Granuloma Faciale Associated with Sinonasal Tract Eosinophilic Angiocentric Fibrosis

Patricia A.V. Chinelli, Milka Y. Kawashita, Mirian N. Sotto and Marcello M.S. Nico
Department of Dermatology, Hospital das Clinicas, Faculdade de Medicina, Universidade de Sao Paulo, l. Av dr Eneas de Carvalho Aguiar, 255, 05403-090 São Paulo, Brazil. E-mail: patchinelli@hotmail.com
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Sir,

Granuloma faciale (GF) is a chronic form of leukocytoclastic vasculitis of unknown origin and poorly understood pathogenesis characterized by red, violaceous or brownish macules, papules, plaques or nodules usually in the face. The course of the disease is chronic, there is no uniform response to any of the proposed therapies and spontaneous remission sometimes occurs (1, 2).

Eosinophilic angiocentric fibrosis (EAF) is an unusual fibrotic condition affecting the mucosa of the upper respiratory tract. It is characterized by a history of progressive nasal obstruction, affecting both nostrils and external swelling of the nose. Holmes & Panje (3) reported this entity for the first time in 1983 and called it 'intranasal granuloma faciale' and in 1985 Roberts & McCann (4) coined it as EAF. There are 15 case reports of EAF in the literature, 9 women and 6 men (4–12). The simultaneous occurrence of GF and EAF has been reported in only four cases (3, 4, 9, 12).

CASE REPORT

A 31-year-old Brazilian white woman had a 2-year history of cutaneous lesions. Clinical examination revealed severe brownish-red, infiltrated, indurated, well-circumscribed plaques with telangiectatic vessels on her back and face (Fig. 1), which had been slowly progressing in size. Bilateral nasal obstruction began in the previous year with progressive external swelling of the nose. The patient had no history of respiratory disease; she was taking no medication and was otherwise healthy.

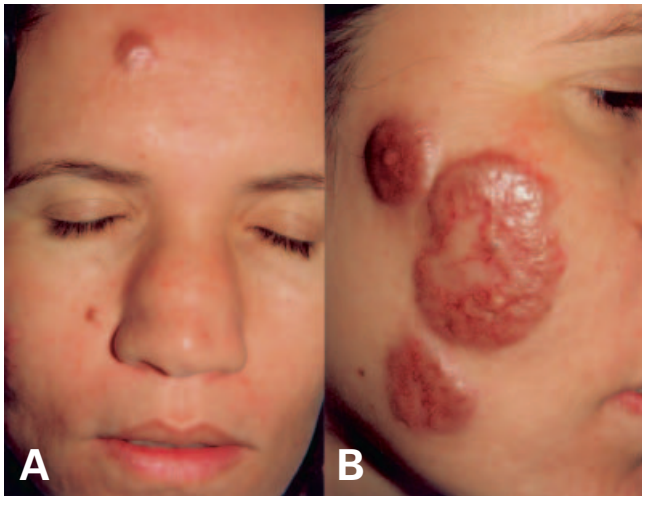

Fig. 1. Granuloma faciale lesion on the forehead and facial appearance of eosinophilic angiocentric fibrosis with external swelling of the nose (A) and chin (B). Permission given by the patient.

Biopsy specimens from cutaneous lesions showed a very dense polymorphous inflammatory infiltrate composed of polymorphonuclear leukocytes, lymphocytes, plasma cells and eosinophils distributed diffusely throughout the involved dermis with a narrow uninvolved grenz zone beneath the epidermis and around pilosebaceous follicles. There was also vessel wall damage and in places fibrinoid necrosis could be seen (Fig. 2A). In addition, some focal fibrosis was observed. Special stains for acid-fast bacilli and fungi were negative. Tissue cultures for mycobacteria, fungi and aerobic and anaerobic bacteria were also negative. The diagnosis of GF was made on the basis of the clinical picture, the histological findings and the negative microbial stains and culture results.

A video-assisted nasofibroscopic examination of the upper respiratory tract demonstrated partial obstruction of both nasal sinuses, with a thickened nasal septum. A CT scan showed thickening of soft tissues at the nasal fossa without osteolytic lesions. A nasal mucosal biopsy was performed and the histopathologic examination of the nasal mucosa showed a mild inflammatory infiltrate with lymphocytes and scant eosinophils in the subepithelial stroma and perivascular onion-skin fibrosis (Fig. 2B). Granulomas were absent. The

