ORIGINAL ARTICLE

# Nasopharyngeal Angiofibroma — Staging and Selecting a Surgical Approach: Changing Trends

**[1]Ashok K Gupta, [2]Sandeep Bansal**

[1]Professor and Head, Department of Otolaryngology and Head and Neck Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh, India

[2]Assistant Professor, Department of Otolaryngology and Head and Neck Surgery, Postgraduate Institute of Medical Education and Research, Chandigarh, India

**Correspondence:** Ashok K Gupta, Professor and Head, Department of Otolaryngology (Unit II) and Head and Neck Surgery Postgraduate Institute of Medical Education and Research, Sector-12, Chandigarh-160012, Phone: 91-172-2747837 Fax: 91-172-2744401, 2747837 e-mail: drashokpgi@hotmail.com

**Abstract**

*Objective:* The purpose of this study was to propose a new staging system which would allow for an accurate preoperative tumor assessment, appropriate surgical planning and to look for any changing trends in the surgical management of JNA.

*Study design and methods:* Retrospective chart review of 91 patients operated between Jan.1998 to Dec. 2007 at the Postgraduate Institute of Medical Education and Research, Chandigarh. The tumor location and extent was based on preoperative imaging reports and operative notes to review the staging system and surgical approaches.

*Results:* All patients were initially staged according to the modified sessions staging system. The revised staging system led to the reclassification of 36 (39.6%) in stage I A, I B and II A in the old staging system to stage I according to the new staging. Before 2003, none of the patients had undergone an endoscopic excision of the JNA, whereas 32 (35%) of patients underwent an endoscopic excision in group II (after 2002) with acceptable recurrence rates.

*Conclusions:* The choice of the surgical approach should be based on precise tumor location and in young patients the approach should be tailored to minimize potential for facial growth retardation. We recommend endoscopic resection for tumors confined to nasopharynx, nasal cavities, sinuses and minimal extension to PMF. Excision via lateral rhinotomy and its extensions and modifications is recommended for tumors having significant involvement of pterygomaxillary fossa, infratemporal fossa, cavernous sinus or minimal intracranial extension according to the surgeon's comfort and expertise.

**Keywords:** Angiofibroma, nasopharyngeal tumors, endoscopic excision, maxillofacial development.

## INTRODUCTION

Juvenile nasopharyngeal angiofibroma (JNA) is a highly vascular and locally aggressive tumor seen almost exclusively in adolescent males associated with significant morbidity and occasional mortality. JNA accounts for only 0.05% of all head and neck tumors.[1] Histologically they are benign, nonencapsulated highly vascular tumors[2] which originate most frequently in the region of sphenopalatine foramen.[3] The tissue of origin is uncertain. Although JNA's may occur at any age between infancy and old age, they are commonly diagnosed between ages of 14 and 25 years.[4] These patients usually present with a painless, unilateral nasal obstruction and epistaxis. Other symptoms would depend upon the direction and spread of the tumor. During evaluation, contrast enhanced computed tomography (CECT) and magnetic resonance imaging (MRI) can reliably assess tumor extent. Preoperative angiography is helpful in the evaluation of feeding vessels and allows embolization of the tumor.

Surgery remains the treatment of choice for JNA, although radiotherapy, hormonal therapy, cryotherapy, electrocoagulation and chemotherapy have all been described in literature. Many surgical approaches have been used, including transpalatal, transzygomatic, trans-mandibular, transantral approach, lateral rhinotomy, midfacial degloving and concomitant craniotomy.[5] In 1996, Kamel described an endoscopic transnasal procedure for confined lesions, leading way for endoscopic transnasal route to be popularized.[6]

Since its first description, the staging and treatment of JNA has undergone an evolution. There have been different staging systems (Chandler's, Sessions, Andrews, etc.)[3] given for JNA, but none of these staging systems gives an idea about the surgical options for a particular stage.

