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CASE REPORT

Nasal Eosinophilic Angiocentric Fibrosis with Orbital Extension

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Abstract Eosinophilic angiocentric fibrosis (EAF) is an extremely rare, chronic, benign, idiopathic disorder that mostly affects the upper respiratory tract, particularly the nasal cavity, and features progressive submucosal perivascular fibrosis. To the best of our knowledge, only seven cases of EAF with orbital involvement have been reported. We report a case of sinonasal EAF with orbital extension that presented with left nasolacrimal duct obstruction. A 35-year-old man presented with left epiphora, proptosis, anterolateral globe displacement and nasal obstruction. Endoscopic sinus examination showed a firm, gritty, creamy, yellow, fibrous, adherent mass of maxillary sinus. Diagnosis was established with histopathological examination of excisional biopsy of the lesion. Although EAF is very rare, it should be considered in the differential diagnosis of lesions of upper airway tract, particularly the nasal cavity. Biopsy is necessary for diagnosis and treatment planning. Resecting of the involved tissues completely is essential for prevention of recurrence.

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Introduction

Eosinophilic angiocentric fibrosis (EAF) is an extremely rare disease that typically presents in young to middle-aged individuals as a slowly progressive upper airway obstruction. It is a submucosal inflammatory, fibrosing tumor-like lesion [1, 2]. This condition is most commonly present in the nasal septum, sinus mucosa and the upper respiratory tract. The symptoms are related to the local progression of the disease [3, 4]. To our knowledge, this is the first recognized EAF case report from Iran.

We report a case of EAF where the lesion was primarily localized to the lateral nasal cavity with involvement of orbit and is unique because the patient presented primarily with epiphora. The main purpose of the current case report is to review the literature and discuss the clinical, radiological, and histological features and diagnosis as well as the management options of this rare entity.

Case Report

A 35-year-old man presented with history of slowly progressive orbital swelling over medial cantus of the left eye for 1 year and epiphora of 5 years duration. There was a relatively long history of on and off left side nasal obstruction. He denied diplopia, pain, visual disturbance, epistaxis, or atopic symptoms. He gave a history of left external dacryocystorhinostomy because of epiphora 5 years ago in a local hospital without significant improvement.

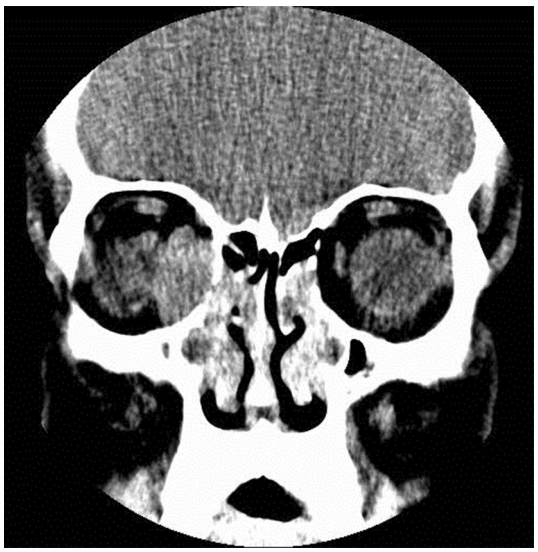

Fig. 1 Soft tissue mass density in the roof of left maxillary sinus with destruction of lamina papyracea. Computed tomography without enhancement

On examination a soft, non-tender mass of the left medial canthal region, as well as mild proptosis and lateral globe displacement was noted. A moderate septal deviation, normal size turbinate was seen on anterior rhinoscopy. Endoscopic nasal examination revealed a firm, gritty, creamy, yellow, fibrous, adherent mass of maxillary sinus. Ophthalmologic examination revealed a nonaxial proptosis and anterolateral globe displacement. No limitation of left eye movement was noted. Visual acuity was normal. General physical examination was unremarkable.

Computed tomography without enhancement showed a soft tissue mass density in the roof of left maxillary sinus with destruction of lamina papyracea. Left globe was displaced anterolaterally (Fig. 1).

