Abstract
Juvenile nasopharyngeal angiofibroma (JNA) continues to provide clinical and surgical challenges consequent to its high vascularity, tendency to intracranial extension, and propensity to postsurgical recurrence. This review encompasses the current concepts with regard to its origin and progression, and discusses the principles of pre-surgical preparation and surgical excision.

Keywords
Juvenile Nasopharyngeal Angiofibroma.

Introduction
Juvenile nasopharyngeal Angiofibroma (JNA) is a benign, but locally aggressive and extremely vascular Head and Neck neoplasm, occurring almost exclusively in the nasopharynx of adolescent males. JNA accounts for only 0.05% of all head and neck tumours. It is a fibrovascular tumour with a tendency to erode and remodel adjacent bone. The common clinical presentation is with nasal obstruction and recurrent, spontaneous and intractable epistaxis. Tissue biopsy may be associated with life threatening hemorrhage. This necessitates that a high degree of suspicion be maintained for this tumour - so that it is clinically suspected and an inadvertent biopsy avoided. Once the diagnosis is suspected it may be easily confirmed today by the characteristic radiology (CT and MR). Surgery remains the main modality of treatment. Surgery may prove challenging due to the tumour's rich vascularity, difficulty access, proximity to vital structures, invasion of the skull base, and the tendency for recurrence.

The advent of adjuvant pre-operative embolisation, anti-androgen therapy, and the development and refinement of endoscopic techniques for tumour removal have led to a significant reduction in the morbidity of surgery. Radiation has been advocated in cases wherein surgical resection is either not considered possible or considered high risk but poses significant potential risks when applied to an adolescent population.

Despite advancements in the surgical armamentarium, angiofibroma continues to remain a challenge to both rhinologists and Head and Neck surgeons.

Basic Sciences considerations
Genetics
The nearly exclusive predilection of the tumour towards adolescent males naturally indicates to an endocrine / hormonal influence in its pathogenesis. The presence of androgen, estrogen and progesterone receptors, and also strong expression of VEGF, TGF-β, and FGF receptors has been noted in the vessels and the stroma of these tumours. An association with adenomatous polyposis has been noted along with mutations in the related gene (APC gene). Further genetic analysis has indicated towards partial or complete losses of the Y chromosome and gains of the AR gene in chromosome X. Gains at chromosomes 4, 6, 8, and X and losses on chromosomes 17, 22, and Y are the most frequent chromosomal abnormalities detected.

Histology
Histologically, the tumour is unencapsulated and consists of a collagenous tissue stroma interspersed with wide vascular spaces lined by a single endothelial layer. These arterial vascular channels are unusual in not manifesting a complete muscular layer in the vessel wall (Figure 1). The lack of muscle layer in the vessels is claimed to be the histological explanation for the profuse epistaxis noted in this condition, as it is supposed that these arterial vessels are deficient in their ability to contract in response to tissue injury and bleeding.

Anatomical patterns of spread:
The advent of sectional imaging with CT and MR has indicated that JNA originates in the pterygo-palatine fossa at the apertura of the pterygoid canal. From its origin in the pterygo-palatine fossa the tumour usually expands medially into the posterior nose and nasopharynx, typically positioned submucosally and causing nasal obstruction (Figure 2A). Further it may spread superiorly into the sphenoid and the orbit, laterally into the infra temporal fossa, and anteriorly into the nasal cavity and sinuses (Figure 2B).

Modern radiology has also indicated to frequent erosion of the base of the pterygoid process, and tumour extension into the vidian canal / pterygoid canal. Tumour in the
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pterygoid canal shows two patterns of extension. In the first there is a simple pressure erosion of the pterygoid base and the vaginal process of the sphenoid, often with backward and lateral displacement of the pterygoids but without invasion of the pterygoid base or body of the sphenoid (Figure 3A). In the second variety there is deep extension into the cancellous bone at the base of the pterygoid process often with expansion and invasion of the diploe of the greater wing of the sphenoid (Figure 3B). Lateral extension into the pterygo-palatine fossa causes its expansion with anterior / forward bowing of the posterior wall of maxilla (Holman-Miller sign, Figure 4A). Further lateral extension leads to the tumour exiting via the pterygo-maxillary fissure to expand into the infra temporal fossa. Superior extension to the sphenoid sinus is by erosion of its floor and anterior wall (Figure 4B). The tumour may also extend posteriorly into the vidian canal and therein erode medially into the sphenoid sinus.