In young patients, the potential for craniofacial surgery to affect facial growth should also be considered when planning its surgical approach.[7,8]

This retrospective analysis was undertaken to propose a new staging system which can help to select an ideal surgical approach and to look for any changing surgical trends in the surgical management of JNA.

The proposed revised staging system would allow for accurate preoperative tumor assessment, appropriate surgical planning and would help in interinstitutional comparison of treatment outcomes.

## MATERIALS AND METHODS

The medical records of 91 patients with histologically proved nasopharyngeal angiofibroma between Jan. 1998 to Dec. 2007 operated by the corresponding author at Department of Otolaryngology, Postgraduate Institute of Medical Education and Research (PGIMER), Chandigarh, India were retrospectively reviewed. A minimum follow-up of 12 months was necessary for inclusion into the study. The tumor location and extent was based on preoperative imaging findings and operative notes.

Available preoperative contrast enhanced computed tomography and magnetic resonance imaging were revaluated to confirm the documented staging. All patients were initially staged according to the modified sessions staging system (Table 1). Modifications were made in the staging of JNA based on the result of this retrospective analysis.

## RESULTS

Ninety-one patients were treated at Department of Otolaryngology, PGIMER, Chandigarh between Jan. 1998 to Dec. 2007. All patients were males within the age range of 9 to 28 years with a median age of 14 years. Ten of these patients who had been surgically treated elsewhere and presented with recurrence were included.

**Table 1:** Staging of juvenile nasopharyngeal aniofibroma (Modified sessions, et al)

| | |
|---|---|
| I A: | Limited to nose and/or nasopharyngeal vault |
| I B: | Extension into ≥ 1 sinus |
| II A: | Minimal extension into PMF |
| II B: | Full occupation of PMF with or without erosion of orbital bones |
| II C: | ITF with or without cheek or posterior to pterygoid plates |
| III A: | Erosion of skull base-minimal intracranial |
| III B: | Erosion of skull base-extensive intracranial with or without cavernous sinus |

**Table 2:** Different site of involvement by the tumor

| *Site* | *No. of patients* | *% patients* |
|---|---|---|
| Nasal cavity | 77 | 85 |
| Nasopharynx | 88 | 97 |
| Pterygopalatine fossa | 81 | 89 |
| Sphenoid sinus | 67 | 73 |
| Ethmoids | 61 | 67 |
| Infratemporal fossa | 57 | 63 |
| Cavernous sinus | 24 | 26 |
| Middle cranial fossa | 14 | 15 |
| Total | 91 | |

Follow-up ranged between 1 year and 4 and 1/2 years (median, 22 months). Preoperative radiography(CT and MRI) the tumor location and extent, and operative notes are summarized in Table 2.

### Radiological Investigations

Contract enhanced computerized tomography (CECT) and magnetic resonance imaging (MRI) were done to determine tumor size and location (Fig. 1). All the patients who had been operated outside and had presented with residual/ recurrence to us, had undergone a gadolinium enhanced MRI. Angiography with preoperative embolization of the principal feeding vessel was performed in 46 patients.

The remaining 45 patients who were not planned for Embolization underwent a CT angiography to know the status of the feeder vessels. On the basis of preoperative radiology and operative notes, the tumor location and extent is summarized in Table 3. All the patients underwent post-operative CT scan to look for any residual tumor at 3 months postsurgery (Fig. 2).

## STAGING

Initial staging was based on the modified sessions staging system. There was no patient in the stage I A, 23% patients in stage I B, 17% patients in II D, 9% in II B and 38% in stage II C. 14% patients had stage III A and III B disease (Table 3).

The revised staging system led to a reclassification of 36 (39.6%) patients out of the 91 patients. These 36 patients in stage I A, I B and II A in the old staging were reclassified in stage I according to the new staging.

## SURGICAL APPROACHES

Out of the 91 patients in the series, 85 patients underwent surgery where as the remaining 6 patients due to extensive intracranial involvement were treated with radiotherapy.

