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EXTRA NASOPHARYNGEAL ANGIOFIBROMA OF THE SPHENOID SINUS: A RARE CASE REPORT

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ABSTRACT

Aim: This study aimed to find the effect of preoperative tumor embolization and transnasal endoscopic excision of a sphenoid sinus angiofibroma on the postoperative outcome.

Case Report: A thirteen year old male presented to the Outpatient ENT Department of a tertiary care centre, with a history of recurrent left-sided spontaneous painless epistaxis since one year. Diagnostic Nasal Endoscopy showed a pink globular mass seen occupying the left choana and extending into the left nasal cavity. CT Paranasal sinuses (Plain and Contrast) revealed a well-defined moderately enhancing soft tissue lesion seen predominantly at the base of sphenoid sinus on the left side, extending into the choanal region with destruction of the base of the sphenoid bone. Patient underwent Diagnostic Angiography in view of pre-operative embolisation and the patient later underwent embolisation of feeder artery supplying the massusing Gelfoam. Subsequently the patient underwent Transnasal Endoscopic Excision of the tumour under general anaesthesia and the specimen was sent for histopathological examination, which revealed features suggestive of Angiofibroma.

Discussion: Extranasopharyngealangiofibromas are vascular fibrous nodules occurring outside the nasopharynx and are rare, benign neoplasms characterized by a different biological history and clinical features with respect to nasopharyngeal tumours, and for these reasons, should be regarded as a separate clinical entity.

Conclusion: Although it sometimes involves the sphenoid sinus, angiofibroma rarely originates from this site and only five such cases have been reported in the literature.

Keywords: Extranasopharyngealangiofibroma, sphenoid sinus, epistaxis, transnasal endoscopic excision

INTRODUCTION

Angiofibromas are benign, highly vascular and locally aggressivetumours observed most frequently in adolescent males and are the most common benign tumours of the nasopharynx, accounting for 0.5% of all head and neck neoplasms[1]. It originates from the sphenoid foramen at the junction of the root of the sphenoid process of palatine bone, horizontal ala of vomer and pterygoid process of sphenoid bone[2,3]. When it occurs out of this site, it is termed as extranasopharyngealangiofibroma (ENA) or atypical angiofibroma. In 1980, De Vincentiis and Pinelli[7] reviewed a series of 704 cases of angiofibroma and found that 13 cases manifested outside the nasopharynx, thus suggesting that extranasopharyngeal localization of the tumour is possible, although rare. This rare tumour usually displays variable clinical presentation and was recently termed as “atypical angiofibroma” due to its distinct characteristic features[4-6]. Thus, extranasopharyngealangiofibromas can constitute

a challenge in terms of diagnosis and treatment. A high level of suspicion is needed for prompt diagnosis and treatment of such lesions. A review of world literature revealed more than 65 cases of extranasopharyngealangiofibromas. The maxilla was the most common site, followed by ethmoids, nasal septum, nasal cavity and sphenoid sinus[8].

CASE REPORT

A 13-year-old male presented to the Department of Otorhinolaryngology of a tertiary care centre with the complaints of intermittent episodes of bleeding from the left nasal cavity since one year. The nasal bleed was intermittent, profuse and which started and stopped spontaneously and was associated with left-sided nasal obstruction. There was no history of any aggravating or relieving factors. There was no previous history of trauma or bleeding diathesis. The patient was not on any regular medication. There was no history in the family of similar illness or of bleeding disorders. A complete ENT examination was done. Anterior rhinoscopy revealed the nasal septum to be deviated to the left with thick mucopurulent discharge in the left nasal cavity. Neurologic examination was normal. Diagnostic nasal endoscopy revealed a pink, globular mass seen occupying the left choana with thick mucopurulent discharge (Fig. I). Contrast enhanced computerized tomography (CT) of the paranasal sinuses showed a well-defined moderately enhancing soft tissue lesion noted predominantly at the base of the sphenoid sinus on the left side, extending inferiorly upto the choanal region, with destruction of the base of the sphenoid bone (Fig. II, III). The patient was referred to the Department of Cardiology and subsequently underwent Diagnostic Angiography in view of pre-operative embolisation to identify the feeder vessels. Selective gel foamembolisation of the feeder artery (Internal Maxillary Artery) was done using 2.8F Progress microcatheter with good results. Following this, the patient was taken up for Transnasal Endoscopic Excision of the tumour. Intraoperatively, the size of the tumour was found to have reduced significantly post-embolisation. The sphenopalatine artery was identified and cauterized. Partial middle turbinectomy was performed on the left side, to aid better exposure and visualization of the tumour. Subsequent to the entry into the sphenoid sinus, the smooth-surfaced tumour was seen seen arising from the base of the sphenoid sinus on the left side, measuring 3.5 x 2.5 cm. The tumour was elevated from the underlying periosteum and was removed en bloc under endoscopic guidance. Intraoperative bleeding was minimal. The left sphenoid sinus and the left nasal cavity werepacked for 48 hours. Post-operative recovery was uneventful. The pack was removed after 48 hours and the patient was discharged without any complications. The histopathological examination of the excised specimen revealed tissue bits lined by respiratory epithelium with squamous metaplastic change focally. The underlying stroma was variable from loose oedematous with stellate cells and mast cells to densely packed spindle cells and collagenous stroma. Numerous vessels ranging from dilated stellate shaped venules with flattened epithelium to small capillaries with plump endothelial lining were present (Fig. IV). These findings were consistent with angiofibroma. At 6 months follow-up, the patient was free of recurrence.

