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CASE REPORT

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EXTRA-NASOPHARYNGEAL ANGIOFIBROMA IN THE NASAL VESTIBULE- A SURPRISE!

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A B S T R A C T

Angiofibromas are highly vascular tumors that predominantly arise from the Nasopharynx. Extra-nasopharyngeal Angiofibromas are rare, vascular or fibrous nodules occurring outside the Nasopharynx. We present a 14 year old boy who presented with mass in the left nasal cavity of 1 month duration. Examination revealed a smooth, pinkish mass arising from the lateral aspect of left nasal vestibule. The mass was excised in toto and sent for histopathological examination which confirmed it to be Angiofibroma. This case is being reported due to its atypical origin from an extremely rare site.

INTRODUCTION

Angiofibromas commonly arises from the Nasopharynx. They are histologically benign but potentially locally aggressive vascular neoplasms that commonly affect adolescent males. The earliest Greek, Roman and Arabian literature used the term nasal polyp to designate all tumors or swellings within the nasal cavity or nasopharynx which caused obstruction to breathing. The specific form of Nasopharyngeal Angiofibroma was first recognized in 1847 when Chelius described a fibrous nasal polyp that "commonly occur in persons about the time of puberty."

In 1980, De Vincentiis and Pinelli studied a series of 704 cases of Angiofibroma, in which 13 cases manifested outside the Nasopharynx. Compared to Nasopharyngeal Angiofibromas, Extra-Nasopharyngeal Angiofibromas differ in that patient affected are older[3], symptoms develop more quickly, incidence in female is comparatively more and hypervascularity is less common. Most common site for atypically localized Nasopharyngeal Angiofibroma is the Maxillary Sinus with an incidence of 32%. Other sites of origin includes Ethmoid Sinus[2], Nasal Septum, Nasal Cavity, Larynx, Sphenoid Sinus, Conjunctiva, Oropharynx, Hypopharynx, Facial Nerve, Lacrimal Sac, Retromolar Area, Middle Turbinate, Inferior Turbinate, Trachea and Esophagus[8]. A case is being reported where the mass originated from the lateral aspect of left nasal vestibule which is one of the rarest sites that has been reported so far in medical literature[3].

Case Report

A 14 year old boy presented to the ENT Department at Rajah Muthiah Medical College with 1 month history of mass in the Left Nasal Cavity. It was insidious in onset, started as a pea-sized mass and progressed to attain the present size of 2 x 1 cm. Patient gave history of recurrent upper respiratory tract infection and two episodes of nasal bleed. Examination revealed a smooth mass arising from the left nasal vestibule and obscuring the entire left nasal cavity. On probing the mass was insensitive to touch, did not bleed on touch and was attached to lateral aspect of left Vestibule. Examination of the right nasal cavity was normal. Posterior Rhinoscopy examination revealed a normal Nasopharynx. Computed Tomography (CT) Scan of the nose and paranasal sinuses showed an ill-defined polypoidal soft tissue dense mass of size 1. 9 x 1.07 cm arising from lateral aspect of left vestibule. The mass was excised in toto using bipolar cautery under general anesthesia. The left nasal cavity was packed with merocel nasal pack, which was removed the following day. Post-operative period was un-eventful. Histopathological examination revealed irregular dilated vascular spaces lined with endothelial cells with elongated spindle shaped cells with plump nuclei and scanty cytoplasm arranged in sheets which are present with blood vessels, some of

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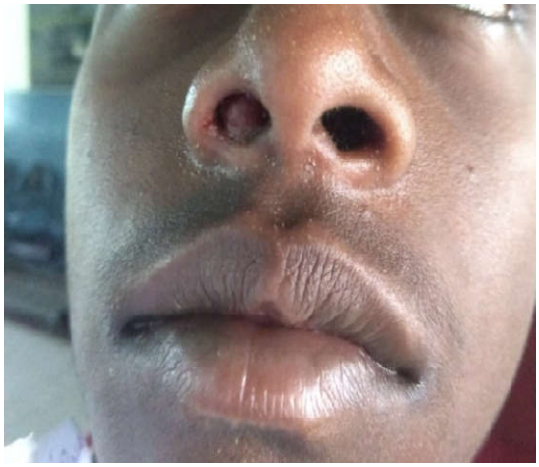
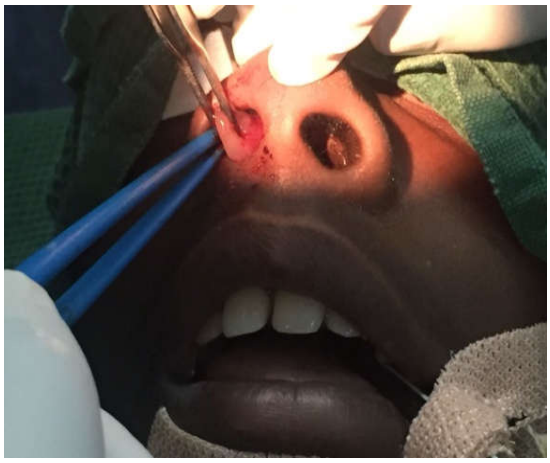
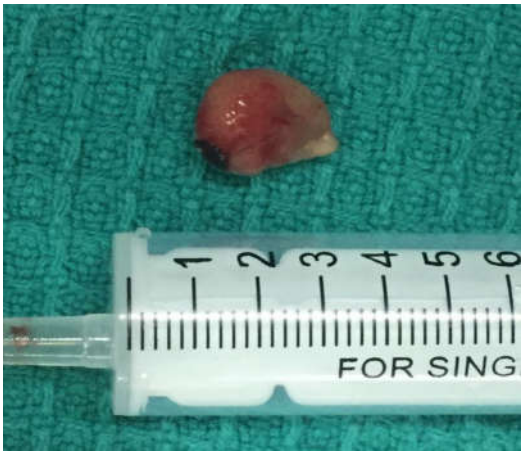
which are collapsed. Focal areas of myxoid changes are seen. The histopathological findings are consistent with the diagnosis of Angiofibroma.

