the tongue and once in the hard and soft palate.[3–6] We report the first case of a dedifferentiated sarcomatoid ACC occurring in the lacrimal gland, that of a 52-year-old man.

CASE REPORT

A 52-year-old man with no medical history presented with diplopia and decreased vision in the right eye for 4 months. The patient noted right upper eyelid swelling and pain in the superolateral orbit. Best-corrected visual acuity was 20/40 in both eyes. Motility in the right eye was restricted most markedly in upgaze followed by lateral and downgaze. CT scan revealed a soft tissue mass involving the right lacrimal gland, and further imaging was obtained to better define the lesion. MRI of the orbits revealed a 3.2 cm (transverse) by 3.2 cm (anteroposterior) by 2.9 cm (craniocaudal) soft tissue mass in the extraconal compartment superolateral to the right eye. The lesion replaced the right lacrimal gland causing inferior globe displacement. There was no bone invasion. Zygomatico-sphenoidal bone thinning was noted (Fig. 1). An incisional biopsy was performed, and the biopsy was composed of cribiform-type ACC.

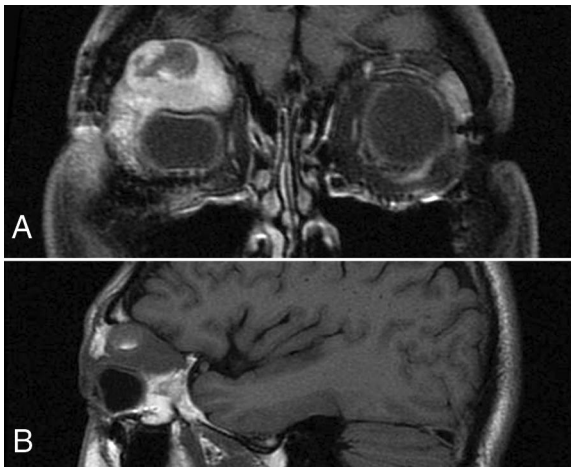

FIG. 1. **A,** T1-fat suppressed coronal image demonstrating a firm soft tissue mass in the extraconal compartment superolateral to the right eye compressing the adjacent sclera. **B,** T1-weighted sagittal image showing the lesion replacing the right lacrimal gland causing inferior globe displacement.

The patient elected to undergo surgical debulking of the tumor with proton beam radiation to follow. A lateral orbitotomy approach with an osteoplastic flap was performed. The lesion was located superotemporally and was adherent to surrounding periosteum and orbital tissue. Healthy lacrimal gland was not identifiable. The tumor extended through the extraconal and intraconal surgical planes and was multilobular in morphology. It did not appear infiltrative to the surrounding extraocular muscles and was completely removed. As there was no clear capsule and the tumor was friable, the mass was removed in its entirety. Representative sections were processed.

Histologically, the tumor consisted of 2 distinct but admixed morphologies. The carcinomatous component showed classic cribiform ACC and glands lined by a myoepithelial layer also representing ACC (Fig. 2A). These areas showed immunohistochemical reactivity for epithelial markers including pancytokeratin (AE1/AE3) and Cam5.2 and weak reactivity for epithelial membrane antigen (EMA). The myoepithelial component was reactive for S-100, smooth muscle actin, and p63. The sarcomatous component consisted of malignant spindle cells and fusiform cells arranged in whorls (Fig. 2B); focal osteoid formation was present (Fig. 2C). The spindle cells were immunohistochemically reactive for vimentin, S-100 and, focally, smooth muscle actin; of the aforementioned, only S-100 stained diffusely and strongly (Fig. 3). They were negative for epithelial markers, including Cam5.2, AE1/AE3, and the myoepithelial marker p63, indicating loss of immunophenotypic features of ACC. The morphologic and immunophenotypic findings were diagnostic of dedifferentiated ACC, with the dedifferentiated component being sarcomatoid with focal myoepithelioid dedifferentiation.

Proton beam therapy was instituted 41 days following surgical removal of the tumor. The patient received a total dose of 72.0 Gy using a combination of photon and proton radiation

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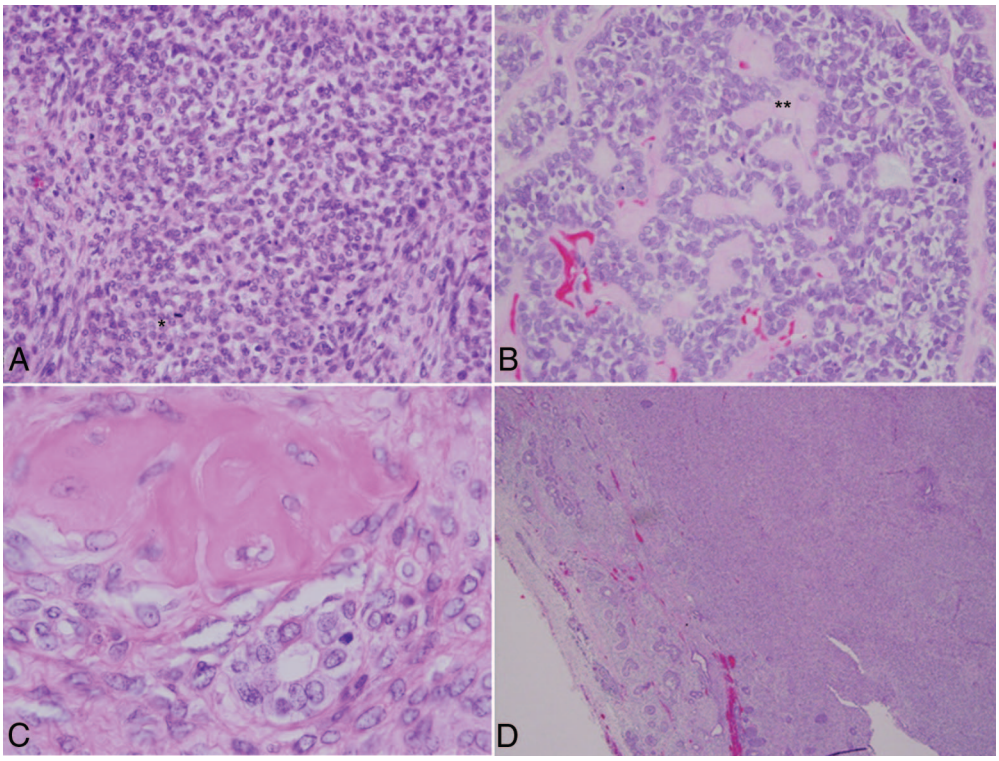

FIG. 2. **A, B,** Sections of the tumor demonstrated areas of malignant spindled cells with solid and whorled architecture and brisk mitotic activity (asterisk) **(A)** and areas of neoplastic epithelial cells with cribiform growth pattern and deposition of abundant, eosinophilic basement membrane material (asterisks) **(B)**. **C,** Focal osteoid formation was present. Hematoxylin-eosin, ×200. **D,** Areas of classic adenoid cystic carcinoma (left) merged with sheets of sarcomatoid carcinoma. Hematoxylin-eosin, ×40.

therapy. One year later, the patient remained asymptomatic and a routine MRI revealed a 25-mm × 3-mm × 17-mm lesion along the undersurface of the right orbital roof with no adjacent dural extension or perineural extension. An incisional diagnostic biopsy revealed a predominantly sarcomatous lesion. The patient will undergo a right orbital exenteration.

DISCUSSION

The concept of dedifferentiation was first proposed by Dahlin and Beabout[7] in 1971, when they described a dedifferentiated chondrosarcoma as a distinct clinicopathologic entity characterized by a low grade chondrosarcoma juxtaposed to a histologically different high grade sarcoma. Dedifferentiated tumors have since been described in bone and soft tissue pathology and in acinic cell carcinomas and carcinoma ex pleomorphic adenoma of the salivary gland. Dedifferentiation may be present in an otherwise low grade tumor at the time of diagnosis or it may arise in a recurrent, previously treated, initially low grade tumor.

We believe the sarcomatoid elements of these tumors represent mesenchymal transdifferentiation of the neoplastic epithelial cells. However, the possibility that the distinct carcinomatous and sarcomatous morphologic components of these neoplasms actually represent a collision of separate malignant processes remains a subject of debate.

Thus far, in the literature only 11 cases of dedifferentiated ACC have been reported and none involved the lacrimal gland or other sites in the orbit.[3–6] Nagao et al.[6] described 6 cases of dedifferentiated ACC; the primary tumor sites included the maxillary sinus twice, the submandibular glands

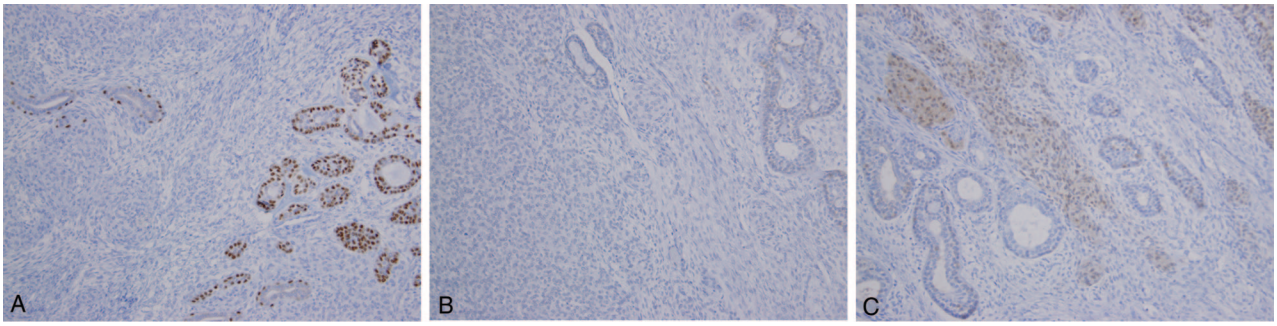

FIG. 3. **A–C,** The sarcomatous component did not stain for p63 **(A)** but did stain weakly for smooth muscle actin **(B)** and strongly for S-100 **(C)**.

twice, and the nasal cavity twice. Before the 6 cases reviewed by Nagao et al., dedifferentiated ACC had been reported twice involving the tongue and once in the hard palate, soft palate, and submandibular gland.[3–5] In all but one previously reported case, the dedifferentiated component of these tumors has been composed of pleomorphic carcinoma or poorly differentiated adenocarcinoma.[3–6] The remaining case occurred in the soft palate and contained sweeping fascicles of pleomorphic spindled cells with brisk mitotic activity.[5] Of 9 patients with long-term follow-up, 6 died of disease within 3 years of the initial diagnosis, indicating that dedifferentiated ACC is a highly aggressive tumor similar to ACC with a predominantly solid growth pattern.[6]

Since a dedifferentiated ACC tumor of the lacrimal gland has not been reported, the ultimate clinical course is difficult to predict; however, all reported dedifferentiated tumors to date have demonstrated aggressive biology. The current case underscores the importance of extensive sampling and thorough histologic examination of salivary gland-type neoplasms of the orbit as dedifferentiation, and thus more aggressive behavior are possible occurrences with important clinical implications.

REFERENCES

1. Gamel JW, Font RL. Adenoid cystic carcinoma of the lacrimal gland: the clinical significance of a basaloid histologic pattern. *Hum Pathol* 1982;13:219–25.
2. Ahmad SM, Esmaeli B, Williams M, et al. American Joint Committee on Cancer classification predicts outcome of patients with lacrimal gland adenoid cystic carcinoma. *Ophthalmology* 2009;116:1210–5.
3. Moles MA, Avila IR, Archilla AR. Dedifferentiation occurring in adenoid cystic carcinoma of the tongue. *Oral Surg Oral Med Oral Pathol Oral Radiol Endod* 1999;88:177–80.
4. Chau Y, Hongyo T, Aozasa K, et al. Dedifferentiation of adenoid cystic carcinoma: report of a case implicating p53 gene mutation. *Hum Pathol* 2001;32:1403–7.
5. Cheuk W, Chan JK, Ngan RK. Dedifferentiation in adenoid cystic carcinoma of salivary gland: an uncommon complication associated with an accelerated clinical course. *Am J Surg Pathol* 1999;23:465–72.
6. Nagao T, Gaffey TA, Serizawa H, et al. Dedifferentiated adenoid cystic carcinoma: a clinicopathologic study of 6 cases. *Mod Pathol* 2003;16:1265–72.
7. Dahlin DC, Beabout JW. Dedifferentiation of low-grade chondrosarcomas. *Cancer* 1971;28:461–6.

Orbital Roof Encephalocele Mimicking a Destructive Neoplasm

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Abstract: **The purpose of this case report is to report an orbital roof encephalocele mimicking a destructive orbital neoplasm. Orbital roof encephalocele is uncommon but can mimic neoplasm. One potential mechanism for the orbital roof destruction is a post-traumatic "growing orbital roof fracture." The growing fracture has been reported mostly in children but can occur in adults. Alternative potential etiologies for the encephalocele are discussed, including Gorham syndrome. Orbital roof encephalocele is uncommon in adults, and the findings can superficially resemble an orbital neoplasm. Radiographic and clinical features that might suggest the correct diagnosis include a prior history of trauma, overlying frontal lobe encephalomalacia without significant mass effect or edema, and an orbital roof defect. The "growing fracture" mechanism may be a potential explanation for the orbital roof destruction in some cases.**

Orbital roof encephalocele is uncommon but may mimic an orbital neoplasm. Orbital imaging might show a mass-like lesion and bony destruction that might suggest the diagnosis of an aggressive orbital neoplasm. One mechanism for the bony orbital roof destruction is a "growing skull fracture."[1,2] It is estimated that this occurs in about 0.05% to 1% of all skull fractures and 90% of the reported cases are children 3 years of age or younger.[1] Growing skull fractures typically occur in the cranial convexity but may occasionally develop in the skull base. Frontal, parietal, and occipital bones are the most frequent sites for growing skull fractures.[2] Post-traumatic leptomeningeal cyst, post-traumatic porencephaly, and post-traumatic bone absorption are other terms used to describe the same condition. Searching English language literature for growing orbital roof fracture (GORF) revealed fewer than 30 cases reported in children,[3] one case reported in an adolescent,[2] and one case reported in an adult.[4] Here we report an adult patient with an orbital roof encephalocele to emphasize the potential differentiating clinical and radiographic features that might allow preoperative consideration for the diagnosis. We hypothesize that the patient developed a GORF after prior head trauma but pre-existing congenital or acquired orbital roof encephalocele cannot be totally excluded.

CASE REPORT

A 50-year-old man presented with intermittent episodes of nonspecific "dizzy" spells and transient numbness involving the left side of his face for 3 months. The origin of these nonspecific symptoms was not clear, and the symptoms later resolved spontaneously. A CT scan of the head showed a left frontal orbital lesion, and the patient was referred for further evaluation and management. Six months before this CT scan, the patient was involved in a motor vehicle accident. He was driving at highway speeds and struck his left forehead on the steering wheel. At the time of the accident, he had some visible bruising over the left brow. The patient was evaluated at an outside emergency department and was discharged without any imaging study.

Past medical history was significant for diabetes mellitus, well controlled on oral medication, and the patient was a nonsmoker. His medical history, surgical history, medication list, social history, review of systems, and family history were otherwise unremarkable. His general physical examination was

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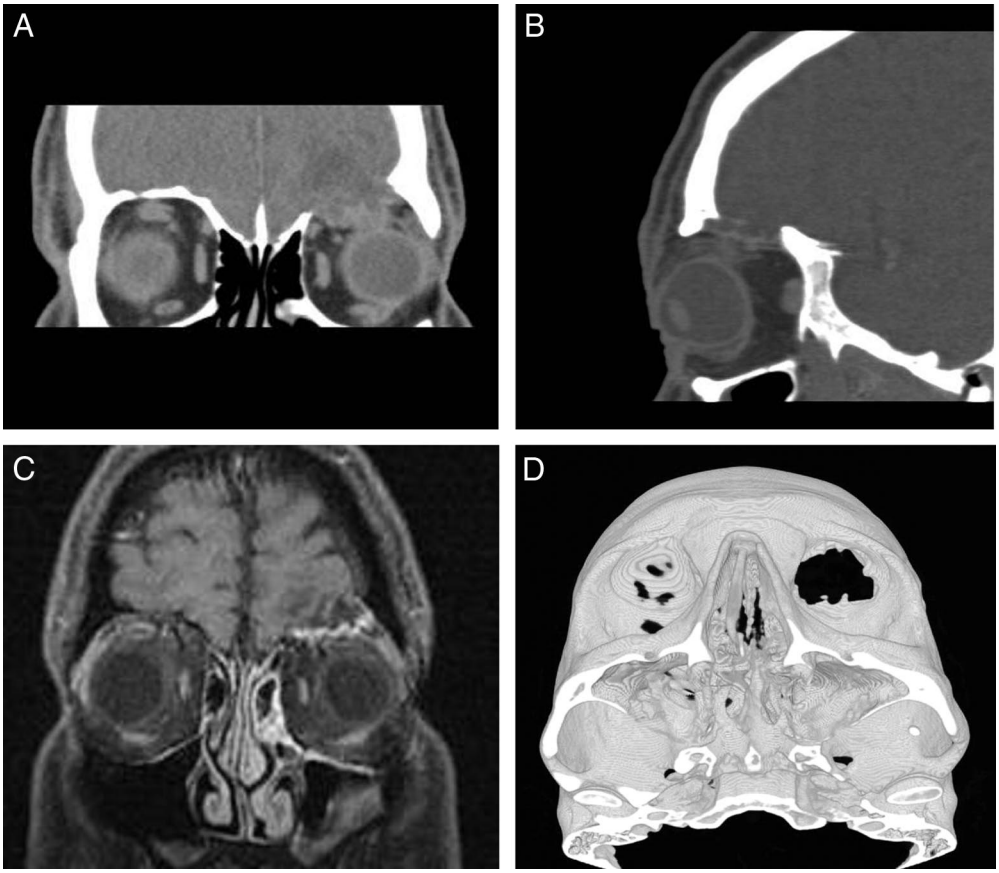

FIG. 1. **A, B,** Coronal **(A)** and sagittal **(B)** orbital CT images show a large left orbital roof defect with a hypodense front orbital lesion. **C,** Coronal MRI after contrast injection demonstrates the left intraorbital encephalocele, left inferior frontal lobe encephalomalacia, and linear enhancement along the margin of the intraorbital encephalocele. The dura cannot be traced over the orbital roof defect, indicating an associated dural defect. **D,** Three-dimensional CT demonstrates the left orbital roof defect.

normal. Neurologic evaluation was unremarkable. Formal neuropsychological testing showed impairment of verbal fluency and scattered, variable mild insufficiencies on verbal reasoning, novel problem solving, attention, and visual spatial organization. His ophthalmic examination was completely normal without evidence of proptosis or ocular motility impairment but a questionable mild hypoglobus of the left eye. CT and MRI of the head showed a large left orbital roof defect with an overlying dural defect, a left intraorbital mass, and left inferior frontal lobe encephalomalacia (Fig. 1A–D). There was no intracranial brain edema, mass effect, or hemorrhage. A metastatic evaluation was negative.