Intracranial Extension Intracranial extension of JNA has been reported to occur in about 20%. Intracranial JNA is almost always extradural, and usually to the middle cranial fossa (peri-cavernous). Extension into the middle cranial fossa is usually by one of three routes (Figure 5 A, B, C): 1. Tumour extension via the inferior orbital fissure into the orbit (Figure 2B), and subsequent posterior extension of orbital tumour via the superior orbital fissure into the middle cranial fossa. This is the most common route for intracranial extension. 2. Expansion of tumour in the sphenoid sinus with erosion of its lateral wall. (Figure 5B- tumour location lateral to cavernous sinus) 3. Expansion of tumour into the pterygoid base and the cancellous diploe of the sphenoid bone, and further superior erosion of the greater wing of sphenoid with extension into the middle cranial fossa. (Figure 5C- tumour location lateral and inferior to cavernous sinus).

The tumour may further erode the floor of middle cranial fossa along the Vb and Vc foramina (F.Rotundum, F.Ovale). The tumour may push and displace the ICA posterolaterally. Encasement of the ICA is most unusual. Occasionally the anterior cranial fossa may be involved through erosion of the cribriform plate, fovea ethmoidalis and planum sphenoidale.

Figure 1: Photomicrograph (H-E) typical of juvenile angiofibroma demonstrating the fibrous tissue stroma and the intervening vascular spaces with a thin walled endothelial layer lining but no muscular layer.

Figure 2: (A, B) Axial and Coronal MR scans demonstrating the “salt and pepper” appearance of JNA, and tumour extensions from the pterygopalatine fossa into the nasopharynx, nose, infratemporal fossa and posterior orbit.
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Clinical features
The tumour is restricted to prepubescent or peri-pubescent males with the tumour being unusual before the age of 8 and after the age of 25. JNA in a female is most unusual, and such a situation should perhaps prompt an evaluation for endocrine abnormalities and serum androgen levels.

Clinical presentation is usually with nasal obstruction or recurrent severe epistaxis in an adolescent male. Clinical features vary depending upon the extent of disease, but extensions into the sphenoid and other sinuses, orbit, infratemporal fossa and middle cranial fossa are often silent. Alternately, such extensions may manifest with cheek swelling, proptosis or visual loss. Nasal obstruction may progress to cause snoring and symptoms of Obstructive Sleep Apnea.

The gross appearance of the neoplasm is of a lobulated, pink to purplish, smooth surfaced mass - which may be occasionally

Figure 3: CT images demonstrating posterior displacement of the pterygoid base (3A) and erosion of the pterygoid base (3B). Figure 3A also displays anterior bowing of the posterior maxillary wall and expansion of the pterygo-palatine fossa (Holman Miller sign), and figure 3B additionally displays the erosion of the ipsilateral vidian canal.

Figure 4: Typical and diagnostic radiological signs of JNA include tumour enhancement with contrast, (4A), expansion of the pterygopatine fossa with anterior bowing of the pterygoid (4A); erosion of the sphenoid and middle cranial fossa (4B); and erosion of the vidian canal by the tumour (4B). The contra-lateral vidian canal is well visualized (white arrow).
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ulcerated and demonstrate superficial slough in situations with epistaxis and nasal packing. In previous years, when trans palatal palpation for an adenoidal or nasopharyngeal mass was advocated, it was noted as being firm in consistency but current practitioners have had no occasion to put this to test.

The advent of nasal endoscopy and of routine sectional imaging, and the regular use of these diagnostic modalities for cases presenting with epistaxis has aided early detection. Due precautions should be undertaken during office endoscopy of suspected Angiofibroma, lest it precipitate significant epistaxis.

