Localized Chronic Fibrosing Vasculitis Causing a Rhinophymatous Eruption

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We describe a patient with localized chronic fibrosing vasculitis who presented with a rhinophymalike eruption of the nose. To our knowledge, the manifestation of this condition on the nose has not been previously reported. The differential diagnosis also is examined.

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The term *localized fibrosing vasculitis* originally was used by Carlson and LeBoit[1] to describe an inflammatory reaction with histologic similarities to erythema elevatum diutinum (EED) and granuloma faciale (GF) but with unusual clinical presentations of nodules, papules, or masses. The patient described in this case report had localized fibrosing vasculitis presenting as a rhinophymalike enlargement of the nose. The differentiating features of EED, GF, and rhinophyma will be discussed.

Case Report

A 60-year-old white man with long-standing alopecia areata presented to the otolaryngology unit with a 10-year history of progressive enlargement and induration of the nose, causing complete obstruction of the airways. The patient was evaluated and diagnosed as having bilateral pansinusitis and nasal polyps. He underwent a nasoseptoplasty with bilateral ethmoidectomies, antrostomies, sphenoid sinusotomies, and polypectomies. Resultant tissue specimens showed nasal polyps, chronic inflammation, and tissue eosinophilia.

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The authors report no conflict of interest.

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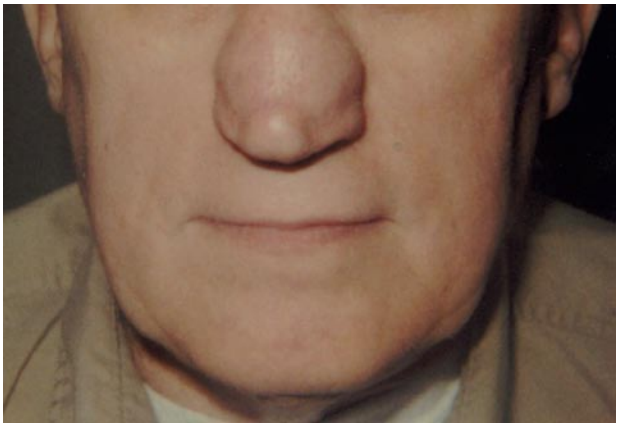

Figure 1. Patient with enlarged nose.

For 13 to 18 years, the patient continued to complain of severe nasal obstruction and an inability to breathe through his nose. When he presented for dermatologic evaluation, his nose was notably enlarged (Figure 1). Skin biopsies from the superficial and deep tissues of the nose were performed. Histopathologic examination of the skin showed fibrosing dermatitis with eosinophilia. Areas of "onion-skinlike" fibrosis interspersed with infiltrate composed of numerous eosinophils, lymphocytes, and a cluster of plasma cells were present (Figure 2). Gomori methenamine-silver, Fite, Steiner, Giemsa, and periodic acid-Schiff fungal stains were all negative for infectious organisms. The superficial portion of the biopsy showed actinic damage. A CD1a stain was negative, excluding eosinophilic granuloma. Prior sinus surgery specimens were reviewed in which findings of onion-skinlike fibrosis with mixed inflammatory infiltrate and numerous eosinophils were found. A workup for systemic vasculitis including antineutrophil cytoplasmic antibodies (cytoplasmic and perinuclear) was negative. A diagnosis of localized chronic fibrosing vasculitis was made. The patient's clinical appearance and ability to breathe through his nose improved somewhat with intralesional corticosteroid injections.

Differential Diagnosis of Rhinophymatous Eruption

Differential Diagnosis	Case Report (publication year)
Inflammatory Dermatoses	
Localized chronic fibrosing vasculitis	Current report
Granuloma faciale	Gomez-de la Fuente et al[2] (2000)
Acne rosacea	Thiboutot[3] (1994)
Benign Cutaneous Lesions	
Angioma	Marsili et al[4] (1993)
Sebaceous adenoma	Acker and Helwig[5] (1967)
Malignant Cutaneous Lesions	
Squamous cell carcinoma	Lutz and Otley[6] (2001); Ross and Davies[7] (1992)
Adenoid squamous cell carcinoma	Acker and Helwig[5] (1967)
Intranasal squamous cell carcinoma	Kornblut and Evers[8] (1973)
Basal cell carcinoma	Acker and Helwig[5] (1967); Brubaker and Hellstrom[9] (1977); Keefe et al[10] (1988); Plenk[11] (1995); Silvis and Zachary[12] (1990)
Basosquamous carcinoma	Brubaker and Hellstrom[9] (1977)
Sebaceous carcinoma	Motley et al[13] (1991)
Microcystic adnexal carcinoma	Bewer et al[14] (2004)
Angiosarcoma	Aguila and Sanchez[15] (2003); Gallardo et al[16] (2000)
Lymphomas	
Cutaneous B-cell lymphoma	Stanway et al[17] (2004)
Systemic lymphocytic lymphoma	Wilson[18] (1982)
Metastasis	
Metastatic primary squamous cell carcinoma of the lung	Nesi and Lynfield[19] (1996)
Paraneoplastic Syndrome	
Paraneoplastic rhinophyma secondary to retroperitoneal malignant hemangiopericytoma	Mayou et al[20] (1989)
Multisystem Disease	
Sarcoidosis	Leonard[21] (2003); Goldenberg et al[22] (1998)
Inherited Disorders	
Dermal cylindroma	Given et al[23] (1977)
Tuberous sclerosis	Bernstein[24] (1978)
Benign symmetric lipomatosis	Izu et al[25] (1994)