Pre-Operative Image

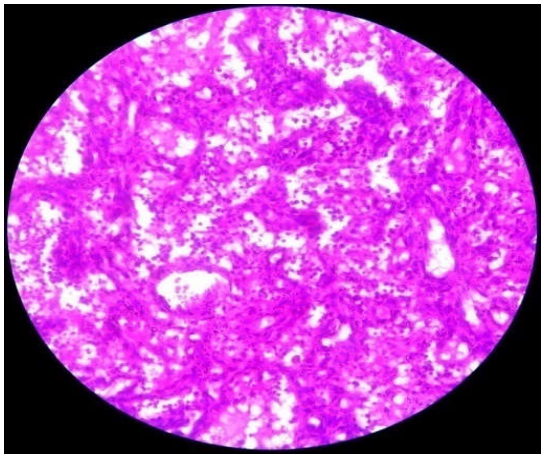


Intra-Operative – Excision of the mass using Bipolar Cautery





Specimen



Histopathological Image

DISCUSSION

Nasopharyngeal Angiofibroma is a well defined entity that usually arises from posterolateral wall of Nasopharynx near the superior margin of Sphenopalatine Foramen[4]. It may later extend into the nasal cavity, paranasal sinuses, spheno-palatine foramen and infra-temporal fossa[6]. It is a rare vascular tumor which represents 0.05% of all head and neck tumors[9]. It is the most common benign neoplasm of the Nasopharynx. The presence of numerous stag-horn vessels within its fibroblastic stroma due to lack of a complete muscular layer in the blood vessels is responsible for catastrophic bleeding during surgery or following biopsies. In 10-20% of these cases, tumor will invade the cranial cavity.

Recently the term Extra-nasopharyngeal Angiofibroma has been applied to describe Angiofibroma arising from sites other than the Nasopharynx[5]. Windfuhr studied a total of 65 patients with Extra-nasopharyngeal Angiofibroma from 16 different countries, where a total of 48 patients (73%) were male. Both Nasopharyngeal Angiofibroma and Extra-Nasopharyngeal Angiofibroma are more common in males but the incidence in females is comparatively more in Extra-Nasopharyngeal Angiofibroma with a mean age of presentation at 22 years. Maxillary Sinus was the most common site.

Other sites of origin includes Ethmoid Sinus, Nasal Septum[7], Nasal Cavity, Larynx, Sphenoid Sinus, Conjunctiva, Oropharynx, Hypopharynx, Facial Nerve, Lacrimal Sac, Retromolar Area, Middle Turbinate, Inferior Turbinate, Trachea and Esophagus[10]. Occurrence of Extra-Nasopharyngeal Angiofibroma in the Vestibule is very rare.

Unlike Nasopharyngeal Angiofibroma, the clinical presentation of these tumors is non-specific and depends on the localization and extent of the tumor. Tumors originating from the nasal cavity typically present with complaints of nasal obstruction and epistaxis and may be diagnosed early due to limited space for tumor growth. Histopathological appearance consists of numerous wide irregular vessels with a single layer of endothelial cells, embedded in a fibrous stroma[1].

Surgical excision of the mass is the treatment of choice and recurrence is rare. Unlike Juvenile Nasopharyngeal Angiofibroma, Extra-Nasopharyngeal Angiofibroma does not necessarily require a pre-operative embolisation as the tumor is comparatively smaller and less vascular. Surgical approach is determined by the size, location, vascularity or blood supply of the tumor. Despite the benign

histopathological nature of the tumor, it is always a challenge to the surgeon.

CONCLUSION

Extra-nasopharyngeal Angiofibromas differ from Nasopharyngeal Angiofibroma in that they lack typical clinical and radiological features, develop in older age group, with a comparatively higher incidence in females, are less vascular and present with a variety of symptoms depending on the site of origin. This case highlights the importance that Angiofibroma should be considered as a differential diagnosis of any nasal mass. Nasal vestibule should be regarded as a potential, although an exceptional, location of these tumors.

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