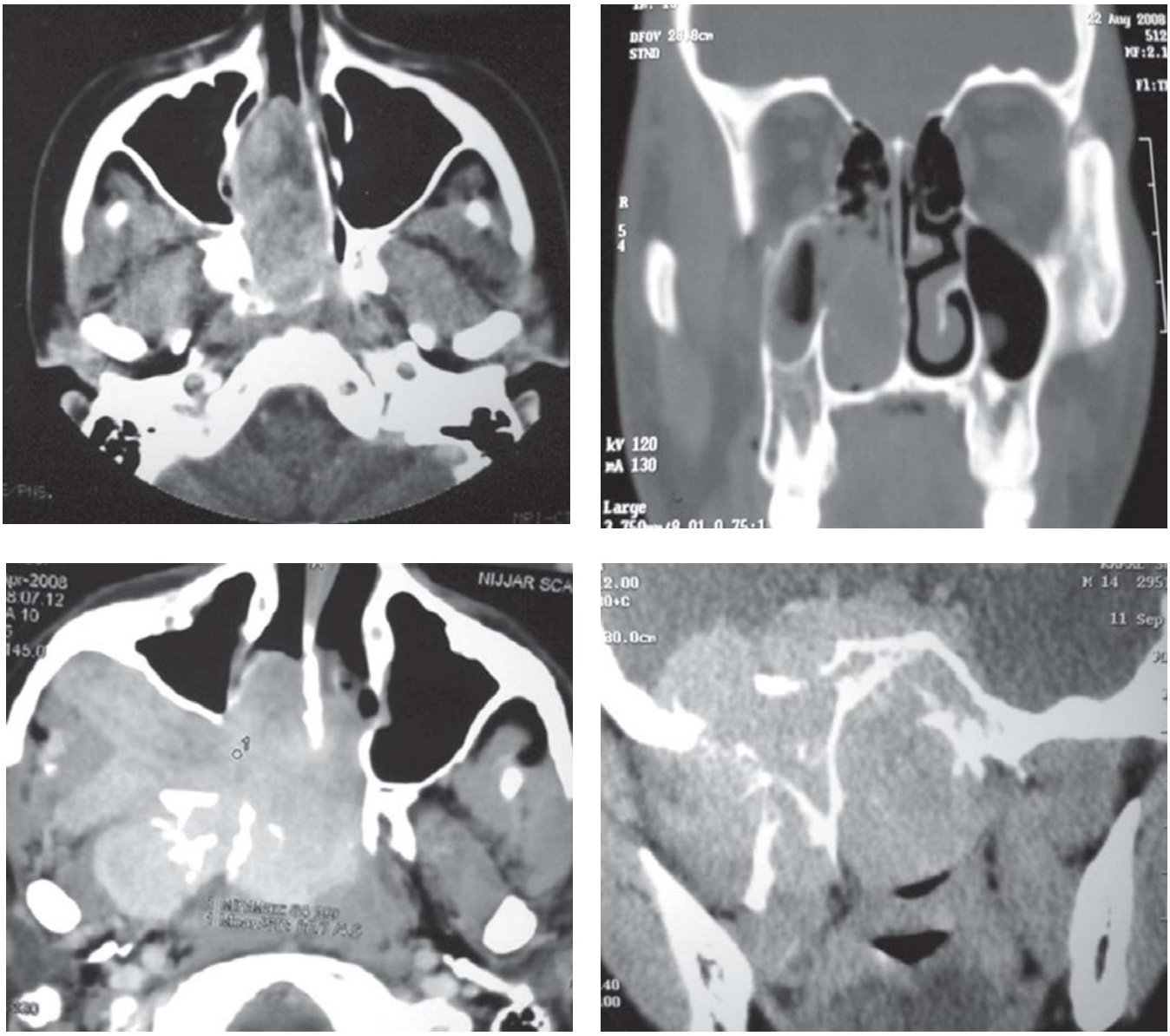


**Figure 1:** Stage I, II, III A, III B (starting from right upper corner clockwise)

**Table 3:** Staging system

| *No. of patients (%)* | *Stage* | *Proposed staging* |
|---|---|---|
| 0 (0) | I A | |
| 21 (23) | I B | I |
| 15 (17) | II A | |
| 8 (9) | II B | II A |
| 34 (38) | II C | II B |
| 5 (5) | III A | III A |
| 8 (9) | III B | III B |

