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CONTINUING EDUCATION PROGRAM: FOCUS...

Tumor pathology of the orbit

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Abstract The term orbital tumor covers a wide range of benign and malignant diseases affecting specific component of the orbit or developing in contact with them. They are found incidentally or may be investigated as part of the assessment of a systemic disorder or because of orbital signs (exophthalmos, pain, etc.). Computed tomography, MRI and Color Doppler Ultrasound (CDU), play a varying role depending on the clinical presentation and the disease being investigated. This article reflects long experience in a reference center but does not claim to be exhaustive. We have chosen to consider these tumors from the perspective of their usual presentation, emphasizing the most common causes and suggestive radiological and clinical presentations (progressive or sudden-onset exophthalmos, children or adults, lacrimal gland lesions, periorbital lesions and enophthalmos). We will describe in particular muscle involvement (thyrotoxicosis and tumors), vascular lesions (cavernous sinus hemangioma, orbital varix, cystic lymphangioma), childhood lesions and orbital hematomas. We offer straightforward useful protocols for simple investigation and differential diagnosis. Readers who wish to go further to extend their knowledge in this fascinating area can refer to the references in the bibliography.

Orbital tumors may be discovered on imaging in many circumstances [1]. They may be found incidentally on a CT or MRI of the brain or facial bones, performed for a non-ophthalmological disease. Alternatively, the lesion may be looked for directly in a patient with ophthalmological signs such as exophthalmos, unilaterally reduced visual acuity, diplopia, and orbital or periorbital pain.

These imaging modalities play a major role in the diagnostic and staging assessment of the lesion, guide therapeutic decision and can be used for simple monitoring or follow up on treatment.

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Imaging: what to use and what protocol?

Radiologists have three methods of examining the orbit [2]. These can be used either in isolation or in combination, depending on the clinical and anatomical features. Imaging is used to confirm exophthalmos and define its grade from the external bicanthal/eye globe line ratio. A line crossing the posterior third defines the grade I, one next to the posterior pole defines the grade II, and one passing behind the posterior pole defines the grade III. The grade is one of the factors used in monitoring the disease, as the risk of the lesion damaging the optic nerve as a result of stretching increases with grade.

Color Doppler Ultrasound (CDU) is extremely useful if a vascular lesion is expected (blue-colored mass, varying with head position, concomitant conjunctival abnormalities), for an anterior lesion (lacrimal gland or eyelid), and when evidence is needed to distinguish between inflammation and lymphoma. A high frequency probe is used, applied directly to the lesion or examining the lesion through gel applied to the closed eyelid. The Valsalva maneuver and patient hyperextension method improve the diagnosis of an orbital varix.

Computed tomography is used to examine the orbital wall, to demonstrate calcifications inside lesions and to describe their morphology (small bead appearance of phleboliths, or lumps shaped calcifications in cartilaginous lesions). It can be useful for positional maneuvers (imaging in prone position is used to assess an orbital varix). The investigation is started with an unenhanced study unless orbital cellulitis is suspected, when contrast injection at the outset is sufficient to confirm the diagnosis and for the staging assessment. Either conventional enhancement or arterial or venous CT angiography may be used depending on the suspected disease. When an orbital varix is investigated, conventional images with the patient in supine position are supplemented with a second CT acquisition with the patient in prone position in order to demonstrate the increase in the volume of the lesion which is typical of this pathology.

MRI remains the reference method to type the lesion, assess extension and investigate for any concomitant cerebral abnormalities. In addition to patients with pacemakers, it is contraindicated in patients with intra or periorbital metal foreign bodies. It can be performed in patients who have had local radiotherapy for a choroid melanoma and patients can continue to wear their contact lenses. Makeup however may cause very troublesome artifacts. We recommend that centers that carry out orbital MRI have their secretaries inform patients to avoid wearing any makeup and to have access to makeup remover and cotton wool pads for patients who did not follow their advice! MRI provides information about the site, morphology and structure of the lesion.

We propose the following protocol. The patient is asked to relax and to keep his eyes shut.

After a scout view focused on the orbit, we perform thin (3 mm) axial T1 and diffusion sequence following the anteroposterior axis of the orbit (grading the exophthalmos, analyzing the orbital apex), thin (2 mm) coronal T2 sequence perpendicular to the former one, from the anterior part of the ocular bulb to orbital apex. Injection is either classical or performed following a dynamic protocol (study of the tumor microvascularization) and followed by thin axial and coronal and/or sagittal T1 FATSAT sequences. Depending on the results of this first step, complementary sequences may be proposed such as prone position axial T1 GD FATSAT acquisition (to depict an orbital varix), axial and coronal T2 FATSAT study of the face and upper cervical area (extension of an hemangiolymphoma), brain study. If a vascular malformation or an aneurysm is suspected, MRA (either TOF or dynamic angiography) is added.

A few important points must be remembered. Fat suppression (FATSAT) after injection is used routinely, as it improves the detection of contrast enhancement and offers a clear detection of the boundaries of the lesion and its relationships with orbital structures (optic nerve, apex muscles). This is required particularly to assess posterior lesions, as identification of extension towards the superior orbital fissure is seen far more clearly than without fat suppression. The contrast injection may be given during a T1 weighted perfusion sequence (or dynamic injection) which assesses the tumor microvascularization. This perfusion weighted image has been used for several years to characterize particularly salivary ENT lesions, and also for breast and liver tumors. Its use in ophthalmology is more recent [3]. Results correlate well with those found for other sites of lesion. The results of the T1 perfusion weighted image are described in a 2011 article [4] in 16 cases of malignant orbital tumors and 43 cases of benign orbital tumors. The authors showed that all of the tumors which had a persistent pattern or type I curve were benign, those with a steep slope and plateau pattern or type II were either benign or malignant, and those with a rapid rise and peak (washout pattern) or type III were more likely to be malignant. The use of various criteria from these dynamic studies should help to improve distinctions between benign and malignant and improve characterization of masses within each category.

Progressive exophthalmos

Progressive exophthalmos suggests an intra-orbital space occupying lesion. This lesion may be born from anatomical orbital component (muscle, vessels, lacrimal gland, optic nerve, orbital wall) or non-orbital components (tumor or infection).

