Rare Pathology of Sinonasal Tract; Eosinophilic Angiocentric Fibrosis

Case Report

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Abstract

Eosinophilic angiocentric fibrosis (EAF) is a relatively rare, benign and slowly progressive fibroinflammatory disorder, which mainly involves the upper respiratory tract. EAF was first described by Holmes and Panje in 1983. EAF is typically seen in young to middle-aged females and it has a slowly progressive nature. As well as the sinonasal region the nasal septum is mainly the most common localization of EAF, involvement of the larynx and orbita was also described in literature. Symptoms vary with the involved area but nasal obstruction, epistaxis and epiphora are the most common complaints. The etiology of EAF is not clear and it is diagnosed by specific histopathological features. Histologically, the lesion is characterized by a perivascular, eosinophil-rich inflammatory infiltrate and progressive fibrosis which appears as a whirling "onion-skin" pattern. In this paper; a case of sinonasal EAF is reported with the clinical course, radiological and histological features, etiology and treatment of these lesions based on updated literature.

Key Words: Eosinophilic angiocentric, fibrosis, sinonasal

Introduction

Eosinophilic angiocentric fibrosis (EAF) is a rare, benign and slowly progressive fibroinflammatory disorder (1, 2). EAF was first described by Holmes and Panje in 1983 as "intranasal granulome faciale" and in 1985 Roberts and McCann (2, 3) reported 3 similar intranasal cases with detailed histopathological behavior. Although EAF has a clear clinical and histological appearance, its pathogenesis is still controversial (2, 4). EAF typically presents in young to middle-aged females with a submucosal inflammatory, fibrosing tumor-like lesion causing slowly progressive upper airway symptoms. Histologically, the lesion is characterized by a perivascular, eosinophil-rich inflammatory infiltrate and progressive fibrosis which appears as a whirling "onion-skin" pattern (2-4). We report a case of sinonasal EAF and review the clinical course, radiological and histological features, etiology and treatment modalities based on updated literature.

Case Presentation

A fifty-five-year old female patient was admitted to our clinic with a history of swelling and pain in the right premaxillary and alar region and also some attacks of epistaxis from the right nasal passage. On her physical examination; a submucosal pinkish purple mass was detected which was located inferolaterally of the inferior turbinate. The mass was cystic-like on palpation. On her magnetic resonance imaging (MRI), a soft tissue mass was detected originating from the lateral portion of the inferior turbinate and lying laterally to the premaxillary region (Figure 1). Transnasal endoscopic excision of the mass was performed. On pathological examination, dense angiocentric "onion-skin" like fibrosis foci and eosinophil rich inflammation were detected, so it was diagnosed as "eosinophilic angiocentric fibrosis" (Figure 2a, b). She was not given any medical therapy after surgery. During postoperative follow-ups, an MRI was performed on the 3rd month, when there was no sign of recurrence of the intranasal mass and also the residual lesion on premaxilla, which could not be resected, was observed as stable (Figure 3a-c). The patient was informed about the rarity of the diagnosis and the conflicts of the treatment options and she gave an informed consent for this case report.

Discussion

Eosinophilic angiocentric fibrosis is an uncommon inflammatory fibrosing lesion of the upper airway tract (2-5). The sinonasal tract and mainly the nasal septum is the most common location for the EAF, laryngeal and orbital involvements are also described in the literature (1, 2, 4). It has been claimed that the slow progression and nonspecific symptoms account for its delayed presentation, and diagnosis is usually postponed for an average of approximately 5 years (2, 5). Symptoms may vary according to the involved site but are frequently seen as nasal obstruction, pain, epistaxis and epiphora (2, 5, 6). On physical examination, the EAF lesion is usually observed as a pink-purple mucosal surface with submucosal thickening without any specific appearance.

Pathogenesis of EAF is not well understood. Allergic etiology is proposed to be responsible due to predominance of eosinophil rich inflam-

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mation (2, 3, 7, 8). However, only some cases have a history of allergy in the literature. Also, EAF has been found to be refractory to steroids and the other anti-allergic therapies (7, 8). Trauma was also thought to be a predisposing factor but this was not supported by cases in the literature (2). There has been some evidence that trauma may cause acceleration of disease progression. After surgical resection, EAF showed rapid progression in some cases (2, 8-10). Our case did not have any symptoms of allergy and and she did not have prior nasal trauma or surgery either.