For the purpose of comparison and due to the changing trends in the surgical management, the results for these 85 patients have been divided into 2 groups according to the proposed new staging. group I (comprising of patients between 1998-2002) and group II (patients presenting between 2003-2007). The surgical approaches that were employed in each group have been summarized in Table 4.

Before 2003, none of the patients had undergone an endoscopic excision of the JNA, whereas 32 (35%) of patients underwent endoscopic excision in group II.

Transpalatal route was employed in 4 patients in Group I and 2 patients in group II. 11 patients in group I and 6 in group II had undergone excision via lateral rhinotomy .

Excision via Weber-Fergusson approach was done in 19 patients in group I and in 7 patients in group II.

Craniotomy was done in 3 patients in group I where as only 1 patients had undergone craniotomy in group II.

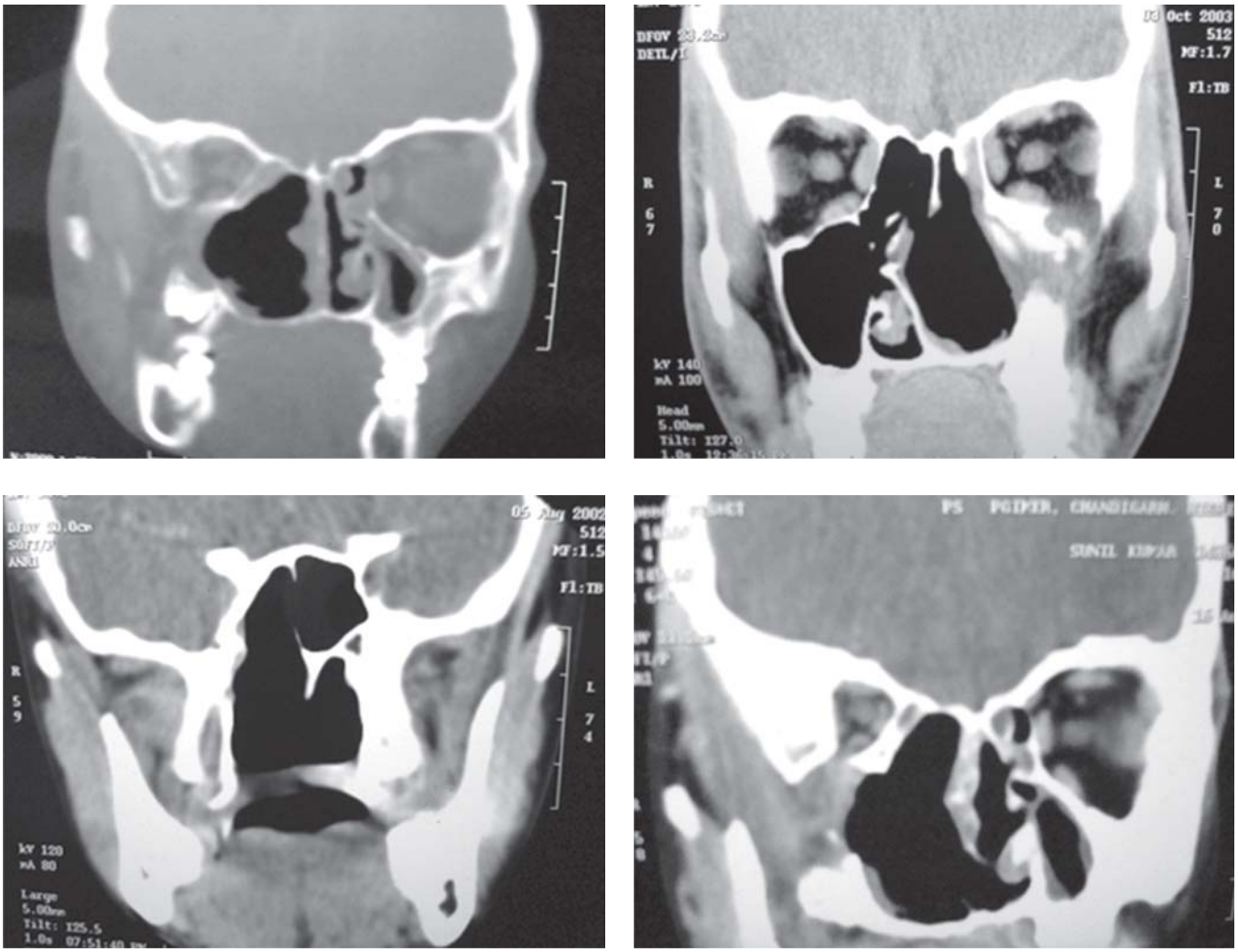

**Figure 2:** Postoperative scans

## COMPLICATIONS

None of the patients had postoperative cerebrospinal fluid leak. One patient had transient diminished vision post-operatively but resolved with intravenous steroids within 48 hours. Two patients required endoscopic dacryocysto-rhinotomy postoperatively.

## RECURRENCES

In group I, 12/37 (32.4%) patients had recurrences, out of which 2 had undergone excision by transpalatal approach. The tumor had involved the pterygomaxillary fossa (PMF). The 3 patients who had undergone excision via lateral rhinotomy presented back with recurrence in the infratemporal fossa. A revision surgery was done and the tumor removed completely. All these 5 patients are free of disease.

Four patients presented back with recurrence involving the infratemporal fossa (ITF) and orbit and 1 patient had extensive intracranial extension. These recurrences in the former 4 patients had been operated by an infratemporal approach. These patients were free of disease for the next 30 months but then were lost to follow-up. Out of the 3 patients who had undergone a craniotomy, two patients came back with tumor invading the middle cranial