Exophthalmos of muscular origin

This is a common cause of exophthalmos, and when bilateral, even asymmetrical the first consideration should be a metabolic cause and, because of its frequency, dysimmune orbital disease usually related to autoimmune hyperthyroidism (Basedow's disease). This disease affects the oculomotor muscles (the superior, medial and inferior rectus muscles, the superior oblique and far more rarely the lateral rectus) and the orbital fat. The exophthalmos is due to hypertrophy of these muscles, the bodies of which contain deposits of mucopolysaccharides, fat and inflammatory cells. These modifications explain the morphology of the disease: spindle shape muscles (Fig. 1), with a globular body, preserved tendon, and its density and appearance: hypodense fatty areas, hyperintense on T1 and T2 weighted

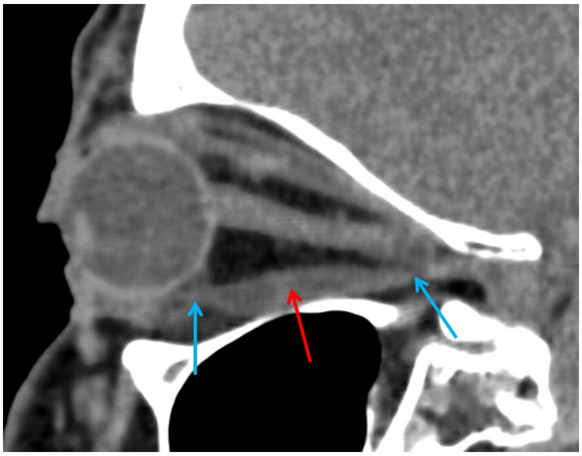

Figure 1. Typical muscle CT appearance in autoimmune thyrotoxicosis (sagittal oblique reconstruction). Thin tendon (blue arrows) and thickened muscle bodies (red arrows).

images, hyperintense inflammation on T2 images persisting in the fat suppression or Fat Sat images (Fig. 2), and then hypointense fibrosis on T1 and particularly T2 weighted imaging. While unenhanced CT is sufficient in most cases to make a diagnosis, MRI however is essential to diagnose inflammation, which is responsible for most of the clinical complications of the disease (diplopia, reduced visual acuity). Concomitant ophthalmological signs (retraction of the upper eyelid, gritty eyes) which are generally due to hyperthyroidism may guide the diagnosis from the outset. This diagnosis is made from biological confirmation of dysthyroid status. It should be remembered that exophthalmos may be unilateral, and may precede the finding of hyperthyroidism (5 % of cases). The radiological signs of this disorder therefore need to be understood and this diagnostic possibility given in the report.

KEY POINT

The investigation MRI protocol for dysimmune orbital disease is simple: scout view, 3 mm axial T2 weighted sections (to assess the grade of the exophthalmos and analyse orbital apex), T2, T1 and T2 Fat Sat coronal images, positioning these from the most anterior part of the globe to the apex, perpendicular to the anteroposterior axis of the orbit.

Figure 3. Lung cancer. Left exophthalmos. Metastatic lesion which has developed in the left medial rectus muscle.

What if the muscle lesion is not autoimmune thyrotoxicosis? Muscles may be affected by myositis (pain, very inflammed appearance of one or more muscles, with tendon involvement), a secondary tumor (local mass, history of breast or lung malignancy, etc.) (Fig. 3) [5].

Orbital wall lesions can also be responsible of progressive exophthalmos. These are mostly spheno-orbital osteomeningioma with a predominantly bone forming component (Fig. 4), or secondary lesions from cancers with a propensity to spread to bone (particularly prostate).

The space occupying effect is due to both tissue and bone. Other bone lesions such as fibrous dysplasia, either isolated or as part of the McCune-Albright syndrome, osteomas and primary lesions, are rarer [6].

Orbital infiltration

This may be caused by a malignant blood disorder (lymphoma or leukemia). The clinical context and extension are very helpful in making the diagnosis (Fig. 5) which also includes inflammatory diseases such as sarcoidosis, or more rarely, Wegener's disease.

An imaging investigation shows an orbital mass

When the mass is isolated, it is important to know how to use the clinical features (past history, ophthalmological signs), the image appearance: diffusion, contrast enhancement (T1

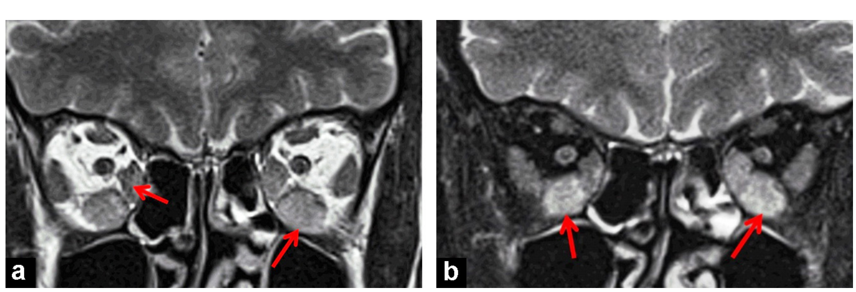

Figure 2. Bilateral exophthalmos due to dysthyroid orbital disease. Orbital MR with coronal T2 weighted sections (a) and T2 Fat Sat sections (b). Muscle hyperintensity (red arrows) persisting with Fat Sat and reflecting acute inflammation.

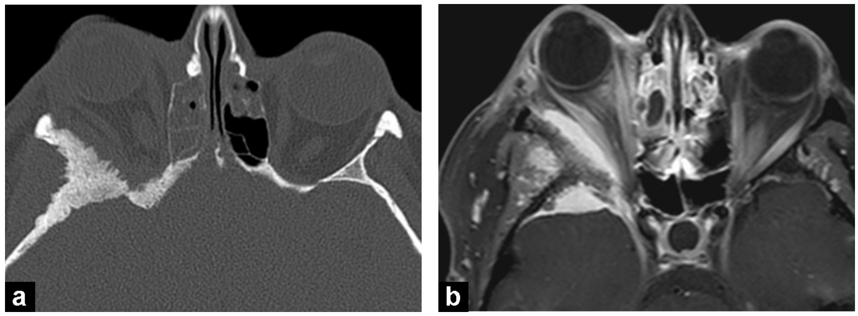

Figure 4. Thickening of the right external orbital wall with spiked edges on CT (a), meningeal contrast enhancement (b), spheno-orbital meningioma.