Imaging modalities, both magnetic resonance imaging (MRI) and computerized tomography (CT) are nonspecific and typically reveal a well circumscribed submucosal soft tissue density mass (2). On MRI, EAF is generally isointense on T1-weighted images with moderate heterogenous enhancement after contrast administration. This heterogeneous contrast involvement is seen because of different stages of inflammation and fibrosis in the same lesion. Most lesions show hypointense signal intensity on T2-weighted images owing to fibrosis (1). On non-enhanced CT, EAF is usually seen as a homogenous, isodense soft tissue mass, rarely with calcifications (1). As lesions progress, they may cause adjacent bone remodelling such as thinning, slight absorption and sclerosis, but not an infiltrative pattern due to its benign nature (1, 2). On our patient's CT images, a homogenous soft tissue mass was observed which did not cause any bony destruction.

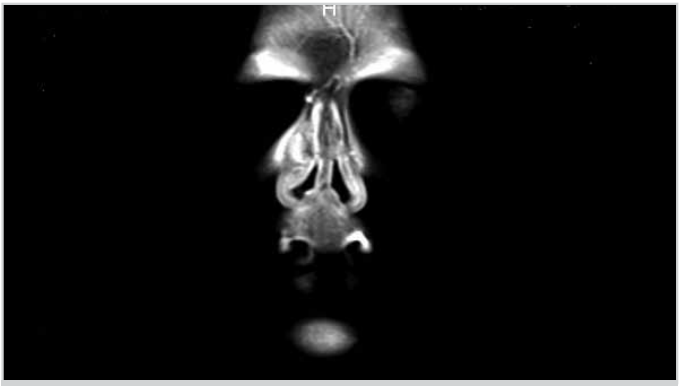

Figure 1. Preoperative MRI showing the lesion
MRI: magnetic resonance imaging

The histology of EAF is pathognomonic and characterized by perivascular inflammatory cell infiltration with progressive fibrosis around small vessels leading to the typical "onion-skin" pattern (3, 4). Eosinophils are the predominant cells with some degrees of plasma cells and lymphocytes of early inflammatory lesions. In the course of EAF, fibrosis becomes more visible around the microvasculature (2). The main differential diagnosis includes lesions with prominent eosinophilic infiltrates (2-4). Granuloma faciale, an inflammatory vascular reaction, presents clinically as papules localized almost on the face. Histologically, there is a polymorphous inflammatory infiltrate and it consists of neutrophils and eosinophils in the dermis (3, 4). Both histological similarities and coexistence in some cases suggest that EAF is a submucosal variant of granuloma faciale affecting mainly the upper respiratory tract, but there is no consensus about this (2, 8, 10). Absence of geographic necrosis, necrotizing vasculitis and granulomatous inflammation excludes Wegener's granulomatosis (WG) and Churg-Strauss syndrome (CS). Also, EAF lacks systemic vasculitis which is a participant of CS and mucosa over the EAF lesion does not show necrosis which is usually seen in WG lesions (4, 10, 11). Blood test positivity for c-ANCA and p-ANCA supports the diagnosis of WG and CS, respectively (4, 11). Both markers were found to be negative in our case.

Treatment of EAF has been a challenge as approximately 70% of patients have persistent or recurrent disease during follow up (2, 5). Medical therapy is found to be ineffective but may cause symptomatic relief (2, 8). Intranasal, intralesional or systemic corticosteroids, antihistamines, dapsone, hydroxychloroquine, azathiopurine and tamoxiphen are trial therapies in the literature but none of them were found to be as effective as surgery (2, 4, 5, 8). Surgical excision of the lesion should be complete resection as far as possible (2, 7). Nevertheless, only 30% of the cases do not have recurrence after surgery, most of the patients need multiple resections (2-4). Some authors suggest medical therapy after surgery but there is not enough evidence that this could prevent any recurrence (2, 4).