DISCUSSION

Angiofibroma is the most common tumour of the nasopharynx and makes up 0.5% of all head and neck tumours. It typically occurs in the nasopharynx of young males in the first and second decades[1,4]. More recently, the term extranasopharyngealangiofibroma has been applied to vascular fibrous nodules occurring outside the nasopharynx. However, extranasopharyngealangiofibroma have virtually nothing in common with nasopharyngeal tumours and the use of the term angiofibroma for these lesions may therefore be confusing. These rare, benign neoplasms are characterized by a different

biological history and clinical features with respect to nasopharyngeal tumours, and for these reasons, they should be regarded as a separate clinical entity. Compared to nasopharyngeal angiofibromas, patients affected are older, females can be involved, symptoms develop more quickly, and hypervascularity is less common[1,3]. Isolated extranasopharyngealangiofibroma is rare. A review of 65 extranasopharyngealangiofibroma cases showed that the average age of these patients was 22.9 and only 25-26% of patients were female[3]. Extranasopharyngealangiofibroma most commonly arises in the maxillary sinus, followed by the ethmoid sinus. The nasal septum, larynx, external ear, cheek, conjunctiva, oropharynx, retromolartrigone and middle and inferior turbinates are other reported sites of occurrence. To the best of our knowledge, only 5 cases originating from the sphenoid sinus have been reported in the literarture previously. The clinical presentation of extranasopharyngealangiofibroma depends mainly on the localization and extent of the tumour.

According to histopathology, extra nasopharyngeal angio fibromas constitute a more heterogenous group. Classic radiological findings characterizing nasopharyngeal angiofibromas are not shared by extranasopharyngealangiofibromas. Most extranasopharyngealangiofibromas enhance after contrast medium injection, however, enhancement is not a constant sign, and may vary from intermediate to strong. Unlike nasopharyngeal angiofibromas, radiological presentation of extranasopharyngealangiofibromas is much more variable due to their various locations. These tumours may spread to adjacent areas by widening of natural foramina and fissures or by erosion of bony structures, which is well demonstrated on CT[2,15,16]. Lack of hypervascularity on angiograms does not exclude the diagnosis of extranasopharyngealangiofibroma. The blood supply to the extranasopharyngealangiofibroma depends on its point of origin and location, whereas in nasopharyngeal angiofibromas, the main feeder is the maxillary artery.[17]

Radiological examination is essential for establishing the correct diagnosis, making an appropriate treatment plan, determining the extent of the lesion and identifying the feeding vessel. Measures such as CT, MRI and angiography can be used. Selective angiography clearly reveals the vascular pattern and hemodynamics of the tumour; however absence of hypervascularity in angiography does not fully exclude ENA.[3] CT and MRI can determine the extent of tumour including the skull base involvement, intracranial spread and its relationship with important vascular and neurological structures.[9] Although bony erosion can be determined with CT, MRI is usually sufficient to show the cortical erosion and trabecular bone formation stimulated by the tumour. Imaging of nasopharyngeal angiofibroma with a contrast agent leads to diffuse and usually homogenous involvement in CT and MRI T1 scans. In contrast, ENA enhances moderate amount of contrast or none due to its weak vascular involvement.[10,14]Embolization can be utilized in cases with increased vascularity. Arteriography before biopsy or removal of tumour may reduce the risk of active bleeding. In this patient, selective embolisation of the feeder artery was done preoperatively to reduce the blood supply to the tumour. Owing to the small size and limited extent of the tumour, transnasal endoscopic excision was preferred.