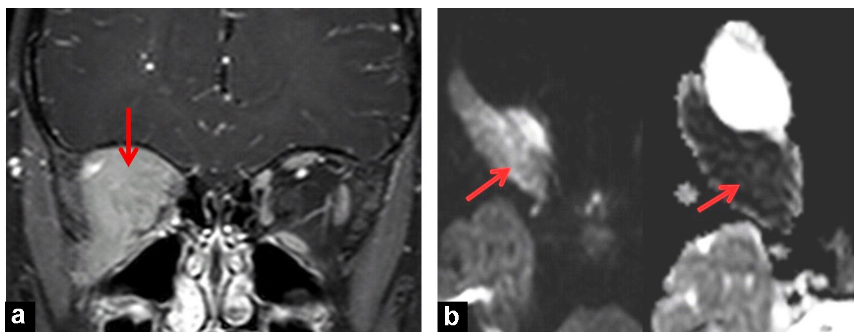

Figure 5. Four year old child with leukemia. Rapidly developing right exophthalmos. Orbital site of the blood disease (red arrows) enhancing with contrast (a), very cellular with B1000 hyperintensity and low ADC coefficient (b).

perfusion weighted image), and positional maneuvers in order to guide the diagnosis.

A logical approach taking account of the incidence of the lesions is needed. A tumor which is clearly delineated, hypointense on T1 weighted imaging, and clearly hyperintense on T2 weighted imaging, dense on CT with heterogeneous contrast enhancement rapidly becoming homogeneous over time, (which is easy to see if enhanced MRI views are taken in two planes) is a vascular tumor and on the basis of incidence, is mostly likely to be a cavernous angioma. Despite the term vascular mass, there is no detectable flow within the lesion on CDU [7] (Fig. 6).

The main differential diagnosis is from a hemangiopericytoma, which is far more aggressive, and often has a weaker hyperintensity on T2 weighted imaging. The diagnosis is often made from progression (rapid growth, unlike a cavernous angioma) [8].

What to consider if it is not a cavernous angioma?

A lymphoma or malignant blood disorder if the lesion is very cellular (low ADC). Diffusion weighted imaging should, therefore, form part of all protocols used to investigate orbital masses [9—11].

A Schwannoma, which develops on the branches of the Vth nerve, or oculomotor nerves, enhances intensely, and grows slowly, and when it becomes bulkier may become cystic (Fig. 7) [12].

A primary malignant tumor, particularly sarcoma, or liposarcoma, in which the fat component is often very minor.

A cystic lesion suggests a lymphangioma, which may be diagnosed late, and also a foreign body granuloma. The cyst content is protein-rich. A lesion with small clearly

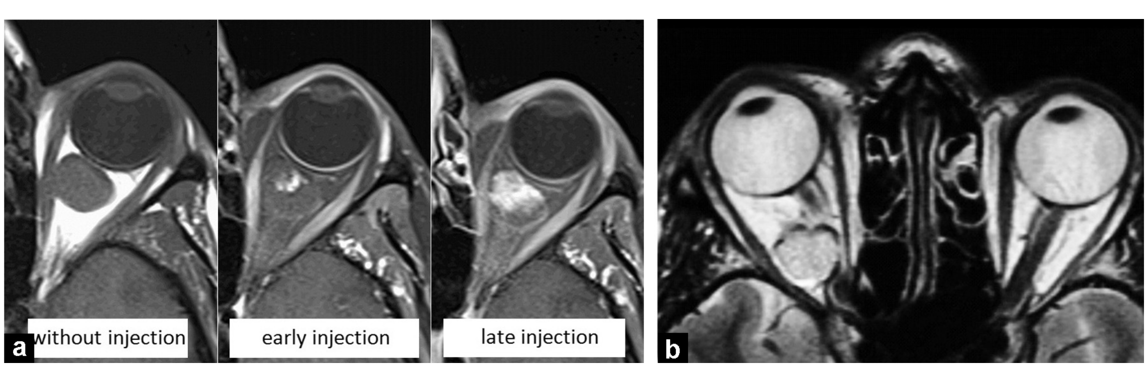

Figure 6. Examples of intra-orbital lesion, hypointense on T1 weighted imaging, enhancing progressively (a), hyperintense on T2 weighted imaging with an incomplete distinct hypointense halo (b). Cavernous hemangiomas.

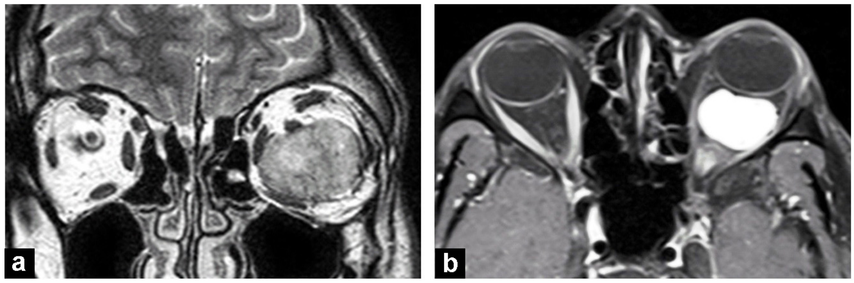

Figure 7. Left intra-orbital mass with moderate hyperintensity on T2 weighted imaging (a), clearly delineated, intensely enhancing with contrast (b). Orbital Schwannoma.

delineated cysts, outside of a clear context of infection, is a metastasis until proven otherwise. This usually develops in a muscle (the second commonest site after the choroid for orbital secondary lesions) (Fig. 3).

Alternatively, it may represent an inflammatory lesion, when the clinical signs are suggestive (orbital pain, chemosis). The lesions enhance intensely with contrast on imaging and have a blurred inflamed appearance in the adjacent fat, which is particularly clearly seen on T1 weighted Fat Sat enhanced and T2 weighted Fat Sat images [13]. The T2 weighted hyperintensity in the acute phase is often replaced when the inflammation becomes chronic by a hypointensity representing fibrous transformation. The preferred sites for this type of inflammation are the lacrimal gland, perioptic meninges fat and oculomotor muscles (myositis). Several sites of disease are often present and the inflammation may even be bilateral (Fig. 8).

Other lesions develop in contact with the optic nerve, and partially or completely surrounds it (rail line appearance on an axial view, or target appearance on a coronal view, formed by the circumferential lesions centered on the optic nerve). These enhance with contrast and often present with a gradual fall in visual acuity, which patients themselves often do not realize. This imaging feature is usually caused by an optic nerve sheath meningioma. Computed tomography may help in the diagnosis if it shows calcifications inside the lesion (Fig. 9). The meningioma has the typical features of this type of tumor, with intense contrast enhancement, and clear demarcation. It may be circumferential or form a mass and cause progressive optic atrophy. The major risk from progression is extension through the orbital canal to the endocranial meninges and meninges of the contralateral optic nerve. Enhanced MRI with thin T1 Fat Sat axial sections is the best monitoring method and can be used to plan when surgery may be needed (Fig. 9) [14—16].

