



A case reports of Extra-nasopharyngeal Angiofibroma arising from the nasal septum- A diagnostic challenge.

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ABSTRACT: Nasopharyngeal angiofibroma is a rare, benign, locally aggressive tumour of vascular origin. The most common site described for the origin of nasopharyngeal angiofibroma is at the sphenopalatine foramen. When this angiofibroma originates from a different site other than the conventional sphenopalatine foramen, it is termed as Extra-nasopharyngeal Angiofibroma. This is a rare finding in comparison to conventional nasopharyngeal angiofibroma. We are reporting a rare case of Extra-nasopharyngeal angiofibroma, in a 31 year old male patient, who presented with complaints of right sided nasal obstruction. The nasal obstruction progressed over 1 month, which was associated with episodes of unprovoked epistaxis. On examination, a pinkish mass was seen protruding from the right nasal cavity, occupying whole of the cavity anteriorly arising from the nasal septum. The polypoidal mass was surgically excised, and sent for histopathological examination, which was reported to be Angiofibroma arising from the Nasal septum (Extra-nasopharyngeal Angiofibroma). There has been no evidence of recurrence of the lesion. Though very rare, it is always better to consider Extra-nasopharyngeal angiofibroma as a differential diagnosis, while evaluating a nasal or a nasopharyngeal mass.

I. INTRODUCTION:

Angiofibroma is a histologically benign and locally aggressive vascular tumour that typically arises from the nasopharynx¹. The most common site for the origin of nasopharyngeal angiofibroma is at the sphenopalatine foramen in the nasopharynx. It comprises of <1 % of all the head and neck neoplasms. The term Extra-nasopharyngeal angiofibroma is applied to the angiofibroma arising from a site other than the typical site of its origin, such as maxillary sinus, ethmoidal sinus, nasal septum, tonsil, retromolar

trigone area etc². These comprise of 0.05% of the head and neck masses^{3,4}.

Though structurally similar to the typical nasopharyngeal angiofibroma, Extranasopharyngeal angiofibroma, are considered to be different from nasopharyngeal angiofibroma due to the varied presentation. There have been multiple studies that suggest that these group of angiofibroma can be referred to as "atypical angiofibroma"^{3,4}. Here, is a rare case reported of angiofibroma originating from the nasal septum.

II. MATERIAL AND METHOD:

Patient came to the outpatient department of otorhinolaryngology at MGM institute of health sciences, who was identified and evaluated in detail. After obtaining history, detailed examination and radiological evaluation was done. Written and informed consent was taken prior to the surgery and surgical excision was planned under general anaesthesia. Patient has also been explained regarding reporting and publishing of the case.

III. CASE:

A 31 year old male patient came to our outpatient department with complaints of right sided nasal obstruction since 1 month insidious in onset, gradually progressive, associated with mass in the right sided nostril since 20 days and 2-3 episodes of unprovoked epistaxis since 20 days from the same nostril, that stopped spontaneously. The patient had no significant past complaints, trauma or infection.

On ENT examination, it was found that the external nasal framework was normal, nasal septum was normal and a pinkish mass protruding from the right nasal cavity which was touching the right inferior turbinate and the lateral nasal wall. Left nasal cavity was found to be normal. Probe test and decongestion test were performed, that revealed that the mass was attached to the cartilaginous nasal septum anteriorly, just posterior to the Little's area (3/4th of an inch above the



mucocutaneous junction) and the mass failed to decongest/shrink in size with 4% lignocaine.. Paranasal sinuses were found to be normal. Oral cavity and ear examination were found to be within normal limits.

A detailed contrast enhanced computed tomography of the nose and paranasal sinuses revealed an enhancing mass measuring 1.9cm in right nasal cavity arising from the nasal septum and just anterior to the middle turbinate extending anteriorly to fill up the right nasal cavity from the level of inferior turbinate to the vestibule, with no erosion of bone or cartilage and normal paranasal sinuses. There was no extension posteriorly in the nasal cavity or the nasopharynx.



Image 1: contrast enhanced CT scan showing mass occupying the right nasal cavity.

After pre-operative anaesthetic workup, the patient was then planned for surgical excision of vascular mass under general anaesthesia. Under all aseptic precautions, apolypoidal, vascular lesion was excised using bipolar diathermy and continuous cauterization from the bed of the nasal septum just posterior to the Little's area, measuring 2x 1.5 cm in toto.