The patient subsequently underwent surgery to confirm the diagnosis and repair the orbital roof defect. A left frontal craniotomy was performed to provide wide exposure to the orbital roof defect. A free bone flap was then reflected, exposing the frontal lobe and leaving the frontal sinus intact. Drill holes were placed in the bony edge and the bone flap. The dura was then incised in horizontal fashion from the midline laterally. The frontal lobe was retracted, exposing a herniated portion of sclerosed gliotic brain that was herniating through the defect in the orbital roof. The portion of the brain that was herniated was amputated and forwarded to the histopathology department (Fig. 2). A pericranial flap was then reflected. This was tacked to the dura posterior to the orbital defect. Six sutures of 4-0 Nurolon were used to tack the pericranial flap posteriorly covering the defect in the orbital roof. Bleeding from the orbital surface of the frontal lobe was controlled with bipolar coagulation and thrombin-soaked Gelfoam. The dura was then sutured, maintaining the integrity of the pericranial flap and allowing it to remain hinged to the scalp. The bone flap

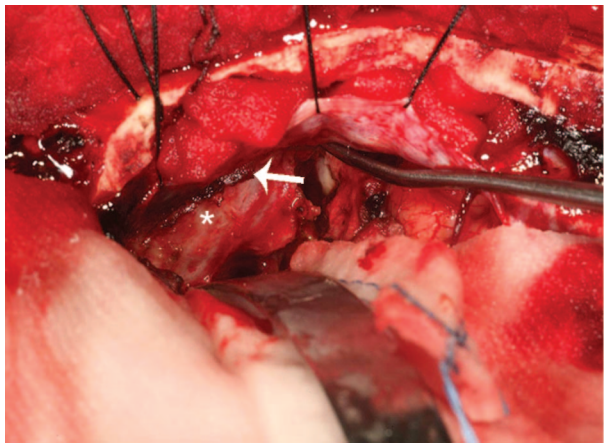

FIG. 2. Intraoperative picture showing the orbital roof defect, defect in the dura over the orbita roof (arrow), and intact superior periorbita (asterisk) after intradural excision of the herniated cerebral gliotic tissue.

was then affixed with 2-0 Nurolon sutures circumferentially. Pathologic examination of the excised intraorbital encephalocele showed glial proliferation. The patient did well postoperatively with no proptosis or eyeball pulsation, but we do not have long-term follow-up results.

DISCUSSION

Orbital encephalocele can be congenital or acquired. Congenital absence of part of the orbital roof, occasionally seen in neurofibromatosis, is one of the causes of orbital roof encephalocele. In this case, congenital absence of the roof can not be totally excluded because no prior imaging was available before his trauma. Acquired orbital roof encephalocele may result from primary orbital roof pathology such as that associated with Gorham syndrome "disappearing bone disease,"[5] from eosinophilic granuloma of the frontal bone, or from inflammatory or malignant bony destructive process originating in the brain, meninges, or adjacent sinuses.[6] Gorham syndrome can also occur following trauma, producing spontaneous, progressive resorption and osteolysis of bone. However, no angiomatosis tissue was identified in the pathologic findings for our patient.

Another potential mechanism of an acquired orbital encephalocele in this case might be a GORF. The exact pathogenesis of GORF is not very clear. An orbital roof fracture with an overlying dural defect is thought to be a prerequisite for its development. In our patient, at the time of surgery a dural defect was identified and repaired. The "mass" seen on the neuroimaging studies was consistent with an encephalocele rather than a neoplasm, and the final pathologic findings were consistent with gliosis from a traumatic encephalocele. Unfortunately, no prior neuroimaging was available for our patient to confirm that the orbital roof encephalocele was not present previously, but we hypothesize that the lesion was acquired rather than congenital.

With continuous pressure and pulsation of the cerebrospinal fluid, the fracture continues to enlarge forming an orbital roof defect ideal for intraorbital encephalocele formation. GORFs were typically found to be present 26 months from the time of the trauma, on average, in a population with a mean age of 6 years.[1] Growing skulls, along with commonality of orbital roof fractures in children, may explain the prevalence of GORF in this age group.

Pulsatile proptosis is the most common presentation of GORF.[1] Diplopia and eyelid swellings are other presenting complaints. Headache and seizures were the presentation of GORF in a 16-year-old girl.[2]

GORF often occurred after minor head trauma for which no imaging was indicated.[1] Months later after the trauma, CT head scans for reported patients with GORF showed diastatic orbital roof fracture with peripheral hyperostotic margins, hypodense or cystic brain parenchymal lesions, and intraorbital masses with cystic appearance. MRI showed more delineation of the intraorbital and inferior frontal lobe cystic lesions, which were surrounded by gliotic tissue and cerebrospinal fluid fistulas.[1]

Spontaneous healing of the growing skull fracture is generally considered to be rare without surgical intervention.[7] Surgical treatment of GORF involves excision of the herniated cerebral gliotic tissue, repair of the dural defect after release of the dural adhesion to the bone defect, and cranioplasty for the orbital roof defect. Cranioplasty may be done with bone graft or pericranial flap. In children, GORF with small orbital roof defect can be managed with duraplasty alone after excision of the intraorbital encephalocele.[3]

In summary, orbital roof encephaloceles may present with or without symptoms in adulthood. The mass-like appearance and bony defect might suggest the diagnosis of orbital neoplasm. Clinical and radiographic clues to the diagnosis include a prior history of trauma, a dural defect, and overlying frontal lobe encephalomalacia without significant intracranial mass effect, brain edema, or enhancement. The GORF may be a potential explanation for the findings even in an adult, but Gorham syndrome, prior congenital encephalocele, and orbital roof defect are additional possible alternative etiologies.

REFERENCES

1. Caffo M, Germanò A, Caruso G, et al. Growing skull fracture of the posterior cranial fossa and of the orbital roof. *Acta Neurochir (Wien)* 2003;145:201–8.
2. Colak A, Akbaçsak A, Biliciler B, et al. An unusual variant of a growing skull fracture in an adolescent. *Pediatr Neurosurg* 1998; 29:36–9.
3. Mohindra S, Mukherjee KK, Chhabra R, Gupta R. Orbital roof growing fractures: a report of four cases and literature review. *Br J Neurosurg* 2006;20:420–3.
4. Giuffrida M, Cultrera F, Antonelli V, et al. Growing-fracture of the orbital roof with post-traumatic encephalocele in an adult patient. Case report. *J Neurosurg Sci* 2002;46:131–4.
5. Krohel GB, Freedman K, Peters GB 3rd, Popp AJ. Gorham disease of the orbit. *Am J Ophthalmol* 2002;133:729–30.
6. Paufique L, Allegre G, Durand L, Magnard G. [3 cases of unilateral exophthalmos caused by tumor of the orbit in children: an eosinophilic granuloma of the frontal bone, a meningo-encephalocele of the orbit, and a mucocele of the roof of the orbit. Neurosurgical approach.] *Bull Soc Ophtalmol Fr* 1968;68:477–81.
7. Jamjoom ZA. Growing fracture of the orbital roof. *Surg Neurol* 1997;48:184–8.

Postoperative Maxillary Sinusitis Mimicking Tumor Recurrence on Positron Emission Tomography

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Abstract: **A significant overlap of standard uptake values occurs in a variety of pathological processes, potentially causing misdiagnosis for tumor recurrence. The authors present a case of acute maxillary sinusitis masquerading as local recurrence of malignancy.**

Since its introduction in the 1970s, F18-fluorodeoxyglucose positron emission tomography (FDG-PET) has become increasingly important in the detection and staging of malignant disease, and due to its ability to differentiate post-treatment changes from residual and/or recurrent tumor, FDG-PET has assumed an important role in the long-term surveillance of cancer survivors. However, FDG uptake reflects glucose metabolism in all living cells and therefore is neither totally specific nor totally sensitive for the identification of tumor cells, with a significant overlap in stan-

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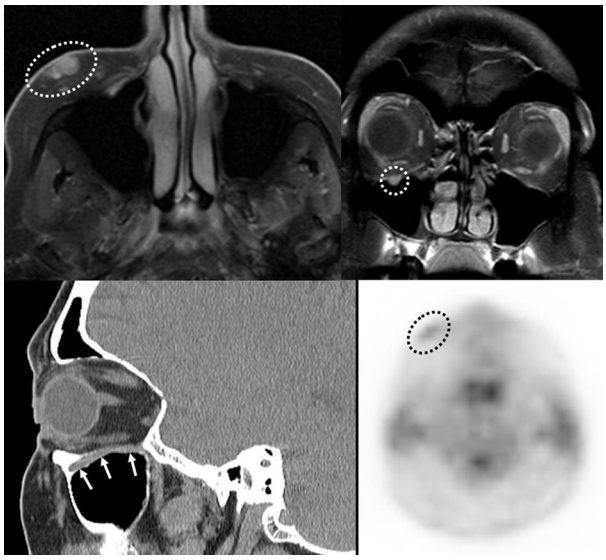

FIG. 1. Top: Magnetic resonance images, T1 weighted after contrast with fat saturation demonstrating a soft tissue mass in the right cheek (dotted line, left) and an enlarged infraorbital nerve (dotted line, right). **Bottom left:** Parasagittal CT image showing an enlarged infraorbital nerve within its canal (arrows). **Bottom right:** Axial positron emission tomography image with an increased standard uptake value (2.75) in the right cheek (dotted line).

dard uptake values (SUVs) between postoperative scarring, infection, noninfectious inflammation, low-grade malignancy, and high-grade malignancy.[1] False-positive studies are therefore a distinct possibility.

We present a case of acute maxillary sinusitis masquerading as local recurrence of a periorbital malignant peripheral nerve sheath tumor.

CASE REPORT

A 68-year-old woman presented for evaluation of a lesion of her right anterior malar eminence. Biopsy demonstrated a malignant spindle-cell neoplasm. In addition to the mass, examination revealed hypesthesia in the distribution of the inferior orbital nerve. MRI demonstrated an enlarged and enhancing inferior orbital nerve without apparent intracranial spread, and PET/CT showed a focal hypermetabolic lesion in the area of known tumor (SUV 2.75) (Fig. 1). The patient underwent wide surgical extirpation of the tumor with resection of the infraorbital rim and anterior maxillary wall. The inferior orbital nerve was resected to the foramen rotundum for negative margins. Orbital reconstruction involved a porous polyethylene and titanium mesh orbital floor implant, and a radial forearm free flap was used to reconstruct the soft tissue of the cheek.

Because final pathologic findings revealed a malignant peripheral nerve sheath tumor, the patient underwent adjuvant radiation therapy. Several months after completion, a mass was noted over the medial canthus. MRI revealed interval enhancement and opacification of the right sinonasal cavity (Fig. 2), obscuring in-office endoscopic evaluation for recurrent tumor. PET/CT demonstrated increased metabolic activity in the right maxillary sinus and orbital floor extending to the skull base (SUV 6.82), which raised concerns of tumor recurrence (Fig. 3).

The patient was taken back to the operating room for exploration. Mucosal scarring was noted intraoperatively and thickened maxillary sinus mucosa was debrided with drainage of a loculated abscess cavity in the pterygopalatine fossa and inferior orbital apex. The orbital floor and rim hardware were removed. Soft tissue of the maxillary sinus, pterygopalatine fossa, and orbital apex was resected to the underlying bone. Frozen and permanent pathologic findings were negative for tumor recurrence, showing acute and chronic inflammatory changes only. The patient was treated for bacterial sinusitis postoperatively.

Follow-up PET/CT several months later demonstrated decreasing activity (SUV 6.53), suggesting a benign etiology, such as postoperative change and resolving inflammation (Fig. 4). The patient has since been observed for 12 months without clinical or radiographic evidence of tumor recurrence.

DISCUSSION

Malignant peripheral nerve sheath tumors are uncommon but aggressive sarcomas that most commonly arise in young and middle-aged adults.[2] While there is some disagreement in the literature as to whether a threshold SUV can distinguish malignant peripheral nerve sheath tumor from benign counterparts,[3,4] some suggest that FDG-PET can be a useful technique in detecting recurrence of the tumor, especially in patients who have undergone radiation treatment.[5] In the present case, MRI suggested a local recurrence at the skull base, and FDG-PET demonstrated an SUV that increased from 2.75 to 6.82 after surgical extirpation and

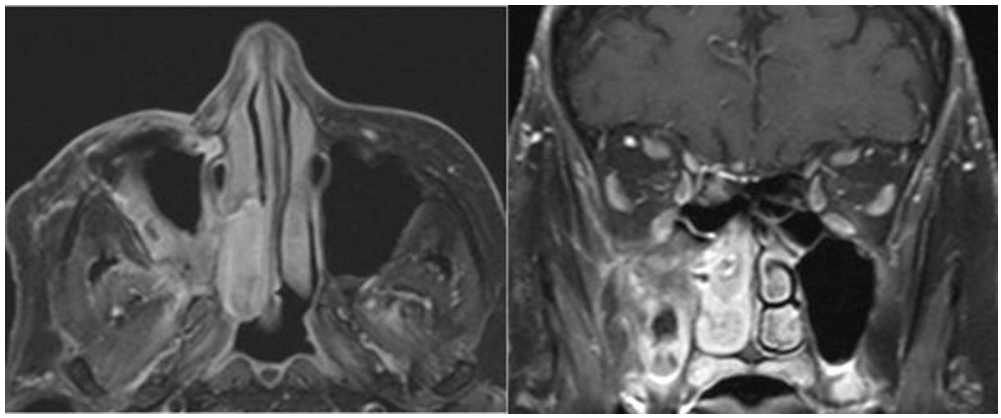

FIG. 2. Magnetic resonance images, T1 weighted after contrast with fat saturation, axial (left) and coronal (right). Soft tissue opacification is noted in the right maxillary sinus extending in the pterygopalatine fossa.

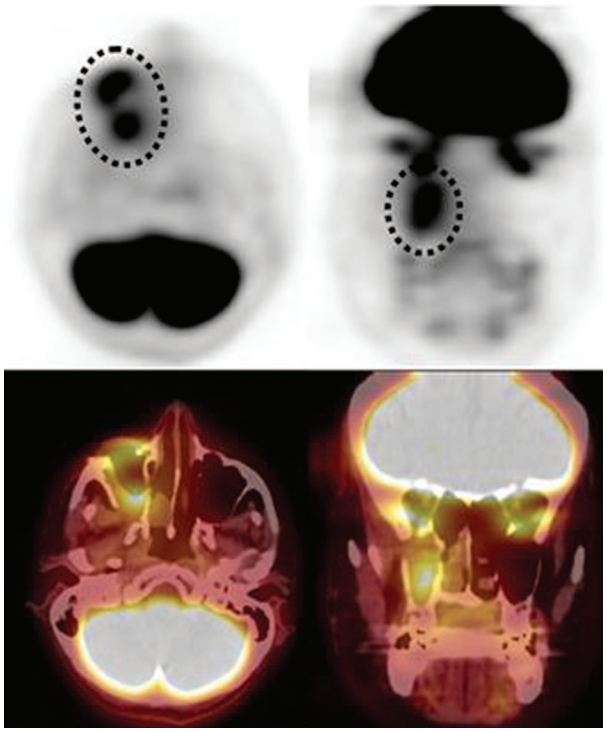

FIG. 3. Positron emission tomography (top) and positron emission tomography/CT fusion (bottom) images with a marked uptake in the left maxillary sinus and pterygopalatine fossa (standard uptake value = 6.82) (dotted lines) suggestive of tumor recurrence.