Classification

Various systems of classification exist for angiofibroma. The Radkowski’s classification (Table 1) is currently popular, and increasing stages have been correlated with incremental rise in tumour recurrences.

Investigations

Imaging

The imaging characteristics of JNA are typical and diagnostic, and recourse to a biopsy or to angiography for confirming the diagnosis is not required in current times. CT and MR offer complimentary information.

Computed Tomography: Computed tomography remains the initial investigation of choice as it may demonstrate a multitude of characteristic radiological signs, and also delineates the extent of bony expansion and destruction. An enhancing mass lesion in the nasopharynx is usual but not invariably - especially in cases with recurrence. Characteristic CT findings include:

- a. Anterior bowing of posterior maxillary wall (Holman-Miller sign) - Figures 2A, 3A, 4A.
- b. Erosion of floor of sphenoid sinus and contiguous tumour extending from the nasopharynx to the sphenoid sinus - Figures 4B, 5B, 5C.
- c. Erosion of the base of pterygoid - Figures 3B, 5C.
- d. Characteristic tumour distribution with a lobulated enhancing and well demarcated tumour involving infra temporal fossa, expanding through inferior orbital fissure, posterior orbit and superior orbital fissure - Figures 2B, 5A.
- e. Erosion and widening of vidian canal - Figures 4B, 5C.

MRI: MR scanning offers improved soft tissue delineation. The tumour may display a “salt and pepper” appearance (Figures 2A, 2B) with the fibrous tumour components appearing white, and the angiomatosus vascular components

Table 1: Radkowski staging system for Nasopharyngeal Angiofibroma.

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<th>Stage I:</th>
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<tr>
<td>IA-</td>
<td>Tumour confined to posterior nares and/or nasopharyngeal vault</td>
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<tr>
<td>IB-</td>
<td>Tumour involving posterior nares and/or nasopharyngeal vault with involvement of at least one Para nasal sinus</td>
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<th>Stage II:</th>
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<td>IIA-</td>
<td>Minimal lateral extension into the pterygo-maxillary fossa</td>
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<tr>
<td>IIB-</td>
<td>Full occupation of the pterygo-maxillary fossa with or without superior erosion of orbital bones</td>
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<tr>
<td>IIC-</td>
<td>Extension into infra temporal fossa or posterior to pterygoid plates</td>
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<th>Stage III:</th>
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<td>IIIA-</td>
<td>Erosion of the base of skull (middle cranial fossa/ base of pterygoids)-minimal intracranial extension</td>
</tr>
<tr>
<td>IIIB-</td>
<td>Extensive intracranial extension with or without extension into the cavernous sinus.</td>
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appearing as dark flow voids. The tumour enhances strongly with gadolinium and tumour extensions are clearly demonstrated. MR is especially useful for assessment of intra cranial tumours abutting the cavernous sinus and the ICA, and for follow up evaluations for residual/recurrent tumour.

**Angiography:** Angiography is of diagnostic value with its characteristic tumour blush. It provides information about the major vascular supply and allows for pre-operative embolisation. The major arterial supply to these tumours is typically the ipsilateral internal maxillary artery, with occasional additional vessels from ascending pharyngeal artery, and branches from cavernous ICA or contra lateral external carotid system (Figure 6).

**Treatment**
As for other benign tumours, surgery is the main modality of treatment. The tumour has been reported to occasionally regress after adolescence but this is most unusual and not a reliable modality for a vascular tumour in the nasopharynx with potential for significant hemorrhage. Radiation has been advocated for advanced or unresectable tumours but is not the preferred treatment for surgically resectable lesions.

**Surgical Treatment**
Surgery requires proper planning. Consideration is also given to pre-treatment tumour shrinkage with anti-androgen therapy, and to tumour devascularisation by tumour embolisation.

