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Case Report

Eosinophilic angiocentric fibrosis of nasal septum: a case report

Gayatri Ravikumar*, Anuradha Ananthamurthy, Usha Kini

Department of Pathology, St. John's Medical, College, Bangalore, Karnataka, India

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***Correspondence:**
Dr. Gayatri Ravikumar,
E-mail: gayatri.ravikumar@gmail.com

ABSTRACT

Eosinophilic Angiocentric Fibrosis (EAF) is an uncommon benign condition involving the sinonasal tract and rarely the larynx and orbit. Its etiology still remains unclear, although some studies have hypothesized it as a form of IgG4-related systemic disease. Histologically EAF is characterised by the presence of mixed inflammatory cells with eosinophil predominance in the early stage, followed by concentric layers of sclerosis around the small caliber vessels in the late stage. The rarity of this condition, tumor like presentation and oblivious etiology, makes it an interesting entity. We hereby report one such rare case of EAF, since the awareness of this entity is essential to make an accurate diagnosis.

Keywords: Eosinophilic angiocentric fibrosis, Fibrosis, IgG4 disease, Sinonasal tract

INTRODUCTION

Eosinophilic Angiocentric Fibrosis (EAF), a rare tumor-like proliferative fibrosing lesion involving the sinonasal tract and upper airways, presents clinically as an obstructive lesion. It was for the first time described by Robert and McCann in 1985.[1,2] Its exact etiopathogenesis and its association with other diseases are still a matter of debate although more recently some data has emerged linking EAF to the spectrum of IgG4 related diseases.[3] It is a slow and progressively evolving disease arbitrarily divided into an early and a late stage, the features of both of which may be seen on the same biopsy.

A review of medical literature has shown not more than 40 cases being reported in the sinonasal tract[4] and occasional ones involve the orbit. This indicates that the lesion is either rare or is being under reported. Its close clinical resemblance to a neoplasm or a vasculitis makes it an interesting entity and should be kept in mind in the differential diagnosis of proliferative lesions of the sinonasal tract.

CASE REPORT

We report here a case of EAF in nasal cavity, in a 53 year old male, who presented with complaints of a swelling in the left nostril with itching, sneezing and occasional episodes of epistaxis for the past two months.

On examination, a nodule, 1 cm in diameter, was seen on the left side of the cartilaginous nasal septum involving the Little's area. He had no other co-morbidities like diabetes mellitus, hypertension, asthma, or tuberculosis. The nodule was excised under local anaesthesia and sent for histopathological examination.

On gross examination, the specimen consisted of multiple irregular fragments of soft tissue which on microscopy, were fragments of nasal mucosa with a diffuse infiltrate of lymphoid cells in the sub mucosal stroma. A striking feature was the presence of numerous eosinophils amidst the lymphocytic infiltrate. A sprinkling of plasma cells was also noted (Figure 1). Also seen was extensive fibrosis with whorls of fibrosis around blood vessels giving an 'onion skin' appearance (Figure 2), highlighted

by Masson's trichrome stain. Neither granulomas nor necrosis were noted. There was no evidence of vasculitis in the entire tissue studied.

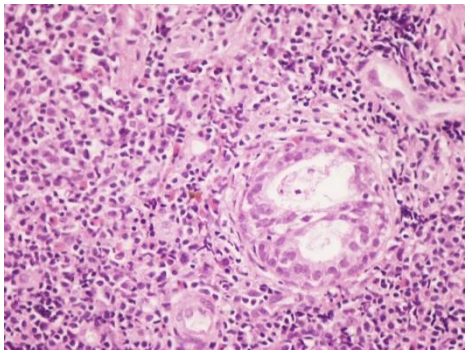

Figure 1: Photomicrograph showing nasal mucosa with a dense infiltrate of chronic inflammatory cells admixed with numerous eosinophils (H&E stain, original magnification x400).

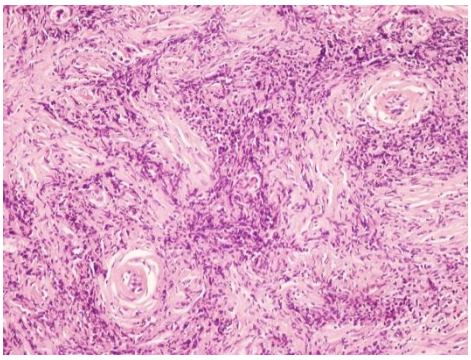

Figure 2: Note the lesion with extensive fibrosis with whorls of fibrosis around blood vessels (H&E stain, original magnification x200).

Immunohistochemistry showed an admixture of CD20 and CD3 positive lymphocytes, indicative of the reactive nature of the infiltrate. The vimentin stain highlighted the fibrotic component while the CD34 highlighted the vascular component.

The patient did well after the surgery and remained asymptomatic for the past one year of post-operative follow up.

DISCUSSION

EAF is a benign fibrosing lesion of the sinonasal tract, and upper respiratory tract. The majority of cases have been reported in women with a mean age of presentation at 44 years.[5] The clinical symptoms and radiological evaluations are nonspecific and include nasal obstruction, epistaxis, breathing difficulties and opacification of nasal cavity or sinuses on radiology. More recently it is suggested that EAF may be a IgG4 related systemic sclerosing disease in view of the elevated levels of serum IgG4 and the presence of IgG4 positive plasma cells seen predominantly in biopsies of these patients.[3] As it is an inflammatory and fibrosing disease process, recognition of this entity is essential as the clinical and radiological features may mimic malignancy and that many cases do respond to systemic steroids.

Both the early eosinophilic and late fibrotic stages of the disease may be encountered in the same biopsy as seen in the current case. During this fibrotic phase, the inflammatory infiltrate may be scanty and tissue IgG4 levels are typically low. Absence of necrosis is a conspicuous feature. The histopathological findings are usually characteristic as seen in the present case and clinch the diagnosis.

A host of reactive and neoplastic proliferations need to be considered in the differential diagnoses including Wegener's granulomatosis, Chrug Strauss syndrome, Kimura disease, granuloma faciale, fibromas, nodular fasciitis and lymphoma. The absence of necrosis, giant cells and granulomas exclude Wegener's and Churg Strauss disease. The absence of dense lymphoid aggregates renders Kimura disease unlikely. Although granuloma faciale and EAF have a few overlapping features, the concentric fibrosis is very characteristic of EAF and is not seen in the former.

Surgical excision remains the treatment option for EAF, although medical management with corticosteroids, dapsone and antihistaminics have been tried with less favorable outcomes. The recurrence rate for this disease is reported to be approximately 70% and disease free survival with surgical excision is reported in only 30% of the diagnosed cases.[4]

In conclusion, a case of EAF, which is a benign proliferative fibrosing disorder of the upper airways, is being reported for the first time in India, to the best of our knowledge, to bring awareness of this entity among the medical personnel and its propensity for recurrence after surgical excision. The classic histopathology of an eosinophilic infiltrate around vascular channels and concentric onion skin fibrosis should clinch the diagnosis. Its accurate diagnosis is essential in order to distinguish it from other more common neoplastic and vasculitic conditions of the nasal septum. Recent data linking EAF to one of the IgG4 related diseases points towards a pathogenetic mechanism that may aid better management in the future.

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