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CASE REPORT

A Case Report of the Natural Course of Eosinophilic Angiocentric Fibrosis of the Nasal Septum

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Abstract

We present the case of a 47-year-old female patient with eosinophilic angiocentric fibrosis (EAF): a rare, benign, fibroinflammatory condition of uncertain etiology in the sinonasal cavity. Her autoimmune profile was normal, and she had a history of adult-onset atopic asthma and atopic dermatitis. Histological and magnetic resonance imaging findings were typical for EAF. The patient was not treated with surgery, and there was no significant worsening of the condition over a 5-year follow up. There have been no recorded cases of malignant changes or mortality related to EAF. Additionally, surgical treatment typically results in recurrence, and EAF has been observed to stabilize over time. Therefore, we conclude that based on the natural course of the disease, surgical treatment should be re-evaluated as the first-choice treatment of this rare condition.

Keywords: *Angiocentric Fibrosis, Case Report, Nasal Septum*

Introduction

Eosinophilic angiocentric fibrosis (EAF) is a rare, benign, fibroinflammatory condition of uncertain etiology, which usually affects the sinonasal cavity. It most typically presents as a fibroinflammatory lesion located in the nasal septum and sinus mucosa, and it causes stenosis of the upper respiratory tract [1]. To date, less than 60 cases have been described in the literature. Presenting symptoms are usually nonspecific and related mostly to the local disturbances caused by the growing mass. Nasal obstruction, swelling, epistaxis, and epiphora are among the most common complaints [2].

A history of atopy or allergy has been observed in about 25% of EAF cases, with a similar proportion affected by granuloma faciale (GF), a skin condition with similar histologic findings, which often presents after EAF has been diagnosed [3]. Diagnosis is based chiefly on histopathological examination to determine the presence of a characteristic dense perivascular inflammatory infiltrate

dominated by eosinophilic granulocytes. The fibroinflammatory lesion consists of a whorled fibrosis, which typically surrounds blood vessels [1]. EAF is a gradually progressive condition, but some evidence suggests that the lesion may stabilize over time. No EAF-related mortalities or malignant alteration cases have been recorded [3]. Herein, we describe a case of EAF of the nasal cavity in a 47-year-old female patient who refused to undergo surgical removal of the lesion, and therefore has provided a rare example of the natural course of the disease. We present the clinical, radiological, and histopathological findings of this case.

Case Presentation

A 47-year-old woman visited our ENT clinic with a 7-year history of progressive nasal obstruction and swelling, epistaxis, and epiphora. Her medical history was significant for adult-onset bronchial asthma and atopic dermatitis. Physical examination showed hard and non-tender midline dorsal nasal swelling. Nasendoscopy revealed mucosal thickening involving the nasal septum and both lateral walls. Her autoimmune profile, including cytoplasmic and perinuclear anti- neutrophil cytoplasmic antibodies, was normal. Serum immunoglobulin A and erythrocyte sedimentation rate were elevated.

A non-contrast computed tomography scan of the paranasal sinus demonstrated abnormal thickening and enhancement in the midline involving the nasal dorsum and lacrimal ducts bilaterally with evidence of right dacryocystitis (Figure 1). Surgical biopsy of septal cartilage and the left and right dorsum was performed. Histopathological assessment showed fibrosis with a whorling angiocentric pattern and mixed, predominantly eosinophilic, inflammation (Figures 2–4). Neither necrotizing vasculitis nor granuloma formation was observed. The diagnosis was consistent with EAF. The patient refused surgery. In the subsequent 7-year follow-up, there was no significant change in the patient's symptoms.

