Intranasal Eosinophilic Angiocentric Fibrosis: A Rare and Suspicious Appearing Lesion

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Objective: The aim of this report is to describe a case of eosinophilic angiocentric fibrosis (EAF) and raise its awareness in the head and neck pathology by discussing its clinical course, histological features, diagnosis, and management options.

Methods: The medical records of a patient with EAF were reviewed at a tertiary medical center. The PubMed database was searched for keywords “eosinophilic angiocentric fibrosis” and “intranasal.”

Results: Here we report a case of intranasal EAF in a 49-year-old male who presented with a several month history of right-sided nasal skin swelling and a 10-year history of chronic congestion. On physical examination, patient had a non-pigmented, rubbery right-sided nasal skin lesion. Nasopharyngoscopy revealed a submucosal intranasal lesion of the right middle vault. Punch biopsy and fine needle aspiration revealed no malignancy. Final pathological diagnosis after surgical excision revealed extensive perivascular “onion skin” fibrosis, consistent with EAF. A comprehensive review of the literature revealed less than 60 reported cases of EAF.

Conclusion: EAF is a benign fibrovascular lesion of unknown etiology that affects mucosal surfaces of the sinonasal and upper respiratory tracts. Patients commonly present during the fifth and sixth decades of life with chronic symptoms of nasal obstruction, sinusitis, foetor, and breathing difficulties. The non-specific presentation and rarity of disease should raise suspicion of more common differentials, including Wegener’s granulomatosis, Churg-Strauss syndrome, and granuloma faciale. Although the clinical course is slow growing, management remains challenging due difficulties in establishing diagnosis and high risk of recurrence with a propensity for progressive local destruction.

**INTRODUCTION**

First described in 1983 by Holmes and Panje as intranasal granuloma faciale (GF), it was later re-named EAF by Roberts and McCann in 1985 after reporting similar cases with idiopathic swelling of the upper features.1,2 EAF is a rare tumefactive lesion affecting mucosal surfaces of the upper respiratory and sinonasal tracts. It represents a benign, non-fatal fibrovascular vasculitis of unknown etiology that is characterized by two distinct histological stages with pathognomonic perivascular “onion skin” fibrosis.3,4 Men and women are equally affected and present during the fifth and sixth decades of life with symptoms of nasal obstruction or swelling, epistaxis, breathing difficulties, and sinusitis.5,6 The clinical course is slowly progressive and locally destructive with a tendency to recur.5,6 Originally thought to be a mucosal variant of GF, the etiology of EAF remains unknown with many suggesting an allergic or autoimmune etiology.7 The most common presenting symptom is progressive nasal obstruction in 78.8%, however, presentation is often nonspecific with an average of 60 to 70 months from time of symptom onset to diagnosis.8,9,10 Other symptoms may include epistaxis, breathing difficulties, sinusitis, nasal tenderness, and facial pain.9,10 Approximately 25.0% have associated nasal deformity or swelling, often overlying the nasal lateral wall, as in our patient.10 EAF is limited to the nasal cavity in over 90% and classically involves the nasal septum.9,10 Differential diagnoses to consider include: Wegener’s granulomatosis, Churg-Strauss syndrome, Kimura’s disease, sarcoidosis, and GF.5,7,10 Lab work-up consists of CBC, ESR, ANCA, IgE tiers, and peripheral eosinophils.9,10 Radiographic imaging is recommended to evaluate extent of disease and exclude pulmonary involvement, a key feature of Wegener’s granulomatosis and Goodpasture’s syndrome. Rheumatological referral may be beneficial, but did not reveal any abnormalities in our patient. A punch biopsy and FNA of the right lateral nasal skin were useful in excluding malignancy, but failed to establish a diagnosis in our patient.

EAF is diagnosed by two distinct and progressive histological stages: vasculitis and fibrosis.11 Early lesions represent an inflammatory phase with an active necrotizing vasculitis of the dermis with a tendency to develop slow-growing reddish-brown plaques on the face.8 However, GF rarely affects mucosal surfaces and lacks “onion skin” fibrosis.8 Other proposed etiologies include prior history of nasal trauma or surgery, atopy, or parasitic infection.5,10 In a study by Fang et al., only 26.9% of cases studied had a history of nasal trauma or surgery. The thought is that these predisposing factors result in an inciting event to an abnormal proinflammatory reaction in response to trauma or surgery. However, immunosuppressive and steroid therapies have been unsuccessful in treating EAF and eosinophilia is the only apparent evidence supporting atopy or parasitic disease.8 Our patient had no history of nasal trauma, surgery, or atopy. Additional studies suggest that EAF may be due to underlying IgG4-RSD, a systemic disease characterized by an elevated number of IgG4 plasma cells and T-cell infiltration producing tumefactive lesions in numerous organs.12,13 Like EAF, it is associated with allergic manifestations and shares similar histology with a rich lymphocytic infiltrate and fibrosis.14,15 This correlation was reported in a series of five cases showing increased IgG4 levels in 4 out of 5.16 However, a subsequent case series by Rimmer et al. failed to show such an association and our case further revealed no such findings.10 Other diseases categorized as IgG4-RSD include: Reidel’s thyroiditis, Mikulicz disease, chronic sclerosing siadaldenitis and orbital pseudotumor.7,10

**DISCUSSION**

While epidemiology is elusive, EAF most commonly presents during the fifth and sixth decades of life in 1:1 male-to-female ratio.4,5,6,9 The most common presenting symptom is progressive nasal obstruction in 78.8%, however, presentation is often nonspecific with an average of 60 to 70 months from time of symptom onset to diagnosis.13,5,6,9 Other symptoms may include epistaxis, breathing difficulties, sinusitis, nasal tenderness, and facial pain.5,6,9 Approximately 25.0% have associated nasal deformity or swelling, often overlying the nasal lateral wall, as in our patient.10 EAF is limited to the nasal cavity in over 90% and classically involves the nasal septum.5,10 Differential diagnoses to consider include: Wegener’s granulomatosis, Churg-Strauss syndrome, Kimura’s disease, sarcoidosis, and GF.5,7,10 Lab work-up consists of CBC, ESR, ANCA, IgE tiers, and peripheral eosinophils.9,10 Radiographic imaging is recommended to evaluate extent of disease and exclude pulmonary involvement, a key feature of Wegener’s granulomatosis and Goodpasture’s syndrome. Rheumatological referral may be beneficial, but did not reveal any abnormalities in our patient. A punch biopsy and FNA of the right lateral nasal skin were useful in excluding malignancy, but failed to establish a diagnosis in our patient.

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**REFERENCES**


Pathology images provided by Dr. Blythe Bowman, Ochsner Department of Pathology, New Orleans, LA.