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CASE REPORT
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A RARE CASE OF EXTRANASOPHARYNGEAL ANGIOFIBROMA OF MAXILLARY SINUS

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ABSTRACT

INTRODUCTION

Extra nasopharyngeal angiofibroma's (ENA) are rare vascular tumour arising outside the nasopharynx. They typically present in older patients with a broader range of symptoms unlike nasopharyngeal angiofibroma (NA).

CASE PRESENTATION

This case report details a 27-year-old male with atypical presentation, including nasal obstruction, epistaxis, and facial swelling with a left sided nasal mass originating from the maxillary sinus. He underwent excision of mass and post operative Histopathology was suggestive of extra nasopharyngeal angiofibroma

DISCUSSION

ENA tend to be localized and less aggressive. The treatment of choice is surgical excision with recurrence rarely reported. Weighing the morbidity of incomplete surgical resection via endoscopic approach, the open approach was selected to ensure complete surgical resection.

CONCLUSION

The clinical and radiological divergence from NA, as demonstrated in this case, emphasizes the need for high index of suspicion, precise preoperative assessment to diagnose ENA accurately so as to guide proper management and prevent recurrence.

KEYWORDS

Extra nasal, Angiofibroma, Maxilla

INTRODUCTION

Nasopharyngeal angiofibroma is the most common benign neoplasm of the nasopharynx. It accounts for 0.05 % of all head and neck tumor, making it a rare clinical entity more often so when the presentation is extra nasopharyngeal. It mostly originates from superior aspect of sphenopalatine foramen at the junction of the sphenoid process of the palatine bone , horizontal ala of the vomer and root of pterygoid process of sphenoid at posterolateral wall of nasal cavity. Angiofibroma's that do not originate from area around sphenopalatine region are rare and referred to as Extra nasopharyngeal angiofibroma (ENA). De Vincentis et al., in his review of 704 cases of angiofibroma found that 13 cases originated outside nasopharynx which makes it a rare although a possible occurrence. Celik et al postulated that patients who have varying clinical characteristics from those of classic angiofibroma should be referred to as atypical angiofibroma. The typical clinical characteristics of ENA like age, sex, and symptoms do not conform to a great extent

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with that of nasopharyngeal angiofibroma. These contradicting characteristic features of ENAs make them a diagnostic challenge, therefore meticulous evaluation and high index of suspicion is required for establishing correct diagnosis. A 27-year-old male had presented to the outpatient department of the Department of Otorhinolaryngology and Head and Neck Surgery at our tertiary care Hospital with the complaints of unilateral left sided nasal obstruction since the last two years which was gradually progressive and associated with two episodes of nasal bleed which resolved with digital pressure. The patient also noticed a diffuse swelling below the left eye for the last 15 days. Examination revealed slight left sided facial fullness and prominence of the maxillary region with obliteration of the left nasomaxillary groove and mild proptosis of the left eye. Anterior rhinoscopy revealed a grey irregular mass filling the left nasal cavity and extending posteriorly, which on gentle probing was found to be attached laterally and did not bleed to touch. The right nasal cavity was found to be free of disease. The patient underwent a contrast enhanced computed tomography scan of the nose and para nasal sinuses which showed a soft tissue density completely filling left maxillary, frontal, ethmoidal, and sphenoid sinuses along with the left nasal cavity with patchy post contrast enhancement and bone remodelling in the form of widening of infundibulum of maxillary sinus, expansion of maxillary sinus with bulging walls causing narrowing of left pterygopalatine fossa and erosion of left superior, middle and inferior turbinates. Focal erosions of the lamina papyracea were also noted with soft tissue extending into extraconal space of the left orbit.

The patient was then taken up for a contrast enhanced magnetic resonance imaging which revealed a heterogeneously enhancing irregular polypoidal soft tissue lesion in left maxillary antrum and the left nasal cavity involving turbinates and projecting into posterior choana obscuring left OMC, nasal cavity and left nasopharynx, further extending into left ethmoid sinus and obliterating the frontal recess without extension into masticator space or infratemporal fossa. Normal left sphenopalatine foramen and effacement of pterygo-palatine fossa and pterygo-maxillary fissure was also noted with the mass bulging into the left medial rectus muscle. (Figure 1)

Fig. 1 – T2 Hyperintense mass lesion filling the left maxillary sinus and nasal cavity with bulging into the medial rectus

Considering the imaging and clinical findings the patient was planned for a biopsy of the nasal mass under local anaesthesia. The histopathological report showed a polypoidal tissue with abundant fibrinosuppurative exudate with the subepithelial region showing fibrinocartilagenous tissue with interspersed variably sized thin-walled channels, some showing fibrinoid change lined by plump endothelial cells, which were SMA and CD34 positive and Betacatenin negative, features suggestive of vascular tumor.

