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Case Report

Pathology Section

Eosinophilic Angiocentric Fibrosis of Sinonasal Region: A Rare & Under Reported Entity

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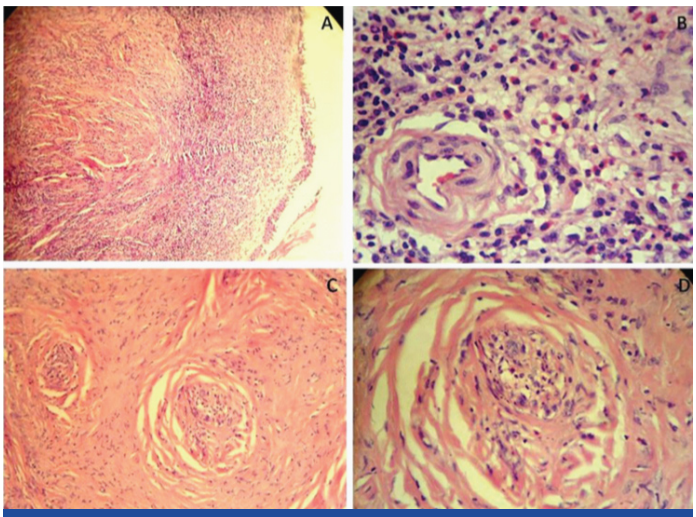
ABSTRACT

Eosinophilic angiocentric fibrosis is a rare pathology of the sinonasal tract and the upper respiratory system characterised by fibrosis with poorly understood pathogenesis. A 47-year-old male presented with a swelling over the dorsum of the nose. The possibility of fungal granuloma was being suggested on Magnetic Resonance Imaging (MRI). Histopathology showed thick collagen bundles whorling around vessels giving an onion skin appearance with focal area of vasculitis. An inflammatory reaction rich in eosinophils along with a fibrotic stroma was seen which was highly characteristic of eosinophilic angiocentric fibrosis. Clinically & microscopically it mimics Granuloma faciale, Wegener's Granulomatosis, Churg-Strauss Syndrome, Kimura's disease and few other granulomatous conditions thus making diagnosis difficult. A probable allergic origin is being suggested because of the typical eosinophil-rich inflammatory reaction. Finally the diagnosis of Eosinophilic Angiocentric Fibrosis was given. It is a diagnosis of exclusion having characteristic histomorphological findings thus biopsy is always required to distinguish it from other lesions whose treatment differs.

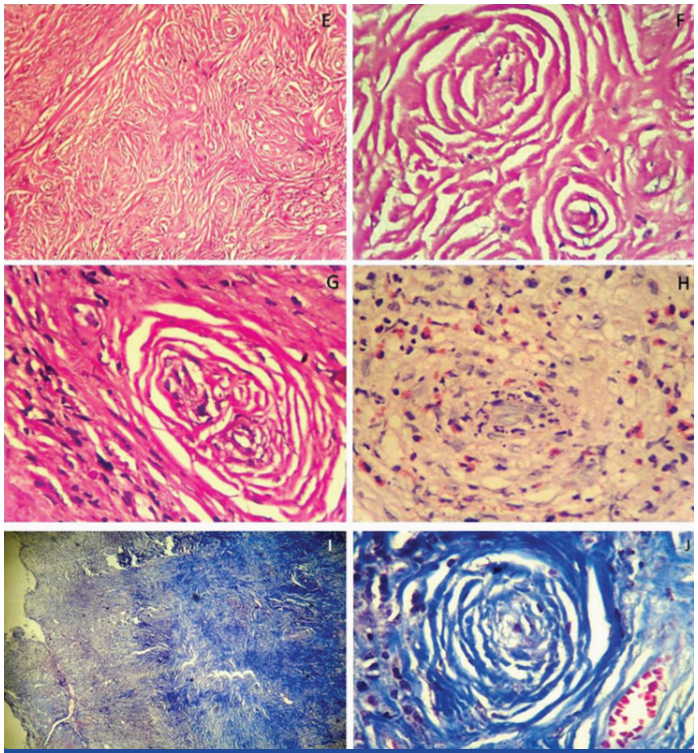
Keywords: Inflammatory cells, Onion skinning stroma, Upper respiratory tract

CASE REPORT

A 47-year-old male presented with a swelling over dorsum of nose since two and a half years. The swelling was insidious in onset & gradually progressive, first appeared on the right side and then involved left side as well. There was no history of any pain, nasal obstruction, discharge, post nasal drip, trauma, bleed, previous surgery, drug allergies, chronic illness and immunocompromised Viral markers, C-ANCA & P-ANCA were negative. Haematological investigations were within normal range with absolute eosinophil count of 0.1 x 10^9/l. Magnetic Resonance Imaging of paranasal sinuses showed a well defined thickening of anterior part of nasal septum extending antero-laterally to involve nasal cartilages with extension up to subcutaneous tissue/fat. The possibility of fungal granuloma was being suggested on MRI. Clinically one of the differentials was chordoma. Excision biopsy was done from the maxilla & some part of the nasal septum. Histopathology showed thick collagen bundles whorling around vessels giving an onion skin appearance with focal area of vasculitis. An inflammatory reaction rich in eosinophils along with a fibrotic stroma was seen which was highly characteristic of eosinophilic angiocentric fibrosis [Table/Fig-1,2].