Rapid or sudden-onset exophthalmos

In children, this may be due to a malignant tumor, when no signs of inflammation or pain are present. Urgent imaging is needed as it may reflect a rhabdomyosarcoma, which is the commonest childhood mesenchymal tumor, orbital sites of which are usually embryonic tumors. The tumor grows extremely quickly and on imaging appears as a clearly demarcated mass, which is dense on CT, isointense on T1, and hyperintense T2 weighted MR images, enhancing with contrast, and often containing small areas of hemorrhage. They may occur in isolation or in conjunction with signs reflecting their aggressive nature, including destruction of the orbital wall (30 to 40% of cases) which is seen clearly on CT, and extension to the sinuses and meninges. They require urgent management with biopsy and chemotherapy (Fig. 10) [17].

Orbital sites of malignant blood disorders may produce an identical uni- or bilateral clinical picture. The lesion is more infiltrating, and diffusion weighted imaging is suggestive (low ADC because of the high cellularity of the masses). A clinical context of leukemia helps in the diagnosis [18,19].

Rapid or sudden exophthalmos may reflect a hematoma at any age (Fig. 11).

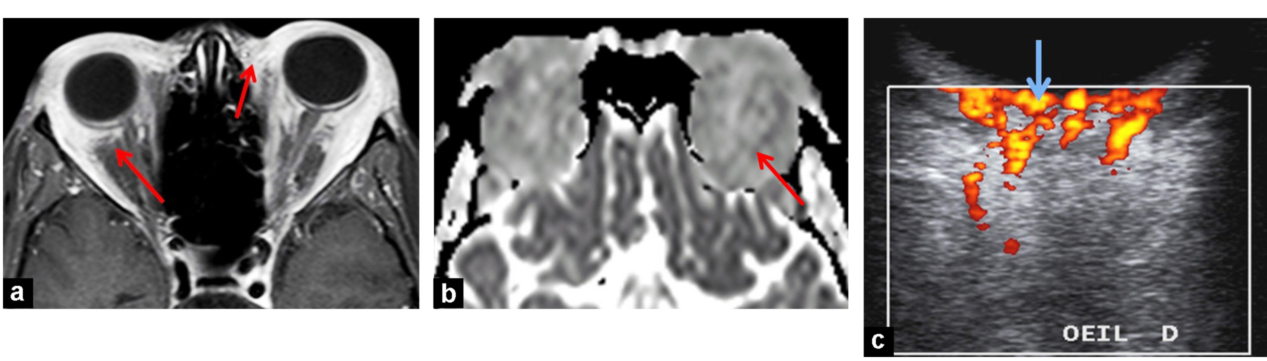

Figure 8. Bilateral orbital inflammation (red arrows). Intense contrast enhancement (a), ADC not reduced (b). Overall hypervacularization on CDU (blue arrow, c).

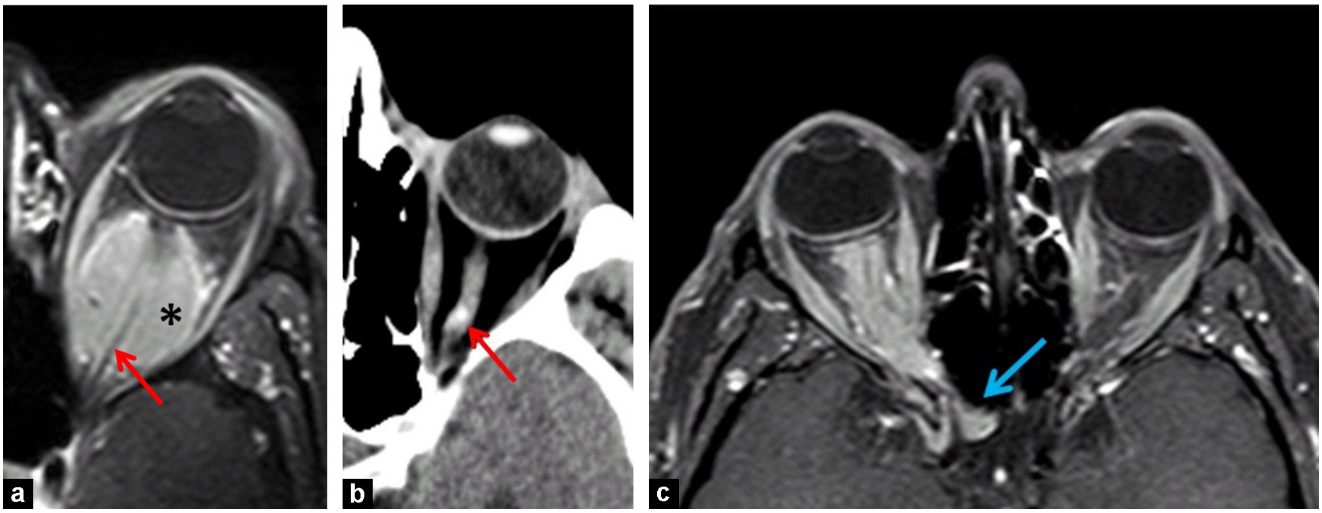

Figure 9. Examples of intra-orbital optic nerve sheath meningiomas (asterisk) with atrophic optic nerve (red arrow, a), calcified on CT (red arrow, b), and extending to the apex of the orbit (blue arrow, c).

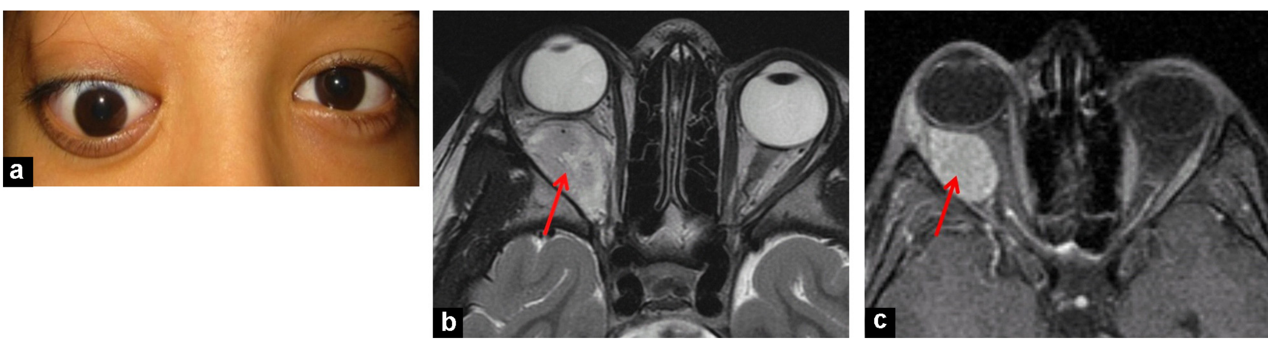

Figure 10. Rapidly progressive exophthalmos with right-sided wasting (a). T2 weighted MR (b) and T1 weighted enhanced Fat Sat (c): lesion (red arrows) hyperintense on T2 weighted imaging and enhancing with contrast. Transferred to a specialist center and biopsied on the same day: rhabdomyosarcoma.