**Pre treatment tumour volume reduction by anti-androgen treatment:** Estrogen treatment was initially proposed in the mid-twentieth century but gradually abandoned with obvious collateral feminization and no clear evidence of efficacy. The availability of current generation anti-androgens with limited side-effects has however led to re-evaluation of anti-androgen treatment as an adjuvant to surgical treatment. Flutamide, a non steroidal androgen antagonist, leads to receptor site competitive inhibition of testosterone and dihydrotestosterone, but with increased serum levels of the same and no suppression of libido. Studies indicate that a 6 week course of flutamide (10mg/kg/day) is found to be efficacious in reducing tumour volume (mean reduction 16.5%, maximum-40%), and also to be safe. The efficacy is however limited to post-pubertal patients, and the treatment is ineffective in pre-pubertal patients (who have minimal or no testosterone levels).

**Pre-Operative Tumour Embo lisation:** Pre-operative embolisation is now becoming routine practice and is undertaken with gelfoam or PVA particles 1-2 days prior to surgery. Embolisation decreases intra operative blood loss and improves the surgical field, and thereby aids in achieving complete tumour excision. It has however also been noted to be associated with increased recurrence, possibly as embolisation may lead to recession of the devascularised tumour into the cancellous spaces of the sphenoid bone. As per current practice, pre-operative embolisation is preferred in most instances, and is considered essential for endoscopic approaches.

**Surgical Excision:** Various surgical approaches have been proposed. The choice of the surgical approach depends upon the age of the patient, disease extent, stage of surgery (primary/revision), vascularity, effectiveness of embolisation and experience of surgeon. Irrespective of the surgical approach selected, certain basic intra-operative principles of tumour excision apply to all.

**Figure 6:** Pre-embolisation angiogram with angiography catheter in the distal external carotid artery. The internal maxillary artery is seen to supply the tumour with an intense “tumour blush” being noted. The middle meningeal branch of the internal maxillary art. is also noted, as is the superficial temporal artery.

**Table 2.** The nasal endoscopic approach is currently in vogue and experience of surgeon. The trans-palatal approach is being currently supplanted by the endoscopic approach as the indications for both are similar. It however remains an effective approach for situations wherein adequate endoscopic expertise is unavailable, or
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Table 2: Commonly used surgical approaches for excision of Juvenile Nasopharyngeal Angiofibroma.

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<tr>
<th>Nasal Endoscopic</th>
<th>Trans palatal</th>
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<tr>
<td>• Trans palatal approach</td>
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<td>• Trans palatal with sub labial extension</td>
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<tr>
<td>Trans maxillary</td>
<td></td>
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<tr>
<td>• Lateral rhinotomy / Medial maxillectomy</td>
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<tr>
<td>• Midfacial degloving</td>
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<tr>
<td>• Le-fort I osteotomy approach</td>
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<tr>
<td>• Maxillary swing / Facial Translocation approach</td>
<td></td>
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<tr>
<td>Lateral skull base approach</td>
<td></td>
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<tr>
<td>• Pre auricular sub temporal infra-temporal approach</td>
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<tr>
<td>• Infratemporal approach Type C</td>
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Combination of any two approaches

wherein facilities for embolisation are unavailable. Excellent exposure is provided for early stage tumours restricted to the nasopharynx or its vicinity (Stage I, IIA, and IIB). Tumours with lateral extensions (Stage IIC) may also be approached by supplementing the median trans palatal approach with a lateral sub labial incision.27 The trans palatal and sub labial incision may be further connected by an incision curving behind the maxillary tuberosity, and this can provide access to the pterygopalatine fossa and the inferior portions of the pterygoid plates. Further access to the pterygoid fossa may be achieved by removing the medial pterygoid plate.27

