Juvenile Nasopharyngeal Angiofibroma

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Grand Rounds Presentation
The University of Texas Medical Branch
Department of Otolaryngology
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JNA

- Overview
- Anatomy
- Diagnosis
- Radiology
- Staging
- Treatment
Overview
JNA

- Benign highly vascular tumor

- Locally invasive, submucosal spread

- Vascular supply most commonly from internal maxillary artery
  - Also: internal carotid, external carotid, common carotid, ascending pharyngeal
JNA Facts and Statistics

- Up to 0.5% of head and neck tumors
- Occurring almost exclusively in males
- Average age of onset = 15 years old
- Intracranial Extension between 10-20%
- Recurrence Rates as high as 50%
Origin

- Considered to be posterolateral nasal wall at sphenopalatine foramen

- Blood supply
  - Primarily internal maxillary artery off of external carotid
Origin

- Posterolateral nasal wall near sphenopalatine foramen
Routes of Spread

- Medial growth
  - Nasal cavity
  - Nasopharynx

- Lateral growth
  - Pterygopalatine fossa
    - Vertical expansion through inferior orbital fissure to orbit possible
  - Infratemporal fossa
    - Superior expansion through pterygoid process may involve middle cranial fossa
      - Lateral and posterior walls of sphenoid sinus can be eroded
      - Cavernous sinus may be involved
      - Pituitary may be involved
Sphenopalatine Foramen

- Sphenopalatine vessels

- Nerves
  - Nasopalatine
  - Posterior superior nasal
Infratemporal fossa exposed by removal of zygomatic arch and mandible

- Pterygomaxillary fissure
- Inferior orbital fissure
- Infratemporal surface of maxilla
- Alveolar foramina
- Tuberosity of maxilla

Sphenoid bone
- Greater wing
- Infratemporal crest
- Lateral plate of pterygoid process
- Pterygoid hamulus (of medial plate of pterygoid process)

Temporal bone
- External acoustic meatus
- Mandibular fossa
- Articular tubercle
- Styloid process
- Pterygopalatine fossa
- Foramen ovale
Histology

- Myofibroblast is cell of origin
- Fibrous connective tissue with abundant endothelium-lined vascular spaces
- Pseudocapsule of fibrous tissue
- Blood vessels lack a complete muscular layer
Diagnosis
Midface and Anterior Skull Base Tumors

- Juvenile Nasopharyngeal Angiofibroma
- Osteoma
- Craniopharyngioma
- Olfactory Neuroblastoma
- Chordoma
- Chondrosarcoma
- Rhabdomyosarcoma
- Nasopharyngeal Carcinoma
Diagnosis

- History
- Physical Exam
- Radiological study
  - CT Scan
  - MRI
  - Angiogram
Characteristic Presentation

- Teenage or young adult male
- Recurrent epistaxis
- Nasal obstruction
Additional Findings at Presentation

- Conductive hearing loss
- Rhinolalia
- Hyposmia/Anosmia
- Swelling of cheek
- Dacrocystitis
- Deformity of hard and/or soft palate
- Orbital proptosis
Appearance

- Smooth lobulated mass in the nasopharynx or lateral nasal wall
- Pale, purplish, red-gray, or beefy red
- Compressible
WE'LL DO AN MRI TO BE SURE, BUT I'M FAIRLY CERTAIN IT'S A SWANNOMA
Radiological Studies

- **CT Scan**
  - Excellent for bone detail
  - Lesion enhances with contrast on CT

- **MRI**
  - Differentiate tumor from other soft tissue structures

- **Angiogram**
  - Evaluation of feeding blood vessels

**Holman-Miller Sign**
Characteristic anterior bowing of posterior maxillary wall
Coronal CT: Bone Window

- Widening of left sphenopalatine foramen
- Lesion fills left choanae
- Extends into sphenoid sinus
Axial CT: Soft Tissue Window with Contrast

- Homogenous enhancement
- Widening of left sphenopalatine foramen
- Extension into
  - Nasopharynx
  - Pterygopalatine fossa
Axial CT: Soft Tissue Window with Contrast

- Homogenous enhancement
- Widening of right sphenopalatine foramen
- Extension into
  - Nasopharynx
  - Pterygopalatine fossa
Axial MRI: T1