The treatment of choice for angiofibroma is total surgical excision. Other treatment modalities include radiotherapy, cryosurgery, embolization, hormone therapy, chemotherapy, arterial ligation, sclerotherapy and watchful observation with the hope of spontaneous regression. Radiotherapy is less effective for ENA compared to nasopharyngeal angiofibroma.[3]

Tillaux[11] suggested that nasopharyngeal angiofibromas originated from the fibrocartilaginous barrier in the lower border of the sphenoid bone, in front of the atlas. Brunner[12]

described this structure as 'fascia basalis' because he found no cartilage in it. Later, Hiraide and Matsubana[13] reported a case of angiofibroma located in the anterior third of the nasal septum and showed that this tumour originated from the periosteum of the perpendicular lamina of the ethmoid bone, away from the fascia basalis. These reports support the presence of ectopic tissue as the most common theory to explain the site of origin of extranasopharyngealangiofibromas.

CONCLUSION

A strong index of suspicion is required to diagnose an extranasopharyngealangiofibroma. CT and MRI are sufficient to diagnose such lesions. Biopsy is to be avoided as it may cause profuse bleeding. Preoperative tumour embolization is an important measure to reduce intraoperative bleeding and to reduce tumour size. Surgical excision is the treatment of choice for such tumours. Transnasal endoscopic excision is a useful surgical modality in the case of limited lesions. Angiofibroma should be included in the differential diagnosis of vascular tumours of the nasal cavity and paranasal sinuses. Whether these lesions represent an angiofibroma or a variant of another lesion is debatable.

CONFLICT(S) OF INTEREST: NONE

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ILLUSTRATIONS

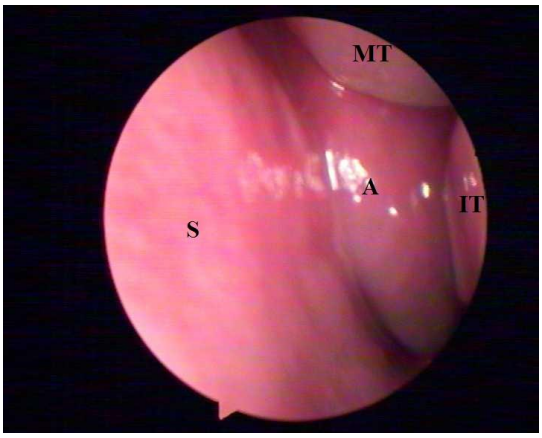
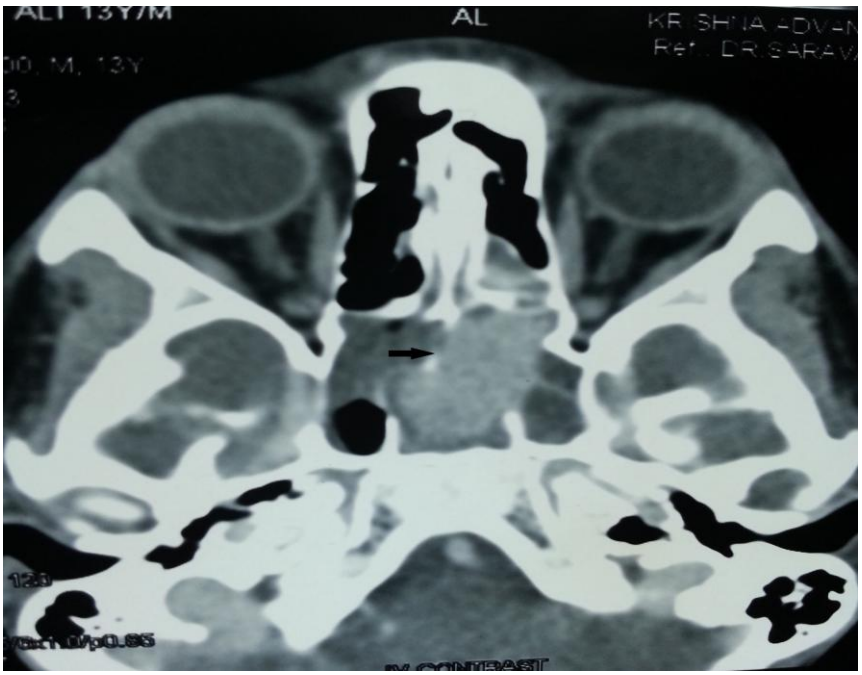
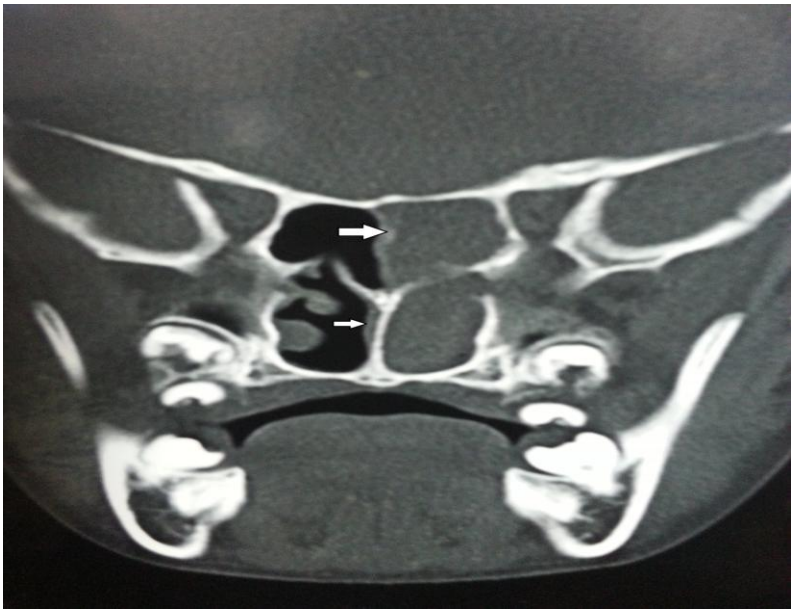