The patient subsequently underwent a CT Angiography of the neck to identify a feeding vessel. The lesion showed multiple vascular channels on arterial phase getting their supply from the maxillary artery ,branch of left external carotid artery with heterogeneous enhancement on venous phase with absent tumor blush, suggestive of a sinonasal vascular polypoidal mass. (Fig 2)

Fig.2– CT Angiography showing the mass deriving blood supply from the left maxillary artery

He was subsequently taken up for excision of the nasal mass under general anaesthesia after due consent, risk and consideration, via the Weber Ferguson approach along with an anteromedial maxillectomy. Prior to the surgery an embolization of the feeding vessel was attempted but no major feeding vessel was noted. The mass was seen filling the maxillary sinus and left nasal cavity with extension posteriorly till choana and erosion of the lamina papyracea into the medial rectus and was removed in-toto (Fig 3) and the final histopathological report revealed the mass to be an angiofibroma. (Fig 4 and Fig 5)

Fig. 3– 7 x 4 cm greyish red soft-firm nasal mass was excised in toto

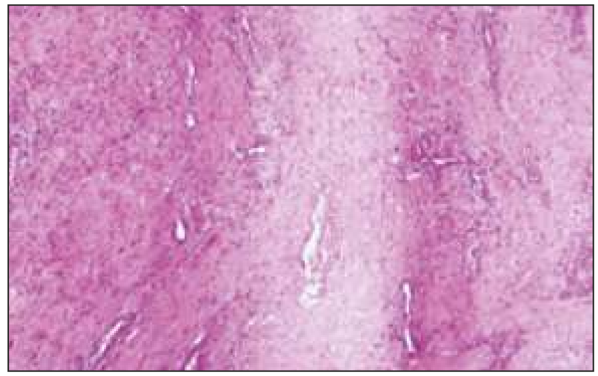

Fig 4. Variably sized vascular channels embedded in fibro collagenous stroma. (H&E x 100)

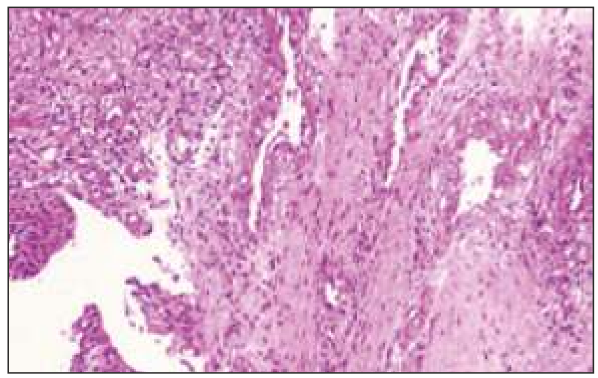

Fig 5. Nasal mucosa with underlying blood vessels embedded in fibro collagenous stroma. (H&E x 200)

The patient was followed up 1 month and 6 months post operative and is doing well with relief from all symptoms and no signs of recurrence.

DISCUSSION

Angiofibroma's are non-encapsulated, highly vascular tumors usually occurring in the nasopharynx and affecting mostly patient at an average age of 14-17 years. (1) It is a unique fibrovascular tumor with characteristic endothelial lined vascular spaces embedded in a fibrous stroma (2). In recent times, the term extra nasopharyngeal angiofibroma has been applied to vascular, fibrous nodules occurring outside the nasopharynx. (3) In comparison to

NA, patients affected are older, female involvement is seen, progression of symptoms is faster and lesser hypervascularity is noted in the lesion. (4–6)

The mean age of presentation of ENA is 22 years (7,8) whereas in our case the patient presented at 27 years of age which is atypical from the existing literature. The male to female ratio is 3:1(7,8) which is corroborated with this report.

Windfur et al. in his recently concluded review of literature showed that nasal septum was the most common site (22.4%) of origin. (1) Other reported sites of ENA include, sphenoid sinus, maxillary sinus, middle and inferior turbinate, larynx, external ear, cheek, carotid bifurcation, esophagus and infratemporal fossa. This report highlights the atypical site of origin of ENA, that is the maxillary sinus.

Maximum patients with ENA present with nasal obstruction either in combination with epistaxis (28.5%) or other symptoms (12.5%). Other symptoms include painless facial swelling (3.4%), dysphagia (3.4%).(1,4) In our case too the patient had primarily nasal obstruction with a few episodes of nasal bleed and a painless left sided facial swelling, most likely due to the mass effect of the tumor. Although histopathologically similar to NA the clinical presentation is very different (9), as is evident from this case report.

Clinical and radiological examination are the mainstay to preoperatively asses the site of origin in cases of ENA as well as to look for its extensions. Alvi et al. considered CT to be sufficient for the diagnosis of ENA since it clearly delineates and identifies the tumor. (10) ENA usually enhances with contrast, however a failure in enhancement is seen in tumors with low vascularity. In our case we had a tumor mass which had a patchy contrast enhancement, due to its hypo vascularity which is a typical feature of ENA.

The internal maxillary artery was identified as the feeder vessel on CT Angiography, however when embolization was attempted via DSA, no major feeding vessel was noted. The characteristic tumor blush of NA was also not seen in this patient hence corroborating the diagnosis of ENA.

Since ENA tend to be localized and less aggressive, the treatment of choice is surgical excision with recurrence being rarely reported (11). Weighing the morbidity of incomplete surgical resection via endoscopic approach against the morbidity of open approach, the open approach was selected due to the extensive spread of the tumor as well as to ensure complete surgical resection.

CONCLUSION

Although ENA are extremely rare, they must be considered as a differential diagnosis of an unclear nasal and sinonasal mass, particularly in adult patients presenting with rapidly developing nasal obstruction.

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- All authors reviewed the manuscript and approved it before submission.

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