[Table/Fig-1a-d]: (a) Pseudostratified columnar epithelium with underlying stroma having acute & chronic inflammatory infiltrate with prominence of eosinophils along with fibrotic stroma underneath (H&E 100X),(b) Thickened vessel wall with perivascular eosinophilic rich infiltrate (H&E 400X), (c) Perivascular fibroblastic proliferation giving an onion skin appearance (H&E 100X), (d) High power depicting same features (H&E 400X)



[Table/Fig-2e-j]: PAS stain highlighting the classical concentric onion skinning under low power x100, (e) and high power view 400X, (f,g). Focal area of vasculitis; (h) is evident without fibrinoid necrosis along with rich eosinophilic infiltrate surrounding the vessel (H&E 400X),(I) Low power showing blue fibrotic area deep in to the stroma (Masson Trichrome 100X), (J) Stain highlighting the perivascular concentric fibrosis (Masson Trichrome 400X)

DISCUSSION

Eosinophilic angiocentric fibrosis is a very rare lesion of the sinonasal area of the upper airways characterised by submucosal fibrosis. The pathogenesis of eosinophilic angiocentric fibrosis is not well known. It should be differentiated from eosinophilic rich inflammatory & allergic conditions. The indexed patient had no significant past history or any known allergic condition. First known cases were

highlighted by Roberts and McCann in 1985 but according to few authors it was first described by Holmes and Panje in 1983 [1,2]. Sevgen Onder et al., stated that not more than 13 cases have been reported in the English literature since that initial description [3]. The unresolving fibrosis and consequent stenosis requires surgical intervention more than once. Eosinophilic angiocentric fibrosis is also considered to be a mucosal variant of granuloma faciale, owing to the histological similarities between these 2 entities [4,5]. The latest research literature till 2014 talked about only 52 cases so far thus making this entity one of the rare pathologies [6,7]. Some authors described it as a neoplastic while other thinks that it's only an inflammatory fibrotic lesion. The histological appearance of this entity seems to be unique. Thick collagen bundles that whorl around vessels, an inflammatory reaction that is rich in eosinophils, and a fibrotic stroma are highly characteristic of this lesion [3,8]. The usual differentials are Granuloma faciale, Wegener's Granulomatosis, Churg-Strauss Syndrome, Kimura's disease and few other granulomatous conditions. Microscopically; in Granuloma faciale there is a polymorphous dermal inflammatory infiltrate consists of neutrophils and eosinophils. Vasculitis, with deposition of fibrinoid material within the vessel wall is seen which has never been reported in eosinophilic angiocentric fibrosis but in our case a focal area showing vasculitis was noted. It means vasculitis per se can be seen in eosinophilic angiocentric fibrosis even in the absence of c-ANCA & P-ANCA. On the other hand, perivascular collagen whorling is not a feature of granuloma faciale. Many authors argue whether eosinophilic angiocentric fibrosis is a mucosal variant of granuloma faciale, or they are distinct entities but till now no definite answer is obtained [9,10]. Wegener granulomatosis and Churg-Strauss syndrome are the other possibilities which should also be excluded. One of the cases of eosinophilic angiocentric fibrosis in the literature was reported to be associated with Wegener granulomatosis. Absence of geographic necrosis, necrotizing vasculitis, and granulomatous inflammation along with negative results for c-ANCA and p-ANCA excludes Wegener's granulomatosis and Churg-Strauss syndrome like in our case. In Kimura disease, there is seen proliferation of thin-walled vessels along with tissue eosinophilia but it differs histologically. Absence of dense lymphoid aggregates with prominent germinal centers excludes Kimura's disease [3,10]. The sinonasal tract is frequently the site of involvement for many granulomatous diseases, but no granuloma formation, giant cell histiocytic reaction, or necrosis is present in eosinophilic angiocentric fibrosis like in our case [3]. The aetiological factors remain elusive despite the consistency of pathological findings in these cases. But we think that if characteristic morphology is there then it must be characterised as chronic inflammatory lesion even if it is associated with some other diseases. Radiographic evaluations are usually non-specific and can be misleading. In our case radiological diagnosis was fungal granulomas. No definitive treatment of choice has been recognized. Most patients had to undergo multiple surgical resections. Although role of using steroids is still debatable, many authors demonstrated beneficial effects of using steroids [11]. Radiology images can give clue but it's not confirmatory [12]. Bony destruction may or may not be seen on CT or MRI [13,14]. Our patient had undergone surgical intervention and is on follow up now.

CONCLUSION

It is concluded that eosinophilic angiocentric fibrosis is a rare lesion, with predilection for the upper respiratory tract, of unknown aetiology. Radiographic investigations are non-specific but it has a characteristic microscopy thus biopsy is always required to distinguish it from other mimickers. Surgical resection is the treatment of choice, though multiple procedures are often required.

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