The patient underwent an endoscopic sinus surgery under general anesthesia. Septoplasty, left uncinectomy, middle meatal antrostomy, and anterior ethmoidectomy were performed. The tumor was severely adherent to lamina papyracea and orbital periosteum. It was removed completely with a part of lamina papyracea. Left endoscopic dacryocystorhinostomy (EDCR) was carried out.

Histological examination of the biopsies of the nasal cavity and sinus showed a dense fibrous connective tissue with chronic inflammatory cells surrounding small blood vessels. An obliterative and concentric perivascular fibrosis with eosinophilia was also present (Fig. 2).

Sections showed a dense fibrotic stroma with concentric onion-skin perivascular fibrosis and eosinophilic vasculitis. Adjacent parenchyma consisted of a mixed inflammatory infiltrate (Fig. 3).

Epiphora, ocular abnormalities, and nasal obstruction improved post-operatively. The patient remained well thereafter, with no evidence of recurrence of the mass during 1 year follow-up visits.

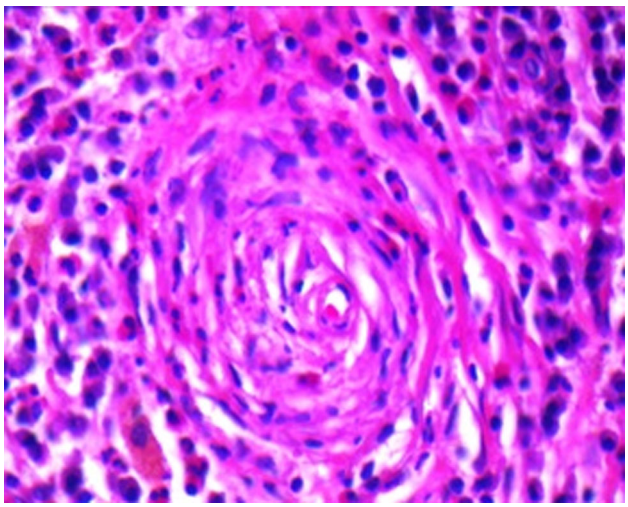

Fig. 2 Dense fibrous connective tissue with chronic inflammatory cells and eosinophils surrounding small blood vessels

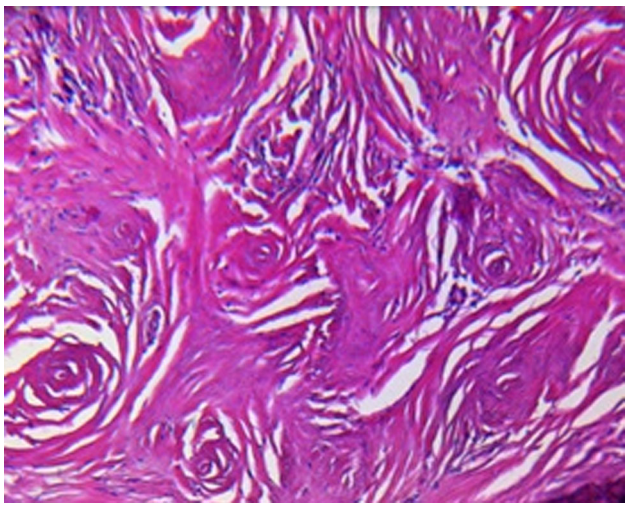

Fig. 3 Dense fibrotic stroma with concentric onion-skin perivascular fibrosis and eosinophilic vasculitis

Discussion

In 1983, the first of case of EAF was reported by Homes and Panje who called that condition ‘intranasal granuloma faciale’. It is believed to represent a mucosal variant of granuloma faciale [5]. Two years later, Roberts and McCann gave the term ‘EAF of the upper respiratory tract’ to this condition distinguished by inflammatory cell infiltration, predominantly eosinophils, and progressive perivascular fibrosis [6]. To the best of our knowledge, 54 EAF

cases [7–9] and only seven cases with orbital involvement including our case have been reported so far [2, 8, 10–13].

