Juvenile Nasopharyngeal Angiofibroma

TREATMENT UPDATE
Introduction

- Rare Benign vascular Tumor

- The management of JA is challenged by the abundant vascular blood supply of the lesion, along with the complex anatomy of the skull base and the young age of the affected population

- Intracranial involvement has been reported to occur in 10% to 37% of all cases

- The prognosis for this disease is extremely good if diagnosed well in time and if the tumor has not extended intracranially
JA typically affects the male population, most commonly between 9 and 19 years of age.

The most frequent symptoms are nasal obstruction and epistaxis. Epistaxis is usually brisk and intermittent.

Nasal obstruction may be bilateral despite the unilaterality of the lesion, due to nasopharyngeal extension as well as deviation of the nasal septum by the expansile lesion.

Purulent nasal discharge and facial pain can be due to sinus drainage pathway obstruction, and conductive hearing loss indicates obstruction of the eustachian tube.
Diagnosis

- Diagnosis is made by typical clinical and imaging findings, while tissue biopsy is unnecessary and may lead to brisk hemorrhage.

- At imaging, JA appears as a highly vascularized and expansile lesion centered on the pterygopalatine fossa (PPF) in both contrast-enhanced computerized tomography (CT) and gadolinium-enhanced magnetic resonance imaging (MRI);

- CT and MRI are complementary in the diagnosis of JA as CT emphasizes skull base bony involvement while MRI is superior in the demonstration of intracranial, orbital, and cavernous sinus CT and MRI are
Differential Diagnosis

- Hemangioma
- Choanal polyp
- Nasopharyngeal carcinoma
- Angiomatous polyp
- Nasopharyngeal cyst
- Hemangiopericytoma
- Rhabdomyosarcoma
- Chordoma
- Adenoid Hyperthropy
Holman Miller sign
Routes Of Spread

- Medially, towards nasopharynx and nasal cavity
- Distort the nasal septum, turbinates and soft palate
- It may erode the posterios ethmoidal and sphenoidal sinuses allowing direct extension of tumor into the orbit, cavernous sinus and the parasellar region
- Laterally, sphenopalatine foramen → Pterygopalatine fossa → Pterygomaxillary fissure → Infratemporal fossa → check
- Can also extend along the inferior orbital fissure, across the apex of the orbit into the superior orbital fissure
- Continued tumor expansion → Pressure erosion of the base of the pterygoid plate and greater wing of sphenoid → tumor comes against the dura of the middle cranial fossa
Treatment Challenge

Patient
- Young Age
- Present at late stage of the disease

Tumor
- Location
- Extension
- Vascular Tumor

Facilities
- Imaging (CT / MRI)
- Angiography

Surgeon
- High Risk Surgery
- Experience
Treatment Option

- Anti VGEF
- Anti Estrogen
- Flutamide

- Conventional
- IMRT

- Open Surgery
- Endoscopy

- Surgery

- Radiotherapy

- Hormonal Therapy
- Chemotherapy
- Anti VGEF

- Chemotherapy

- Anti Estrogen
- Flutamide

- Surgery
General surgical principles

- JNAs may be resected by endoscopic, open or combined (endoscopic & open) techniques
- • The surgical approach is dependent on:
  - o Tumour location and extent
  - o Pattern of vascular supply
  - o Effectiveness of embolisation
  - o Facial skeletal maturity
  - o Experience of the surgical team
- • Carefully plan surgical approach(es) according to the imaging studies
- • In expert hands and with effective preoperative embolisation, most JNAs can be removed endoscopically with reduced morbidity
General surgical principles

- Complete all bone work and ensure good access to the tumour before attempting resection, because once bleeding is encountered, the surgery may become very difficult and hazardous, and haemostasis may only be possible after completing the tumour resection.

- In young patients, avoid excessive soft tissue and bone dissection of the mid-face to minimise the risk of causing growth abnormalities.
General surgical principles

- External approaches are associated with high rate of morbidity. These approaches usually involve extensive osteotomies which are associated with increased blood loss, increase operative time, and which may interfere with the normal facial growth of the adolescent patient. Facial scars are another concern of utmost importance. Cerebrospinal fluid leak, facial and infraorbital nerve damage, lacrimal dysfunction, facial deformities, trismus, and dental malocclusion have been reported with transfacial approaches.
Carotid External artery Ligation