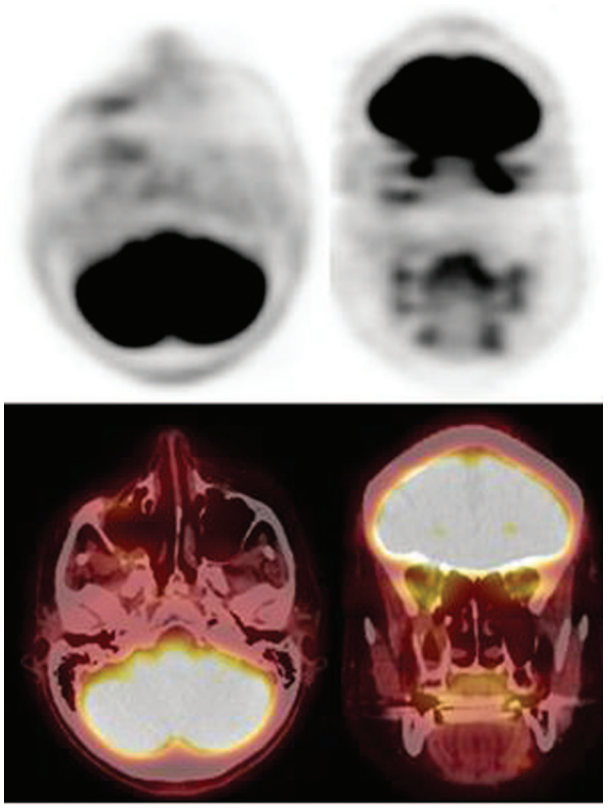

FIG. 4. Positron emission tomography (top) and positron emission tomography/CT fusion (bottom) images following exploration and drainage of infection. The area of increased uptake is markedly reduced, with a slight decrease in metabolic activity (standard uptake value = 6.53).

adjuvant radiotherapy, but tissue histology showed infectious and inflammatory tissue only.

In addition to the role of FDG-PET in the surveillance of patients with known malignancy, there is a growing body of evidence suggesting that it may be useful in the diagnosis and management of infectious disease, particularly in immunosuppressed patients, who may harbor clinically silent infections,[6] and in patients with metallic implants,[7] as FDG uptake, in contrast to MRI and CT, is not hampered by metallic artifacts. FDG uptake in infection is mediated by mononuclear cells and granulocytes, which use large amounts of glucose in a "respiratory burst" while fighting infection.[7] The amount of uptake can be intense, depending on the presence of certain infectious humoral stimuli.[7] In a large group of immunosuppressed patients with a known infection, the mean SUV was 6.07 (range 0.8–22.1), comparable to what was seen for our patient.[6] In a case similar to ours, a patient with nasopharyngeal carcinoma with an SUV of 6.37 on PET/CT demonstrated simultaneous acute sinusitis (SUV 5.24), simulating tumor invasion.[8]

While there is ongoing research in the applicability of PET/CT in the evaluation of infectious disease, there is some evidence that "dual-time-point" (i.e., serial) measurement of SUVs may aid in distinguishing inflammatory from malignant causes. Zhuang et al.[9] demonstrated that the SUVs of infectious processes were either stable or decreased over time, while malignant lesions showed an increase in SUV between measured time points. The authors suggested that in difficult cases, such dual-time-point measurement may provide a better way to differentiate benign and malignant processes in FDG-PET imaging.

Our case of acute sinusitis simulating a recurrence of a malignant peripheral nerve sheath tumor highlights the possibility of false-positive studies in patients undergoing surveillance PET/CT after cancer treatment of the orbit and periorbital anatomy. Despite the ever-growing indications for and popularity of noninvasive studies such as FDG-PET in the evaluation of such patients, the role of clinical, surgical, and histopathologic evaluation remains crucial.

REFERENCES

1. Bakheet SM, Powe J. Benign causes of 18-FDG uptake on whole body imaging. *Semin Nucl Med* 1998;28:352–8.
2. Lyons CJ, McNab AA, Garner A, et al. Orbital malignant peripheral nerve sheath tumours. *Br J Ophthalmol* 1989;73:731–8.
3. Benz MR, Czernin J, Dry SM, et al. Quantitative F18-fluorodeoxyglucose positron emission tomography accurately characterizes peripheral nerve sheath tumors as malignant or benign. *Cancer* 2010;116:451–8.
4. Beaulieu S, Rubin B, Djang D, et al. Positron emission tomography of schwannomas: emphasizing its potential in preoperative planning. *Am J Roentgenol* 2004;182:971–4.
5. Halac M, Cnaral F, Sait S, et al. FDG PET/CT findings in recurrent malignant schwannoma. *Clin Nucl Med* 2008;33:172–4.
6. Mahfouz T, Miceli MH, Saghafifar F, et al. 18F-fluorodeoxyglucose positron emission tomography contributes to the diagnosis and management of infections in patients with multiple myeloma: a study of 165 infectious episodes. *J Clin Oncol* 2005; 23:7857–63.
7. Srobel K, Stumpe K. PET/CT in musculoskeletal infection. *Sem Musculoskelet Radiol* 2007;11:353–64.
8. Chan SC, Ng SH, Tzu-Chen Y, et al. False-positive findings on F-18 fluoro-2-deoxy-D-glucose positron emission tomography in a patient with nasopharyngeal carcinoma and extensive sinusitis. *Clin Nucl Med* 2005;30:62–3.

9. Zhuang H, Pourdehnad M, Lambright ES, et al. Dual time point 18F-FDG PET imaging for differentiating malignant from inflammatory processes. *J Nucl Med* 2001;42:1412–7.

Preoperative Radiation Therapy in the Management of Recurrent Orbital Hemangiopericytoma

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Abstract: **A 77-year-old man presented for evaluation of a recurrent right orbital hemangiopericytoma. It had been excised twice over the past 6 years. After the second resection, the tumor rapidly recurred and was insensitive to systemic chemotherapy, and the patient was thus referred to the authors' institution. The patient had proptosis, restricted ocular movement, and binocular diplopia on presentation. Orbital imaging confirmed a well-circumscribed right extraconal mass in the medial orbit. Preoperative radiation therapy was given, which reduced the tumor volume considerably and allowed a successful complete surgical excision of the mass.**

Hemangiopericytoma is a rare mesenchymal tumor that can affect the orbit. Surgical resection is the treatment of choice for localized tumors. Complete excision of this hypervascular tumor can be challenging, as intraoperative bleeding may be profuse, and tumor recurrence following incomplete excision is common. Adjuvant chemotherapy and radiation therapy have historically been used in the management of recurrent cases of hemangiopericytoma in anatomic locations other than the orbit.[1] We herein report what may be the first successful use of preoperative neoadjuvant radiation therapy for recurrent orbital hemangiopericytoma to help reduce tumor volume and facilitate complete surgical excision.

CASE REPORT

A 77-year-old man was referred to our institution for management of a recurrent hemangiopericytoma of the right orbit. Six years prior, the patient began experiencing decreased vision and proptosis of the right eye. Imaging showed a right extraconal medial orbital mass, and excisional biopsy confirmed the tumor to be a malignant hemangiopericytoma, with positive surgical margins. He remained stable for the next 4 years until he developed binocular diplopia. CT showed a recurrent mass in the right orbit. Surgical excision of the recurrent mass again showed hemangiopericytoma with tumor present at the surgical margin. Postoperative imaging studies showed residual tumor. Four months later, there was radiographic evidence of further tumor growth. Two cycles of systemic chemotherapy consisting of ifosfamide and epirubicin were given but failed to yield a clinical response. The patient was then referred to our institution for further recommendations and treatment options.

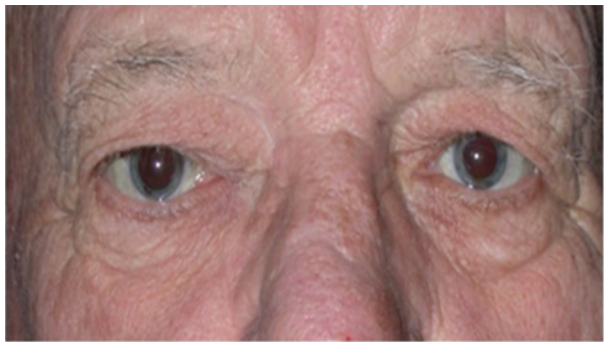

FIG. 1. Patient at presentation with right globe proptosis and superior periorbital fullness.

On examination, visual acuity was 20/30 and 20/25 for the right and left eye, respectively. There was no relative afferent pupillary defect. He had restricted supraduction and adduction of the right globe and binocular diplopia in left gaze. He had 4 mm of right proptosis and superior periorbital fullness (Fig 1).

Orbital CT showed a 2.0- × 2.5- × 1.6-cm, enhancing, well-circumscribed ovoid mass in the superomedial right orbit interposed between the superior and medial recti (Fig 2A, B). Systemic workup was negative for metastatic disease.

After input from the medical and radiation oncology services, in light of the tumor's poor response to prior chemotherapy, and 2 prior failed attempts at surgical excision, the patient received high-dose neoadjuvant external beam radiation of 50 Gy to the right orbit. The rationale for giving radiation therapy preoperatively as opposed to postoperatively was because preoperative delivery of radiation meant that the total radiation dose would be 10 Gy less than that required to adequately treat with radiation therapy in the postoperative setting. This is based on common radiation treatment planning guidelines that recommend a higher dose of radiation for a recently surgically treated bed due to concerns for compromise of the blood supply to the bed of tissue after surgery. As a general rule, 10 Gy more of total radiation dose is delivered in the postsurgical setting compared with the presurgical situation. The 60 Gy of radiation that would have to be given to our patient in the postoperative setting would put the patient at an unacceptably higher risk for optic neuropathy and consequent visual loss in the involved eye. While shrinkage of tumor was not necessarily an expected outcome from presurgical delivery of radiation, the orbital tumor decreased in size to 1.1 × 1.2 × 0.8 cm, an 87% decrease in total volume (Fig 2C, D). The tumor was then completely excised through a transcutaneous anterior orbitotomy. Histopathology confirmed the lesion to be malignant hemangiopericytoma (Fig 3). Postoperatively, the patient's ocular motility, diplopia, and proptosis improved. There was no clinical or radiographic evidence of local recurrence at last follow-up, 12 months after the most recent surgical resection (Fig. 4).

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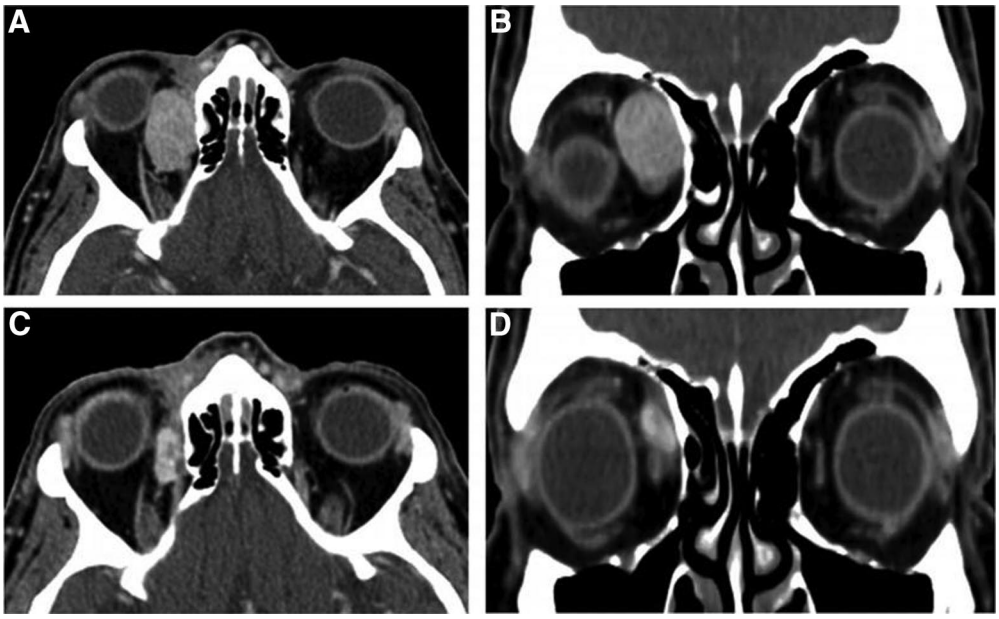

FIG. 2. **A, B,** Axial **(A)** and coronal **(B)** CT with contrast demonstrate an enhancing, well-circumscribed, ovoid, extraconal, superomedial right orbital mass without bony erosion. **C, D,** Axial **(C)** and coronal **(D)** CT with contrast after radiation therapy show a drastic decrease in size of the tumor.

DISCUSSION

Hemangiopericytoma is a rare, hypervascular mesenchymal tumor that arises from pericytes. It accounts for 1% to 3% of all orbital biopsies.[2] It is a well-circumscribed tumor containing a pseudocapsule, most commonly found in the superior orbit.[2] The middle aged are most commonly affected, with a median age of 42 years (range, 0–90 years).[2] Histologically,

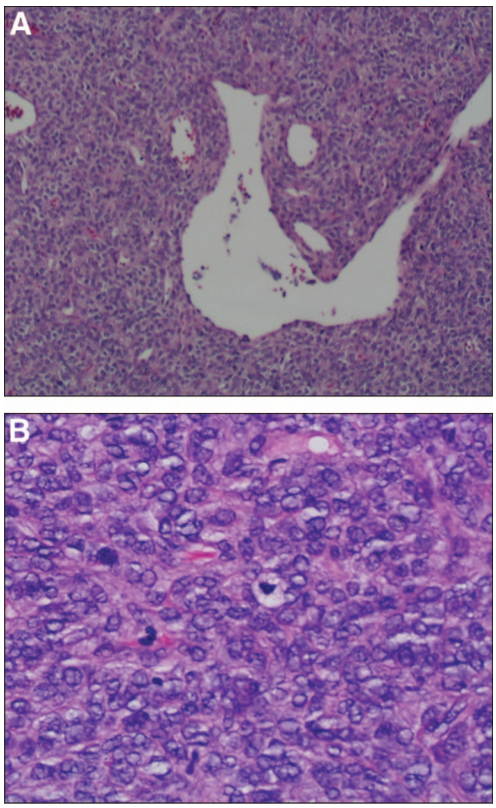

FIG. 3. Histopathologic findings. **A,** Staghorn vascular channels surrounded by densely packed spindle cells (hematoxylin-eosin, ×10). **B,** Mixture of spindle and epithelioid cells with mitotic figures (hematoxylin-eosin, ×40).

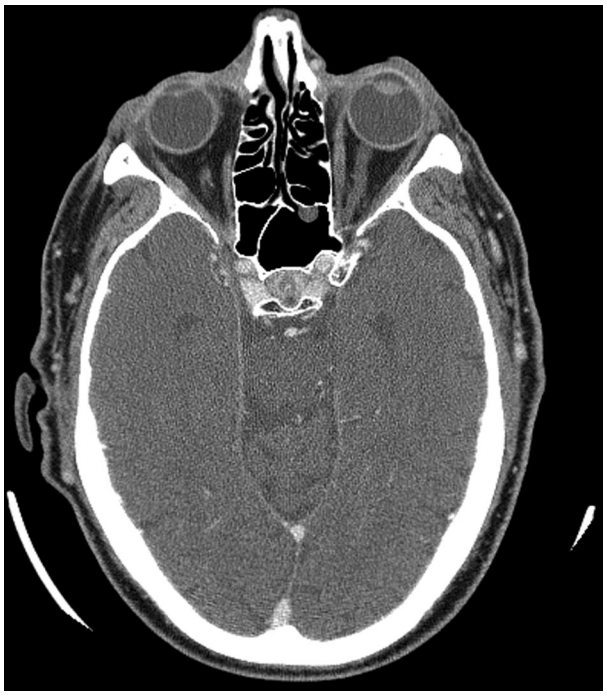

FIG. 4. Axial CT with contrast does not show any signs of recurrence of orbital hemangiopericytoma at 12-month follow-up.

hemangiopericytomas typically display high cellularity, a small amount of interstitial collagen, and a high number of mitotic figures. These tumors typically follow a benign indolent course, causing slowly progressive and painless proptosis. Malignant potential does exist, and the 5-year rate of metastasis is estimated to be approximately 15%.[3] Orbital imaging is warranted to investigate the extent of the tumor, and biopsy is necessary to establish the diagnosis.

Surgical resection is the treatment of choice for symptomatic tumors. Complete excision can be challenging owing to the tumor's vascularity, friable nature, and lack of a true capsule in most cases. Tumors have a recurrence rate of 33%.[3] The relatively high rate of local recurrence is largely thought to be due to incomplete surgical excision. Treatment options previously described for recurrent hemangiopericytomas include the following: orbital exenteration, further attempts at surgical excision, excision combined with adjuvant radiation therapy, and chemotherapy.[4] We found no other reports in the literature describing preoperative neoadjuvant radiation therapy in the treatment of these tumors.

There are multiple advantages to preoperative radiation therapy, and this modality has been used successfully in the past for the treatment of soft tissue sarcomas in other locations.[5] Local control outcomes for adjuvant radiation therapy in management of soft tissue sarcomas are the same whether the radiation is delivered preoperatively or postoperatively. However, studies have shown that postoperative radiation therapy may be associated with worse long-term functional outcomes for patients treated for soft tissue sarcomas of the extremity.[6] This is likely a result of the increased volume of tissue irradiated and the higher dose needed (60 Gy) when patients are treated postoperatively. Preoperative radiation therapy for soft tissue sarcoma requires only 50 Gy to be administered to the tumor plus a margin rather than an entire surgical field plus a margin. In this case, the 10-Gy difference in dose was significant in that the postoperative dose of 60 Gy would exceed the tolerance dose of the optic nerve (50 Gy) for an unacceptably high risk of optic neuropathy and visual loss.[7] Furthermore, although shrinkage of tumor was not the goal of radiation therapy, the tumor did shrink considerably, and this facilitated surgical excision. This case therefore illustrates another potential benefit of preoperative radiation therapy: the potential to downsize a tumor in the orbit.

There are risks associated with the administration of radiation therapy before surgery. Investigations of its use for soft tissue sarcoma occurring in the extremity show that preoperative radiation therapy was associated in some cases with higher risks of wound complications after surgery.[5] Additionally, preoperative treatment may delay definitive surgical resection. It is recommended that surgical resection be undertaken no sooner than 4 weeks after the completion of radiation therapy to allow resolution of the acute inflammation associated with radiation therapy. However, surgery should be performed within a 4- to 8-week window, before significant scarring of orbital soft tissues has occurred.

