

# PATTERNS OF VASCULARIZATION AND SURGICAL APPROACH IN JUVENILE NASOPHARYNGEAL ANGIOFIBROMA: A CASE SERIES

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## ABSTRACT

### BACKGROUND :

Juvenile Nasopharyngeal Angiofibroma affecting exclusively young, adolescent males ; is a highly vascular tumour arising close to the sphenopalatine foramen, having locally invasive potential. Radiographic imaging is required to determine the extent and vascularity of the tumour, which aid in planning the surgical modality of the tumour.

### MATERIALS AND METHOD

A retrospective review of all 11 patients undergoing surgical resection of JNA, from the time period between July 2020 and February 2023 in the Department of ENT , Assam medical college was carried out. CT and MRI Angiography was done to know the extent and vascularity of the tumour to determine the staging and planning of the surgical approach .

### RESULTS

All 11 cases were male with a mean age of 16 years. Majority of the cases received multiple feeding vessels of which 9 cases received primary supply from ipsilateral Internal Maxillary Artery and the remaining 2 cases received primary supply from Ascending Pharyngeal Artery.

Both endoscopic and open approaches were carried out depending on the Radkowski staging of the tumour. 6 cases (54.54%) underwent endoscopic approach , 4 cases (36.36%) underwent open approach and one case was operated upon by Combined endoscopic and open approach.

### CONCLUSION

Successful management of JNA tumour depends on a comprehensive and meticulous pre-operative clinical and radiological evaluation and correlation, in order to effectively deal with the myriad patterns of vascularisation of the tumour, and thereby minimise the intraoperative blood loss, and enhance the prognosis of the patients.

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## INTRODUCTION

Juvenile Nasopharyngeal Angiofibroma (JNA) is a rare, unique, benign tumour of the nasopharynx, occurring almost exclusively in young, adolescent males. It is a vascular, fibrous tumour with locally invasive potential. <sup>1</sup>Accounting for around 0.5% of all head and neck tumours, upto 20% of these tumours present with intracranial extension.<sup>2</sup>

The juvenile nasopharyngeal angiofibroma was formerly considered to originate from the nasopharynx. However, the recent theory is that JNA originates from the postero-lateral wall of the nasal cavity, close to the superior aspect of the sphenopalatine foramen where the posterior part of the middle turbinate attaches<sup>3</sup>. It

is a locally invasive tumour which expands to nasopharynx and nose, and subsequently through the sphenopalatine foramen to the infratemporal fossa, and to the middle cranial fossa through the skull based foramina and superior orbital fissure<sup>4</sup>.

Pre-operative CT scan serves as an important tool to know the extent of JNA. Angiographic evaluation demonstrates heterogenous vascularization patterns of the tumour, the most common feeder being the internal maxillary artery. <sup>2</sup>Other feeding vessels of the tumour include the ascending pharyngeal artery, the ascending and descending palatine arteries, the sphenopalatine artery, the posterior superior alveolar branches and, less commonly, the pterygoid canal (vidian) artery, the ophthalmic artery. Bilateral supply is possible in JNAs due to which bilateral carotid systems angiography is

recommended by some authors.. Angiography is more of a therapeutic tool rather than a diagnostic tool to perform embolisation as an adjunct to surgical excision.<sup>5</sup>

In an adolescent male, unprovoked, painless, recurrent episodes of profuse epistaxis suggest juvenile nasopharyngeal angiofibroma, until proven otherwise.<sup>6</sup> Unilateral nasal obstruction, epistaxis, and lobulated mass which extends into the nasopharynx, constitutes the classic characteristic triad for diagnosis.<sup>7</sup> Anterior rhinoscopy and endoscopic examination reveals a smooth, rubbery, lobulated, polypoid, yellowish to red/purple, hypervascularized mass arising from behind the middle turbinate with variable size, extending into choana and nasopharynx.<sup>8</sup>

Amongst the various proposed theories regarding the origin of the tumour, the preponderance amongst adolescent males concludes the presence of androgen receptors, strongly suggestive of hormonal influence on growth of the tumour.<sup>9</sup>

Different modalities of treatment include surgery, radiotherapy, electrocoagulation, oestrogen therapy, embolization, injection of sclerosing agents and cryotherapy. Due to the negative effects of radiotherapy on craniofacial growth and its potential carcinogenic effects which limits its use, surgery is considered the gold standard in the management of JNA<sup>10</sup>.

Several surgical approaches are currently available, ranging from microendoscopic techniques, to open techniques like transpalatal approach, lateral rhinotomy, medial maxillectomy, midfacial degloving and infratemporal fossa resection, or a combination of the above approaches, like Denker's (transpalatal+ transantral), Sardana's (sublabial + transpalatal), Triple approach Hiranandani (transpalatal+ lateral rhinotomy + Caldwell Luc) depending upon the extent of the tumour.

