Juvenile nasopharyngeal angiofibroma (JNA) is a rare, benign, vascular neoplasm that accounts for less than 0.5% of all head and neck tumors. JNAs occur almost exclusively in the nasopharynx of adolescent males.

The site of origin of JNA remains controversial. Some believe that it takes origin from the superior lip of the sphenopalatine foramen at the junction of the pterygoid process of the sphenoid bone and the sphenoid process of the palatine bone. Others claim that it arises from the bone of the vidian canal.

JNAs are slow growing and initially expand intranasally into the nasopharynx and nasal cavity and then into the pterygomaxillary space. Over time, JNAs will eventually erode bone and invade the infratemporal fossa, orbit, and middle cranial fossa.

The blood supply to these benign tumors is most commonly from the internal maxillary artery, but may also be supplied by the external carotid artery, the internal carotid artery, or the ascending pharyngeal artery.

Histologically, JNAs originate from myofibroblasts. The tumor lacks a capsule and spreads submucosally. It is composed of a fibrous abundance of single endothelial cell lined vascular spaces or channels. These channels are surrounded by a collagenous tissue network and lack a complete muscular layer.

Extension and Spread

- Medial: into the nasopharynx and nasal cavity
- Lateral: the sphenopalatine fossa opens into the pterygopalatine fossa laterally, advancing into the infratemporal fossa
- Superior: expansion into the sphenoid sinus.
- Intracranial extension in 20-36%. Superior spread through the pterygoid process may extend into the middle cranial fossa. As the lateral and posterior walls of the sphenoid sinus are eroded, the cavernous sinus (which contains CN III, IV, V1, V2, VI, and the internal carotid artery) or pituitary and sella turcica may be involved.
Staging

Dr. Sessions and Dr. Alford described the radiologic staging of JNA.

- IA correlates with tumors confined to the nose and nasopharynx;
- IB with extension into the sinuses; IIA with minimal extension onto the pterygopalatine fossa;
- IIB for occupation of the pterygopalatine fossa with or without erosion of the orbital bones;
- IIC extending into the infratemporal fossa;
- Stage III with intracranial extension.

Ugo Fisch developed his own staging system a couple of years after that paper and, as you can see here, ranging from stage I, with tumor occupying the nasal cavity and nasopharynx to stage IV, representing massive invasion of the cavernous sinus, optic chiasm or pituitary fossa.

Radkowski developed another staging system. This was taken directly from Dr. Sessions’ previous staging system.

<table>
<thead>
<tr>
<th>Stage</th>
<th>Features</th>
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<tbody>
<tr>
<td>IA</td>
<td>Limited to nose or nasopharynx</td>
</tr>
<tr>
<td>IB</td>
<td>Extension into at least one paranasal sinus</td>
</tr>
<tr>
<td>IIA</td>
<td>Minimal extension into sphenopalatine foramen, includes minimal part of medial pterygomaxillary fossa</td>
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<tr>
<td>IIB</td>
<td>Full occupation of pterygomaxillary fossa with Holman-Miller sign, lateral or anterior displacement of maxillary artery branches, may have superior extension with orbital bone erosion</td>
</tr>
<tr>
<td>IIC</td>
<td>Extension through pterygomaxillary fossa into cheek, temporal fossa, or posterior to pterygoids</td>
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<tr>
<td>IIIA</td>
<td>Skull base erosion with minimal intracranial extension</td>
</tr>
<tr>
<td>IIIIB</td>
<td>Skull base erosion with extensive intracranial extension +/- cavernous sinus</td>
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Surgical approach

Surgical approach is dictated by the stage of the tumor with maximized exposure of the tumor while, of course, limiting the morbidity and mortality.

Surgical Planning

Smaller tumors (Class IA, IB, IIA, IIB, IIC)
- Trans-nasal
- Transpalatal
- Transantral, for lesions extending laterally up to pterygopalatine fossa

Larger tumors (IIIA, IIIB)
- Lateral rhinotomy
- Midface degloving

More extensive resections have higher associated morbidity, however, limited resections have a higher rate of recurrence

Conventional approaches include transpalatal or transantral approaches; however, modern approaches include endoscopic approaches. In advanced JNA, there is considerable debate as to the optimal surgical approach.

The options for approaching JNA include:
- Maxillary Swing approach
- Medial maxillectomy
- Bilateral rhinotomy or midfacial degloving
- Le Fort I osteotomy,
- The endoscopic approach.
- Infratemporal fossa approach
- Tyagi's combined transmaxillary, transpalatal approach
- Cranial facial resection
- Frontotemporal craniotomy approach
Maxillary Swing Approach:

This approach is a variation of trans-maxillary approach and provides wide access to nasopharynx as well as pterygopalatine fossa and pterygomaxillary fissure. It also preserves soft palata function which is so vital for speaking and swallowing.

