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CASE REPORT : A CASE OF JUVENILE NASOPHARYNGEAL ANGIOFIBROMA.

ENT

Dr. Chandani D. Bhatt	M.S. ENT
Dr Ishita Shah	M.S. ENT
Dr. Niral Modi	M.S. ENT

ABSTRACT

Nasopharyngeal Juvenile angiofibroma is a benign , locally invasive and aggressive tumor, most commonly seen in adolescent male. It is highly vascular and comprises less than 1 % of all head and neck neoplasms. Here we are presenting a case of a 24 years old male patient presented to us with chief complaints of right sided nasal blockage for 2 to 3 years and occasionally blood stained watery nasal discharge.On further evaluation by clinical and radiological investigations ,the patient was diagnosed with Juvenile Nasopharyngeal Angiofibroma.

KEYWORDS

Juvenile Nasopharyngeal Angiofibroma, endoscopic resection

Case:

A 24-year-old male patient presented with chief complaints of right sided nasal blockage which was continuous, progressive, with occasional epistaxis since 2 years.There was no complaint of headache, fever or any other significant history. Past history reveals no episodes of nasal bleeding before two years but intermittent nasal discharge. Patient had no clinical pallor but mouth breathing was evident, especially during sleep. Endoscopic examination of the nose reveals that the mass was congested, compressible, filling the right inferior and middle meatus, extending posteriorly filling the right sided Choana and extending to nasopharynx obstructing the left choana as well .There was no tenderness on palpation and no external facial deformity was found.The oro-dental examination was normal. General examination revealed no significant abnormality. Routine haematological investigations were found to be within normal limits. Computed tomography with angiography revealed a well defined soft tissue density (HU- 40) lesion of size(42 x 36 x 39) mm arising from right sphenopalatine foramen with erosion of the floor of right sphenoid sinus superiorly and extending to it and engulfing the right inferior and middle turbinates and causing erosion of posterior superior part of bony nasal septum medially and reaches upto left inferior turbine along with complete blockage of bilateral posterior Choana with no any intracranial or intraorbital extension .The above findings suggestive of nasopharyngeal Juvenile angiofibroma.

Surgical procedure: The main goal of surgery was to decrease the amount of blood loss as much as possible by the right approach.Hence, endoscopic resection was done using coblater and clamping the main feeding vessel which was sphenopalatine artery.

To reach sphenopalatine artery, Modified Denker's approach was taken and posterior wall of right maxillary sinus was removed and all branches of sphenopalatine artery were identified and ligated and controlled hemostasis was achieved and 4 units of whole blood was given intraoperatively and rest of tumor was resected with the help of coblater and removed from nasopharynx . Post- operative nasal packing was done.

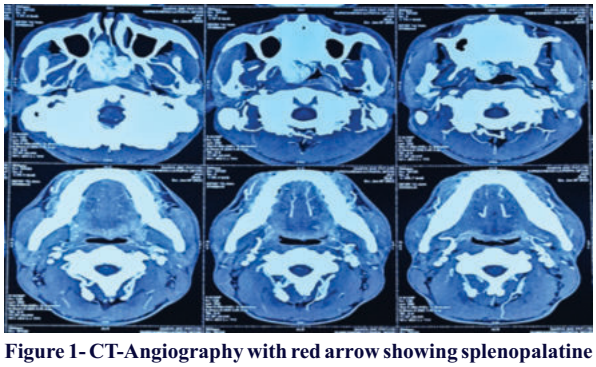

Figure 1- CT-Angiography with red arrow showing splenopalatine artery supplying the juvenile nasopharyngeal angiofibroma.

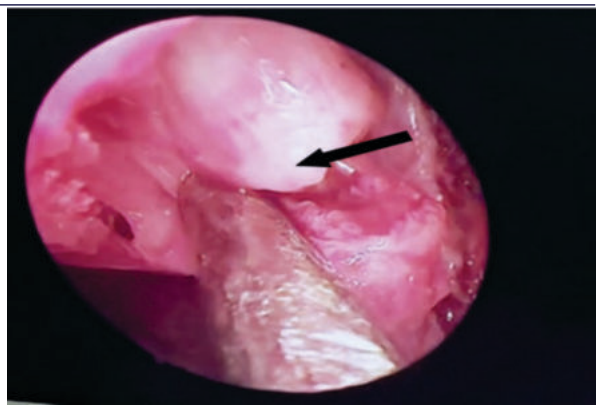

Figure 2- Nasal endoscopy with black arrow showing mass (juvenile nasopharyngeal angiofibroma).