Differential Diagnosis	Case Report (publication year)
Infectious Disorders	
Mycobacterium kansasii	Klotch et al[26] (1992)
Cryptococcus neoformans	Mares et al[27] (1990)
Post-kala-azar dermal leishmaniasis	Ramesh et al[28] (1999)
Rhinoscleroma	Hadders et al[29] (1970)
Dermatoheliosis	
Actinic rhinophyma	Kligman[30] (1996)
Medication	
Phenytoin	Jaramillo et al[31] (2000)

Comment

To our knowledge, this is the first case of localized chronic fibrosing vasculitis presenting with a rhinophymalike appearance. In previous reports of this condition, the patients presented with solitary plaques or nodules on the neck, thighs, arms, back, vulva, and penis. EED and GF were considered in the differential diagnosis. EED characteristically presents as persistent red, violaceous, or yellow plaques, papules, or nodules most commonly seen on the extensor surfaces of bilateral joints and extremities, while GF presents as brownish-red plaques, papules, or nodules, most commonly on the face. A case of GF mimicking rosacea has been described, but unlike our case, the infiltrate was localized to the dermis and not subcutaneous tissue.[2] Clinically, our patient presented with a rhinophymalike appearance involving the entire nose, unlike GF, which usually presents as localized plaques or nodules. Also unlike GF, the histology did not show a grenz zone and hemosiderin deposition, which imparts the brownish-red color indicative of GF. Although a variant of GF cannot be entirely excluded, we felt the clinical and pathologic findings were best classified as localized chronic fibrosing vasculitis.

Rhinophyma stems from the Greek *rhinis* meaning nose and *phyma* meaning growth.[3] Because of the clinical appearance, we reviewed the literature to see what other entities clinically mimic rhinophyma. Rhinophymalike lesions have been described as occurring in a wide array of processes, including various cutaneous and metastatic tumors,

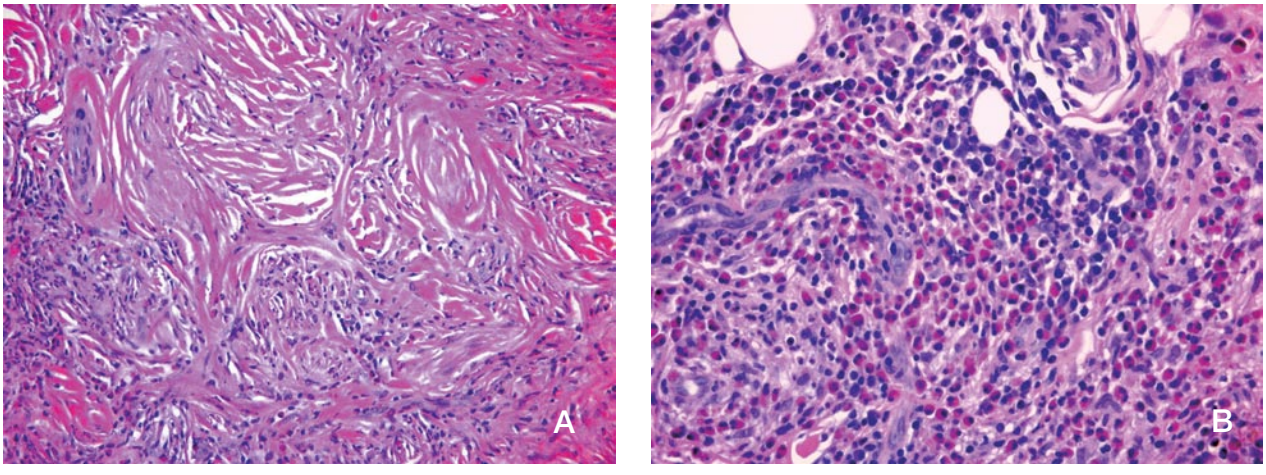

Figure 2. Skin biopsy demonstrated "onion-skinlike" fibrosis (H&E, original magnification ×20)(A) and inflammatory infiltrate containing numerous eosinophils (H&E, original magnification ×40)(B).

as a part of a paraneoplastic syndrome, cutaneous and systemic lymphoma, as a manifestation of vasculopathic reactive processes, granulomatous disease, infectious disease, and as a side effect of medications such as phenytoin (Table).[2-31] Genetic diseases with cutaneous tumors or disorders such as dermal cylindroma or tuberous sclerosis also may produce a rhinophymatous appearance.[23,24]

Conclusion

Although rhinophyma is a relatively common late manifestation of rosacea, rhinophymatous change can be caused by a variety of different processes. Patients with unusual presentations of rhinophyma should undergo diagnostic biopsy.

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