The trans-maxillary route is currently the most commonly used surgical approach. Its main advantages are in providing a) improved exposure of lateral tumour in the infratemporal fossa (Stage IIC), and b) in affording direct access to the pterygopalatine fossa and vidian canal to facilitate drilling at the vidian canal after tumour removal.22 The mid facial degloving approach avoids a facial scar and provides good access for limited lateral extensions, but access to the superior and posterior tumour surfaces can be limiting, and it may therefore be inappropriate in cases with significant sphenoid involvement27 or dural exposure. A lateral rhinotomy incision with a maxillary maxillectomy provides better exposure of the sphenoid, infra temporal fossa, anterior skull base and orbit. The facial translocation (maxillary swing) approach28 provides a dramatically wider field of exposure, and is the preferred approach for large tumours (stage IIIa) with significant skull base erosion and tumour extensions medial to the cavernous sinus, or with limited tumour extension lateral to the cavernous sinus. The wide exposure afforded allows for a safe dissection of the tumour from the dura and the orbit. The approach however is associated with increased morbidity consequent to the facial incision, the requisite osteotomies and plating, and also the risk of a palatal fistula.

Lateral approaches (i.e. Preauricular sub temporal-infratemporal or Fisch Type C & D) are useful primarily in being easily combined with a temporal craniotomy and so providing access to tumours in the superior orbital fissure and tumours lateral to the cavernous sinus. A facial incision is also avoided. In centers with adequate expertise the lateral infratemporal fossa approach has been reported to provide for a high rate of radical tumour removal (80%) and the lowest rate of recurrence (6%).29 The lateral approaches sometimes need to be supplemented with a simultaneous endoscopic approach to release the medial compartments of tumour.

Endoscopic excision of JNA

The endoscopic technique to excision of JNA is currently the preferred surgical approach for most early lesions, and the procedure is therefore being described in some detail. The illuminated, magnified and panoramic views provided ensure that excellent access is obtained without the extensive bone removal and osteotomies of previous conventional techniques.30 Pre-operative embolisation is routinely undertaken.

Technique: As with any trans-nasal approach, initial dissection includes ethmoidectomy with partial resection of the middle turbinate and a wide maxillary antrostomy. Further dissection is as per the disease extent. Bipolar cautery on the tumour surface may help to slightly shrink the tumour and to minimise tumour ooze. A large antrostomy facilitates exposure of the postero-lateral maxillary wall, which is then removed from medial to lateral so as to expose the pterygopalatine fossa and the medial infratemporal fossa.10,18

As tumour is dissected out from the pterygopalatine and infra temporal fossae, the terminal portion of internal maxillary artery is identified within the fat pad and clipped along with its branches.31 Tumour extensions superiorly to the orbit may be followed and dissected free. The lateral tumour in the infratemporal fossa and orbit is easily dissected as it is not adherent to any of the surrounding tissues.31,35 The medial tumour however is not so easily dissected as it is often adherent to the pterygoid process at its base wherein it may erode into the cavernous bone, or be attached to the vidian nerve and its adjoining artery. Further medially in the nasopharynx, the tumour is often located submucosally and needs to be freed with sub periosteal dissection. Tumour in the sphenoid is not always adherent and may often be easily delivered.18

In addition to these standard techniques, further modifications may be required for specific disease extensions. Significant infratemporal fossa extensions may be mobilized by an additional sub labial incision and digital dissection to push the lateral tumour into the endoscopically accessible medial surgical field.32 The same may also be accomplished by an endoscopically created antral window.25 Presence of disease behind the pterygoids necessitates removal of pterygoid plates.

The risk of perioperative bleeding has been noted to be greater at certain sites of dissection - i.e. adjacent to the internal maxillary artery, the sphenoid sinus, the root of the pterygoids, the interpterygoid fossa as well as in the region of the cavernous sinus.34 Tumour once dissected off all attachments and adhesions is ideally removed in one piece, but the limitations of the nasal aperture sometimes necessitate that it be removed by delivering it posteriorly into the nasopharynx and oral cavity, or that it be sectioned and then removed. Section of the tumour is conventionally avoided as it may precipitate bleeding, but may be undertaken after the tumour is completely dissected and devascularised, and especially so if it has been embolised.