Figure 11. Left orbital hematoma with T2 weighted hypointensity reflecting its acute nature (red arrow).

Apart from injury, the main cause to consider is rupture of an orbital varix, which is occasionally difficult to identify in the acute phase [20]. A varix is the most common orbital vascular malformation and is seen on imaging views taken on prone position (MRI, CT) or hyperextension (ultrasound views) positions. The varix increases in volume with these positional maneuvers on all imaging methods. Additional features supporting the diagnosis include a description of exophthalmos varying with position given by the patient (the patient's eye ''comes'' out when he/she leans forward), phleboliths on CT, bidirectional flow on CDU, and a lesion with serpiginous morphology (Fig. 12).

Bleeding within a cystic lymphangioma should be considered in children. Cystic lymphangioma, a hemodynamically independent hamartoma, is one of the angiodysplasias and consists of branching abnormal lymphatic vessels present from birth, accounting for 1% of orbital tumors and 12% of vascular tumors [21]. It is diagnosed early, usually before the age of 10 years old. Complications develop shortly after the diagnosis, at around the age of 13. The tumor has pleomorphic clinical features, with both superficial and deep lesions causing, occasionally severe, progressive exophthalmos, which may develop suddenly. This is due to reactive lymphoid hyperplasia, as a result of an ENT infection, or hemorrhage within the lesion, and may require urgent surgery because of the risk of damage to the optic nerve (by compression or stretching). Surgery involves draining the largest hemorrhagic cysts and less commonly, excision. Some believe that surgery increases the risk of recurrent hemorrhage. The heterogeneous appearances

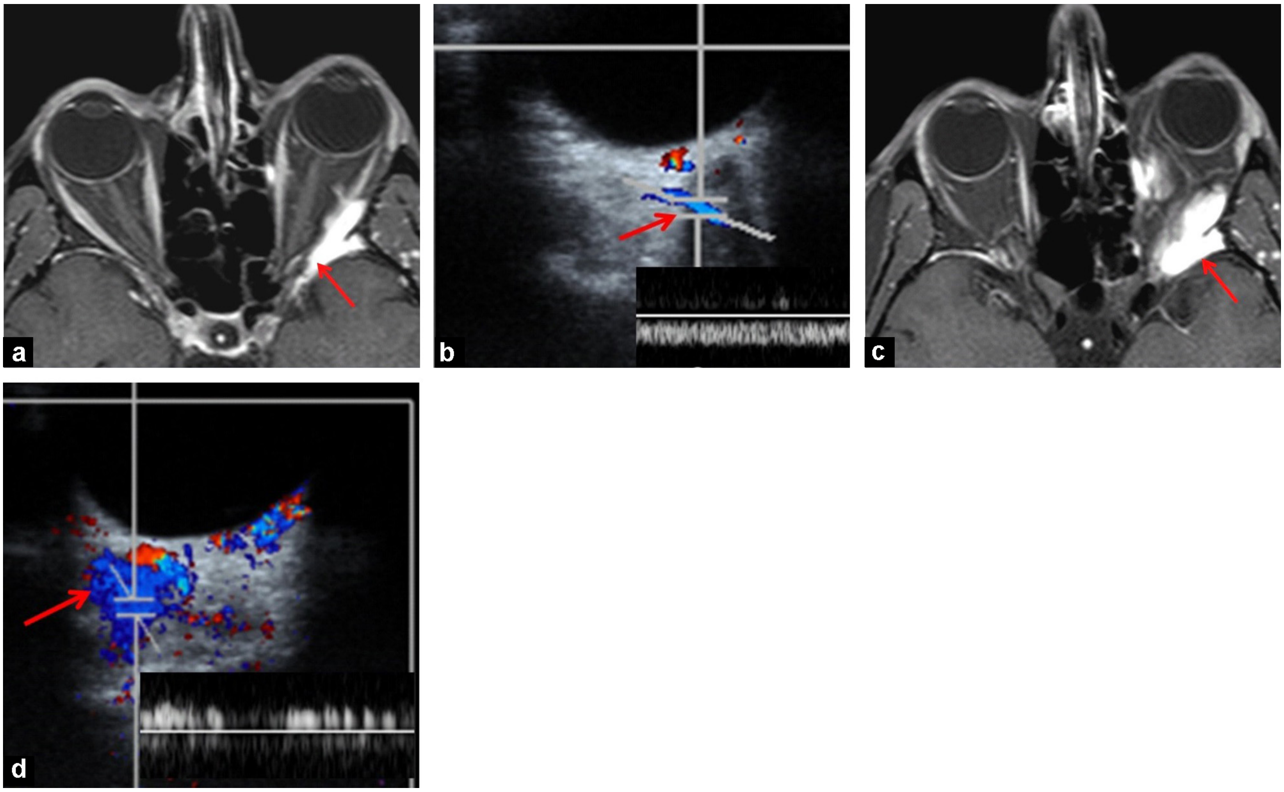

Figure 12. Left orbital varix (red arrow). T1 weighted MR with enhanced Fat Sat and CDU with the patient in decubitus (a and b) and in prone/hyperextended (c and d) position. The lesion increases in volume after changing position.

of the lesion are explained by its type and progression. Imaging shows more or less extensive orbital infiltration by cystic lesions of variable volume and number, and/or hemorrhagic lesions with fluid levels in the acute phase (Fig. 13). The lesion may be localized to the orbit or may extend to the deep facial bones, neck or pharynx and therefore requires a staging assessment, which is easier with MRI than CT. The most ''productive'' view is T2 weighted Fat Sat either 3D or axial and coronal plane imaging covering all of the regions potentially involved [22—24].

Far more rarely, an orbital hematoma is caused by hemorrhage within a tumor [25], or represents a complication of another intra-orbital vascular malformation (AVM, ophthalmic artery aneurysm).

