

# JUVENILE NASOPHARYNGEAL ANGIOFIBROMA IN AN ADULT - A CASE REPORT

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## ABSTRACT

### BACKGROUND

Juvenile nasopharyngeal angiofibroma is a benign lesion which is characterized by three classical features: it occurs mostly in males, over one period of age and in one location. Juvenile nasopharyngeal angiofibromas are benign but locally aggressive tumours of nasopharynx seen exclusively in adolescent males. Angiofibromas arising in sites other than nasopharynx is very rare. It is characterized by episodes of painless profuse epistaxis.

JNA constitutes of less than 0.5% of all head and neck neoplasms. In very few cases it has been reported in men over 25 years old or females. Median age of onset of symptoms is 14 to 18 years. Most common symptoms are painless episodes of epistaxis, nasal obstruction, headache. Here we report a case of adult JNA.

### CASE PRESENTATION

A 29 years old Male presented with complaint of bleeding from right nasal cavity for last five years associated with on and off nasal obstruction. Contrast enhanced CT scan shows large enhancing soft tissue density mass lesion in nasopharynx extending into posterior nasal cavity abutting the fossa of rosenmuller. Mass was excised using bipolar cautery and was removed in toto. HPR showed features of Nasopharyngeal angiofibroma with high fiber elements.

## CONCLUSION

Nasopharyngeal angiofibroma in adult male population is extremely rare. Adult male presenting with painless repeated bleeding should be investigated. One should always try to correlate symptoms with examination and take assistance of radiological investigations wherever necessary. As clinicians we are forced to research more on pathogenesis of this disease.

## KEYWORDS

Juvenile Nasopharyngeal Angiofibroma, Angiofibroma, Vascular Nasopharyngeal Tumor

## INTRODUCTION

Less than 0.5% of all head and neck neoplasms are JNA<sup>1</sup>. It is benign non-encapsulated highly vascular tumour with single endothelial cell lining of vessels without muscularis layer on histological examination<sup>2</sup>. Very few reports of it in women or men over 25 have been reported<sup>3</sup>. The origin is believed to be a vascular nidus in the posterolateral wall of nasal cavity near superior margin of sphenopalatine foramen<sup>4</sup>. Median age of onset of symptoms is 14 to 18 years.

Most common symptoms are painless episodes of epistaxis, nasal obstruction, headache and rest other symptoms depends on the spread of tumour which includes facial, ophthalmological

and neurological symptoms. Imaging techniques like Contrast Enhanced Computed Tomography and Magnetic Resonance Imaging are useful in the assessment of staging of tumour<sup>5</sup>. Preoperative embolization is preferred and along with angiography is done to reduce intraoperative bleeding.

Treatment methods, such as hormone therapy, cryotherapy, electrocoagulation, surgery combined with radiotherapy, and chemotherapy.

### CASE REPORT

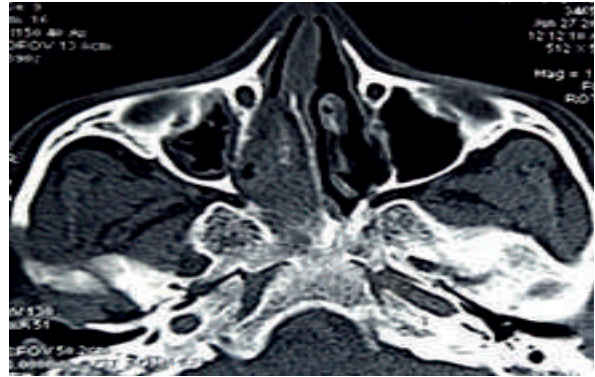
A 29 years old Male presented with complaint of bleeding from right nasal cavity for last five years associated with on and off nasal obstruction, there was no history of massive bleeding. There was no history of allergies, discharge, or trauma that was linked. Contrast enhanced CT scan shows large enhancing soft tissue density mass lesion in nasopharynx extending into posterior nasal cavity abutting the fossa of rosenmuller as shown in Fig1 and Fig 2.

Laboratory test did not show any abnormality and coagulation profile was within normal limits. Patient was planned for endoscopic approach excision mass was seen to be originating from lateral nasal wall in the region of sphenopalatine foramen which was extending to basisphenoid and posterior nasal septum as shown in Fig 3 and Fig 4. Mass was excised using bipolar cautery and was removed in toto as shown in Fig 5.

Mass with dimensions around 2.8\*3.7cms in greatest dimension and weighing 10.9gm & volume of 10mL was removed as a whole and send for histopathology, on macroscopic assessment the mass showed features of Nasopharyngeal angiofibroma with high fiber elements.

Still total surgical excision with radiotherapy is treatment of choice. Various approaches like Transpalatine, Mid Facial Degloving, Lateral Rhinotomy are described.

In 1996, Kamel popularized endoscopic trans nasal endoscopy route for surgery. Newer techniques like Gamma knife are now also used for small size tumor<sup>6</sup>.