Multidisciplinary management of orbital mesenchymal tumors is critical. The involvement of medical and radiation oncologists with experience in treating such tumors is needed to consider all treatment options and the appropriate timing of treatment modalities. As demonstrated in the patient described in this report, preoperative radiation therapy can be considered in patients with radiosensitive tumor subtypes and can be a useful adjunct to surgery. Further studies with larger numbers of patients and with follow-up time of longer than 12 months are needed to ascertain whether preoperative radiation therapy can decrease local recurrence rates for orbital hemangiopericytomas.

REFERENCES

1. Park MS, Araujo DM. New insights into the hemangiopericytoma/solitary fibrous tumor spectrum of tumors. *Curr Opin Oncol* 2009; 21:327–31.
2. Karcioglu ZA, Nasr AM, Haik BG. Orbital hemangiopericytoma: clinical and morphologic features. *Am J Ophthalmol* 1997;124:661–72.
3. Croxatto JO, Font RL. Hemangiopericytoma of the orbit: a clinicopathologic study of 30 cases. *Hum Pathol* 1982;13:210–8.
4. Bernardini FP, de Conciliis C, Schneider S, et al. Solitary fibrous tumor of the orbit: is it rare? Report of a case series and review of the literature. *Ophthalmology* 2003;110:1442–8.
5. O'Sullivan B, Davis AM, Turcotte R, et al. Preoperative versus postoperative radiotherapy in soft-tissue sarcoma of the limbs: a randomised trial. *Lancet* 2002;359:2235–41.
6. Davis AM, O'Sullivan B, Bell RS, et al. Function and health status outcomes in a randomized trial comparing preoperative and postoperative radiotherapy in extremity soft tissue sarcoma. *BJ Clin Oncol* 2002;20:4472–7.
7. Danesh-Meyer HV. Radiation-induced optic neuropathy. *J Clin Neurosci* 2008;15:95–100.

Topical Antiglaucoma Treatment With Prostaglandin Analogues May Precipitate Meibomian Gland Disease

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Abstract: **The popular use of topical prostaglandin analogues (TPAs) in glaucoma patients has a well-documented association with changes in number, length, thickness, and growth pattern of eyelashes. A link between meibomian gland disease and seborrheic blepharitis resulting from topical administration of prostaglandin analogues has not been made to the authors' knowledge. A retrospective study of 43 patients presenting for incision and curettage of chalazion to the operating theatre over a 2-year period was performed. The patients' histories were reviewed to establish whether they were using TPAs for glaucoma at the time of surgery. Eight patients (19%) were using TPAs at the time of surgery. No patient had a history of eyelid margin disease before the commencement of TPAs. The exact pathogenesis of how meibomian gland disease is linked to TPA use is unclear. We postulate that the topical application of prostaglandin analogues may contribute to the formation of chalazion by acting directly to stimulate meibomian gland secretion. The authors present the findings of a preliminary case series showing a higher-than-expected incidence of patients using TPAs requiring surgical intervention for chalazion.**

Synthetic prostaglandin analogues are manufactured to bind to a prostaglandin receptor. The popular use of topical prostaglandin analogues (TPAs) in glaucoma patients has a well-documented association with changes in number, length,

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thickness, and growth pattern of eyelashes.[1] Its exact pathogenesis, however, still remains unclear. Houssay et al.[2] demonstrated the modulation of hair growth by prostaglandins in 1976, long before the introduction of TPAs. Meibomian gland disease and seborrheic blepharitis resulting from topical administration of prostaglandin analogues have not to our knowledge been studied or reported. It came to our attention that a significant number of patients presenting for management of blepharitis and meibomian gland disease were using a variety of topical agents within the prostaglandin analogue family of drugs. This study aimed, therefore, to evaluate whether the use of TPAs contributed to a higher incidence of patients presenting for chalazion surgery to the operating theatre.

CASE REPORT

A retrospective study of 43 consecutive patients presenting for incision and curettage of chalazion to the operating theatre of 2 oculoplastic surgeons over a 2-year period was performed. The patients' histories were reviewed to establish whether they were using TPAs for glaucoma at the time of surgery in the operated eye.

Forty-three patients were sampled. The male-to-female ratio was 23:20. Patients ranged in age from 1 to 90 years with a mean of 48. Eight patients (19%) were using TPA monotherapy to manage their glaucoma at the time of surgery. No patient had a history of eyelid margin disease symptoms or documentation of its treatment before the commencement of TPAs. All 8 patients using TPAs at the time of surgery were doing so for a minimum period of 1 year and a maximum of 5 years. Four patients of the 8 were using latanoprost, and 4 patients were using bimatoprost.

DISCUSSION

The exact pathogenesis of how meibomian gland disease is linked to TPAs remains unclear. The presence of prostaglandin receptors within the meibomian gland itself has not been discussed to date to our knowledge. Human sebocytes, however, express prostaglandin receptors,[3] and the induction of prostaglandin pathways has been demonstrated in sebocytes of facial skin and implicated in troublesome facial acne.[4] Sebaceous gland function is influenced by androgens, estrogens, glucocorticoids, and prolactin. Sebaceous gland lipids are known to have important antibacterial effects, and perhaps this property is affected by abnormal gland functioning, encouraging bacterial overgrowth on the eyelid margin.[5]

We postulate that topical application of prostaglandin analogues may contribute to the formation of chalazion by acting directly to stimulate or alter meibomian gland secretion. This can result in a seborrheic-type blepharitis with meibomian gland outlet obstruction, chalazion, and secondary tear film abnormalities typical of meibomian gland dysfunction previously studied by Jackson[6] in Canada. Li et al.[7] found that travoprost and latanoprost were equivalent with respect to intraocular pressure lowering, but conjunctival hyperemia was less with latanoprost. Of the patients in this study using TPAs, the incidences of chalazion were equal in those patients using latanoprost and bimatoprost preparations. Interpretation of the data in this study is limited by small numbers, the lack of an age-matched control group, and the lack of previously published data postulating the link between TPAs and chalazion occurrence.

We present the findings of a preliminary small case series highlighting a higher-than-expected incidence of patients on TPAs with blepharitis requiring surgical intervention for chalazion. Further study is required to elucidate the precise link of TPAs to meibomian gland disease and the mechanisms involved.

REFERENCES

1. Johnstone MA, Albert DM. Prostaglandin-induced hair growth. Review. *Surv Ophthalmol* 2002;47(suppl 1):S185–202.
2. Houssay AB, Arias NH, Davison TA, Epper CE. Effects of prostaglandins upon hair growth in mice. *Acta Physiol Lat Am* 1976;26:186–91.
3. Chen W, Tsai SJ, Wang CA, et al. Human sebocytes express prostaglandin E2 receptors EP2 and EP4 but treatment with prostaglandin E2 does not affect testosterone production. *Br J Dermatol* 2009;161:674–7.
4. Alestas T, Ganceviciene R, Fimmel S, et al. Enzymes involved in the biosynthesis of leukotriene B4 and prostaglandin E2 are active in sebaceous glands. *J Mol Med* 2006;84:75–87.
5. Zouboulis CC. Acne and sebaceous gland function. *Clin Dermatol* 2004;22:360–66.
6. Jackson WB. Blepharitis: current strategies for diagnosis and management. *Can J Ophthalmol* 2008;43:170–9.
7. Li N, Chen XM, Zhou Y, et al. Travoprost compared with other prostaglandin analogues or timolol in patients with open-angle glaucoma or ocular hypertension: meta-analysis of randomized controlled trials. *Clin Experiment Ophthalmol* 2006;34:755–64.

Tuberculosis of the Nasolacrimal Duct

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Abstract: **Nasolacrimal tuberculosis is rare. The authors present a young Peruvian-born female with subacute onset of right eye epiphora, isolated right inferior turbinate enlargement, and ipsilateral cervical lymphadenopathy. Turbinate and neck mass incisional biopsies demonstrated histopathological findings consistent with tuberculosis. QuantiFERON-TB Gold-In-Tube testing was positive. Complete resolution of symptoms occurred after dacryocystorhinostomy and 9 months of standard 4-drug antituberculosis therapy. Tuberculosis of the nasolacrimal duct is highly unusual but should be considered in patients with tuberculosis risk factors who present with nasolacrimal obstruction from an inferior turbinate mass.**

Active tuberculosis disease of the head and neck is rare.[1] We describe an extremely unusual presentation of postprimary tuberculosis of the nasolacrimal system with inferior turbinate enlargement and cervical adenopathy.

CASE REPORT

A 39-year-old Peruvian-born female living in Portland, Oregon for 9 years had onset of right-sided epiphora in Feb-

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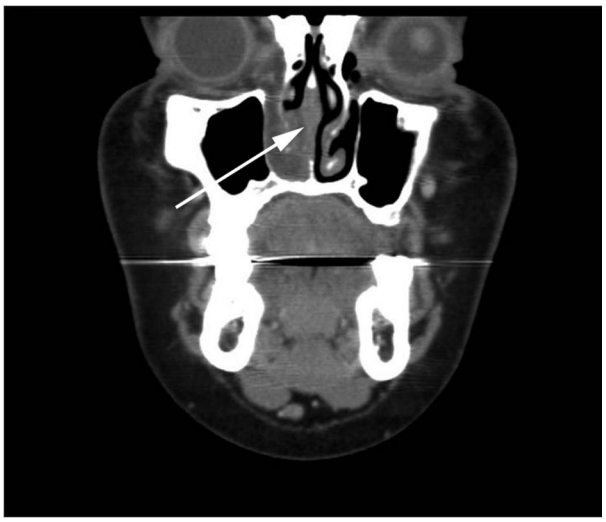

FIG. 1. CT examination confirming a mass (arrow) involving the right inferior nasal turbinate.

ruary, 2008. Within several weeks, she also noted an ipsilateral swollen, painful, cervical lymph node that continued to enlarge. Her epiphora continued to worsen until she presented to an otolaryngologist 7 months later. The patient reported palpitations, 10-lb weight loss, and right nasal obstructive symptoms but denied having pulmonary or constitutional symptoms. She could not recall any exposure to sick contacts or pets and reported no significant travel history besides her immigration from Peru. She was otherwise treated for hypertension. Physical examination revealed an enlarged, indurated right inferior turbinate with multiple enlarged, nontender, right submandibular and upper jugular lymph nodes. CT examination confirmed a right inferior nasal turbinate mass (Fig. 1) and extensive enlargement of the right cervical lymph nodes with necrotic appearance (Fig. 2). A complete blood count with differential revealed a mildly elevated red cell count. An antineutrophil cytoplasmic antibody evaluation was negative and the angiotensin-converting enzyme level was normal. Acute antibodies to cytomegalovirus, Epstein-Barr virus, mycoplasma, and bartonella were not identified on serologic testing. Fine-needle aspiration biopsy of the right cervical lymph node and direct biopsy of the inferior turbinate were inconclusive.

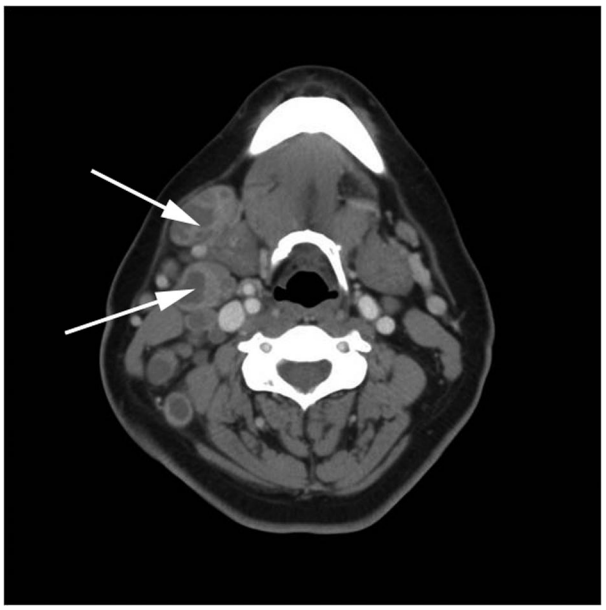

FIG. 2. CT depicting extensive enlargement of the right cervical lymph nodes (arrows) with necrotic appearance.

Subsequent incisional biopsy of the turbinate and a cervical lymph node demonstrated extensive fibrosis, necrosis, and caseating granulomas. Stains, cultures, and polymerase chain reaction analysis of tissue were negative for acid-fast bacilli and fungus. The patient was then referred to an infectious disease specialist. On further testing, the patient's QuantiFERON-TB Gold-In-Tube (Cellestis, Victoria, Australia) result was positive, consistent with previous tuberculosis exposure. Her chest radiograph was normal. She began empiric 4-drug antituberculous therapy with isoniazid, rifampin, pyrazinamide, and ethambutol.

After 2 months of therapy, she had vast improvement of her right cervical lymphadenopathy but reported continued epiphora. Nasal and lacrimal examination revealed no residual mass but suggested a narrowed, scarred duct, which was successfully treated via right endoscopic dacryocystorhinostomy. The patient subsequently continued twice-weekly isoniazid, rifampin, and ethambutol for a total of 9 months, with a full resolution of lymphadenopathy.

DISCUSSION

In the United States, tuberculosis presents as pulmonary disease 80% of the time, with fewer than 2.7% of infections occurring in the head or neck.[2] Of head and neck infections, cervical adenopathy is the most common finding (87%) and occurs primarily in young, immunocompetent individuals.[1,3] Cultures from the head and neck are frequently negative or not obtained for tuberculosis.[1,4] Appropriate diagnosis usually occurs through histologic evaluation. Tuberculous cervical lymphadenitis is frequently accompanied by other disease sites, especially pulmonary. Because tuberculosis is associated with immunosuppressed conditions such as underlying malignancies, a neoplastic process should be considered.

A literature review revealed only a small number of reported nasolacrimal tuberculosis cases. Some of these cases were initially diagnosed histopathologically.[4,5,6] In 2 of these cases, active pulmonary disease was discovered following identification of tuberculosis in the lacrimal gland.[4,5] Both of these patients developed cervical lymphadenopathy and were from areas of tuberculosis endemicity.[4,5] One reported patient had no other sites of active disease and presented only with epiphora and recurrent lacrimal passage infections.[6] Isolation of tuberculosis from the nasolacrimal duct is uncommon, and diagnosis is often made by the appearance of granulomas on histopathological evaluation.[4] Our tissue biopsies failed to isolate *Mycobacterium tuberculosis*, which is also not uncommon in scrofula.[1] QuantiFERON-TB Gold-In-Tube testing and tuberculin skin testing are potentially useful adjuncts to making the diagnosis.

CONCLUSION

We present a patient with tuberculosis of the right inferior turbinate and associated lacrimal ductal obstruction. It is likely that this focus of disease drained to regional neck lymph nodes, resulting in scrofula. Tuberculosis of the nasolacrimal duct is a highly unusual cause of turbinate enlargement but should be considered in the differential diagnosis in patients with tuberculosis exposure and mass obstruction of the nasolacrimal system.

REFERENCES

1. Menon K, Bem C, Gouldesbrough D, Strachan DR. A clinical review of 128 cases of head and neck tuberculosis presenting over a 10-year period in Bradford, UK. *J Laryngol Otol* 2007;121:362–8.
2. Centers for Disease Control and Prevention. Reported tuberculosis in the United States, 2008. September 2009. Available at: http://www.cdc.gov/tb/statistics/reports/2008/pdf/2008report.pdf. Accessed February 2010.
3. Polesky A, Grove W, Bhatia G. Peripheral tuberculous lymphadenitis: epidemiology, diagnosis, treatment, and outcome. *Medicine (Baltimore)* 2005;84:350–62.
4. van Assen S, Lutterman JA. Tuberculous dacryoadenitis: a rare manifestation of tuberculosis. *Neth J Med* 2002;60:327–9.
5. Sen DK. Tuberculosis of the orbit and lacrimal gland: a clinical study of 14 cases. *J Pediatr Ophthalmol Strabismus* 1980;17:232–8.
6. Al-Malki AF, Issa TM, Riley F, Karcioglu ZA. Nasolacrimal tuberculosis in a patient with conjunctivodacryocystorhinostomy. *Ophthal Plast Reconstr Surg* 1999;15:213–6.

Treatment of a Lacrimal Pneumocele With Punctal Plug Insertion

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Abstract: **A 37-year-old man presented with symptoms of air and mucus reflux from the right medial canthus on blowing his nose. Examination showed crepitus in the region overlying the right lacrimal sac, and he was able to produce bubbles in his conjunctival lake on instillation of topical 2% fluorescein. Dacryoscintigraphy showed symmetrical slow passage of tracer from the lacrimal sac in the nasolacrimal duct indicating delay at the postsac level. Dacryocystography showed that the right lacrimal system contained round filling defects, suggesting the presence of soft tissue tumors. However, a CT scan showed an enlarged right lacrimal fossa with air in the nasolacrimal duct and no evidence of a mass. Following lower punctal plug occlusive canaliculoplasty on the right, using a preloaded silicone plug, the patient noted an immediate resolution of reflux symptoms.**

Air reflux from the nasolacrimal system is an uncommon but potentially troublesome feature following dacryocystorhinostomy. It is particularly rare in the absence of previous lacrimal surgery. We describe a case of lacrimal pneumocele presenting with air reflux that was successfully managed with punctal plug insertion.