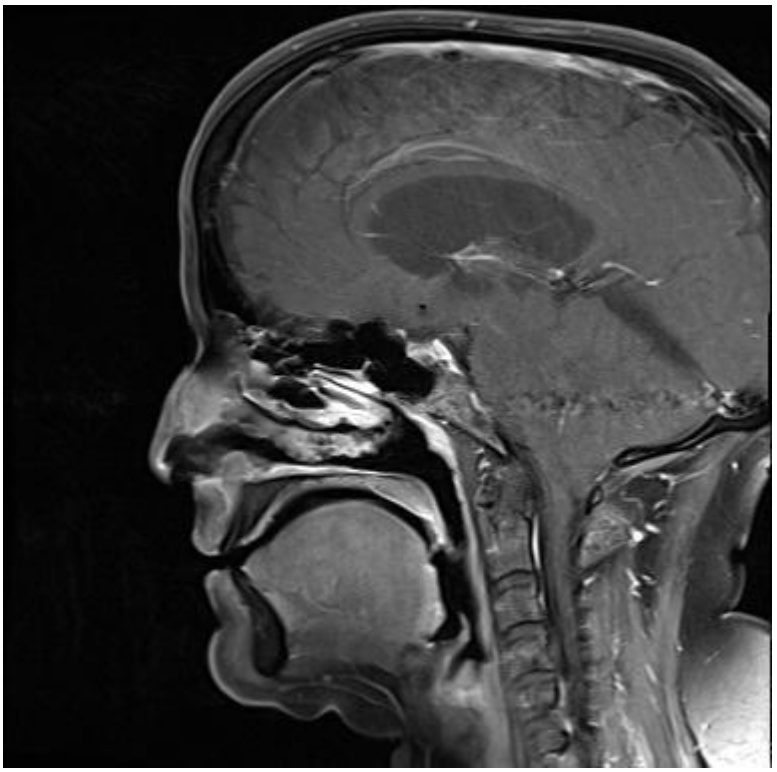

Figure 1. Multi-planar MRI images show an enhancing mass involving the nasal dorsum. There is also an extension into the pre-maxillary region on both sides of the anterior maxillary spine.

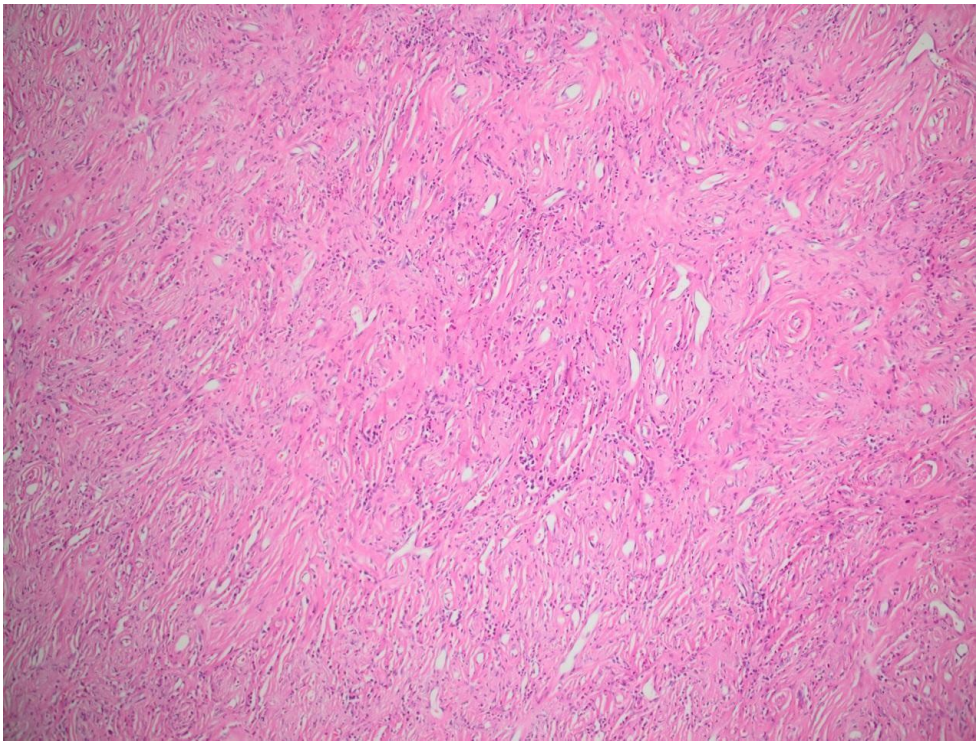

Figure 2. Low power view of a hematoxylin and eosin (H&E) stained section shows mainly spindle cell lesions that are hypocellular.