fossa and extensive involvement of the cavernous sinus. In group II, i.e. after 2002, 5 patients out of the 32 who had undergone an endoscopic transnasal excision of the tumor recurred. Two out of these were managed again by an endoscopic excision while 2 underwent an external procedure. Two out of the 2 patients operated via a transpalatal route had recurrence. This patient also underwent an endoscopic transnasal removal of tumor and is free of disease.

Only 2 patients out of 13 who had undergone a medial maxillectomy via a lateral rhinotomy or a Weber-Fergusson approach had recurrences in group II. The only patient who underwent craniotomy for stage IIIB in group II had recurrence. He was referred for radiotherapy and was lost to follow-up after 2 years.

The 6 patients who were sent for primary radiotherapy due to a very extensive intracranial extension had decrease in size of the tumor after radiotherapy and 3 out of them are still alive with persistent of disease with a follow-up ranging between 2 ½ years to 4 ½ years. The other 3 have been lost to follow-up.

## DISCUSSION

Juvenile nasopharyngeal angiofibroma is a benign, vasoformative neoplasm arising almost exclusively in adolescent males from a broad base on the posterolateral wall of the nasopharynx. The occurrence of these tumors almost exclusively in adolescent males supports the hypothesis that an alteration of the pituitary androgen estrogen axis contributes to the pathogenesis of JNA.[9] But many studies of the pituitary gonadal axis in these patients have failed to identify any endocrinologic abnormality.[10,11]

The most widely accepted theory of origin of JNA is that the tumor is derived from the embryologic chondrocartilage during the development of the cranial bones. The specific point of origin is at the superior margin of the sphenopalatine foramen formed by the bifurcation of the palatine bone, horizontal ala of the vomer and the root of the pterygoid process.[11,12]

The Modified Session's classification is a systematic classification but we cannot have a clear cut demarcation about the treatment modality to be followed for each stage.