Our study aims to characterize the patterns of vascular supply to the JNA and study the various surgical approaches that have been employed in the management of the aforementioned tumour.

**MATERIALS AND METHODS**

A retrospective review of all 11 patients undergoing surgical resection of JNA, from the time period between July 2020 and February 2023 in the Department of ENT, Assam medical college were studied. A retrospective, radiographic review of our institutional experience of JNA cases to provide detailed characterization of vascular pattern and thereafter, the surgical approaches have been studied.

The surgical approaches had been planned as per the radiographic extensions of the tumour and comprised of both open and endoscopic techniques as mentioned above. The surgically excised tumour specimens had been sent for histopathological examinations, and all the 11 samples tested positive for JNA. The patients have now been retrospectively reviewed on their subsequent follow up visits.

**PATIENT CHARACTERISTICS**

We identified 11 patients who underwent operative management of JNA. Ten cases were primary presentations and one was referred to our medical center for management for recurrence (following surgery done twice). One patient was lost to follow-up at the time of this study review.

The presenting patient characteristics and the line of management have been summarised and tabulated below.

Characteristics	Number(n)	Percentage(%)
<b>Number of patients</b>	11	100
<b>Sex</b>	Male	100
<b>Age</b>		
Mean	16 years	
Range	11- 26 years	
<b>Presenting symptoms</b>		
Epistaxis	9	81.81
Nasal obstruction	6	54.54
Facial deformity	5	45.45
Nasal discharge	2	18.18
Anosmia	1	9.09
<b>Laterality</b>		
Right	6	54.54
Left	5	45.45
<b>Imaging</b>		
MRI	11	100
CT	10	90.90
Both	10	90.90
<b>Surgical Approach</b>		
Endoscopic	6	54.54
Open	4	36.36
Combined	1	9.09

Fig 1: Table Representing Clinical Characteristics

**Patterns Of Vascularity and Management Approaches**

Serial no.	Age(yrs)	Radkowski staging	Approach	Estimated blood loss	Follow up (months)
1	15	Ib	Endoscopic medial maxillectomy	500	12
2	16	Ia	Transpalatal	350	12
3	15	Ia	Endoscopic Transnasal excision	450	12
4	16	Ic	Lateral rhinotomy	3000	Loss to follow up
5	17	Ic	Combined(Endoscopic medial maxillectomy with Lateral Rhinotomy with Right ECA ligation)	3000	15
6	11	Ib	Endoscopic medial maxillectomy	1000	24
7	14	Ic	Lateral rhinotomy	1500	10
8	26	Ib	Endoscopic transnasal excision (coblation assisted)	500	2
9	16	Ib	Lateral rhinotomy with medial maxillectomy	1000	1
10.	16	Ia	Endoscopic transnasal excision	500	10
11	16	Ib	Endoscopic medial maxillectomy	750	12

Fig 2: Table Representing Management Approaches

**DISCUSSION**

All patients of Juvenile Nasopharyngeal Angiofibroma, who attended the Department of ENT and were subsequently operated during the study period, were included in this retrospective study. There was a total of 11 patients of JNA who were operated during this study period.

All the cases in our study were exclusively males ranging in the age group of 11 to 26 years, the mean age being 16 years. In similar case

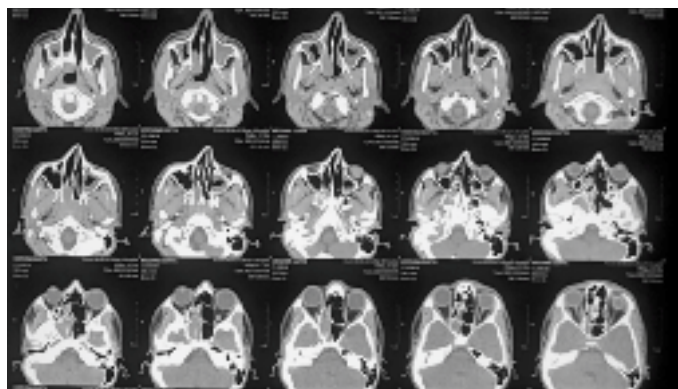
series done by Jonathan B. et al, the mean age was around 16 years, which is comparable to our study.<sup>2</sup> Almost all the cases (n=10) were primary presentations of the disease, however one case (aged 26 years) was a post-operative recurrent case, who had undergone surgery twice for excision of JNA via transpalatal (aged 13 years), and transnasal (aged 14 years) approach on the respective two occasions.

Around 81.81% patients (n=9) had presented with the chief complaints of epistaxis which was spontaneous in onset, painless, recurrent and profuse in amount. Furthermore, patients also presented with complaints of nasal obstruction (54.54% cases, n=6); facial deformity (45.45% cases, n=5), blood-stained nasal discharge (18.18%, n=2) and anosmia (9.09%, n=1). It is noteworthy to mention that the recurrent case did not present with epistaxis, instead he presented with nasal obstruction and blood stained nasal discharge.