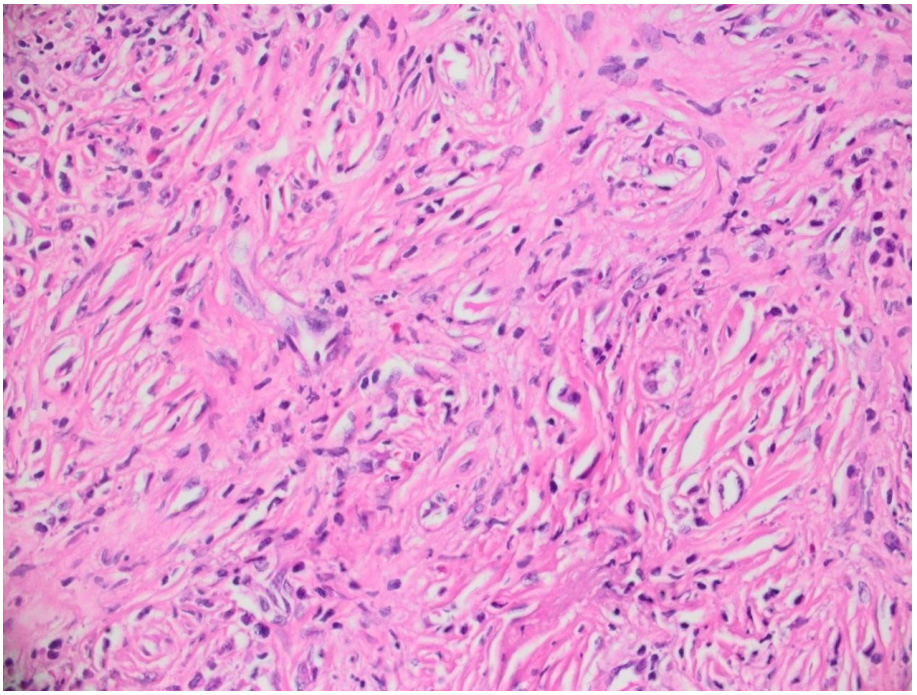

Figure 3. High power view of a hematoxylin and eosin (H&E) stained section shows spindle cells in fascicles with abundant collagen arranged concentrically around thin blood vessels and scattered inflammatory cells. No evidence of vasculitis or granuloma is seen. Spindle cell nuclei are bland.

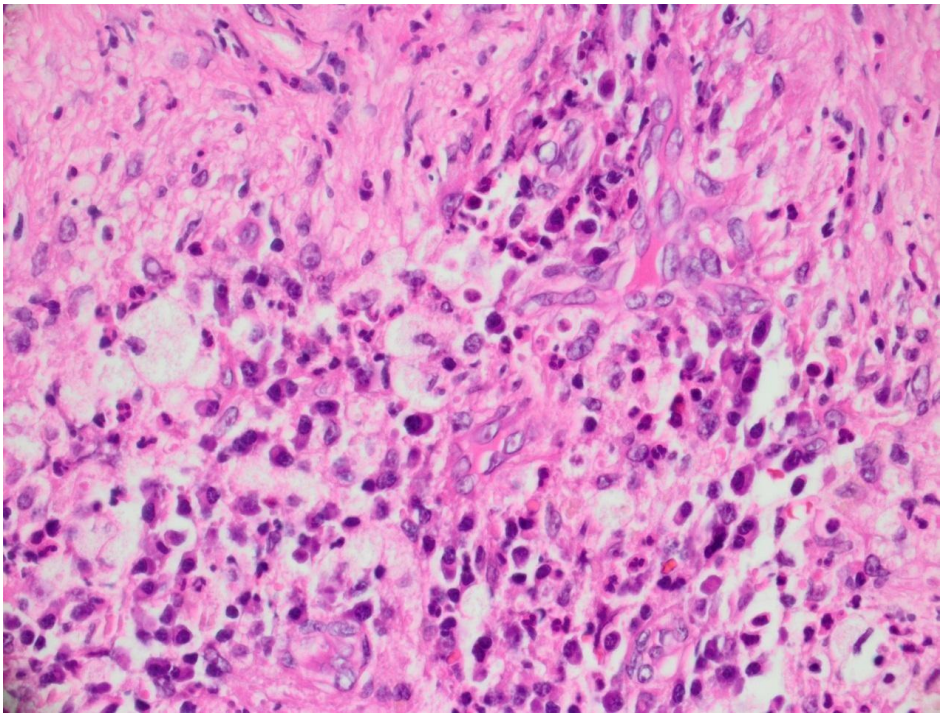

Figure 4. Moderate mixed inflammation that consists of plasma cells, eosinophils, neutrophils, and lymphohistiocytic cells. Staining for specific organisms gave negative results (Gomori methenamine silver stain, Ziehl–Neelsen stain, and periodic acid–Schiff stain).

Discussion

Because EAF is a rare condition, case studies are the most common type of reports in the literature on this topic. However, we were unable to find any report of a case in which surgical treatment was strictly indicated but not performed owing to a lack of patient consent. Our patient's refusal to undergo surgery provides us with a rare glimpse into the natural course of the disease. Most clinicians still regard a wide surgical resection of the lesion as the treatment of choice. However, disease recurrence has been reported in most of the cases for which satisfactory follow-up information is available [2].

The treatment of EAF has proven challenging, with persistent disease in many cases in spite of surgical resections. Corticosteroid therapy has also been proven ineffective [3]. From the initial diagnosis in 2010 to a final follow-up 7 years later, we found no significant worsening of the patient's condition. Because no cases of malignant alteration or EAF-associated mortality have been recorded, and because EAF may have a tendency to stabilize over time [3], our case presents an argument against routine surgical treatment of this condition. The nature of a case report is such that this particular natural disease course may not apply to all patients. However, owing to the rarity of this disease, information on clinical outcomes is scarce, and determining a first choice of treatment may not be as straightforward as previously believed.

There have been no recorded cases of malignant changes or mortality related to EAF. Additionally, surgical treatment typically results in recurrence, and EAF has been observed to stabilize

over time. Therefore, we conclude that based on the natural course of the disease, surgical treatment should be re-evaluated as the first-choice treatment of this rare condition.

Conflict of interest

The authors declare that there is no conflict of interest regarding the publication of this paper.

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