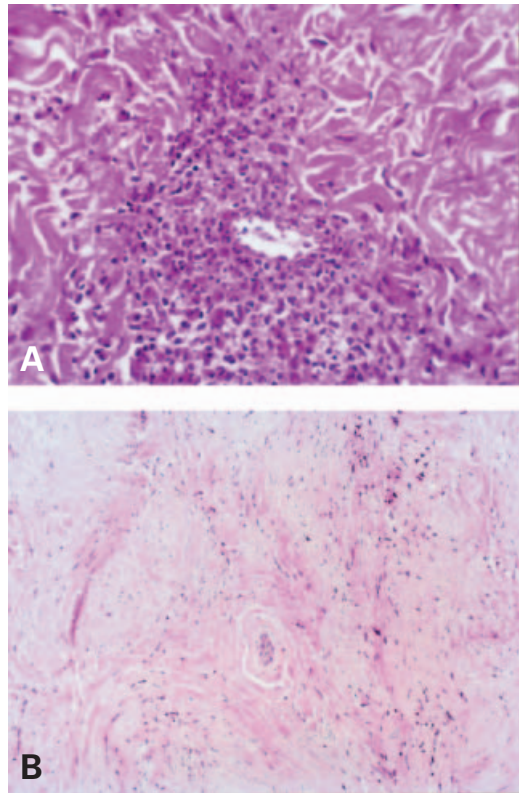

Fig. 2. Granuloma faciale: a dense polymorphous perivascular inflammatory infiltrate rich in eosinophils (A). Eosinophilic angiocentric fibrosis: inflammatory infiltrate with lymphocytes and scant eosinophils in the subepithelial stroma and perivascular hyalinizing onion-skin fibrosis (B). (H&E, ×200 original magnification.)

septal cartilage was spared. These histological findings correspond to a later stage in the evolution of the EAF (5–8).

All laboratory tests, including differential blood counts, coagulation tests, ANCA, ANA, ESR, and serum electrophoresis were unremarkable.

Oral dapsone therapy was initiated at a dosage of 100 mg/day. Monthly intralesional injections of 5 mg/ml triamcinolone were administered to the cutaneous lesions. After eight injections the lesions showed significant improvement, leaving only red-brown atrophic residual lesions with a few telangiectatic vessels.

The patient did not want to be submitted for nasal surgery for the EAF and nasal injection of steroids was not done. Improvement in nasal obstruction was remarkable during dapsone treatment, and nasal breathing became possible. She has been followed up for 1 year now with no relapse.

DISCUSSION

GF occurs almost exclusively in the face. To our knowledge, only 14 previous cases of extrafacial lesions have been reported (13). Treatment with intralesional steroids provided good cosmetic and functional outcome in our case.

EAF is an extremely rare fibrosing lesion that mainly affects the sinonasal tract of young to middle-aged patients, leading to obstruction of the upper airways. There might also be epistaxis and pain. Clinical differential diagnosis of EAF includes Churg-Strauss syndrome, Wegener's granulomatosis, sarcoidosis, infectious granulomatous conditions and juvenile nasopharyngeal angiofibroma. Most of the above conditions have characteristic clinical and laboratory featur7es (1, 8–10).

The aetiology of EAF remains obscure. The presence of eosinophils in the lesions, its predominance in women and a clinical history of allergy in some patients may suggest an allergic aetiology. Some patients have had a history of surgical procedures such as septal surgery before the diagnosis (4, 5) but no histopathological studies were performed prior to surgery. Besides, EAF generally does not appear to be a response to trauma (4, 6–10). The histopathologic studies of EAF show rich inflammatory infiltrate with eosinophilic vasculitis without necrosis in the early lesion and dense fibrosis, thickening of the subepithelial stroma with a characteristic obliterative perivascular onion skin whorling of collagen fibres and reticulin in the late lesion. There is thus considerable morphological similarity between GF and EAF.

Various treatments have been tried in EAF with only partial success, including surgical excision, which tends to lead to recurrence, and oral or intralesional steroid injections, with little or no response (5, 6, 8–10). Dapsone is known to improve neutrophil-mediated disorders such as GF and acute leukocytoclastic vasculitis (1, 14, 15). Dapsone interferes with neutrophil chemotactic migration, and suppresses local production of toxic respiratory and secretory products, including oxygen-derived radicals (14, 15). In addition, this drug reduces the release of prostaglandins and leukotrienes, blocking their inflammatory effects (15).

The obstructive symptoms of EAF in our patient significantly improved with dapsone. These findings may also suggest an association between EAF and GF, which both run a chronic, indolent course, with considerable histological similarity, but occurring in different sites. If patients with GF have sinus symptoms, the diagnosis of EAF is very likely; conversely, if patients with suspected destructive sinopulmonary disease (i.e. Wegener or lymphoma) have cutaneous lesions, a biopsy is essential as the answer may be EAF and GF.

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