This is one of the common approaches to nasopharynx. It exposes the nasopharynx and surrounding areas from the anterolateral aspect. Through Weber Ferguson incision maxilla is separated from its bony attachments and swung laterally intact with the masseter muscle and cheek flap. Access to opposite side can be established by removing the posterior portion of nasal septum. After tumor resection, the maxilla is swung back and fixed to facial skeleton.

Indications for maxillary swing include:

- Surgical excision of residual or recurrent nasopharyngeal malignancy, nasopharyngeal carcinoma
- Resection of recurrent JNA
- Covering exposed internal carotid artery following radio necrosis of nasopharynx after radiotherapy for nasopharyngeal malignancy with pedicled or free flap.

Lateral rhinotomy

This is appropriate for stages IIA to IIC or IIIA tumors, and these are tumors confined to the nasopharynx, nasal cavity, paranasal sinuses, pterygopalatine fossa, and specifically, the medial temporal fossa.

Advantages: It does give direct exposure of the surgical field

Disadvantages: include intranasal crusting, which occurs in almost every patien; scarring; numbness; and nasolacrimal duct obstruction as well as a propensity for facial asymmetry and the development of problems with growth of the facial bones.

The midfacial degloving approach

This approach is appropriate for stages IIA through IIIA. These are tumors in the same locations, as those accessible via lateral rhinotomy approach.

Advantage of midfacial degloving is that it has no external incisions, and it does have direct exposure of the tumor.

Drilling the base of sphenoid via the pterygoid canal resulted in removal of all residual tumor and therefore minimizes recurrence rate.
The complications include the fact that it could affect facial development. It also could develop vestibular stenosis and, as mentioned previously, intranasal crusting.

The Le Fort I approach

This approach is appropriate for stages IIIA and IIIB tumors. These are tumors with extensive skull base as well as medial cavernous sinus invasion. It can involve unilateral or bilateral osteotomies with a resultant palatal drop and you can also resect the posterior maxillary sinus wall, allowing you to control tumor vascular supply.

The disadvantage of this approach is the extensive bony disruption that it is warranted and results in facial skeletal abnormalities.

The infratemporal fossa approach is appropriate for stages IIC and IIIB tumors, particularly tumors that invade the lateral cavernous sinus. This approach is a temporal modification of the infratemporal fossa approach that permits early ligation of the internal maxillary artery as well as early visualization of the internal carotid artery. Several modifications of this approach have been given, specifically the Fisch type C or postauricular approach, as well as the Fisch type B or preauricular approach.

Craniofacial resection is appropriate for stage IIIB tumors. This approach is for extensive intracranial involvement. This is a stage IIIB JNA tumor invading the left cavernous sinus and displacing the internal carotid arteries laterally.

Complications included nasal crusting, serous otitis media, and facial numbness.

The intranasal endoscopic approach.

This approach has recently been shown to be appropriate for stages up to IIIA, excluding stage IIC as there is limited exposure of the lateral infratemporal fossa. The reason why these tumors, which are somewhat extensive, are accessible via the endoscopic approach is because the tumors, although nonencapsulated, often come out in one whole total piece. Briefly, the technique starts with a middle turbinectomy, and goes on to removing the posterior maxillary sinus wall and tumor resection.

The advantages are reduced blood loss, complication rate, and hospital stay, and more importantly, this approach has been shown to have comparable recurrence rate.

The craniotomy approach is appropriate for stage IIIB tumors, as in our case. It is often combined with anterior or lateral approaches, and it likely requires neurosurgical intervention. It does have increased risk of surgical morbidities, specifically to the cranial nerves, and it may be performed as Tyagi suggested, as a staged approach to minimize morbidity.
Transpalatal Approach to Nasopharynx (Without Palatal Split)

Indications

- Juvenile nasopharyngeal angiofibroma (JNA), stage I
- Small nasopharyngeal tumors

Contraindications

- Tumors extending lateral to nasopharyngeal sidewalls

Transpalatal Approach:

- Soft palate is split and retracted
- Hard palate resection for increased exposure
- Palatine bone and interior aspect of pterygoid plate resected

Denker approach:

- Wide anterior antrostomy
- Removal of ascending process of maxilla
- Removal of inferior half of lateral nasal wall