Advantages: The angle of vision provided by endoscopy may be better than with a lateral rhinotomy approach.30 The avoidance of a facial incision and of osteotomies is an obvious benefit to the young adolescent, and would be expected to diminish the impact on future midfacial skeletal growth.32 Improved illumination and the magnified view enables better identification of the anatomy and the tumour-tissue interface,
and further may aid disease clearance from the recesses in the sphenoid bone.\textsuperscript{22,32}

\textbf{Limits of endoscopic surgery:} The endoscopic approach limits the free space to work around the periphery of tumour. The limited access through the nasal aperture restricts the role of an assistant in providing a clear surgical field and in providing traction on the tumour to aid its dissection.\textsuperscript{22} Even minimal bleeding can obscure the surgical view. Involvement of the cavernous sinus, or tumour extension toward the internal carotid artery through the foramen lacerum are associated with a risk of uncontrollable bleeding and probably represents the limits of endoscopy.\textsuperscript{31,32,34}

\textbf{Intra cranial JNA}

The routes whereby JNA extends to the intracranial compartment have been listed previously. The surgical approach to such a tumour is largely dependent on its relation to the cavernous sinus. Tumour lateral to the cavernous sinus is best approached by a lateral skull base approach (Pre-auricular subtetmal approach / Fisch Type D approach) and complete excision may be achieved in 80\% of cases.\textsuperscript{29} Tumour situated medial to the cavernous sinus is however not accessible by such a lateral route,\textsuperscript{29} and is best addressed by an anterior trans-maxillary approach. The maxillary swing / facial translocation technique provides wide exposure and enables meticulous dissection of the tumour from the dura, cavernous sinus and optic nerve under direct vision.\textsuperscript{30,39}

A formal craniotomy may be avoided in tumours with lesser degrees of intracranial extension and not in a major relation to the cavernous sinus or the ICA. Access to the tumour may then be gained by following the tumour along its route of spread. This may be undertaken by open trans nasal approaches, and the wide access afforded by the maxillary swing approach is preferred in such situations. Centers with dedicated and experienced endoscopic skull base teams may also undertake the same by endoscopic or endoscopic assisted techniques.\textsuperscript{36}

\textbf{Radiotherapy}

Although radiation has been reported to be effective therapy for JNA,\textsuperscript{30-38} the general view has been that it is not entirely appropriate for a benign tumour in the adolescent population. The potential for long term complications has discouraged its use – reported complications include malignant transformation, thyroid carcinoma, bone and soft tissue sarcomas, basal cell carcinoma, hypopituitarism, cataract, optic nerve atrophy, osteoradionecrosis, osteomyelitis of skull base, and facial growth retardation.\textsuperscript{38}

Radiation may however be used as primary therapy for advanced and unresectable lesions, and has also been used for recurrent / residual tumours. Moderate radiation of 3000-3500 rads over conventional fractionation has been reported to control tumour progression and symptoms in up to 80\%. Tumour regression is slow over many months, and 10-63\% of cases treated primarily with radiotherapy may continue to demonstrate a radiologically visible mass on follow up.\textsuperscript{36,37} Non-progressive tumour in asymptomatic individuals (post radiotherapy) usually requires no further treatment.\textsuperscript{30-38}

\textbf{Residual/recurrent tumour}

“Recurrence” is a conspicuous feature of the natural history of juvenile Angiofibroma. Though these tumours have been traditionally labeled as “recurrent”, they are almost always consequent to residual tumour tissue left behind at surgery. Involvement of the infratemporal fossa, sphenoid sinus, base of pterygoids, clivus, medial aspect of the cavernous sinus, foramen lacerum, and anterior fossa have all been associated with an increased risk of residual / recurrent disease.\textsuperscript{30} Imaging evidence of invasion of the sphenoid diploe through the pterygoid canal is also associated with increased risk of residual / recurrence.\textsuperscript{8} Pre-operative embolisation too has been associated with increased recurrence.\textsuperscript{20}

\textit{Minimizing residual / recurrent disease:} Attention to the surgical principles as outlined above with pre-surgical optimization, adequate surgical exposure, control of vascular feeders, and systematic tumour removal would serve to minimize residual disease. Radiological evaluations have indicated that involvement of the basisphenoid and pterygoid canal (vidian canal) is frequent (Figures 3b,4b, 5c ), and specific attention is directed to these areas by drilling on them with the intent of removing incipient tumour tissue in these sites.\textsuperscript{22}

\textit{Post treatment surveillance:} It is currently recommended that follow up endoscopy and radiology be undertaken at 3-4 months post-surgery.\textsuperscript{21} Gadolinium enhanced MRI is the preferred modality for this situation. If no residual mass is identified on the first MR evaluation, further surveillance evaluations may be limited to clinical examination and nasal endoscopy alone.