- Ligation of the external carotid artery (ECA) can be performed with the patient under local or general anesthesia
- A horizontal skin incision is made between the hyoid bone and the superior border of the thyroid cartilage
- Subplatysmal skin flaps are then raised, and the sternocleidomastoid muscle is retracted posteriorly
- The ECA is identified by following the internal carotid artery (ICA) for a few centimeters and dissecting the ECA beyond its first few branches.
- After the ECA has been positively identified, it is usually ligated just distal to the superior thyroid artery
- Continued bleeding after ligation may be from anastomoses with the opposite carotid system or the ipsilateral ICA.
Carotid External artery Ligation
Preoperative Embolization

- Preoperative embolisation is generally undertaken 24-72 hours prior to resection.
- Early stage juvenile nasopharyngeal angiofibroma (JNA) has a distinct blood supply from the ipsilateral ECA, most commonly the internal maxillary artery, ascending pharyngeal, sphenopalatine artery, and descending palatine artery.
- Advanced lesions receive multiple blood supplies from the ipsilateral ECA as well as the contralateral one and from the ICA, most commonly through the vidian artery, ophthalmic artery, and inferolateral and meningohipophyseal trunks.
Hormonal Therapy

- The exclusivity of JNA to males has pointed toward an obvious hormonal influence, and early workers noted increased tumor growth with testosterone and a regression with estrogens.
- Recent years have seen a greater understanding of the sex-hormone receptors present on the tumor.
- Specific receptors to both testosterone and dihydrotestosterone (DHT) have been noted.
- Hormonal therapy used for tumor regression.
- Side Effect
  - Feminizing effects and a perceived risk of cardiovascular disease.
  - Further, estrogen treatment led to suppression of gonadotropin release by the pituitary and consequent suppression of testosterone secretion.

- Flutamide: 10 mg.kg.day in 3 divided doses x 6 weeks → 44% tumor shrinkage
- Diethylstilbestrol → 5 mg TID
- Bevacizumab → monoclonal antibody against VEGF
- Sirolimus/ Rapamycin
Conclusions. Prepubertal and postpubertal patients differ in their response to flutamide. In postpubertal patients, 6 weeks preoperative use is safe and leads to partial tumor regression. Tumor regression from adjacent vital structures may facilitate surgical excision and limit morbidity.

Radiotherapy

- Reserved for selected patients → inoperable intracranial extension of recurrent tumours
- Local control rates of 80-85%
- 30-55 Gy dose
- Disadvantages
  - Skin, thyroid and nasopharyngeal malignancies
  - Growth retardation
  - Temporal lobe necrosis
  - Panhypopituitarism
  - Cataracts
  - Radiation induced keratopathy
Management of locally advanced JNA has long remained a challenge. Gross total excision is always attempted whenever possible for long term disease control. However, the presence of skull base involvement or intracranial extension makes most patients not amenable for complete surgical resection.

Radiotherapy can be considered as a feasible option in stage III JNA with a local control rate of 90.3% at 1 year. The estimated 3- and 5-year median PFS were 91.7 and 70.7%, respectively. Moderate doses of radiation (30-45 Gy) delivered by the conformal technique can achieve durable disease control with limited morbidity.
Conclusions: Surgery is the best treatment for JNA when cure is likely with low morbidity, but the threshold for using radiotherapy should be low because moderate-dose radiotherapy cures about 90% of patients with a low risk of serious complications. We recommend 36 Gy at 1.8 Gy per treatment in most cases. Elective nodal irradiation is not necessary. Radiographic response should be almost complete within a year of radiotherapy. Patients should be followed with cross-sectional imaging every 6 months for at least 5 years. (© 2011 American Society for Radiation Oncology. Published by Elsevier Inc. All rights reserved).
Conclusion: Conformal radiotherapy shows promise as an alternative treatment approach for locally advanced JNA and confers long-term disease control with minimal toxicity.
Endoscopic Surgery

**Indication**

- Tumours involving nasal cavity, para-nasal sinuses, and nasopharynx
- Tumours with only medial infratemporal fossa involvement or extradural parasellar involvement with limited intracranial extension
- Facilitation of open approaches

**Relative Contraindication**

- Lateral infratemporal fossa involvement,
- extensive parasellar extension,
- encasement of the optic nerve,
- intradural spread, or cavernous sinus involvement.