CASE REPORT

A 37-year-old man presented with air and mucus reflux from the right medial canthus on blowing his nose. He produced a "squeaky" sound on palpation of that area. There was no history of hemolacria, previous surgery, or trauma, and the patient reported no preceding features of nasolacrimal duct obstruction. Examination showed crepitus in the right medial canthal region overlying the lacrimal sac. Following topical instillation of 2% fluorescein, the patient could blow out air bubbles in the conjunctival lake on holding his nose (Fig. 1). There was no palpable mass or swelling and ocular examination was unremarkable. Lacrimal syringing revealed a patent system, although fluid entry was extremely easy compared with the normal side, indicating a wide-open passage. Nasal endoscopy was normal, although Hasner's valve could not be visualized.

FIG. 1. Left, normal tear film with fluorescein at the medial canthus. Right, tear film with the appearance of air bubbles after lacrimal sac massage (arrow).

Dacryoscintigraphy showed a symmetrical slow passage of tracer from the lacrimal sac in the nasolacrimal duct, indicating a delay at the postsac level (Fig. 2A). Dacryocystography showed that the right lacrimal system contained air pockets (Fig. 2B) in addition to a small, round filling defect within the lacrimal sac, causing it to expand; a further round filling defect was seen 1 cm below the sac. These findings suggested the presence of soft tissue tumors. However, a CT scan showed an

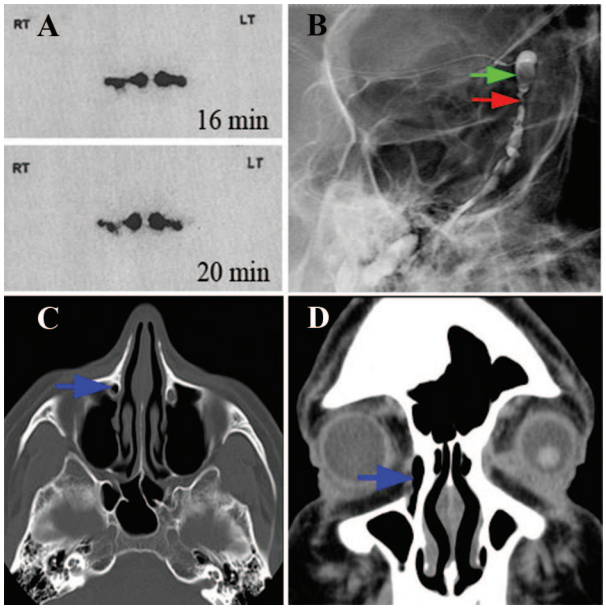

FIG. 2. Dacryocystogram, dacryoscintigraphy, and CT scans. **A,** Dacryoscintigraphy shows symmetrical, slow passage with retention of tracer within the lacrimal sac at 20 minutes, with slight overspill on the right. **B,** Dacryocystogram shows a rounded filling defect within an enlarged lacrimal sac (green arrow). The red arrow shows a narrowed portion of the nasolacrimal duct below which lies an irregularly dilated nasolacrimal duct with a further rounded filling defect approximately 1 cm below the sac. **C, D,** Axial and coronal scans respectively showing air within the right nasolacrimal duct (blue arrows) compared with an opacified duct on the left, the latter consistent with duct mucosa. No extrinsic mass is seen surrounding the duct or lacrimal sac.

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enlarged right lacrimal fossa with air in the nasolacrimal duct and no evidence of a mass (Fig. 2C, D). The left nasolacrimal duct was opacified, consistent with normal nasolacrimal duct mucosa. Following lower punctal plug occlusive canaliculoplasty on the right, using a preloaded silicone plug (Bouchon Meatique; Issy-les-Moulineaux, France), the patient noted an immediate resolution of reflux symptoms.

DISCUSSION

Originally described by Purkinje in 1823, the phenomenon of air reflux from the lacrimal sac to the canalicular system—or "ocular whistling" as termed by Duke-Elder—was thought to be due to incompetence of the valve of Hasner at the lower end of the nasolacrimal duct.[1] An alternative mechanism has been attributed to incompetence of the valve of Rosenmüller, a flap of mucosa which normally acts as a barrier between the lacrimal sac and canalicular system. This is generally thought to act as a one-way stop valve allowing fluid and air egress from the canaliculus to the lacrimal sac but not vice versa. However, one study investigating the rate of air reflux following external dacryocystorhinostomy with membranectomy showed otherwise. This study reported the incidence of air reflux with membranectomy (36% of patients) as similar to that without membranectomy (50% of patients; $p = 0.30$), meaning that removal of the valve of Rosenmüller did not increase air or tear reflux.[2] This suggested that the valve does not work by a one-way mechanism.

Lacrimal sac enlargement occurs with nasolacrimal duct stenosis, more commonly when completely obstructed. Differential diagnosis for an enlarged lacrimal sac should include an inflammatory or neoplastic mass. Although lacrimal sac tumors are rare, their commonest presenting symptoms include epiphora, recurrent dacryocysitis, and lacrimal sac mass. Bleeding from the lacrimal punctum invariably suggests the presence of a lacrimal sac tumor, particularly malignant melanoma. Dacryocystography in the presence of tumor may show an enlarged lacrimal sac with filling defects and delayed emptying. CT features of tumor include a mass in the lacrimal sac or adjacent paranasal sinuses, bony erosion, nasolacrimal duct enlargement, and extension of mass in surrounding tissues.[3] In contrast, CT imaging of inflammatory conditions of the lacrimal sac will not show these features but may be characterized by lacrimal sac dilatation with a cystic mass centered at the lacrimal fossa.

Definitive diagnosis is obtained through lacrimal sac biopsy. The latter was not performed in our case, since the clinical picture and CT result did not suggest the presence of a solid mass, contrary to the initial dacryocystographic findings.

Clinical features similar to those in our case have been reported following a compression pneumocele of the lacrimal sac. This presented with a recurrent, firm, crepitant swelling in the medial canthal region after coughing and sneezing, albeit with no associated air reflux.[4] During dacryocystorhinostomy surgery, a large, diverticulous pneumocele was noted. This had compressed the lacrimal sac, causing it to distend due to entrapped air. Nasal endoscopy showed a wide nasolacrimal ostium without a Hasner's valve, which therefore allowed air to flow back to the lacrimal system from the nasal cavity. Aside from providing an alternative diagnosis to be considered in such cases, this case again underlines the role of Hasner's valve as a barrier to prevent reflux in the lacrimal sac.

This study reported the possibility of a previous nasolacrimal canal/duct obstruction resulting in raised intraluminal pressure causing chronic dilatation of the lacrimal sac. The raised intraluminal pressure may have been enough to overcome the obstruction hence its absence at presentation. Certainly, this theory has been proposed for enlargement of the nasolacrimal canal itself in a case series of lacrimal obstruction, which were managed surgically due to the suspicion of a neoplastic process.[5]

It is interesting that our patient had resolution of symptoms following insertion of a lower punctal plug, while the upper punctum was not occluded. Perhaps the intraluminal pressure created on blowing the nose was insufficient to overcome the small diameter of the upper canaliculus alone. Importantly, the patient did not experience epiphora following lower punctal plug insertion, suggesting that the upper canalicular system was still patent. In summary, this case shows that punctal plug insertion offers a safe and effective alternative in the management of reflux symptoms associated with a lacrimal pneumocele, thereby obviating the need for surgery.

REFERENCES

1. Duke-Elder S. The ocular adnexa. In: Duke-Elder S, Macfout PA, eds. *System of Ophthalmology.* Vol. 13, Pts. 1 and 2. St. Louis, MO: Mosby, 1974:691–3.
2. Herbert HM, Rose GE. Air reflux after external dacryocystorhinostomy. *Arch Ophthalmol* 2007;125:1674–6.
3. Stefanyszyn MA, Hidayat AA, Pe'er JJ, Flanagan JC. Lacrimal sac tumors. *Ophthal Plast Reconstr Surg* 1994;10:169–84.
4. Rochels R, Bleier R, Nover A. [Compression pneumatocele of the lacrimal sac.] *Klin Monbl Augenheilkd* 1989;195:174–6.
5. Rheeman CH, Meyer DR. Enlargement of the nasolacrimal canal in the absence of neoplasia. *Ophthalmology* 1998;105:1498–503.

Bifocal Orbital and Nasopharyngeal Amyloidomas Presenting as Graves Disease

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Abstract: **A 72-year-old man presented with a slowly progressive left hyperglobus, left infraduction deficit, bilateral lower eyelid retraction, and dysphagia. He had a notable chin-down head position, diplopia in primary position, and 3 mm of left proptosis. He had been diagnosed with Graves disease 3 years before presentation. CT scans showed enlargement of the left inferior and medial rectus muscles with associated stranding of the retrobulbar fat and a low-density heterogeneous mass in the left aspect of the neck protruding in the nasopharynx. Biopsies of the orbit and nasopharynx revealed focal areas of amyloid. This represents the first report of bifocal amyloidomas of the orbit and nasopharynx.**

Amyloidosis is a heterogeneous group of disorders characterized by the deposition of abnormal fibrillar proteins in tissues and organs. On a molecular level, this insoluble protein

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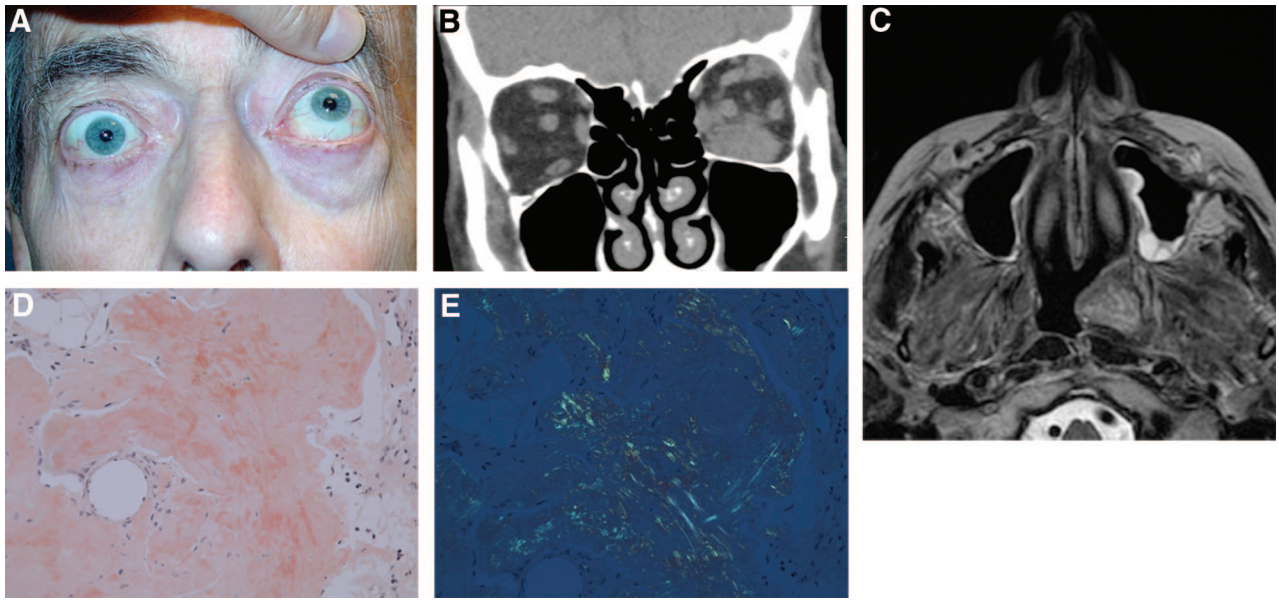

A, Preoperative photo with patient in primary position. **B,** Coronal CT scan showing enlarged inferior and medial rectus muscles. **C,** Axial CT scan showing low-density heterogeneous enhancing mass protruding in the left nasopharynx. **D,** Congo red staining of amyloid within the lesion. **E,** Apple-green birefringence of amyloid upon exposure to polarized light.

is arranged in antiparallel, beta-pleated sheets and produces a distinctive apple-green birefringence upon exposure to polarized light. Amyloid protein stains histologically with Congo red dye. Amyloidosis commonly has been described as acquired or hereditary, systemic or localized, and primary or secondary. Currently, amyloidosis is classified by the type of protein that makes up the amyloid fibril, such as amyloid light chain and amyloid-associated protein. Systemic involvement demonstrates widespread deposition of amyloid with a slowly declining clinical course and poor prognosis. Localized disease is less common but carries an excellent prognosis, and when present, is more commonly found in the head and neck. Localized orbital amyloidosis has been reported as 4% of head and neck amyloidomas.[1] In this paper, we report the clinical and histopathologic findings in a 72-year-old man with an unusual presentation of discrete amyloidomas of the orbit and nasopharynx. The management options and the result of surgical treatment of this case are discussed.

CASE REPORT

A 72-year-old man presented with a slowly progressive left hyperglobus, left infraduction deficit, bilateral lower eyelid retraction, and dysphagia. His examination revealed a notable chin-down head position, left hypertropia, diplopia in primary position, and 5 mm of left proptosis (Fig. A). He had been diagnosed with Graves disease 3 years before presentation at another institution based on the finding of enlarged muscles. There was no history of autoimmune hyperthyroidism, trauma, or infection. Ocular examination was otherwise normal. CT scans showed enlargement of the left inferior and medial rectus muscles with associated stranding of the retrobulbar fat (Fig. B). The low-density heterogeneous enhancing mass measured 6 cm in the transverse, 3.2 cm in the anteroposterior, and 4.1 cm in the craniocaudal dimension involving both the left medial and lateral pterygoid muscles and extending to abut the medial cortex of the left mandibular ramus (Fig. C). Biopsies of the orbit and nasopharynx revealed vascularized stromal tissue with amorphous eosinophilic material staining orange typical of amyloid (Fig. D). The amorphous material was congophilic and produced the distinctive apple-green birefringence upon exposure to polarized light (Fig. E).

The hematology/oncology team performed a screening examination for evidence of systemic disease. The patient's blood showed elevated free light chain κ and λ; however, the κ/λ ratio was normal. All other blood chemistries were normal, and the patient's urine Bence Jones protein was negative. To rule out amyloid involvement of the heart, an echocardiogram was performed, which was normal. CT scans of his chest, abdomen, and pelvis were all negative to rule out the possibility of underlying lymphoma and further amyloidomas. Bone marrow aspiration and biopsy were performed to assess for evidence of multiple myeloma or amyloid involvement of the bone marrow and possible lymphoma. The bone marrow biopsy was normal, showing no lymphoproliferative disorder, plasma cell dyscrasia, or amyloid deposition. Polymerase chain reaction and sequence analysis of peripheral blood showed no amyloidogenic mutation in the genes associated with autosomal dominant hereditary amyloidosis: transthyretin (TTR), fibrinogen α chain (FGA), lysozyme (LYZ), and apolipoprotein A-1 (APOA1).[2] Because the patient's workup for systemic amyloidosis was negative, his biopsy specimen was sent for ionization mass spectroscopy for peptide fingerprinting to further investigate the etiology of his amyloidomas.

DISCUSSION

Amyloidosis in its localized form is rare in the orbit, with only 30 or so reports in the literature.[1] Among localized amyloidosis of the head and neck, only 4% are found in the orbit.[1] Thus, having 2 localized areas of disease, one of which is in the orbit, is even more unusual.

Amyloid may be found anywhere within and around the globe. Corneal involvement may be seen as hereditary localized disease, while eyelid involvement is the hallmark of systemic disease.[1] Localized conjunctival amyloidosis is usually bilat-

eral and chronic in course and is rarely with associated ocular involvement.[3]

The typical clinical picture associated with orbital amyloidosis is of a painless palpable mass or exophthalmos present for years, which distinguishes it from idiopathic orbital inflammation.[4] Calcification on CT and heterogeneous hypointensity on T2-weighted MRI, while seen more commonly in cavernous hemangiomas, may also represent amyloid deposition.[5] Most cases of orbital amyloidosis are found in the superior temporal orbit, with or without displacement of the globe inferonasally.[6] A study of 17 periocular and 7 orbital amyloidosis cases showed the presence of visible or palpable periocular mass or tissue infiltration (95.8%), ptosis (54.2%), periocular discomfort or pain (25%), proptosis or globe displacement (21%), limitations in ocular motility (16.7%), recurrent periocular subcutaneous hemorrhages (12.5%), and diplopia (8.3%).[4]

This patient's polymerase chain reaction and sequence analysis screening did not match any of the known hereditary amyloidoses. Hereditary amyloidosis accounts for only 5% of all cases of amyloidosis with variable penetrance and clinical presentation.[2] Some of the commonly screened genes include TTR, FGA, LYZ, and APOA1 for familial visceral amyloidoses. The likelihood of this patient having a hereditary form of amyloidosis was low, because there are no reports of hereditary amyloid leading to amyloidomas.

The primary treatment for localized orbital amyloidosis is surgical excision, which may be difficult because amyloid usually extends deep in the orbit with an infiltrative pattern. Amyloid has a propensity to deposit histologically near fine arterial vessels, and significant bleeding may occur during resection due its friable vasculature. Thus, the use of CO_2 laser may facilitate removal of these lesions. Although it is rare for systemic disease to develop, local recurrences have been reported in approximately one third of the cases treated surgically.[1] To avoid repeated excisions, radiation has been performed successful with limited follow-up.[1]

This is the first report of bifocal amyloidomas of the orbit and nasopharynx. The ability of amyloid to deposit within any ocular tissue gives rise to its many stigmata. When confined to the orbit, proptosis, diplopia, and eyelid retraction that mimic other orbital disease often ensue. Thus, amyloidomas may masquerade as Graves disease, idiopathic orbital inflammation, malignancy, or benign tumors. Although complete surgical excision is not feasible in many cases, the goal of treatment is to preserve function and to prevent sight-threatening complications.