To facilitate this, we have revised this classification (Table 3) so that the stages I A, I B and II A (tumors involving the nose, nasopharyngeal vault, with extension into sinuses and with minimal extension into PMF) are classified as stage I, stage II B (full occupation of PMF with or without erosion of orbital plates) becomes II A, II C (extension into ITF and pterygoid plates) becomes II B. The implications of skull base erosion in JNA are different from those in malignant lesions. Angiofibromas do not invade the skull base by cellular infiltration as do malignant carcinomas but rather lead to local bone resorption.

We revised the stage III to differentiate between isolated skull base erosion with minimal intracranial involvement into stage III A and an extensive intracranial tumor and cavernous sinus involvement into stage III B. Such a staging system would also help interinstitutional comparison of treatment outcomes as we have proposed a surgical approach deemed fit for a particular stage.

The goal of surgery for JNA is complete tumor resection with minimal morbidity. Due to complexity of the anatomy of skull base and the vascular nature of the tumor, complete resection requires good surgical exposure with direct visualization of both the tumor and the tumor bed. Along with this, the surgical approach and technique should also take into account the fact that the majority of JNA patients are young and that the growth of the male craniofacial skeleton continues until at least 20 years of age.[8,13]

Due to the retrospective nature of this study and a short (median 22 months) follow-up of our patients, it is not possible to draw conclusions about the potential for the various surgical approaches to affect facial growth. A number of studies have examined the factors that may cause growth restriction of mid face, which include elevation of soft tissue[8] and periosteum[14] from the midface, dissection of the mucoperiosteum of the palate,[8] ethmoidectomy,[15] nasal septal trauma[7] and facial osteotomies.[7,8,14]

Therefore, soft tissue dissection from the facial bones, and the number of osteotomies should be kept to a minimum and should be designed in such a way so as to avoid suture lines.

As discussed earlier the surgical approach chosen should allow for an adequate exposure of the tumor. Transpalatal approach was indicated for tumors involving only nasopharynx which can now be more effectively managed by endoscopic transnasal approach; hence transpalatal approach can be considered obsolete.

**Table 4:** Surgical approaches and recurrences

| *Approach* | *Group I 1998-2002* | *Group I 1998-2002 (Recurrence)* | *Group II 2003-2007* | *Group II 2003-2007 (Recurrence)* |
|---|---|---|---|---|
| Endoscopic transnasal approach | 0 | 0 | 32 | 5 |
| Transpalatal approach | 4 | 2 | 2 | 1 |
| Excision via lateral rhinotomy approach | 11 | 3 | 6 | 0 |
| Excision via Weber-Fergusson approach | 19 | 5 | 7 | 2 |
| Excision via craniotomy | 3 | 2 | 1 | 1 |

Endoscopic exposure and excision of juvenile nasopharyngeal angiofibroma can be considered as a standard approach for limited lesions extending upto pterygopalatine fossa.[16] Excision via lateral rhinotomy is still the most useful approach for JNA. It offers an excellent exposure for complete removal of tumor for stage II A and may be combined with Weber-Fergusson or its extensions to give additional lateral exposure when necessary for stages II B, III A.

The management of JNA with intracranial extension remains controversial despite improvements in neurosurgical techniques. Skull base erosion with minimal intracranial extension does not necessitate craniofacial resection to achieve cure as it is not a malignant tumor and often allows for complete tumor removal from below without formal craniotomy. Three out of 4 patients who had been classified into stage III B and had undergone a craniotomy had recurrence while one patient who is disease free should have been classified into stage III A according to the proposed classification. This emphasizes the importance of skull base erosion and it is best to reserve radiotherapy for stage III B tumors.