Note that cavernous sinus involvement is often over-estimated on MRI scan due to hypervascularity of the tumour bed.
Conclusion: Endoscopic approach is possible for varies stages of JNA. Preoperative angiographic embolization has minimised the blood loss intraoperatively. The endoscopic method has reduced the postoperative morbidity.
Conclusions: In this study, endoscopic resection had a significantly lower intraoperative blood loss and lower recurrence rate when compared to open resection. However, there was no difference in recurrence rate when analyzing the IPD and controlling for Radkowski/Sessions grading. Therefore, further large-scale studies may be required to fully elucidate treatment options.
Surgical Technique

- It is essential that a surgeon be familiar with the detailed vascular anatomy of the maxillary artery and its terminal branches, and that of the maxilla, paranasal sinuses, pterygopalatine fossa, orbit and anterior skull base. Studying a cadaver skull and having it available in the operating room is of great value.
Surgical Technique

- **Approach**
  - Transpalatine
  - Transpalatine + sublabial
  - Lateral Rhinotomy with medial maxillectomy
  - Midfacial degloving approach
  - Tranmaxillary (Le fort I) Approach
  - Maxillary swing approach or facial translocation approach
  - Infratemporal fossa approach
  - Intracranial – extracranial approach
# Surgical Approach VS Tumor Location

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Transpalatal Approach

- The inferior transpalatine approach allows wide tumor exposure through the oral cavity with direct access to the nasal cavity, nasopharynx, sinuses and pterygomaxillary fissure with minimal disturbance to soft tissues and the bony anterior face.

- This can be combined with endoscopes or microscopes if additional visualization and documentation is desired.

- Lateral extent of exposure was limited by the need to preserve the greater palatine foramen and palatine arteries.

- The exposure to the nasopharynx is excellent, there are no external incisions, and the complications are few and rarely serious.
Transpalatal Approach

**FIGURE 1.** Palatal incisions include: the U-shaped incision along the gingival margin (solid curvilinear line) and the midline incision with or without a uvula split (dashed line). We favor the U-shaped incision, and this will be depicted in these figures. Several millimeters of mucosa must be left on the gingival side for adequate closure.

**FIGURE 2.** The palate mucoperiosteum is elevated posteriorly, and the greater palatine vessels are preserved. The soft palate musculature is dissected off of the posterior hard palate.
FIGURE 3. Bone from the posterior hard palate is removed with a rongeur or drill. Portions of the posterior vomer are also resected to allow adequate exposure. Bony removal should be as conservative as possible, while still allowing visualization of the tumor margins within the nasopharynx.

FIGURE 4. The tumor is removed with sharp and cautery dissection, being careful to find and remove any obvious lobulations. Hemostasis is achieved by cold packing or suction cautery when needed. The flap will be closed with 4-0 chromic sutures on the nasal side, and 3-0 vicryl sutures on the oral side.
Midfacial Degloving Approach

Surgical approaches to juvenile nasopharyngeal angiofibroma, Author links open overlay panel MB BoazForerMD AriDeroweMD JacobT.CohenMD RoeeLandsbergMD, PhDZivGilMDDan M.Fliss Operative Techniques in Otolaryngology-Head and Neck Surgery Volume 12, Issue 4, December 2001, Pages 214-218
Midfacial Degloving

Surgical approaches to juvenile nasopharyngeal angiofibroma. Author links open overlay panel MD\Kaaz\G\MD\AnDeroweMD\Jacab
T.CohenMD\BaclandsbergMD, PhD\ReG\MD\Dan M Flis Operative Techniques in Otorhinolaryngology Head and Neck Surgery
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Le Fort I osteotomy

Le Fort I osteotomy with down-fracturing of the palate is suited to tumours limited to the nose, nasopharynx, sphenoid, pterygopalatine fossa, medial infratemporal fossa and medial cavernous sinus.

Bilateral Caldwell Luc incision and Le Fort I osteotomy (A). Down fracture of the maxilla and the tumor exposure (B).

Exeresis of the tumor that was firmly adhered to the nasopharynx mucosa (C). Position of the nasal drain plug and the maxilla stabilization with four L shape plaques (D).

Maksilofacial Translocation
Maxillary Swing

- provides wide access to the nasopharynx as well as to the pterygopalatine fossa and pterygomaxillary fissure;

- The operation may be considered in 3 stages:
  - Soft tissue preparation - incisions and limited soft tissue dissection to expose bone for osteotomies
  - Bone work - miniplate preparation, osteotomies and maxillary swing
  - Closure (following tumour resection)
Case
Maxillary Swing

Courtesy Of Agung DP., MD
Conclusion

- Juvenile angiofibroma is a pathology that should be included in the differential diagnosis of unilateral nasal obstruction, associated or not with epistaxis, especially in young adolescent males.

- The finding at nasal endoscopy, which is the first step in the diagnostic algorithm, of a hypervascularized lesion occupying the posterior half of the nasal fossa should immediately raise suspicion. Morphologic imaging confirms the diagnosis.

- Endoscopic surgery after embolization has been demonstrated to be a viable alternative to external techniques for the management of small-intermediate size JA.

- Radiotherapy have a significant role in the advance and inoperable case

- External Approach still the treatment of choice for large extensive and massive tumor
Thank You