\textbf{Management of residual/recurrent tumour:} The advent of routine endoscopy and radiological imaging in the follow-up schedule has probably led to an increase in the detection of residual / recurrent tumours. Small and asymptomatic residual / recurrent disease detected only on follow up imaging may however not necessarily require surgical excision, especially so if the tumour is demonstrated to be non-progressive on serial imaging. Large or symptomatic tumours, and tumours demonstrating progression require revision surgery. Gamma knife treatment and conventional radiotherapy are other alternatives to be considered for intra cranial remnants.\textsuperscript{39,33}

\textbf{Conclusion}

Nasopharyngeal Angiofibroma is a highly vascular tumour of the skull base, with origins from the region of pterygoid canal and frequent extensions to adjacent extra and intra cranial regions. The tumour has a near-exclusive predilection to males in the peri-pubertal age group. Surgery is the prime treatment modality, but poses challenges consequent to difficult access, the possibility of life threatening intra-operative hemorrhage, tumour proximity to skull base structures, and the tendency to recurrence. Advancements in pre-surgical radiological evaluation, the development of pre-surgical adjuvant treatments, and the dramatic improvement in surgical technology have combined to improve surgical resection and reduce surgical morbidity. Further understanding of the genetic and hormonal influences on the tumour is expected to impact on future treatment.

\textbf{Conflict of Interest}

All authors have no conflict of interest to declare. No extraneous funding was obtained.
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**Key Points**
- JNA is a highly vascular benign tumour with characteristic patterns of spread. The peculiarity of it being restricted to adolescent males, has provided for intellectual and surgical challenges with regard to its origins and treatment.
- Current concepts localize its origins to the vidian canal and pterygoid base. The tumour has been demonstrated to manifest receptors for testosterone, and also for estrogen and progesterone.
- Diagnosis is based on the characteristic CT and MR findings demonstrating typical patterns of spread. Tissue biopsy and angiography are not currently considered necessary for establishing a pre-operative diagnosis.
- Surgical excision is the treatment of choice. Radiation therapy may be used for the rare situations with significant intracranial spread wherein surgical excision is deemed as high risk.
- Residual and recurrent disease following surgical excision has been frequent in previous times. Improved radiology in current times allows for a complete assessment of disease extensions, and the selection of appropriate surgical approaches for improved surgical excisions.
- Surgical residual disease may be improved and surgical residual disease minimized by attention to pre-surgical optimization (tumour shrinkage by anti-androgen therapy, and pre-operative embolisation), and the surgical principles of adequate tumour exposure, control of vascular supply, lateral to medial dissection/sub-periosteal dissection, and post excision drilling of the pterygoid base and vidian canal.
- The evolution of endoscopic techniques and instrumentation in recent times has provided for its application as a low morbidity option for early stage tumours. Further evolution of expertise and experience is expanding its role for larger stage tumours.

**References**

Nasopharyngeal Angiofibroma

1. The propensity for intractable and massive haemorrhage in JNA is ascribed to –
   a. Sequestration of platelets in the venous channels
   b. High flow arteriovenous communications
   c. Consumption coagulopathy and metabolic acidosis
   d. Inadequate muscular contraction in intra-tumoral arterial vessels.