Sudden-onset exophthalmos may also reflect intra-orbital drift of a frontal or ethmoid mucocele, particularly after a patient has blown his/her nose, causing a fracture of the sinus wall. This is usually easy to diagnose on imaging (Fig. 14) [26].

What imaging should be carried out for sudden-onset exophthalmos? The investigation to perform in this situation is CT. In rapidly progressing exophthalmos in a child, MRI should be performed if possible as this provides evidence of rhabdomyosarcoma if present and provides a better staging assessment than CT.

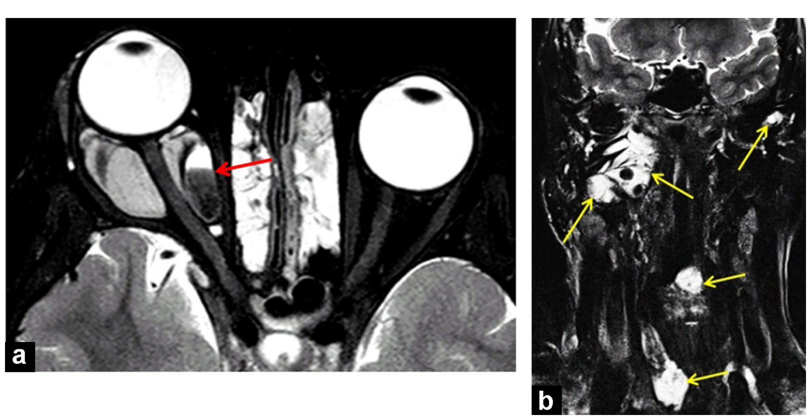

Figure 13. T2 weighted axial Fat Sat orbital MR (a) and cervico-facial coronal MR (b). Left orbital cystic lymphangioma with liquid-fresh blood fluid level (red arrow). Multiple other sites (yellow arrows).

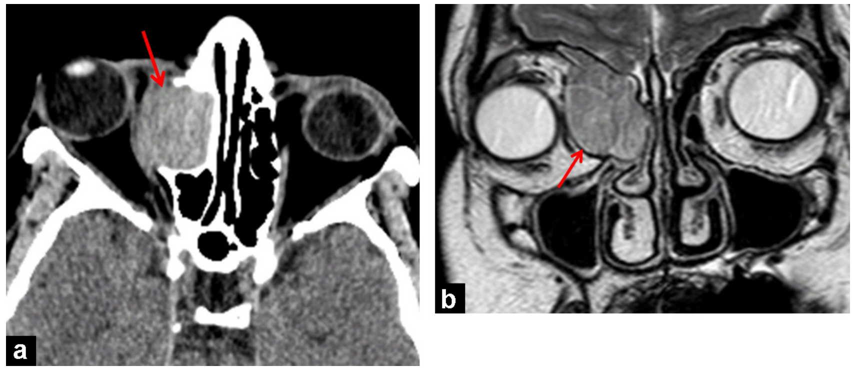

Figure 14. CT (a) and T2 weighted MR (b). Ethmoid mucocele presenting with sudden-onset exophthalmos, hyperdense protein content, hypointense on T2 weighted imaging (red arrow).

Lacrimal gland disease

The clinical presentation of lacrimal gland hypertrophy is highly suggestive with a ''comma'' appearance at the lateral end of the eyebrow (Fig. 15) [27].

Lacrimal gland lesions can be divided into two histological categories, each including both benign and malignant lesions. The epithelial lesions (45 to 50%) include dacryops, and pleomorphic adenomas, and the non-epithelial lesions (50 to 55%) include dacryoadenitis, non-Hodgkin's lymphoma, and benign lymphoid hyperplasia. Two diagnostic approaches can therefore be considered to look for tumor and inflammation. Imaging such as ultrasound confirms the site of the lesion although definition of the lesion relies particularly on MRI and ultrasound. We only consider the common lesions here.

A pleomorphic adenoma or mixed tumor has dual epithelial and myxoid components. When typical, it is characteristic on MRI with an obvious hyperintensity on T2 weighted imaging, and pronounced heterogeneous enhancement (unless it is small), with a type I dynamic curve, which is hyperintense on a B1000 diffusion weighted image with a high ADC (Fig. 16).

Figure 15. Characteristic left lacrimal gland hypertrophy (red arrow).

Dacryops is a dilatation of the intraglandular duct structures, and presents as a uni- or multicystic structure [28].

Non-hematological malignant tumors (adenocarcinoma, cystic adenoid carcinoma) are often heterogeneous, and poorly demarcated, hypointense on T2 weighted imaging with a low ADC and a type II or III enhancement curve. The staging assessment is by MRI. CT is performed in addition in aggressive disease involving the orbital wall.

Inflammatory lesions may be idiopathic or occur as a complication of sarcoidosis, or Wegener's disease. These are uni- or bilateral, and either isolated or associated with more diffuse orbital involvement. They are often hypointense on T2 weighted imaging (fibrous component), with no fall in the ADC (unlike lymphoma) (Fig. 8). In superficial masses, CDU clearly distinguishes between blood malignant disorders (hypoechogenic nodules separated by hypervascularized lines) (Fig. 17) and inflammation (hypervascularized anarchic mass) (Fig. 12) [29].

Periorbital lesions

The diagnostic approach to a periorbital region mass is based on the clinical appearances and age of the patient. The predominant causes of etiologies are vascular lesions, infections, and skin tumors.

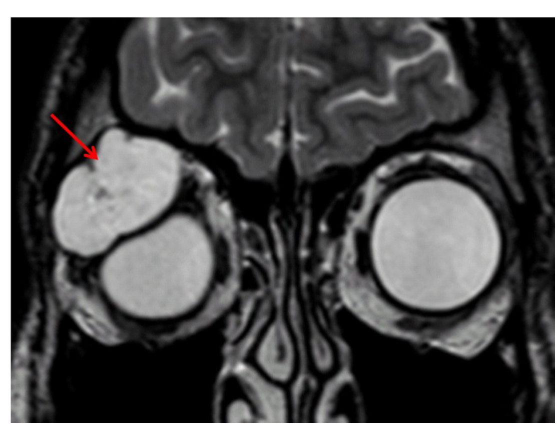

Figure 16. T2 weighted orbital MR. Right pleomorphic lacrimal gland adenoma (red arrow).