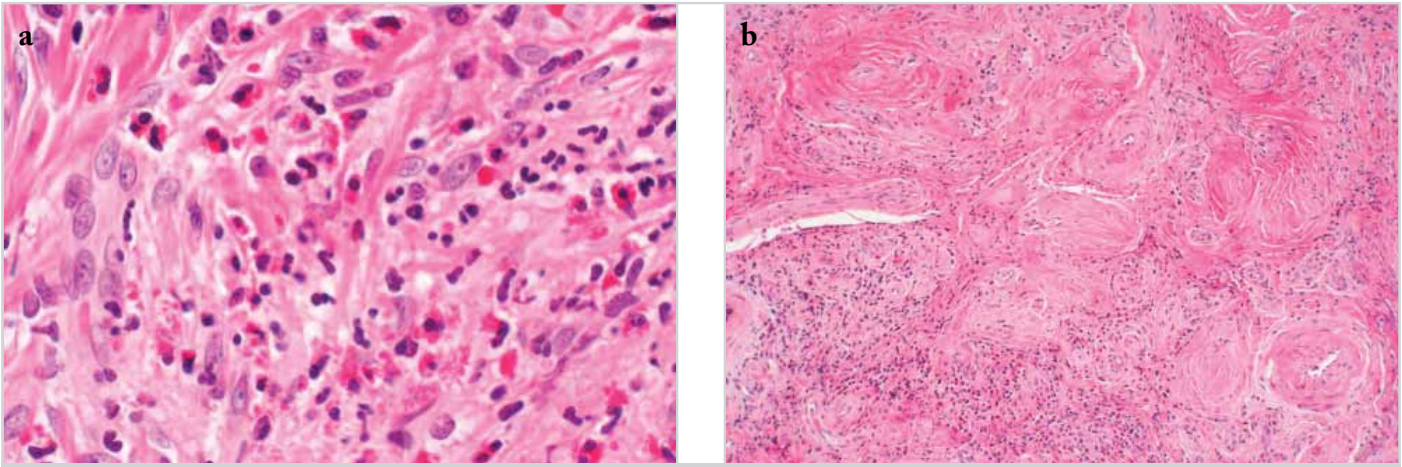

Figure 2. a, b. Histopathologic staining showing the 'onion-skin' appearence of the lesion

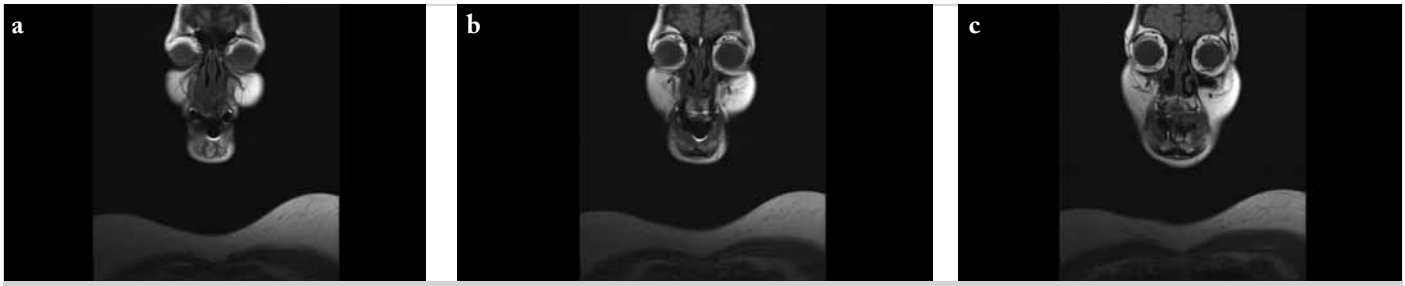

Figure 3. a-c. Postoperative MRI showing that there was not any recurrence of the disease
MRI: magnetic resonance imagining

Conclusion

EAF is a benign, progressive fibrosing disease with an elusive etiology. It has a nonspecific clinical presentation but a typical histological appearance that leads to diagnosis. Treatment remains controversial but as more cases are reported it is likely that a clear understanding of etiology and management will result in better outcomes.

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