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Pathology in Focus

Eosinophilic angiocentric fibrosis

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Abstract
Eosinophilic angiocentric fibrosis (EAF) is a rare inflammatory fibrosing condition of unknown aetiology that involves the nose or larynx producing mucosal thickening and severe obstructive symptoms. We report the first case affecting a male. He presented with nasal obstruction requiring septoplasty. The clinical and histopathological features of the condition are discussed and a comparison is made with the seven previous reported cases.

Key words: Fibrosis, eosinophilic angiocentric; Nasal obstruction; Nasal septum, surgery; Laryngostenosis

Introduction

Eosinophilic angiocentric fibrosis (EAF) of the upper respiratory tract is a rare benign condition of uncertain aetiology. It was first described by Roberts and McCann in 1985 and seven cases have been reported to date in the literature.[1–4] The lesions begin with an infiltration of inflammatory cells, mainly eosinophils, lymphocytes, plasma cells and neutrophils which surround small blood vessels. Gradually there is progress to a perivascular fibrosis which shows a characteristic angiocentric whorling with an 'onion skin' pattern.[1] The unresolving fibrosis and consequent stenosis requires surgical intervention.

Case report

A 51-year-old man presented with a history of progressive nasal obstruction, affecting both nostrils, and increased snoring in the previous few months. Steroid nasal sprays and drops prescribed by the patient's general practitioner resulted in limited improvement. He had no history of asthma but had suffered from hayfever as a young man. There was a history of one episode of trauma to the nose as a child although he had not had any previous septal surgery. He was an ex-smoker, having given up smoking 10 years earlier. Examination revealed hyponasal speech and an unusually thickened septum. There was no evidence of rhinitis or polyposis and the erythrocyte sedimentation rate (ESR) was found to be 7 mm/hr. There was no peripheral eosinophilia and an anti neutrophil cytoplasmic antibody (ANCA) test was negative. A septoplasty was performed two months following the initial presentation, at which time the widened septum was reduced and the unusual appearance again noted. Biopsies from the septum were sent for histopathology. On follow-up, at one year following surgery, the patient continued to report his nose is clearer than it was pre-operatively.

Histopathological features

Histopathological examination of the biopsies of the nasal septum showed all the features of EAF. In EAF, there is progression from inflammation to fibrosis although both usually co-exist. The early lesion consists of eosinophilic vasculitis with leucocytoclasis involving small blood vessels in the submucosa (Figure 1). There is fibrosis and proliferation of spindle-shaped fibroblasts producing a pseudo-granulomatous appearance but there are no true granulomas and epthelioid or multinucleated giant cells are not seen. The overlying mucosa may show a few patches of ulceration. The late lesions show dense fibrosis, thickening of the subepithelial stroma with a characteristic obliterative perivascular onion skin whorling of collagen fibres and reticulin (Figure 2).[1] Immunohistochemical findings confirm the inflammatory rather than neoplastic nature of EAF.[4]

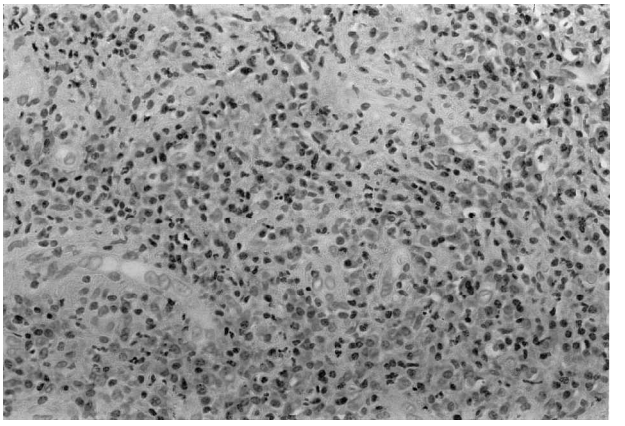

Fig. 1.
Inflammatory cells surrounding small blood vessels with coexisting fibrosis (H&E; ×250).

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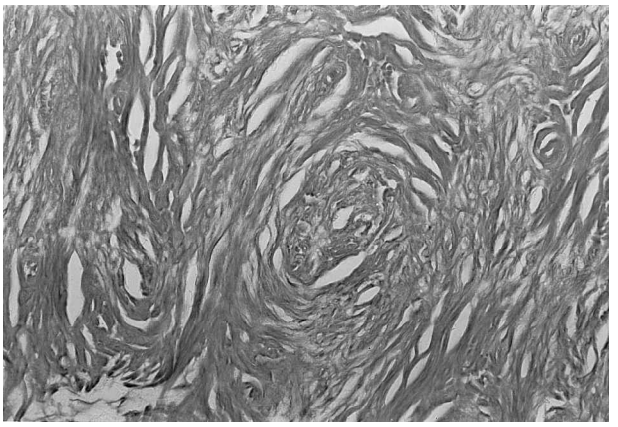

FIG. 2.
Reticulin stain showing perivascular 'onion skin' whorling of collagen and reticulin fibres (H&E; ×250).

Discussion

EAF is an unusual and rare condition. There have been seven cases reported in the literature, all female.[1–4] Our case is the eighth and the first affecting a male. The diagnosis can only be made by histological examination that shows a stereotyped pathognomonic picture as described previously.

Of the eight cases of EAF, seven have been female and ours is the first male. Their ages have ranged from 25 to 59 years. All six nasal cases presented with nasal obstruction the symptoms being present for several years. Topical and intralesion steroids used in at least five cases were found to be ineffective. The lesions were present in the septum and/or lateral cartilage of the nose and required repeated surgical excision, recurring in four of the six cases.

This is also the first case where the diagnosis has been made at the time of the first septal surgery. In the other five cases the diagnosis was made following repeat surgery. The two laryngeal cases presented with subglottic stenosis causing causing shortness of breath and requiring tracheostomy and eventual laryngotracheoplasty. In all the reported cases, the lesions have occurred either in the nose or the larynx, never both in the same patient. Base line investigations of full blood count (FBC) reported in six and ESR in four of the eight cases were normal. Chest X-rays (CXR) in the subglottic lesion patients were also normal.

A clinical differential diagnosis of EAF would include Churg-Strauss syndrome, Wegener's granulomatosis, sarcoidosis, infectious granulomatous conditions and Sjögren's syndrome. Most of the above has a characteristic clinical history with histological diagnosis confirming the glanulomatous lesions of Wegener's, sarcoidosis and infectious granulomatous conditions. Sjögren's shows glandular involvement absent in EAF. Hence, investigation of EAF would include FBC, ESR, ANCA, CXR, endoscopy and biopsy to exclude the above disorders. A computed tomography (CT) scan would help determine the extent of the lesion especially if involving the subglottic region. Any tissue excised from an unusually thickened septum must be sent for histology.

The aetiology of EAF remains obscure. The presence of eosinophils in the lesions and a clinical history of allergy in at least five cases suggests an allergic aetiology although this may be coincidental. Trauma may play a role in the condition. This may be accidental as in our case where the patient had a history of nasal injury as a child or it may be iatrogenic as in all the reported case of septal lesions where the patients had had previous septal surgery.

However, this association may also be coincidental. Roberts and McCann proposed that EAF could be a mucosal variant of granuloma faciale because of the prominent eosinophilic infiltrate and vasculitis seen histologically. However, in granuloma faciale the fibrosis is not extensive and the peculiar angiocentric whorling is not present. Also, coexisting granuloma faciale and EAF has been reported only by Roberts and McCann.[4]

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