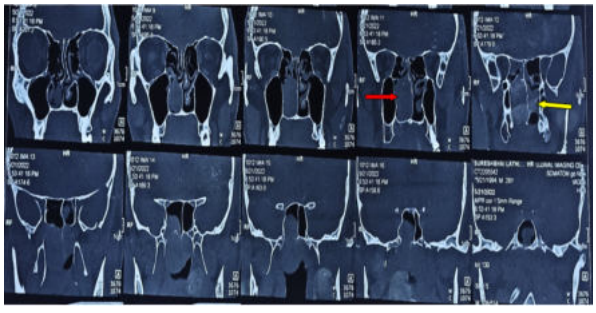

Figure 3- CT-Scan of Paranasal Sinuses with axial view : Red arrow showing mass(Juvenile nasopharyngeal angiofibroma) in right nasal cavity and yellow arrow showing extension of mass in nasopharynx blocking the left sided Choana posteriorly.

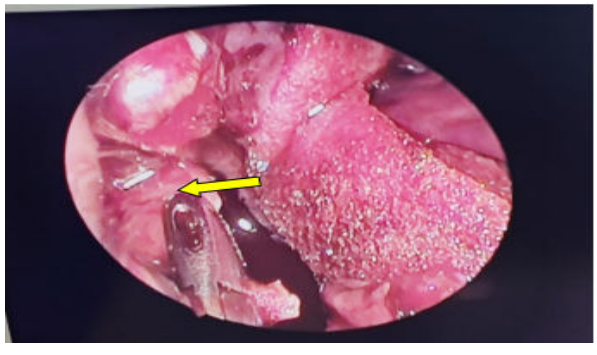

Figure 4- Yellow arrow showing removal of right sided inferior turbinate by Modified Denkers Approach via nasal endoscopy.

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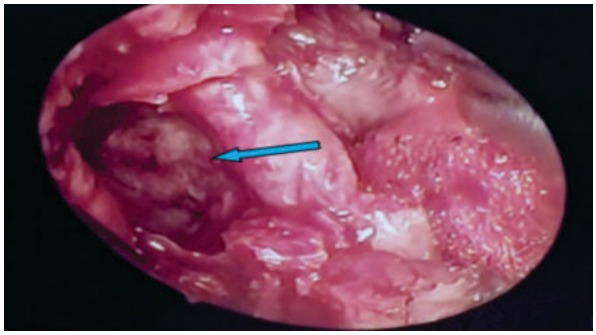

Figure 5- Nasal endoscopic view- Blue arrow showing exposed posterior wall of right sided maxillary sinus .

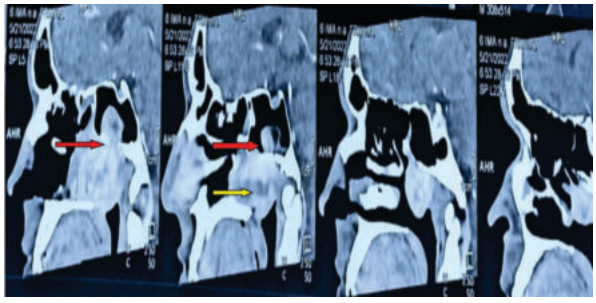

Figure 6- CT Scan of paranasal sinus with Sagittal view : Yellow arrow showing the mass(Juvenile nasopharyngeal angiofibroma) and Red arrow showing extension of mass into sphenoidal sinus respectively.

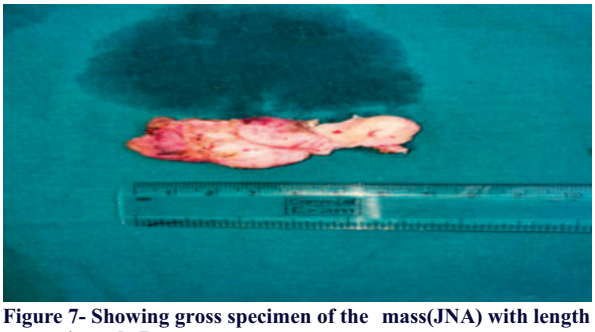

Figure 7- Showing gross specimen of the mass(JNA) with length approximately 7cm.