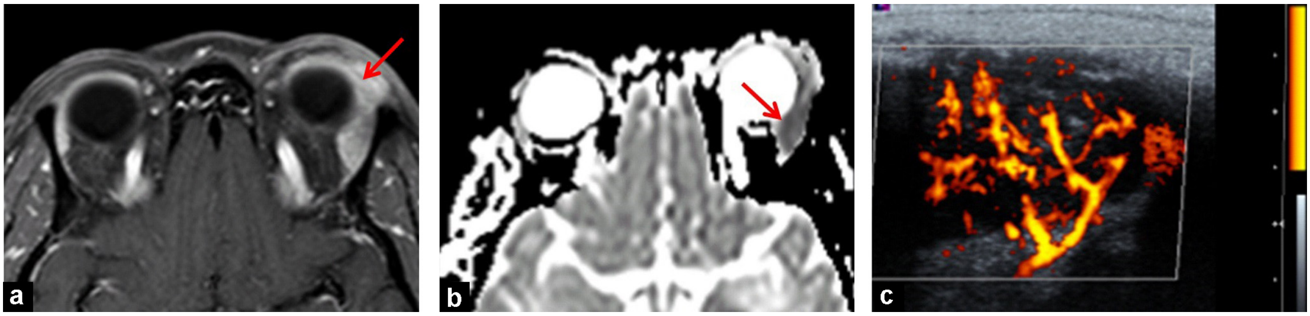

Figure 17. T1 weighted enhanced Fat Sat orbital (a) and diffusion ADC (b) MR, and CDU (c). Left lacrimal gland lymphoma (red arrow).

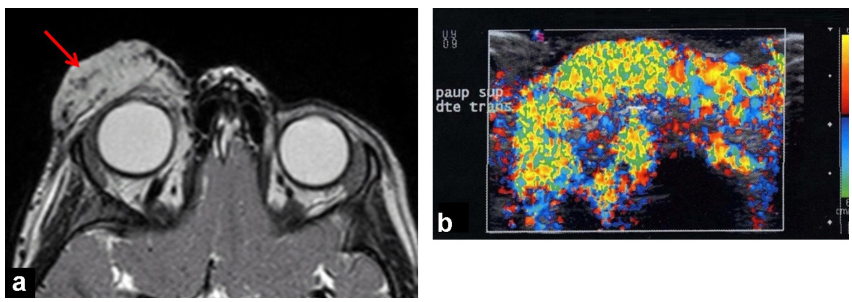

Figure 18. T2 weighted orbital MR (a) and right palpebral CDU (b). Juvenile angioma (red arrow).

A purple colored, renitent palpebral swelling in an infant is highly suggestive of a juvenile papillary angioma. CDU appearances are characteristic (fetal hypervascularization) (Fig. 18). This resolves spontaneously in 95% of cases, between the age of 1 and 6 years old, and may occur in isolation or as part of a malformation syndrome (Sturge Weber, PHACE, Bonnet-Dechaume-Blanc) [30,31].

An external swelling of the canthus in a child is usually a dermoid cyst (a cyst at the end of the eyebrow). Unenhanced CT is sufficient to investigate for intra-orbital extension or confirm the type of lesion before planning surgery (Fig. 19).

If infection is suspected in a child or adult, orbital CT is performed with enhancement from the outset with the purpose of distinguishing preseptal infection (generally originating from the lacrimal gland: dacryocystitis, only requiring antibiotic therapy) or with retroseptal extension (generally of sinus origin or post-traumatic, and requiring intensive management with hospitalization and intravenous therapy). It is also useful to identify the cause (sinusitis, dacryocystitis, foreign body, etc.).

Figure 19. Orbital CT. Right dermoid cyst (red arrow).

Retroseptal extension behind the line passing through the middle of the globe may occur into fat (dense, with perioptic contrast enhancement), and oculomotor muscles (increase in volume, intense contrast enhancement). This may be complicated by an intra-orbital abscess spreading along the walls (a mass with a hypodense center with peripheral contrast enhancement) (Fig. 20).

Clinical features may be suggestive of a malignant tumor of skin origin (for example a basal cell or squamous cells carcinoma), assessment of which is done with MRI.

Enophthalmos and intra-orbital lesions

Enophthalmos is due to enlargement of the orbit (post-traumatic, postoperative or malformation), or to reduction in the volume of the orbital contents (for example after surgery or radiotherapy). Paradoxically, some intra orbital masses also present with enophthalmos. The leading cause to consider with a mass which is fibrous in appearance, hypointense on T2 weighted imaging, and enhances with contrast is a metastasis of a scirrous cancer, above all from the breast, and occasionally from the stomach (Fig. 21). This secondary site may be the presenting feature and these potential causes should be suggested in the report (suggest that markers and mammography be performed if breast cancer is not known) [32—34]. These fibrous masses may also represent a chronic inflammatory lesion. This is an end point of known inflammation, the diagnosis of which raises no difficulties.

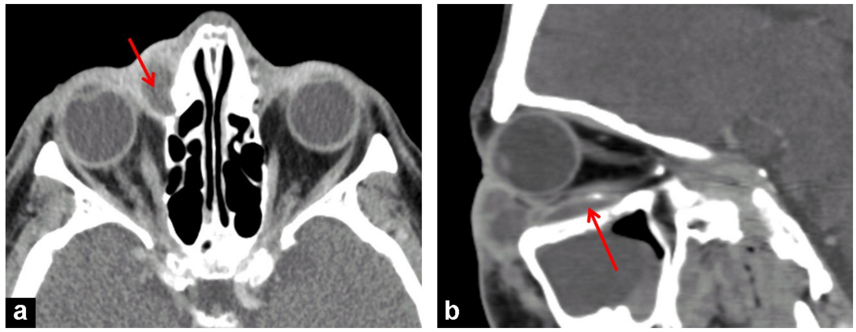

Figure 20. Enhanced orbital CTs in two patients. Preseptal orbital cellulitis (a) with dacryocystitis (red arrow) and retroseptal cellulitis (b) with contiguous subperiostial abscess (red arrow).