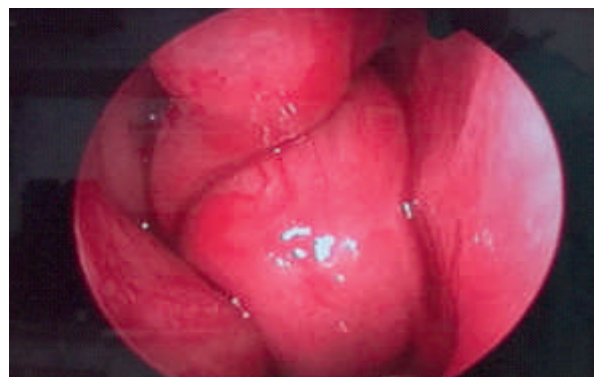


**Fig.1** Axial Section

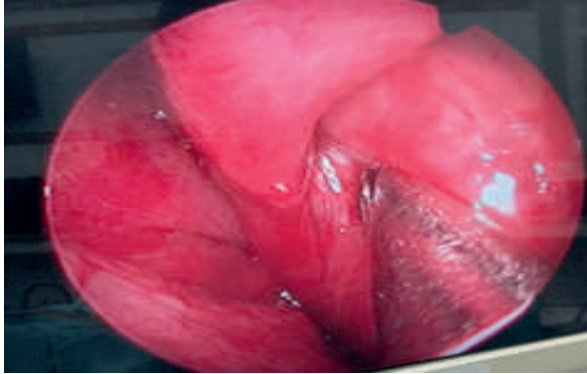


**Fig.2** Coronal section

Fig 1 and Fig 2- Contrast enhanced CT scan ( Fig1 axial section and Fig 2 coronal section) shows large soft tissue density mass lesion in nasopharynx extending into posterior nasal cavity

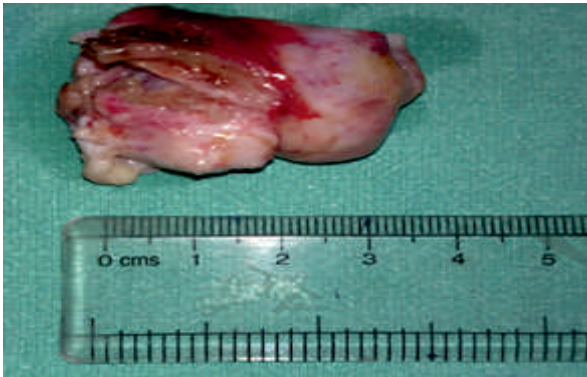


**Fig.3-** Right Nasal Cavity

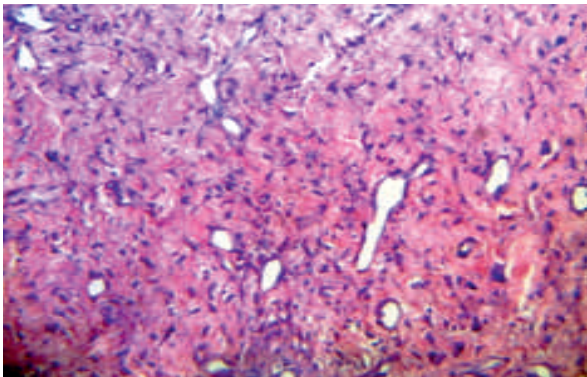


**Fig.4-**Left Nasal Cavity

Fig 3 and Fig 4 :DNE showing mass originating from lateral nasal wall in the region of sphenopalatine foramen



**Fig.5-** Excised mass



**Fig.6-** Histopathology

Histopathological examination large fibrocollagenous stroma containing multiple variable sized thin walled vessel lined by endothelial lining, few blood vessels showed stellate and staghorn pattern, suggestive of nasopharyngeal angiofibroma as shown in Fig 6.

## DISCUSSION

Nasopharyngeal angiofibroma is highly rare and has been reported in literature to occur in elderly persons. In a research, Lukomski et al. found that adults had a low incidence of this illness<sup>7</sup>.

These patients usually presents with nasal obstruction and epistaxis out of which one symptom usually predominate depending on the proportion of elements. There is also evidence of increased type of androgen receptor and successfully tumor regression after anti-androgen therapy<sup>8</sup>. Other theory by Andrade, because of its high frequency in teenage males is due to intranuclear accumulation of androgen receptors and by a high number of growth factors VEGF - endothelial growth factor, TGFβ - transforming growth factor beta<sup>9</sup>.

However, some theories also state it as a vascular malformation due to incomplete regression of the first branchial artery<sup>10</sup>. Some consider it to be vascular malformation originating from endothelial cells<sup>11</sup>.

The tumor having orbital and intra cranial extension are difficult to treat. Pathognomonic radiological sign of JNA is anterior bowing of posterior maxillary wall (Holman miller sign). Imaging-based diagnosis is based on growth pattern, hypervisualization following contrast enhancement, and the site of origin. The clinical history, radiographic examination, and histology report ultimately determine the diagnosis.

## CONCLUSIONS

Nasopharyngeal angiofibroma in adult male population is extremely rare. Adult male presenting with painless repeated bleeding should be investigated. Medical comorbidities like Hypertension, Haemophilia, Liver Disease, Thrombocytopenia, Platelet Dysfunction, Leukemia should be kept in mind. One should always try to correlate symptoms with examination and take assistance of radiological investigations wherever necessary. As clinicians we are forced to research more on pathogenesis of this disease.

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