REFERENCES

1. Taban M, Piva A, See RF, et al. Review: orbital amyloidosis. *Ophthal Plast Reconstr Surg* 2004;20:162–5.
2. Lachmann HJ, Booth DR, Booth SE, et al. Misdiagnosis of hereditary amyloidosis as AL (primary) amyloidosis. *N Engl J Med* 2002;346:1786–91.
3. Pirouzmand F, Hurwitz J, Howarth D. Primary localized bilateral conjunctival amyloidosis and epiphora. *Orbit* 2002;21:139–44.
4. Leibovitch I, Selva D, Goldberg RA, et al. Periocular and orbital amyloidosis: clinical characteristics, management, and outcome. *Ophthalmology* 2006;113:1657–64.
5. Okamoto K, Ito J, Emura I, et al. Focal orbital amyloidosis presenting as rectus muscle enlargement: CT and MR findings. *AJNR Am J Neuroradiol* 1998;19:1799–801.
6. Murdoch IE, Sullivan TJ, Moseley I, et al. Primary localised amyloidosis of the orbit. *Br J Ophthalmol* 1996;80:1083–6.

Orbital Lymphoma in the Setting of Idiopathic CD4+ Lymphocytopenia (HIV-Negative AIDS)

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Abstract: **A 43-year-old Asian man with idiopathic CD4+ lymphocytopenia (human immunodeficiency virus [HIV]-negative acquired immunodeficiency syndrome [AIDS]) noted blurred vision OD. Visual acuity was 20/30 OD and 20/15 OS. The right eye showed features of axial proptosis (2 mm), blepharoptosis, upgaze restriction, conjunctival chemosis, color desaturation, and choroidal folds. The OS was unremarkable. There was no lymphadenopathy. Orbital MRI showed a well-circumscribed mass indenting the globe. Excision revealed diffuse large-B-cell lymphoma. Systemic evaluation was negative and adjuvant external beam radiotherapy to the right orbit was performed. Idiopathic CD4+ lymphocytopenia is a rare hematologic condition characterized by depletion of CD4+ T cells in HIV-negative patients. This condition closely resembles AIDS but HIV serology is negative. Similar to AIDS, patients are at risk for opportunistic infections and neoplasia, including systemic lymphoma. This is the first case, to the authors' knowledge, of orbital lymphoma with idiopathic CD4+ lymphocytopenia.**

In 1992, the Centers for Disease Control and Prevention issued an alert in *Morbidity and Mortality Weekly Report* that a series of 26 patients from around the world manifested a condition of CD4+ lymphocytopenia, serious opportunistic infections associated with classic acquired immunodeficiency syndrome (AIDS), but no evidence of human immunodeficiency virus (HIV) infection.[1] Some initial reports termed this entity HIV-negative AIDS, but it is now known as idiopathic CD4+ lymphocytopenia (ICL). ICL predisposes affected patients to similar complications as classic AIDS including multiple opportunistic infections and neoplasia, particularly lymphoma.[2] Although the coexistence of ICL and lymphoma has been reported, we report herein the first case, to our knowledge, of ICL-related orbital lymphoma.[2–5]

CASE REPORT

A 43-year-old Asian-American man presented with a 3-week history of blurred vision OD. His ocular history was unremarkable, but medical history revealed diffuse large-B-cell lymphoma of the bladder 8 years previous, treated with surgical

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excision and chemotherapy. The patient was diagnosed 15 years earlier with idiopathic CD4+ lymphocytopenia following an episode of cryptococcal meningitis. The patient took acyclovir, fluconazole, and trimethoprim-sulfamethoxazole for prophylaxis against opportunistic infections.

The visual acuity was 20/30 OD and 20/15 OS with normal intraocular pressures OU. The OS was unremarkable. The OD displayed 2 mm of axial proptosis, 1 mm of blepharoptosis, mild chemosis, 1+ restriction in upgaze, and color desaturation with Ishihara color plates (Fig. A). There was no preauricular, submandibular, or cervical lymphadenopathy. Fundus examination revealed choroidal folds suggestive of orbital compression (Fig. B). Orbital MRI disclosed a well-circumscribed, intraconal, enhancing mass in the right orbit with homogenous features and globe indentation posteriorly (Fig. C, D).

Removal of the mass via an inferior transconjunctival forniceal route was performed. Intraoperatively, the tumor was friable, adherent to the inferior oblique and optic nerve, and was grossly removed. Intravenous dexamethasone (Decadron) (16 mg) intraoperatively and intraorbital triamcinolone acetonide (40 mg in 1 cc) at closure were given to decrease inflammation and minimize optic nerve damage. Histopathology of the lesion showed sheets of large, atypical cells demonstrating irregular nuclei (Fig. E). Immunohistochemical stains were positive for CD20 and CD45 and negative for CD3, CD5, and CD10, consistent with orbital diffuse large-B-cell lymphoma.

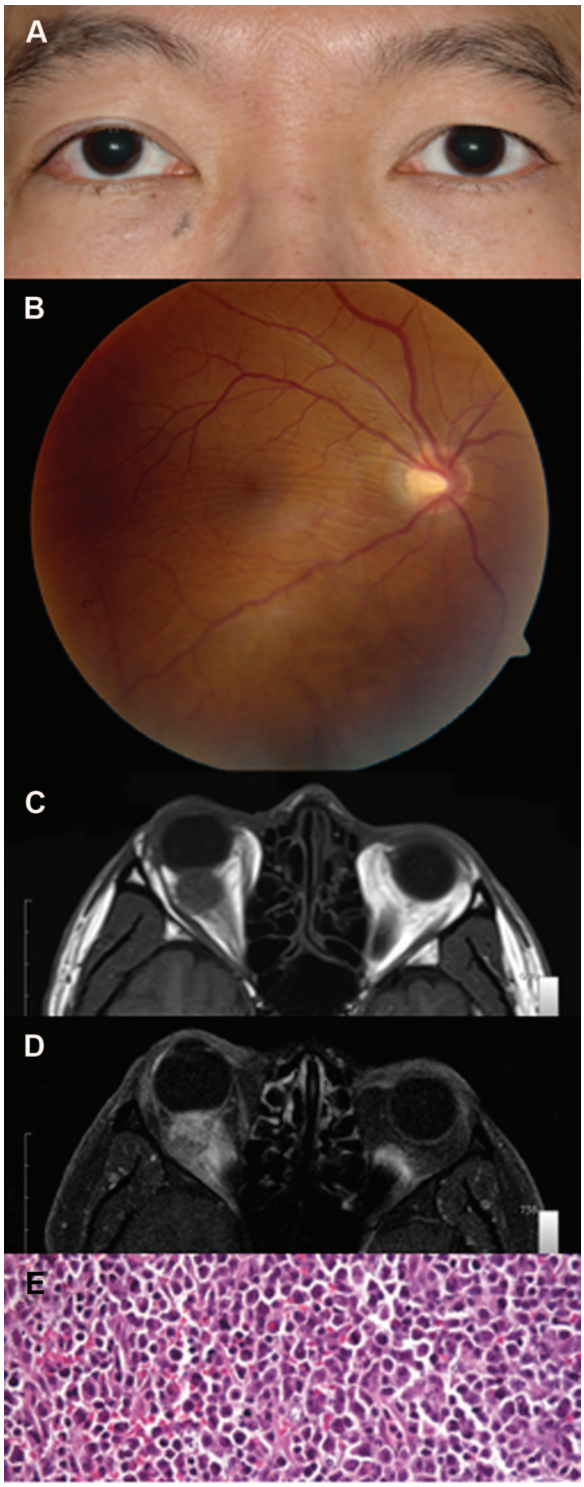
Following surgery, visual acuity was 20/20 OU with resolution of previous proptosis, upgaze restriction, and blepharoptosis. Full body positron emission tomography imaging was negative. Adjuvant external beam radiotherapy (40 Gy) was delivered to the right orbit for microscopic residua. Three months after lesion excision, the patient remained disease free with no evidence of systemic spread based on CT imaging.

DISCUSSION

Idiopathic CD4+ lymphocytopenia is an entity that initially received attention in 1992 for its phenotypic similarity to AIDS but without serologic evidence of HIV infection.[1] These patients usually present with unexplained infections but can present asymptomatically with low CD4+ T-lymphocyte counts.[2] To diagnose ICL, the clinician must document a CD4+ T-lymphocyte count below 300 cells/μl or $<$20% of the total T-cell repertoire on 2 separate occasions separated by at least 6 weeks.[2] Additionally, the patient must have negative serology for HIV-1 and HIV-2 on multiple occasions.[2] In 1996, Laurence et al.[2] reported that the CD4+ cells in these patients aberrantly overexpressed Fas ligand, which leads to apoptosis.[3] This depletion of helper T cells leads to impaired immunity along with immune dysregulation. Other authors have reported increased CD4+ T-cell activation and turnover, suggesting a disturbance in the normal homeostatic mechanisms for CD4+ T cells.

In a prospective, natural history study of 36 patients with ICL, 42% developed opportunistic infections and 14% had an AIDS-defining illness, such as Mycobacterium avium complex infection.[2] An ICL-related malignancy was found in 2 patients: one had B-cell lymphoma and another had metastatic squamous cell carcinoma.[2] Both patients with ICL-related malignancies died during the study period, and 19% of all patients in the study died within 42 months of ICL diagnosis.[2]

There have been numerous reports of ICL-related lymphoma including involvement of the leptomeninges, pleural space, ovaries, and submandibular lymph nodes.[4–5] Most of such lymphomas have been classified as non-Hodgkin's B-cell lymphomas, with diffuse large-cell lymphoma the most common type.[4–5] Six patients with non-Hodgkin's lymphoma have



A 43-year-old Asian man with known idiopathic CD4+ lymphocytopenia (ICL) developed right orbital lymphoma. **A,** Right proptosis with blepharoptosis is noted. **B,** Compressive choroidal folds from the orbital mass are seen in the macular region. **C, D,** Axial T1-weighted MRI showing a well-circumscribed, intraconal right orbital mass indenting the globe **(C)** that enhances with gadolinium and fat-suppression technique **(D). E,** Histopathology of orbital mass lesion showing sheets of large, atypical cells demonstrating irregular nuclei suggestive of lymphoma (hematoxylin-eosin, $\times$40).

been described in case reports in the literature, and 3 of these patients achieved durable remission following treatment with chemotherapy.[4] The pathophysiology underlying the high incidence of lymphoma in these patients has not been elucidated. However, a likely hypothesis is that the perturbations in T-cell populations that occur in ICL lead to impaired immune surveillance. Specifically, the depletion of CD4+ T cells may lead to compensatory cytokine stimulation, which then drives the expansion of B-cell subpopulations in lymphoid tissues.

In contrast to the wealth of information on HIV-positive AIDS-related eye disease, reports of ocular conditions related to HIV-negative AIDS (ICL) are relatively uncommon. Wladis et al.[6] reported a case of ICL in a 52-year-old woman in association with ocular symptoms of Sjogren syndrome, and others have observed an increased prevalence of Sjogren syndrome in patients with ICL. Schuil et al.[7] described a case of transient viral retinopathy following measles-mumps-rubella vaccination in a 4-year-old child with previously undiagnosed ICL. Other authors described a case of cytomegalovirus retinitis confirmed by polymerase chain reaction of aqueous humor in the setting of ICL.[8] To this point, there have been no ocular malignancies reported in association with ICL.

In our case, orbital lymphoma developed after a 15-year history of ICL. Eight years before presentation, the patient was treated for B-cell lymphoma of the bladder. The orbital lymphoma could represent a related lymphomatous deposit, but with negative positron emission tomography imaging, it most likely represents a second de novo site.

Treatment for ICL is focused on managing opportunistic infections, and investigators have attempted to boost the CD4 counts using interleukin-7 and interleukin-2 as experimental therapies.[1] There are no published reports of using antiretroviral therapy in ICL, although initial reports speculated that a viral etiology may exist for the disease.[1] Overall, patients with ICL have poor prognoses from coexistent life-threatening infections and neoplasia. Orbital lymphoma could represent a related neoplastic lesion, and the ophthalmologist should keep this in mind when evaluating such patients.

REFERENCES

1. Centers for Disease Control. Unexplained CD4_T-lymphocyte depletion in persons without evident HIV infection–United States. *MMWR Morb Mortal Wkly Rep* 1992;41:541–5.
2. Zonios DI, Falloon J, Bennett JE, et al. Idiopathic CD4+ lymphocytopenia: natural history and prognostic factors. *Blood* 2008;112:287–94.
3. Laurence J, Mitra D, Steiner M, et al. Apoptotic depletion of CD4+ T cells in idiopathic CD4+ T lymphocytopenia. *J Clin Invest* 1996;97:672–80.
4. Niino D, Tsukasaki K, Torii K, et al. Human herpes virus 8-negative primary effusion lymphoma with BCL6 rearrangement in a patient with idiopathic CD4 positive T-lymphocytopenia. *Haematologica* 2008;93:e21–3.
5. Campbell JK, Prince HM, Juneja SK, et al. Diffuse large cell lymphoma and t(8;22) (q24;q11) in a patient with idiopathic CD4+ T-lymphopenia. *Leuk Lymphoma* 2001;41:421–3.
6. Wladis EJ, Kapila R, Chu DS. Idiopathic CD4+ lymphocytopenia and Sjogren syndrome. *Arch Ophthalmol* 2005;123:1012.
7. Schuil J, van de Putte EM, Zwaan CM, et al. Retinopathy following measles, mumps, and rubella vaccination in an immuno-incompetent girl. *Int Ophthalmol* 1998;22:345–7.
8. Sloan DJ, Taegtmeyer M, Pearce IA, et al. Cytomegalovirus retinitis in the absence of HIV or immunosuppression. *Eur J Ophthalmol* 2008;18:813–5.

Recurrent Infarction of Sphenoid Bone With Subperiosteal Collection in a Child With Sickle Cell Disease

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Abstract: **Infarction of the orbital bone in patients with sickle cell disease is very rare. The authors report a young boy who presented twice with marked acute proptosis and eyelid swelling of the right eye resulting from infarction in the greater wing of the sphenoid bone accompanied by an orbital subperiosteal collection. The time interval between the 2 attacks was 3 years.**

Sickle cell disease is a genetic lifelong blood disorder characterized by red blood cells that assume an abnormal rigid sickle shape. The sickled erythrocytes are relatively rigid and tend to obstruct the microcirculation, causing tissue infarction. All bones with active marrow are potential targets for vaso-occlusive crisis, but a predilection for long bones has been reported.[1] Fewer than 50 cases of infarction of the orbital bones in patients with sickle cell disease have been reported in the literature.[1,2] Here, we report a child with sickle cell disease who had 2 episodes of infarction of the same greater wing of the sphenoid bone with an orbital subperiosteal collection that occurred 3 years apart.

CASE REPORT

An 11-year-old boy with sickle cell disease presented with pain and swelling of the right eye of 2-day duration. The patient had sickle crisis 3 months earlier with infarction of one of the lumbar vertebra.

The patient was in mild pain with a low grade fever. His visual acuity was 20/20 OD and 20/20 OS with no relative afferent pupillary defect. Color vision was normal in both eyes. A mild restriction of ocular motility in superior and lateral fields of gaze in the right, and 4 mm of proptosis, was detected (Fig. 1). A marked right upper eyelid edema, with tenderness over the right lateral orbital rim, was also found. The rest of the ophthalmic examination was normal.

The white cell count was within a normal range. Orbital CT with contrast demonstrated a right subperiosteal collection over the lateral orbital wall with no evidence of sinusitis. MRI with contrast showed infarction of the marrow space of the greater wing of the sphenoid bone with subperiosteal collection and ring enhancement (Fig. 2). The blood culture grew coagulase-negative staphylococci. The patient had full resolution of his ophthalmic manifestations after undergoing a few days of conservative treatment with systemic antibiotics. Three years earlier, the patient had experienced a similar attack of acute proptosis in the same eye, resulting from infarction of the greater wing of the

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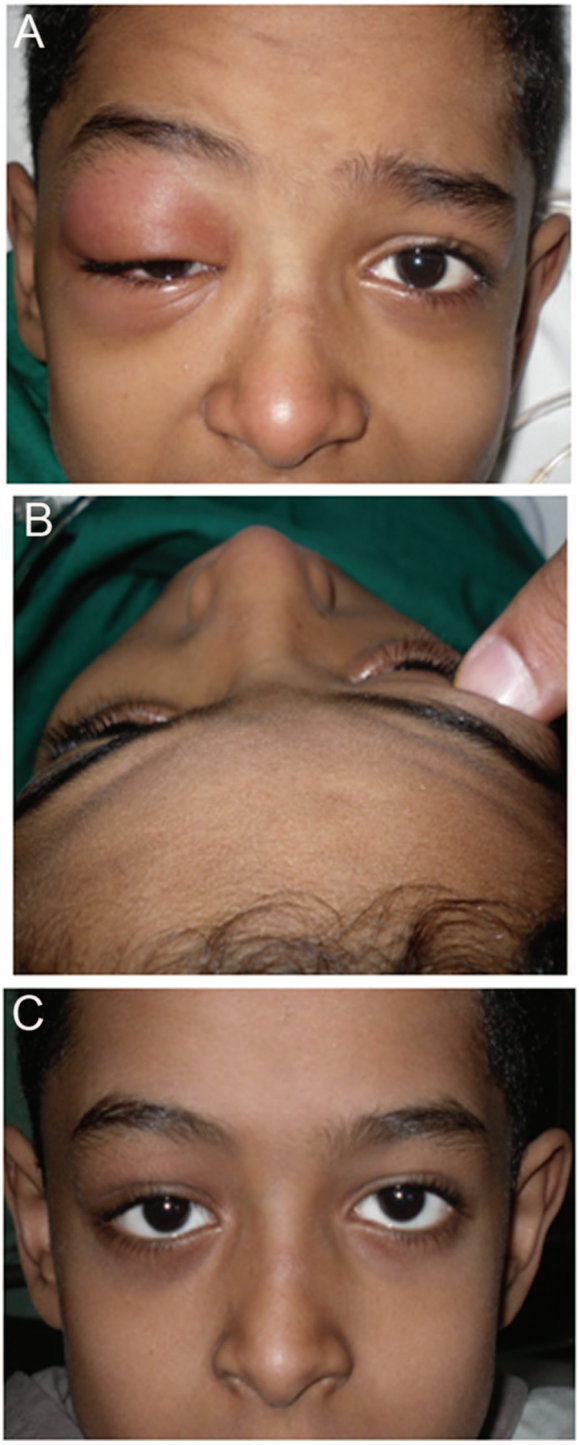

FIG. 1. **A, B,** Clinical photographs were taken at presentation with acute proptosis and eyelid swelling in the right for the second time. **C,** Almost complete resolution after 1 week of treatment with systemic antibiotics.

sphenoid bone with subperiosteal collection seen in orbital CT scan and had responded nicely to conservative treatment.