Based on the evidence regarding the importance of age in selecting a surgical approach, tumor extent and the recurrence patterns, the proposed staging is summarized in Table 5.

## CONCLUSIONS

The choice of the surgical approach should be based on precise tumor location and in young patients the approach should be tailored to minimize potential for facial growth retardation.

We recommend endoscopic resection for tumors confined to nasopharynx, nasal cavities, sinuses and minimal extension to PMF. Tumors that have significant involvement of infratemporal fossa, cavernous sinus or minimal intracranial extension can be resected by Weber-Fergusson approach, infratemporal fossa approach, or craniotomy according to the surgeon's preference and expertise. We need further studies to determine the long-term effects of the various approaches on the facial growth.

**Table 5:** Proposed staging and surgical protocol for JNA

| *Modified sessions* | *Proposed staging* | *Surgical approach* |
|---|---|---|
| I A, I B, II A | I | Endoscopic transnasal |
| II B | II A | Excision via lateral rhinotomy |
| II C | II B | Excision via Weber-Fergusson |
| III A | III A | Excision via Weber-Fergusson and its extensions/maxillary swing/craniotomy |
| III B | III B | Radiotherapy (IMRT) |

## REFERENCES


1. Batsakis JG. Tumors of the Head and Neck: Clinical and pathological consideration (2nd ed). Baltimore: Williams and Wilkins 1979;296-300.
2. Barnes L (Ed). Surgical Pathology of the Head and Neck New York: Marcel Dekker 1985;1:417.
3. Sekhar LN, Janecka IP (Eds). Surgery of cranial base tumors. New York: Raven Press 1993;485.
4. Wiatrak BJ, Koopmann CF, Turrisi AT. Radiation therapy as an alternative to surgery in the management of intracranial juvenile nasopharyngeal angiofibroma. Int. J Pediatr Otorhinolaryngol 1993;28:51-56.
5. Neel HB III. Juvenile angiofibroma. In: Blitzer A, Levoson W, Friedman WH (Eds). Surgery of Paranasal Sinuses. Philadelphia: WB Saunders Company 1985;247-59.
6. Kamel RH. Transnasal endoscopic surgery in juvenile nasopharygeal angiofibroma. J.Laryngol Otol 1996;110:962-68.
7. Ousterhont DK, Vagervik K. Maxillary hypoplasia secondary to midfacial trauma in childhood. Plast Reconstr Surg 1987;80:491-99.
8. Freihofer HPM. The timing of facial osteotomies in children and adolescents. Clin Plast Surg 1982;9:445-56.
9. Schiff A. Juvenile nasopharyngeal angiofibromas: A history of pathogenesis. Laryngoscope 1969;69:981-1016.
10. Grant TD, Figzpatrick PJ, Berman J. Nasopharyngeal Angiofibroma: A twenty years study. Laryngoscope 1976;88:1247-51.
11. Neel HB, Whicker JH, Devine KD, Weiland LH. Juvenile Angiofibroma: Review of 120 cases. Am J Surg 1973;126:547-56.
12. Bremer JW, Neel HB, SeSanto KW, Jones GL. Angiofibroma: Treatment trends in 150 patients during 40 years. Laryngoscope 1986;96:1321-29.
13. Love RJ, Murray JM, Mamandras AH. Facial growth in males 16 to 20 years of age. Am J Orthod Dentofac Orthop 1990;97:200-06.
14. Bachmayer DI. Discussion remaxillary hypoplasia secondary to midfacial trauma in childhood. Plast Reconstr Surg 1987;80:498-99.
15. Mair EA, Balger WE, Bresich IA. Sinus and facial growth after pediatric endoscopic sinus surgery. Arch Otolaryngol Head Neck Surg 1995;121:547-52.
16. Gupta AK, Rajiniganth MG, Gupta AK. Endoscopic approach to juvenile nasopharyngeal angiofibroma: Our experience at a tertiary care centre. JLO 2008;122:1185-89.