2. The major vascular supply of JNA is by the –
   a. Ascending pharyngeal artery
   b. Internal mammary artery
   c. Ascending palatine artery
   d. Internal maxillary artery

3. Intracranial extension of JNA may occur via any of the following routes except—
   (*PPF=pterygopalatine fossa; IOF=Inferior orbital fissure; SOF= Superior orbital fissure; PMF=pterygo-maxillary fissure; ITF= Infratemporal fossa)
   a. PPF—IOF—SOF— middle cranial fossa
   b. PPF—PMF—ITF—SOF—sphenoid sinus—cavernous sinus
   c. PPF—Vidian canal/ Pterygoid base erosion— sphenoid body erosion—middle cranial fossa
   d. nasopharynx—sphenoid sinus—medial cavernous sinus

4. Drilling of bone at following sites at surgery is expected to reduce recurrence in JNA except
   a. postero-superior boundary of the pterygo-palatine fossa
   b. Vidian canal
   c. Posterior ethmoid
   d. Base of pterygoid

5. Basic surgical step(s) to be adhered during surgical resection of JNA includes,
   a. Medial to lateral dissection
   b. Intra capsular dissection
   c. Good surgical exposure
   d. Piecemeal removal

6. Strategies to improve surgical outcome in extensive JNA includes all except,
   a. Pre operative embolisation
   b. Anti androgen therapy
   c. Pre operative radiotherapy
   d. Drilling of bone at basi-sphenoid

7. The efficacy of hormonal therapy with flutamide for JNA is based on its—
   a. estradiol antagonist effect.
   b. gonadotropin regulation effect.
   c. androgen antagonist effect
   d. progesterone agonist effect.

8. Which of the following statements is correct with regard to endoscopic excision of JNA—
   a. Obviates need for extensive osteotomies.
   b. Best indicated for cavernous sinus lesion.
   c. Is associated with Increased recurrence rates.
   d. Has significant impact on maxillary skeletal growth.

9. The prime role of radiotherapy in JNA is towards—
   a. Haemostasis
   b. Rapid control of tumor progression
   c. Pre-surgical treatment to reduce tumor volume
   d. None of the above

10. Which of the factor(s) listed below is associated with residual/recurrent disease—
    a. Intracranial extension
    b. Pre operative embolisation
    c. Invasion of pterygoid and basisphenoid
    d. All of the above

11. The probable site of origin of JNA is,
    a. Superior aspect of the pterygopalatine fossa
    b. Pterygoid canal
    c. Vidian canal
    d. All of the above
Diagnostic Quiz Questions

1. Pre operative imaging of 12 year male with nasopharyngeal angiofibroma revealed localized tumor involving the right posterior nose and nasopharynx and lateral extension posterior to the pterygoid plates.

The staging of tumor as per Radkowski classification is:

a. Stage I b
b. Stage II c
c. Stage I a
d. Stage II a

2. Pre operative imaging of 14 year old male patient with diagnosis of nasopharyngeal angiofibroma revealed tumor involving nasopharynx, pterygopalatine fossa and sphenoid sinus with expansion and erosion of lateral wall of sphenoid and intracranial tumor extension adjacent to the medial cavernous sinus (intra cranial/extra dural extension).

The most appropriate surgical approach among the following options is:

a. Lateral skull base approach
b. Mid facial degloving approach
c. Maxillary swing approach
d. Transpalatal approach

3. A 12 year old boy presented to the A & E with profuse epistaxis from the left nasal cavity and had bilateral nasal packing. Following pack removal, a nasal endoscopic examination reveals a pinkish mucosa covered mass blocking the left posterior choana. Further evaluation for diagnosis and therapy is best undertaken by-

a. an early angiogram to localize the bleeding site, and to consider embolisation of the same.
b. surgical excision of the mass for histological evaluation.
c. An initial incisional biopsy for histological confirmation and to plan subsequent treatment.
d. Radiological evaluation by contrast enhanced CT or MR scanning.

4. Following surgical excision of a Stage III Angiofibroma in a 19 year old male, the 6 month follow-up MRI scan reveals a small residual tumor in the superior orbital fissure/ anterior cavernous sinus. The most appropriate option for further management would be-

a. Surgical excision by the lateral skull base approach
b. Serial MRI at 6 month interval to assess tumor growth pattern
c. Radiotherapy
d. Any of the above.