Symptoms of EAF are nonspecific, with the most prevalent symptoms related to the sinonasal region including progressive nasal obstruction, epistaxis, sinusitis, nasal tenderness, facial pain, swelling, and shortness of breath [1, 3, 7, 14]. Progressive nasal obstruction is the most common symptom [7]. EAF mainly affects the upper airways with approximately 75 % limited to sinonasal tract (90 %) including nasal septum, lateral nasal wall, and paranasal sinuses [1, 3, 7]. The most common involvement site was nasal septum (62 %). There have been cases with subglottic, pharyngeal, soft palate, and lower respiratory tract involvement [1, 7, 9, 14].

We report a rare and remarkable case of nasal EAF with orbital extension. The patient presented primarily with orbital manifestations. Very few EAF cases with orbital involvement and nasolacrimal duct obstruction have been described in the literature. These cases present with periorbital edema and painless proptosis, globe displacement, and limitation in ocular movements, epiphora, diplopia, and eyelid swelling [2, 8, 10–13].

Review of articles show that the slow progressive and nonspecific symptoms explain delayed diagnosis of this entity [1, 7, 13, 14]. There is some evidence that the lesion stabilizes over time [1].

Some authors initially reported that women are commonly affected more than men [14], but the current sex ratio of reported cases is 22 men to 32 women [7–9]. The age range is from 19 to 79 years [1, 3].

The etiology of EAF is unclear, but a number of potential etiologies were described such as previous nasal trauma or surgery, atopic disorders including asthma, allergic rhinitis, urticaria, allergy to environmental factors, nonspecific rhinitis, chronic bowel inflammatory disease, and rheumatic fever were observed among the reported cases. Some authors believe that it may be because of abnormal inflammatory response to a nonspecific stimulus but lack of response to immunosuppressive drugs does not support this opinion [4, 6, 11, 13, 14, 16].

Histological examination shows early and late lesions. The early lesion is a vasculitis with inflammatory infiltrate comprising of lymphocytes, plasma cells, scattered neutrophils, and with numerous eosinophils. The late lesions are composed of dense fibrosis with angiocentric pattern and mixed inflammatory cells. The fibrous component has a concentric layered onion-skin–type perivascular arrangement, which is characteristic of EAF and are useful for making the histologic diagnosis of EAF [1, 2, 6, 17]. EAF is now considered a part of IgG4-related disease spectrum [18, 19].

The differential diagnosis of EAF include Churg–Strauss syndrome, Wegener's granulomatosis (WG), microscopic polyangitis, cocaine associated midline facial destruction, sarcoidosis, infectious granulomatous conditions, Sjogren's syndrome, angiolymphoid hyperplasia with eosinophilia (AHLE), erythema elevatum diutinum (EED), Kimura disease, Erdheim–Chester disease, inflammatory pseudotumor, trigeminal trophic ulcer, and relapsing polychondritis, and two neoplastic conditions are lymphomatoid granulomatosis and NK/T cell lymphoma [7, 14, 17].

Due to this differential diagnosis, any of the serologic and lab data related to differential diagnosis were not significant.

CT and MRI imaging is nonspecific and generally shows a well-circumscribed submucosal soft tissue density mass with sinus opacification. Cartilaginous and bone destruction have been reported in some cases [1].

No definitive treatment has been identified but medical and surgical treatments have been given. Systemic and local corticosteroids, antibiotic therapy, and Dapsone have not been useful. Surgical resection appears to be the treatment of choice, though some authors reported that recurrences are common and multiple excisions are frequently required [2, 15, 16, 20–25]. Incomplete surgical resection at the first time or even biopsy might play a negative role to stimulate and accelerate the tumor growth [7]. Our patient has been symptom free 1 year after surgical intervention.

The current case report indicates a locally aggressive and unusual presentation of an extremely rare disease. Literature review shows only seven cases with orbital involvement, including our case. Otolaryngologists and ophthalmologists need to be aware of this entity in patients suffering from nasal obstruction or epiphora, etc. At present, there is not enough evidence to explain the etiology of EAF. A multicenter research study is essential for recognition of etiology and better treatment of this disease.

Conclusion

Although EAF is very rare in the nasal cavity, orbit, and upper airway tract, it should be considered in the differential diagnosis of lesions of these regions.

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