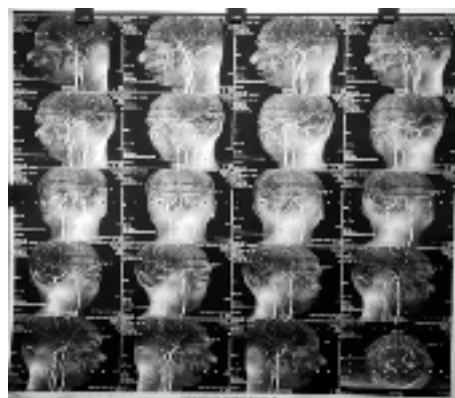
The cases presented almost comparable laterality, with mild preponderance to the right side (54.54%, n=6).

The feeding vessels of the tumour were identified with the aid of MRI Angiography, which coupled with CT scan, also helped in determining the extent of the tumour and subsequent staging. On MRI Angiography, majority of the cases were found to receive multiple feeders from the External Carotid Artery (ECA) branches. 9 cases (81.81%) received primary supply from ipsilateral Internal Maxillary Artery. The remaining 2 of 11 tumors (18.18%) received primary supply from Ascending Pharyngeal Artery.

On the basis of radiological imaging, the cases were staged as per Radkowski staging, and the extent of spread of the tumour was determined which helped in the planning of the surgical approaches. All the cases belonged to Stage I and stage II, with three cases extending to the infratemporal fossa (Stage Iic).



*Fig 3a: In the above CT scan, the JNA tumour is noted to involve the posterior part of right nasal cavity, extending to nasopharynx and right pterygopalatine fossa (Radkowski stage-IIb)*

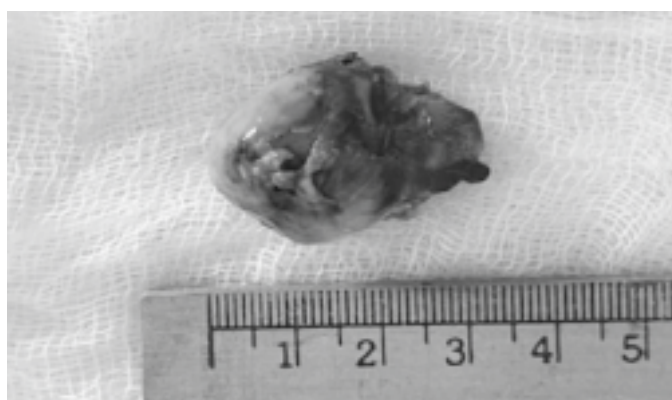


*Fig 3b: Dynamic MR angiogram reveals feeders from right internal maxillary artery supplying the tumour.  
Fig 3: CT scan and Dynamic MR Angiogram, of the nose and paranasal sinuses of a 16 year old JNA patient (Radkowski Stage 2b)*

With the advancements of endoscopic surgical techniques, the management of JNA tumors has evolved significantly. The relatively smaller tumours, belonging to stage Ia, had undergone transnasal endoscopic excision, and in one case, the tumour was excised from the nasopharynx using transpalatal approach. The Ib staged tumours had undergone endoscopic medial maxillectomy. In one redo case, staged Ib, endoscopic transnasal excision (coblation assisted) was performed.

For the tumours invading to the Pterygopalatine Fossa (Stage Iib), two cases underwent resection via endoscopic medial maxillectomy approach and one was excised by open approach via lateral rhinotomy approach.

For tumours encroaching into the Infratemporal Fossa (Stage Iic), mainly open approaches were performed, wherein one case was approached by combined method, including both endoscopic medial maxillectomy with lateral rhinotomy with ligation of right sided External Carotid Artery.



*Fig 4: Post-operative surgically excised specimen of a nasopharyngeal angiofibroma, of the 16 year old male patient, whose CT scan and MRI angiogram have been demonstrated above. The tumour was excised by right sided lateral rhinotomy with medial maxillectomy.*

The amount of blood loss was determined intraoperatively and maximum amount of blood loss was found to be 3000 ml, which occurred in two cases of stage IIc undergoing open approach. In one case, ligation of Right ECA had to be performed to achieve adequate hemostasis.

The surgically excised specimens were all sent for histopathological examination for confirmation of the angiofibroma.

All the patients had been followed up post-operatively, with the first follow up being at one week interval. The subsequent follow-up visits were done at 3 month intervals, continuing upto two years.

## CONCLUSION

Our review suggests that the JNA tumors, being highly vascular tumours require a meticulous pre-operative planning for excision of the same.

Due to the varied patterns of vascularisation, angiography is mandatory to know the feeding vessel and determine the extent of the tumour, on the basis of which the treatment modality is decided by the surgeon. Furthermore, knowledge of the feeders pre-operatively, helps in effectively reducing the amount of intra-operative blood loss, and henceforth decrease the morbidity, and enhance the prognosis of the patients.

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