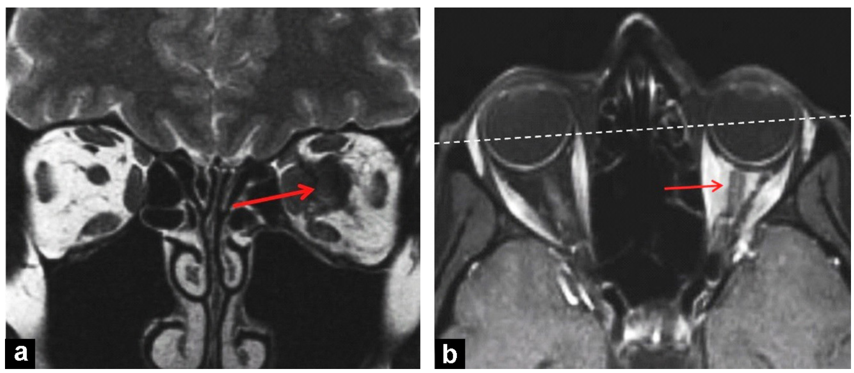

Figure 21. Left orbital metastasis from a breast cancer (red arrow) and T1 weighted enhanced Fat Sat (a) and T2 weighted image (b) MR. Left enophthalmos.

Finally, it should be noted that after long term progression with flares of exophthalmos and hemorrhagic changes, orbital varices may lead to intra-orbital lipolysis and progressive enophthalmos [35].

KEY POINT

Metastasis of a breast cancer or inflammation should be considered in enophthalmos and if the patient has a clinical history of exophthalmos followed by enophthalmos, the imaging assessment should be combined with a leaning forward view, which will confirm the diagnosis of a varix.

TAKE-HOME MESSAGES

- Progressive exophthalmia: dysimmune orbital disease.
- Intra-orbital mass: cavernous hemangioma.
- Cystic lesion: lymphangioma, schwannoma, metastasis (microcysts).
- Infection: distinguish between pre- and retroseptal site, an enhanced CT is sufficient.
- Variable exophthalmos: varix? Consider a leaning forward view.
- Children and rapid exophthalmos: rhabdomyosarcoma, perform urgent imaging.
- Sudden-onset exophthalmos: hematoma (varix?), mucocele.
- Enophthalmos and mass: metastasis from breast cancer, chronic inflammation, varix.

Conclusion

The clinical features (patient age and presentation) are essential factors in the assessment of an orbital tumor. MRI is the preferred investigation except in urgent situations or in exophthalmos suggestive of autoimmune thyrotoxicosis. The investigation will often need to be extended beyond the orbit as many systemic disorders can produce this clinical sign.

Clinical case

A 69 year old patient presents with a periorbital swelling for a month which has gradually been worsening. She has slight local pain and a past history of Hodgkin's disease eight years previously, currently in remission (Fig. 22).

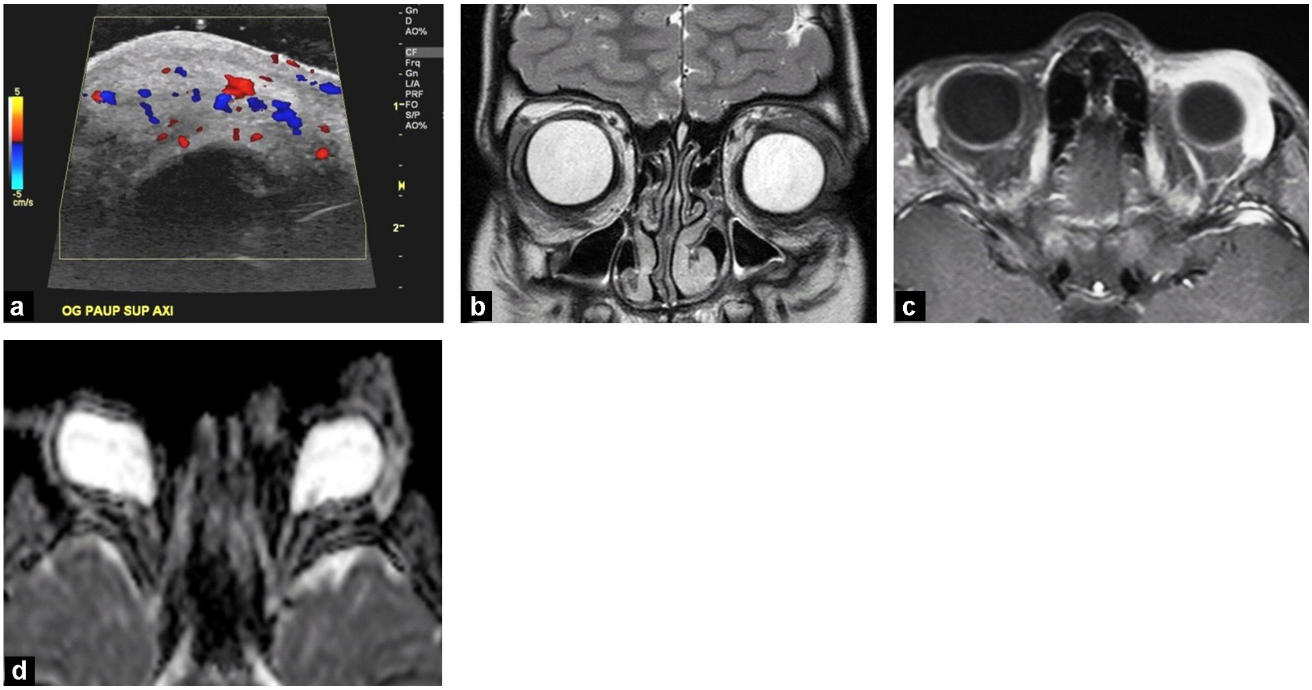

Figure 22. CDU (a), T2 weighted (b), T1 weighted Gd Fat Sat (c) and Diffusion weighted (ADC) MR (d).

Questions

1. What is the site of the lesion?
2. How does this lesion behave on CDU?
3. What is the lesion image signal? What image sequence is particularly useful to characterize it?
4. What is your diagnosis and what is the management?

Answers

1. The lesion has developed in the left lacrimal gland and extends to the adjacent soft tissues. Extension of contrast enhancement is seen on the T1 weighted image to the internal region of the canthus.
2. It is hypervascularized, anarchic, and overall slightly hyperechogenic.
3. The lesion is hypointense on T2 weighted imaging, and intensely enhances with contrast. On diffusion weighted imaging, it is paucicellular and has a relatively high ADC. This latter view provides reassuring evidence in this patient with a past history of a malignant blood disorder. The high ADC argues against the diagnosis of a site of hematological disease, which would have a very low ADC.
4. The most likely diagnosis is inflammation of the lacrimal gland (or dacryoadenitis). A general assessment into the cause is needed (investigating for sarcoidosis, Wegener's disease, etc.). Treatment is with corticosteroid therapy. Clinical and MRI follow up are required and if the lesion persists or increases in volume, a biopsy may be offered.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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