Image 2: A polypoidal, highly vascular specimen excised in toto.

Sample was then sent for histopathological examination. Intraoperative bleed was very minimal. Adequate haemostasis was achieved. An anterior nasal pack was placed bilaterally which was removed after 48 hours. Post-operative period was uneventful without any kind of complications. Histopathological examination revealed multiple dilated, cavernous vascular spaces lined with endothelial cells which were separated by fibrous stroma with stromal cell nuclei. This was found to be consistent with features suggestive of Angiofibroma. At 3 month and 6 month follow, nasal cavity was found to be within normal limits with no features of any kind of residual disease or recurrence.

IV. DISCUSSION:

Angiofibroma of the head and neck in most of the cases originate from the nasopharynx. It has already been established that the angiofibroma are histologically benign and locally aggressive vascular tumours^{1,2,3,4}. The term Extra nasopharyngeal angiofibroma has by far come in practice to identify angiofibroma arising from the site other than the typical site of origin⁴, which is at the sphenopalatine foramen in the nasopharynx. Though, Epistaxis is the most common presenting sign, nasal obstruction, pain, progressive swelling of the nose are also seen.

The variation that we experience are usually in the peak age of clinical presentation, sex and the site of origin. The vascularity also plays an important role, as these lesions seem to be relatively less vascular in comparison to the typical nasopharyngeal angiofibroma. They are mostly known to arise from an ectopic nidus of vascular tissue in the periosteum of the nose. The strong sex and age correlation suggests the contribution of hormonal disorders as well³. However, the aetiology of nasopharyngeal angiofibroma (typical as well as atypical) remains unclear as there is no direct evidence to prove this hypothesis. The Extra nasopharyngeal angiofibroma can evolve with a variety of symptoms and radiological signs, depending on its site. Here, our patient reported with unilateral nasal obstruction due to a rare location in the nasal septum.

The CT scan has been considered the preferred radiological exam for many years and MRI is being used to determine the tumour site and extension. Treatment for the nasopharyngeal angiofibroma includes radiotherapy, endoscopic surgery and pre-operative embolization. Surgical excision is generally considered the best treatment option for extra nasopharyngeal angiofibroma with



recurrences rarely seen. However, management can vary based on the site of origin as well.

V. CONCLUSION:

The variable presentation of Extranasopharyngeal angiofibroma pose as a great diagnostic challenge. A very meticulous evaluation with a high index of suspicion of the disease, is required to establish the diagnosis and treatment. Although a rare diagnosis, it should be kept in mind as a differential diagnosis with any unilateral vascular mass causing nasal epistaxis and nasal obstruction^{2,4}.

CONFLICT OF INTEREST:

None.

REFERENCES:

- [1]. Windfuhr JP, Remmert S. Extranasopharyngeal angiofibroma: etiology, incidence and management. *Acta Otolaryngol.* 2004;124(8):880-9. PMID: 15513521 DOI: [http:// dx.doi.org/10.1080/00016480310015948](http://dx.doi.org/10.1080/00016480310015948) [Links]
- [2]. Shroff Makhasana, Meena A Kulkarni, SuhasVaze, Adil Sarosh Shroff *J Oral MaxillofacPathol.* 2016 May-Aug; 20(2): 330. doi: 10.4103/0973-029X.185908 , PMID: PMC49895741.
- [3]. Amarendrasingh, Kalpana rajivkumar, suman P Rao, A rare case of extranasopharyngeal angiofibroma. *International journal of medical and allied health sciences*, 2014;4,381-382
- [4]. Tasca I, Compadretti GC. Extranasopharyngeal angiofibroma of nasal septum. A controversial entity. *Acta Otorhinolaryngol Ital.* 2008; 28(6):312-314.
- [5]. Harsh Sobhari, Shiv Kumar, Jyoti Sobhari *Indian J Otolaryngol Head Neck Surg.* 2019 Nov; 71(Suppl 3): 2117–2120. Published online 2018 Dec 8. doi: 10.1007/s12070-018-1542-x PMID: PMC6848666
- [6]. Bhargava S. K., Phatak S. Angiofibroma arising from nasal septum in adult male—a rare occurrence. *Indian Journal of Otolaryngology and Head and Neck Surgery.* 1995;47(1):39–41. doi: 10.1007/bf03047916