DISCUSSION

Sickle cell disease has several ophthalmic manifestations, such as anterior segment ischemia, glaucoma, angioid streaks, and retinopathy.[3] Infarction of the orbital bones, which has rarely been reported, can lead to acute proptosis, periorbital pain, limited motility, and, potentially, compressive optic neuropathy.[1,2] Our patient had 2 episodes of infarction of the same greater wing of the sphenoid bone with subperiosteal collection. To the best of our knowledge, this case is the first of its kind to be reported in the literature.

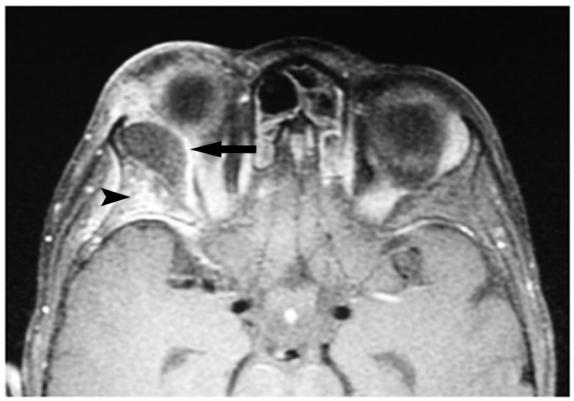

FIG. 2. Axial MRI with contrast shows involvement of the marrow space of the right greater wing of the sphenoid bone (arrowhead) with adjacent subperiosteal collection and ring enhancement around this collection (arrow).

In sickle cell disease, occlusion of the microvasculature is commonly associated with distal infarction, which in turn leads to localized tissue necrosis. Infarction and subsequent necrosis of bone marrow present as a painful episode known as a vaso-occlusive crisis. Orbital infarction is more likely to occur in children because of the larger marrow volume compared with adults.[4] One feature of orbital infarction in sickle cell disease is the formation of hematomas, which results from the extravasation of blood from necrosed vessel walls. These hematomas may be found in the orbit (subperiosteal collection) or intracranially (epidural hematomas).[5] Infarctions of the sphenoid bone can lead to an inflammatory response that spreads to the adjacent subperiosteal space, resulting in acute proptosis, periorbital pain, limited motility, and potentially, compressive optic neuropathy.

Patients with sickle cell disease are at an increased risk for osteomyelitis. Its occurrence may be related to hyposplenism, impaired complement activity, and the presence of infarcted or necrotic bone.[2] The distinction between a painful infarcted but noninfected bone of a vaso-occlusive crisis and osteomyelitis complicating a bone infarct can be extremely difficult on clinical grounds. Blood culture is an important part of managing these patients. MRI with contrast may be of great help in evaluating the soft tissue changes, ischemic changes in the marrow space, and the presence of ring enhancement suggesting an orbital abscess.[1,2] Sometimes, osteomyelitis may not be suspected until the signs and symptoms of a typical painful crisis have failed to resolve after 1 to 2 weeks of standard therapy.

Most patients with infarction of the greater wing of the sphenoid bone accompanied by an orbital subperiosteal collection improve with medical management alone. Close monitoring of optic nerve function is required. Systemic antibiotic treatment is recommended when osteomyelitis is suspected. Corticosteroids may be of help for the inflammatory component, especially for patients with compressive optic neuropathy. If compressive optic neuropathy fails to

respond to medical treatment, surgical drainage of subperiosteal collection is indicated.[1,2]

In summary, infarction in the same orbital bone may occur more than once and present with severe proptosis and subperiosteal collection. This process may be complicated by osteomyelitis and orbital cellulitis.

REFERENCES

1. Sokol JA, Baron E, Lantos G, Kazim M. Orbital compression syndrome in sickle cell disease. *Ophthal Plast Reconstr Surg* 2008; 24:181–4.
2. Ganesh A, Al-Zuhaibi S, Pathare A, et al. Orbital infarction in sickle cell disease. *Am J Ophthalmol* 2008;14:595–601.
3. Nagpal KC, Goldberg MF, Rabb MF. Ocular manifestations of sickle hemoglobinopathies. *Surv Ophthalmol* 1977;21:391–411.
4. Curran EL, Fleming JC, Rice K, Wang WC. Orbital compression syndrome in sickle cell disease. *Ophthalmology* 1997;104:1610–5.
5. Resar LM, Oliva MM, Casella JF. Skull infarction and epidural hematomas in a patient with sickle cell anemia. *J Pediatr Hematol Oncol* 1996;18:413–5.

Bilateral Upper and Lower Eyelid Severe Psoriasiform Blepharitis: Case Report and Review of Literature

Feilin Zhu, A.B., and Jeremiah P. Tao, M.D., F.A.C.S.

Abstract: **The purpose of the present study is to describe a case of severe, psoriasiform blepharitis by means of a case report and literature review. A 44-year-old man developed chronic blepharitis and tearing months after bilateral cataract surgery. Exam showed diffuse quad-eyelid erythema, discharge, edema, madarosis, and scale. He also had insufficient tear drainage due to bilateral upper eyelid cicatricial punctal atresia with bilateral lower eyelid punctal stenosis. Biopsy of the lower eyelids exhibited psoriasiform hyperplasia. Topical 0.1% tacrolimus achieved improvement but caused some subjective eye irritation. Psoriasiform dermatitis manifesting on the eyelids is rare, may be associated with insufficient tear drainage, and may respond favorably to 0.1% tacrolimus.**

Psoriasis is a common inflammatory disease of the skin characterized by thick red skin and silvery scales. It involves hyperproliferation of the keratinocytes in the epidermis and most often occurs on the trunk and extensor surfaces of the body. Eyelid psoriasis is very uncommon and the literature is scarce on this subject. We describe herein a case of severe psoriasiform blepharitis.

CASE REPORT

A 44-year-old man presented with chronic bilateral eye discharge and eyelid inflammation for 7 years. He reported that eyelid inflammation and discharge started within a few months after bilateral cataract surgery. He had a history of allergic conjunctivitis that was treated with steroid eye drops for 4 years before his cataract diagnosis. Steroid medication was thought to have precipitated his cataracts. His use of steroid medications also contributed to his diagnosis of glaucoma in recent years. Antibiotics, corticosteroids, and artificial tears had been utilized previously, without much improvement.

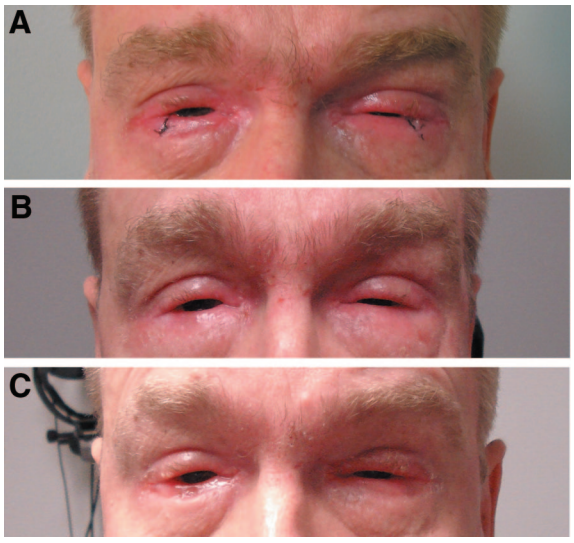

FIG. 1. **A,** Severe chronic bilateral upper and lower eyelid redness, edema, madarosis, and discharge with skin scaling. **B,** Eyelid appearance after 1 week of topical 0.1% tacrolimus twice daily. Improvement in redness and edema. **C,** Eyelid appearance after 1 month of topical 0.1% tacrolimus twice daily. Continued modest improvement in redness and edema.

On initial assessment, visual acuity was 20/50 OD and 20/70 OS without correction and 20/50 OD and 20/60 OS with pinhole correction. Examination was significant for thickened, edematous lids (with secondary ptosis), skin scale, mucous discharge, and madarosis (Fig. 1A). He also exhibited an increased tear lake due to insufficient tear drainage through bilateral upper cicatricial punctal atresia with bilateral lower eyelid punctal stenosis. Bilateral punctoplasty with placement of silicone tube stents and bilateral lower eyelid biopsy were performed.

In the weeks following surgery, he reported subjective improvement of tearing and mucous accumulation. Examination showed an improved tear lake and less mucopurulent discharge.

Differential diagnosis of his eyelid inflammation includes basal cell carcinoma, contact dermatitis, trichiasis, and ocular rosacea. Microscopic analysis of the lower eyelids revealed psoriasiform hyperplasia of epidermis with hyperkeratosis, parakeratosis, and ulceration. Neutrophils were present in the parakeratotic layer. A mixed inflammatory infiltrate including eosinophils was present in the dermis. These findings were consistent with psoriasiform changes (Fig. 2).

Tacrolimus 0.1% cutaneous ointment 2 times a day was initiated. One week later, he described subjective improvement of his eyelid irritation. His symptomatic improvement was corroborated by his exam that showed less eyelid redness and edema (Fig. 1B). On 1 month follow-up, the patient reported the side effect of eye irritation when tacrolimus got in his eyes. He had temporarily stopped the medication. A new regimen of applying the ointment carefully and away from the eyelid margin resulted in satisfactory results with continued improvement (Fig. 1C).

Literature Review

Psoriasis is a common chronic inflammatory disease involving relapsing eruptions of skin in scaly thick patches, most often on the trunk and extensor surfaces of the body. Although forehead, scalp, and eyebrow psoriasis are relatively common,

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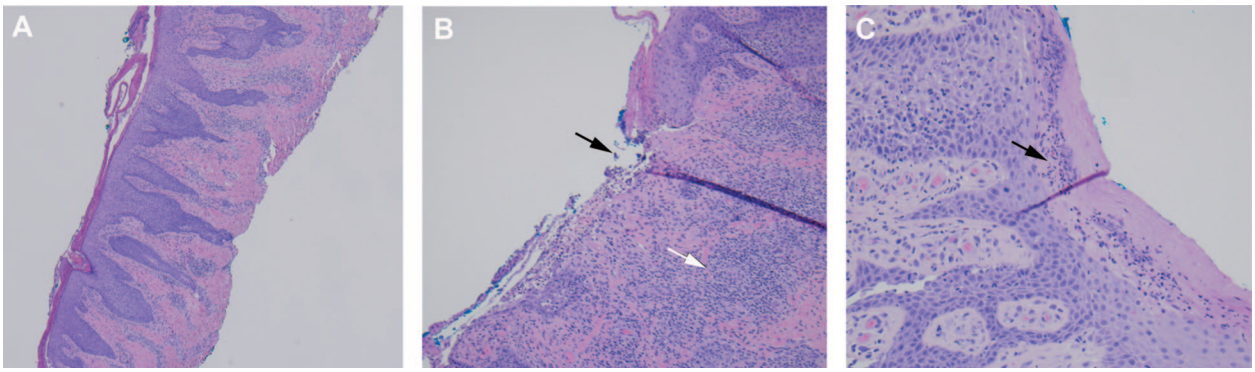

FIG. 2. **A,** Elongated rete pegs typical of psoriatic lesions (hematoxylin-eosin, original magnification ×4). **B,** Ulceration of epidermis (black arrow) and mixed inflammatory infiltrate in the dermis (white arrow) (hematoxylin-eosin, original magnification ×10). **C,** Hyperkeratosis with neutrophils present in the parakeratotic layer (black arrow) (hematoxylin-eosin, original magnification ×10).

facial psoriasis is considered uncommon—estimated to occur in only 17% to 46% of patients with psoriasis. These patients have an earlier onset with a more chronic course.

Even more uncommon is eyelid psoriasis. In a review of 203 cases of causes of eyelid dermatitis, the frequency of psoriasis was 7%.[1] Reported ocular findings in psoriasis involve red to pink scaling plaques along the eyelid margins and eyelid edema and ectropion.[2] Other complications include conjunctivitis, superficial punctate keratitis, trichiasis, symblepharon, photophobia, and increased lacrimation. Stuart[3] listed variable manifestations including nodular excrescences localized at the limbus, surrounded by small stromal opacities and vascularization, and homogeneous axial nonvascularized corneal opacities. Based on our case, we here add madarosis, ptosis, punctal atresia, and punctal stenosis.

There are few case reports of psoriasis involving the eyelids. Aliagaoglu et al.[4] reported a case of periorbital psoriasis developing in a 7-year-old boy after his brother scratched his eyelid. Zug et al.[2] reported that environmental factors such as drugs (lithium, β-blockers, antimalarials), trauma, repeated friction, and streptococcal infection can trigger or exacerbate psoriasis.

The treatment for eyelid psoriasis may include topical ointments, steroids, and radiation. Wolf surveyed 20 dermatologists from the United States and Europe for their opinions on treatment for eyelid psoriasis. Low-potency steroids, preferably not stronger than 1% hydrocortisone, and constant emollition were thought to be the most effective. Long-term, low-dosage retinoid therapy and ultraviolet B phototherapy were also recommended.[5] Several studies have also shown the effectiveness of tacrolimus, an immunosuppressant, in the treatment of facial and intertriginous psoriasis. Unlike steroids, it does not lead to atrophy and rosacea of the face from long-term use. Lebwohl et al.[6] reported a randomized control study of 167 patients treated with either 0.1% tacrolimus ointment or a corticosteroid and found significant improvement in the tacrolimus group by as early as day 8 ($p = 0.004$).

DISCUSSION

Psoriasiform blepharitis is rare. This case is remarkable for the severity and the negative history of psoriasis. Also, a complete dermatological examination did not show psoriasis elsewhere on the body. The precise mechanism and etiology for the psoriasiform changes in this case remain uncertain. However, the condition was somewhat consecutive (onset within months) to cataract surgery, and the Koebner phenomenon, or isomorphic response—skin lesions that develop in areas of local trauma or injury, such as a surgical wound—may offer one explanation. Topical 0.1% tacrolimus was associated with improvement in our patient but was also associated with the side effect of conjunctival irritation when administered too closely to the eyelid margin. This case also serves as reminder of the important ophthalmic side effects of periocular corticosteroids, namely, cataracts and glaucoma. Further investigation is necessary to establish the best treatments for psoriasiform blepharitis.

REFERENCES

1. Guin JD. Eyelid dermatitis: experience in 203 cases. *J Am Acad Dermatol* 2002;47:755–65.
2. Zug KA, Palay DA, Rock B. Dermatologic diagnosis and treatment of itchy red eyelids. *Surv Ophthalmol* 1996;40:293–306.
3. Stuart JA. Ocular psoriasis. *Am J Ophthalmol* 1963;55:615–7.
4. Aliagaoglu C, Yildirim U, Atasoy M, et al. Periorbital psoriasis developing after trauma. *Dermatol Online J* 2006;12:22.
5. Wolf R. How to manage psoriasis of the eyelids. A glance at opinions of other dermatologists. *Dermatologica* 1986;173:105–8.
6. Lebwohl M, Freeman AK, Chapman MS, et al. Tacrolimus ointment is effective for facial and intertriginous psoriasis. *J Am Acad Dermatol* 2004;51:723–30.

Orbital Floor Abscess Secondary to Sinusitis in an Immunocompromised Patient

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Abstract: **A 65-year-old man with a history of renal transplantation presented with facial pain, purulent nasal discharge, and periorbital swelling. Signs of optic nerve compromise developed and persisted despite medial orbital wall**

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decompression. Further imaging revealed an orbital floor abscess secondary to direct communication of a maxillary pseudomonal sinusitis. Full recovery was achieved after further surgical drainage via an endoscopic endonasal approach. Failure to improve after surgical decompression is an indication for repeat imaging. Immunocompromised patients can present atypically. Orbital floor abscess secondary to sinusitis without an underlying odontogenic or traumatic cause has not previously been reported. The authors highlight the importance of clinical vigilance, serial imaging, microbiological investigations, and early surgical intervention in high-risk patients.

Orbital cellulitis is a serious condition that must be recognized and managed promptly to avoid permanent and devastating complications.[1] These include loss of vision from optic nerve compromise, cavernous sinus thrombosis, intracranial abscess, and even death.[1,2]

Orbital cellulitis typically arises secondary to sinusitis, trauma, surgery, or odontogenic infection.[2] An orbital abscess usually develops medially from spread of ethmoidal infection through the thin lamina papyracea.[1,3]

CASE REPORT

A 65-year-old man with type II diabetes mellitus and history of renal transplantation 18 months previously presented to the otolaryngology team with left facial pain, purulent nasal discharge, and epistaxis for 2 weeks. Regular medications included oral immunosuppression with mycophenolate mofetil, tacrolimus, and prednisolone. Despite one week of a broad-spectrum antibiotic (co-amoxiclav), symptoms worsened with development of left periorbital swelling, conjunctival chemosis, and blurring of vision. On ophthalmological assessment, best corrected logMAR visual acuity was 0.0 in the right eye, 0.18 in the left. Ishihara color vision, eye movements, and pupil reactions were all normal.