Figure I: Diagnostic nasal endoscopic view of the left nasal cavity showing a pink globular mass occupying the left choana. S: septum, IT: inferior turbinate, MT: middle turbinate, A: angiofibroma





Figures II and III: CT Nose and Paranasal sinuses (Plain and Contrast) showing a well defined moderately enhancing soft tissue lesion noted predominantly at the base of the sphenoid sinus on left side, extending inferiorly upto thechoanal region with destruction of the base of the sphenoid sinus

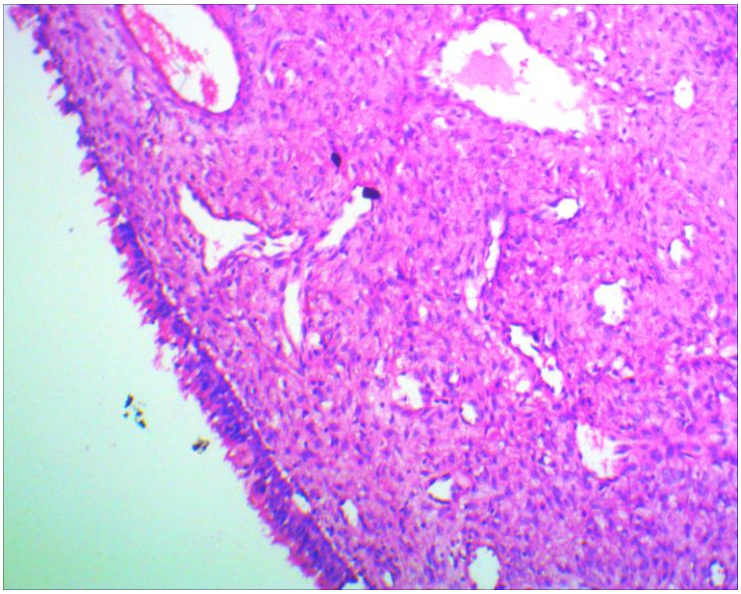


Figure IV: H & E stained section of the specimen showing tissue bits lined by respiratory epithelium with squamous metaplastic change focally, with numerous vessels ranging from dilated stellate shaped venules with flattened epithelium to small capillaries with plump endothelial lining present