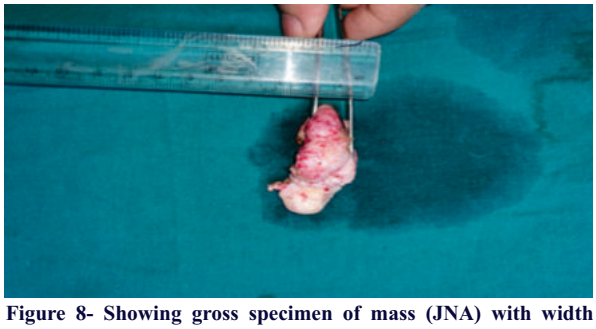

Figure 8- Showing gross specimen of mass (JNA) with width approximately 2.5 cm.

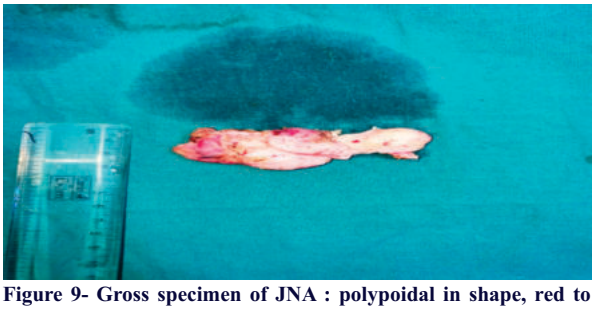

Figure 9- Gross specimen of JNA : polypoidal in shape, red to pinkish yellow in colour, irregular surface and spongy appearance.

DISCUSSION:

Juvenile Nasopharyngeal Angiofibroma (JNA) is a rare tumour presenting mostly in adolescent males. The average age of presentation of this tumour is 14 years. The reason for exclusivity in males is hypothesised as androgen dependence of the tumour. JNA originates within the sphenopalatine foramen, and further spreads locally into the pterygopalatine fossa, nasal cavity and paranasal sinuses. Patients usually present with a triad of progressive nasal obstruction, recurrent epistaxis and mass in the nasal cavity and nasopharynx. It has potency to cause proptosis, affecting both the orbit and with intracranial extension, eventually leading to cranial nerve palsies and loss of vision . However, advanced stages of the tumour can present as locally invasive causing facial dysmorphism, Eustachian tube obstruction causing conductive hearing loss, and diplopia that occurs secondary to erosion of superior orbital fissure and due to third and sixth nerve palsy, intraorbital extension leading to proptosis, or rarely anosmia, recurrent otitis media and eye pain. The most common site of initiation is posterolateral wall of the nasopharynx, precisely at the trifurcation of the sphenoidal process of the palatine bone, the roof of the pterygoid process and the horizontal process of the vomer .

The origin of JNA is from a hamartomatous nidus of vascular tissue in the region of the sphenopalatine foramen, which is being stimulated during early pubertal period by endogenous androgenic hormones. Radiographic findings suggestive of the disease is the anterior bowing of the posterior wall of maxillary sinus of same side (Holman Miller Antral Sign) which is considered as pathognomonic . Diagnosis of an angiofibroma essentially includes preoperative CT-scan, with MRI and Magnetic Resonance Angiography (MRA). MRI is even more important postoperatively to show any residual or recurrent tumour and monitor the effects of radiotherapy.Recurrence results from deeper extension of the tumour resulting in residual tissue after surgery. Commonly, the vascular supply to JNAs is primarily from distal Internal Maxillary Artery (IMA) branches, particularly the sphenopalatine, descending palatine and posterior superior alveolar branches . JNAs may also be supplied by the ascending pharyngeal artery. Histologically, JNAs originate from myofibroblasts. The tumour usually spreads submucosally and is non-encapsulated. It is composed of intricate mixture of stellate and staghorn blood vessels with variable vessel wall thickness ranging from single layer of endothelium to variable smooth muscle coat, irregular fibrous stroma (loose, oedematous to dense, acellular) positive stains include c-Kit/CD117, androgen receptor (75%) .Surgery remains the main stay in management . Endoscopic nasal microsurgery is a viable approach with prospectively lesser postoperative morbidity and intraoperative blood loss, but it is advocated only for limited lesions. But for larger tumours, approaches such as transpalatal, lateral rhinotomy and midfacial degloving have been suggested. Preoperative embolisation of tumour is recommended with all of these approaches to limit the intraoperative blood loss .The advantage of endoscopic approach over open surgery is a shorter mean duration of surgery. Complications of the surgical approach used in our case were mild midfacial pain following two months after surgery that gradually subsided and nasal synechiae which was managed with intranasal stents and nasal douching.

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