A CT scan demonstrated almost complete opacification of the left maxillary antrum and left ethmoidal air cells (Fig. 1). There was also a thin rim of more focal abnormal soft tissue lying adjacent to the left orbital floor (Fig. 2). There was no apparent discrete orbital abscess.

Endoscopic sinus surgery was performed to drain the sinuses and decompress the orbit. A large middle meatal antrostomy was fashioned and tissue was collected from the antrum for histologic diagnosis. Anterior and posterior ethmoidectomies were then performed, allowing exposure of the lamina papyracea. The lamina was opened anteriorly to expose the orbital periosteum, but no pus was encountered. The orbital periosteum was therefore opened to decompress the orbit. No attempt was made to trace the soft tissue swelling on the orbital floor because an abscess in this area has not been encountered previously. Histology showed chronic inflammation with no features of fungal infection or malignancy.

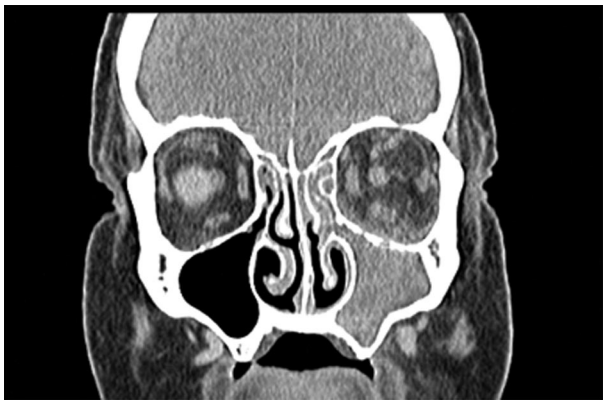

FIG. 2. Coronal CT scan demonstrating opacification of left maxillary and ethmoid sinuses. Note the area of opacification on the floor of the orbit.

The patient's symptoms initially improved, but 2 weeks later, increasing pain, limited extraocular movements, and 4 mm of proptosis prompted a further CT scan. This demonstrated a left inferior orbital abscess communicating through the inferior orbital floor from the maxillary sinus (Fig. 3). Urgent surgical drainage of the abscess was performed via an endoscopic endonasal approach. This was done via subperiosteal dissection of the orbital floor using angled endoscopes and instruments. A copious amount of pus was drained and sent for analysis. The floor of the orbit was inspected and a small area (about 4 × 4 mm) of dehiscence was identified.

Culture identified *Pseudomonas aeruginosa* as the causative organism, resistant to ciprofloxacin and tazocin but sensitive to meropenem. Blood cultures were negative. Histologic examination of surgical biopsies revealed necrotic turbinate bone with no features of fungal infection and no evidence of

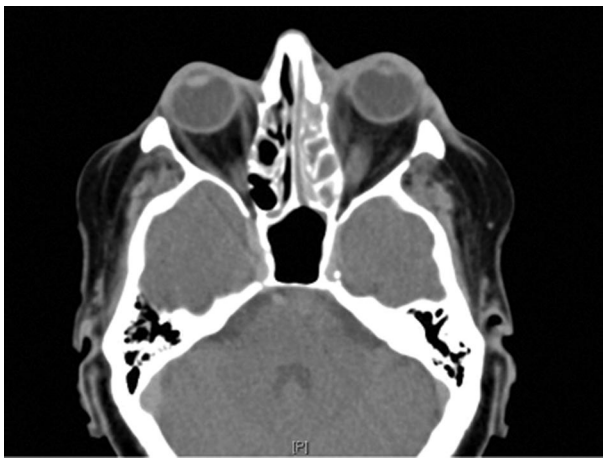

FIG. 1. Axial CT scan demonstrating opacification of left ethmoid sinuses.

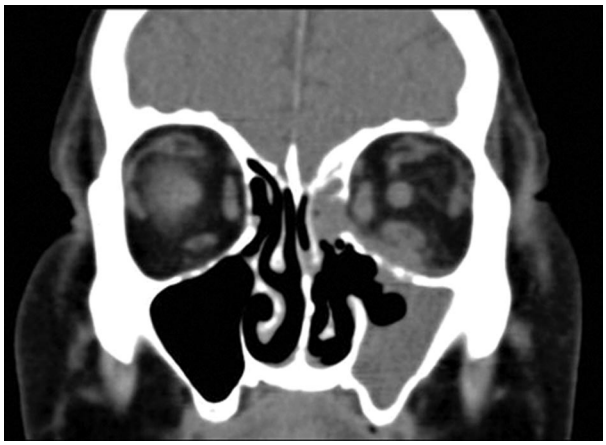

FIG. 3. Coronal CT scan demonstrating left inferior orbital abscess.

malignancy. Symptoms subsequently fully resolved over the next 2 months.

DISCUSSION

Orbital infections were broadly classified by Chandler et al.[4] in 1970 in 5 stages: inflammatory edema, orbital cellulitis, subperiosteal abscess, orbital abscess, and cavernous sinus thrombosis. Orbital cellulitis is a broad term covering diffuse postseptal cellulitis of the orbit, subperiosteal abscess of the orbit, and orbital abscess.[5]

In up to 84% of cases, orbital cellulitis arises secondary to adjacent sinusitis by direct transmission through the sinus wall or via thrombophlebitis of the valveless venous system that communicates between orbit and sinuses.[2,4,5] Other causes include trauma, surgery, periorbital infection, and odontogenic infection. Orbital cellulitis secondary to complications from dental infections or dental surgery has been reported to account for approximately 2% of orbital cellulitis cases, including several reported cases of orbital floor abscess.[6,7] Typically, these cases involve spread from molar teeth to maxillary sinus then directly through to orbit.[6]

Sinusitis leading to orbital complications is well reported particularly between the ethmoid sinus and the orbit via the thin lamina papyracea.[1,3] However, complications from maxillary sinusitis are less common and an orbital abscess secondary to maxillary sinusitis is very rare. There are no reports in the literature of an inferior orbital abscess secondary to sinusitis. There are however 2 cases reported of inferior subperiosteal abscess; the first was inferolateral from direct extension of maxillary sinusitis in the orbit that was not apparent on preoperative CT scan but was identified surgically, and the second was an inferior subperiosteal abscess in a 75-year-old woman with late presentation and loss of vision.[5,8] Neither of these were treated endoscopically.

There is a higher incidence of unusual pathogens including mucormycosis species and *Pseudomonas aeruginosa* in patients who are immunosuppressed, with the latter accounting for 3% of orbital abscess cases.[8,9] The common pathogens in the immunocompetent patient are *Staphylococcus* and *Streptococcus* species.[8,10] Blood cultures tend to have a negative yield in identifying the causative organism, while cultures from infected sinuses or the orbital abscess itself have demonstrated positive results in 50% to 100% of cases.[8,10]

A patient presenting with typical features of orbital cellulitis should be investigated urgently with appropriate imaging. The preferred choice is CT scanning, as this provides identification of disease and the anatomy for presurgical planning.[11] However, if the features are atypical or there is diagnostic uncertainty, MRI can be a useful tool with higher sensitivity for soft tissue changes.[11]

Prompt treatment is vital to protect the patient from serious complications, which include loss of vision in up to 11% of cases, cavernous sinus thrombosis, and intracranial abscess.[2,10] Loss of vision arises due to optic neuritis, ischemia secondary to thrombophlebitis, or pressure ischemia.[1] Early surgical intervention prevents permanent vision loss. When managed appropriately, orbital cellulitis rarely has significant final morbidity.[8,10]

Treatment failure from orbital cellulitis after surgical decompression is an indication for repeat imaging and consideration of further surgical intervention. Inferior orbital abscess is rare but should be considered in patients with orbital symptoms secondary to sinusitis. We have shown that inferior orbital abscess is amenable to endoscopic drainage with orbital decompression like the far more common medial abscess.

REFERENCES

1. Patt BS, Manning SC. Blindness resulting from orbital complications of sinusitis. *Otolaryngol Head Neck Surg* 1991;104:789–95.
2. Kloek CE, Rubin PA. Role of inflammation in orbital cellulitis. *Int Ophthalmol Clin* 2006;46:57–68.
3. Ikeda K, Oshima T, Suzuki H, et al. Surgical treatment of subperiosteal abscess of the orbit: Sendai's ten-year experience. *Auris Nasus Larynx* 2003;30:259–62.
4. Chandler JR, Langenbrunner DJ, Stevens ER. The pathogenesis of orbital complications in acute sinusitis. *Laryngoscope* 1970;80:1414–28.
5. Mills RP, Kartush JM. Orbital wall thickness and the spread of infection from the paranasal sinuses. *Clin Otolaryngol Allied Sci* 1985;10:209–16.
6. Caruso PA, Watkins LM, Suwansaard P, et al. Odontogenic orbital inflammation: clinical and CT findings—initial observations. *Radiology* 2006;239:187–94.
7. Allan BP, Egbert MA, Myall RW. Orbital abscess of odontogenic origin. Case report and review of the literature. *Int J Oral Maxillofac Surg* 1991;20:268–70.
8. Chaudhry IA, Shamsi FA, Elzaridi E, et al. Outcome of treated orbital cellulitis in a tertiary eye care center in the Middle East. *Ophthalmology* 2007;114:345–54.
9. Cheung SW, Lee KC, Cha I. Orbitocerebral complications of pseudomonas sinusitis. *Laryngoscope* 1992;102:1385–9.
10. Ferguson MP, McNab AA. Current treatment and outcome in orbital cellulitis. *Aust N Z J Ophthalmol* 1999;27:375–9.
11. McIntosh D, Mahadevan M. Failure of contrast enhanced computed tomography scans to identify an orbital abscess. The benefit of magnetic resonance imaging. *J Laryngol Otol* 2008;122:639–40.

Vision Loss After Inadvertent Corneal Perforation During Lid Anesthesia

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Abstract: **A 68-year-old woman was referred for glaucoma management after inadvertent corneal perforation during eyelid anesthesia for upper eyelid blepharoplasty. A mixture of 50:50 2% lidocaine with 1:100,000 epinephrine and 0.5% bupivacaine buffered with sodium bicarbonate was injected intracamerally. Decreased vision and uncontrollable intraocular pressure resulted, despite prompt anterior chamber washout. Examination showed corneal edema, inflammation, and secondary angle closure. Intraocular pressure control with seton placement led to an improvement in vision; however, mild corneal haze remained, and specular microscopy showed endothelial cell loss, presumably secondary to local anesthetic toxicity. Inadvertent ocular penetration is a rare but serious complication of local eyelid anesthesia. Prompt recognition is essential to institute appropriate management and minimize subsequent vision loss.**

Visual loss after blepharoplasty is rare and most often secondary to postoperative retrobulbar hemorrhage. Inadvertent globe penetration during eyelid anesthetic injection is

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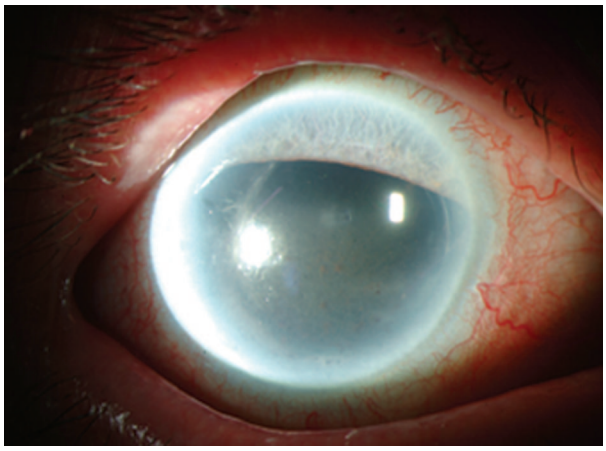

External photograph on initial presentation. The needle tract can be seen centrally, and the iris is retracted superiorly and inferiorly.

an uncommonly reported event that can lead to corneal perforation, traumatic cataract, vitreous hemorrhage, retinal tears or detachment, and optic nerve injury.[1] In addition to needle trauma itself, inadvertent injection of anesthetic may cause damage. Preserved anesthetics routinely used for local subcutaneous anesthesia are not used for intracameral anesthesia because of their potential for corneal endothelial toxicity. Current knowledge about the ocular toxicity of preserved lidocaine and bupivacaine is based on studies using in vitro cell cultures and animal models. We report a case of angle-closure glaucoma and endothelial cell loss after inadvertent intracameral injection of local anesthetic for upper eyelid blepharoplasty.

CASE REPORT

A 68-year-old woman was referred for glaucoma management 5 weeks after inadvertent corneal perforation during local anesthetic injection for upper-eyelid blepharoplasty. A mixture of 50:50 2% lidocaine with 1:100,000 epinephrine and 0.5% bupivacaine buffered with sodium bicarbonate was injected intracamerally. Immediately after injection, the patient reported pain, and examination showed a dilated pupil and corneal edema. The surgeon promptly "washed out" the anterior chamber several times by draining and reforming it with Balanced Salt Solution (Alcon Surgical, Fort Worth, TX). Postoperatively, medical therapy was initiated for elevated intraocular pressure (IOP) as high as 52 mm Hg.

Visual acuity on presentation had decreased from 20/30 to 20/200 in the affected eye. Examination revealed a 0.6 log unit relative afferent pupillary defect and an IOP of 45 mm Hg in the affected eye on dorzolamide-timolol fixed combination, brimonidine, bimatoprost, and oral methazolamide. Slit-lamp examination showed diffuse microcystic corneal edema, dense pigment on the endothelium, a superiorly and inferiorly retracted iris, and a centered posterior chamber intraocular lens (Figure). Central corneal thickness was 532 μm as compared with 485 μm in the unaffected eye. Gonioscopy showed 270 degrees of peripheral anterior synechiae with angle structures visible only in the superior quadrant. Two days later, a superotemporal Ahmed seton was placed between the iris and the intraocular lens. Postoperatively, the IOP stabilized in the mid to high teens, and the vision improved to 20/50. Endothelial examination with Confoscan4 (Nidek Inc., Fremont, CA) showed cell pleomorphism and polymegathism. Postoperative endothelial cell count was 471 cells/mm^2, and slit-lamp examination showed mild corneal haze.

DISCUSSION

Blepharoplasty is one of the most commonly performed oculoplastic procedures. The most dreaded complication is permanent visual loss secondary to retrobulbar hemorrhage, which can occur hours to days postoperatively. Reports of inadvertent ocular perforation are generally associated with retrobulbar or peribulbar anesthetic injections and are much less common with eyelid anesthesia. Severe vision loss secondary to anesthetic injection is usually related to initially unrecognized posterior globe puncture. Consequences include central retinal artery occlusion and optic atrophy, presumably secondary to high IOP.[2] We report a case of visual loss after corneal perforation secondary to anesthetic toxicity.

Routine local anesthetics used for blepharoplasty include lidocaine and/or bupivacaine with or without epinephrine, and contain the preservative methylparaben. Chang et al.[3] compared in vitro corneal endothelial cell toxicity among a variety of commercially available local anesthetics. Cultured endothelial cell damage after 1 minute of exposure to preserved lidocaine 2% with epinephrine was not statistically different from that in a control group. After 1 minute of exposure to bupivacaine 0.5%, however, significantly more endothelial cell damage was found compared with controls. This has been attributed to the higher lipid solubility of bupivacaine. In vivo rabbit models have shown transient corneal edema after intracameral injection of undiluted bupivacaine, suggesting endothelial cell damage.[4] Less cell damage was seen in vivo and in vitro when the bupivacaine was diluted with lactated Ringer solution (1:10 and 1:1, respectively).[4,5] In our patient, inadvertent intracameral injection of a local anesthetic mixture led to inflammation, iris retraction, and secondary angle-closure glaucoma in addition to the endothelial cell loss. Because studies have shown that lidocaine is well tolerated in the anterior chamber, we postulate that bupivacaine and/or the preservative was the inciting factors responsible for angle closure and endothelial damage. Lid anesthetic injection should be performed with caution to avoid ocular perforation. Preventive measures include bending the needle 30 degrees superiorly to avoid posterior penetration, should the patient move or involuntarily sneeze during injection.[1] In addition, a corneal shield may be used to protect the globe. Severe pain during injection, corneal clouding, and a tense globe are signs of ocular penetration. Prompt recognition and examination are essential for appropriate management to minimize subsequent vision loss.[6]

REFERENCES

1. Vagefi MR, Lin CC, McCann JD, Anderson RL. Local anesthesia in oculoplastic surgery: precautions and pitfalls. *Arch Facial Plast Surg* 2008;10:246–9.
2. Shiramizu KM, Kreiger AE, McCannel CA. Severe visual loss caused by ocular perforation during chalazion removal. *Am J Ophthalmol* 2004;137:204–5.
3. Chang YS, Tseng SY, Tseng SH, Wu CL. Cytotoxicity of lidocaine or bupivacaine on corneal endothelial cells in a rabbit model. *Cornea* 2006;25:590–6.
4. Judge AJ, Najafi K, Lee DA, et al. Corneal endothelial cytotoxicity of topical anesthesia. *Ophthalmology* 1997;104:1373–9.
5. Anderson NJ, Nath R, Anderson CJ, Edelhauser HF. Comparison of preservative-free bupivacaine vs lidocaine for intracameral anesthesia: a randomized clinical trial and in vitro analysis. *Am J Ophthalmol* 1999;127:393–402.
6. Rinkoff JS, Doft BH, Lobes LA. Management of ocular penetration from injection of local anesthesia preceding cataract surgery. *Arch Ophthalmol* 1991;109:1421–5.