- Heterogeneous intermediate signal
- Flow voids represent enlarged vessels
- Extension into
  - Nasopharynx
  - Masticator space
Coronal MRI: T1 with Contrast

- Diffuse intense enhancement
- Multiple flow voids within hypervascular mass
- Extension into
  - Nasopharynx
  - Pterygopalatine fossa
Axial MRI: T2

- Heterogeneous intermediate to high signal enhancement
- Multiple flow voids within hypervascular mass
- Extension into
  - Nasopharynx
  - Pterygopalatine fossa
External Carotid Arteriogram

Feeding vessel = Internal Maxillary Artery
Staging
## Radkowski Nasopharyngeal Angiofibroma Staging System

<table>
<thead>
<tr>
<th>Staging</th>
<th>Description</th>
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</thead>
<tbody>
<tr>
<td>IA</td>
<td>Limited to the nose or nasopharynx</td>
</tr>
<tr>
<td>IB</td>
<td>Same as IA but with extension into one or more paranasal sinuses</td>
</tr>
<tr>
<td>IIA</td>
<td>Minimal extension through the sphenopalatine foramen, into and including a minimal part of the medialmost part of the pterygomaxillary fossa</td>
</tr>
<tr>
<td>IIB</td>
<td>Full occupation of the pterygomaxillary fossa, displacing the posterior wall of maxillary antrum forward. Lateral or anterior displacement of branches of the maxillary artery. Superior extension may occur, eroding orbital bones</td>
</tr>
<tr>
<td>IIC</td>
<td>Extension through the pterygomaxillary fossa into the cheek and temporal fossa or posterior to pterygoid plates</td>
</tr>
<tr>
<td>IIIA</td>
<td>Erosion of skull base— minimal intracranial extension</td>
</tr>
<tr>
<td>IIIB</td>
<td>Erosion of skull base— extensive intracranial extension with or without cavernous sinus</td>
</tr>
</tbody>
</table>

Treatment
Treatment Options

- **Surgery**
  - Gold standard

- **Radiation therapy**
  - Reserved for unresectable, life-threatening tumors

- **Chemotherapy**
  - Recurrent tumors with previous surgery and radiation

- **Hormone therapy**
  - Estrogens and antiandrogens used to decrease tumor size and vascularity
Surgical Approaches

- Endoscopic transnasal
- Transpalatal
- Denker approach
- Facial translocation
- Medial maxillectomy
- Infratemporal fossa with or without craniotomy
Preoperative Embolization

- 24 to 72 hours preoperatively
- Gelfoam or polyvinyl alcohol foam
  - Gelfoam: resorbed in approximately 2 weeks
  - Polyvinyl alcohol: more permanent
- Efficacy
  - Stage I patients reduced from 840cc to 275cc blood loss
- Complications
  - Brain and ophthalmic artery embolization
  - Facial nerve palsy
  - Skin and soft tissue necrosis

Embolization
Embolization
Surgical Approaches

- Endoscopic transnasal
- Transpalatal
- Denker approach
- Facial translocation
- Medial maxillectomy
- Infratemporal fossa with or without craniotomy
Endoscopic Transnasal

- Middle turbinectomy may be performed for improved exposure
Endoscopic Transnasal

- Middle meatus antrostomy
- Resection of posterior maxillary wall
Endoscopic Transnasal

- Sphenopalatine artery ligation
- Tumor resection from pterygopalatine fossa
Surgical Approaches

- Endoscopic transnasal
- **Transpalatal**
- Denker approach
- Facial translocation
- Medial maxillectomy
- Infratemporal fossa with or without craniotomy
■ Soft palate is split and retracted
Transpalatal

- Hard palate resection for enhanced exposure
Transpalatal

- Palatine bone and inferior aspect of pterygoid plate resected
Surgical Approaches

- Endoscopic transnasal
- Transpalatal
- Denker approach
- Facial translocation
- Medial maxillectomy
- Infratemporal fossa with or without craniotomy
Denker Approach

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

- Endoscopic transnasal
- Transpalatal
- Denker approach
- Facial translocation
- Medial maxillectomy
- Infratemporal fossa with or without craniotomy
Midface Degloving with Maxillary Osteotomies

- Gingivobuccal incision
- Nasal intercartilaginous incisions with transfixion incision
Midface Degloving with Maxillary Osteotomies

- Soft tissue elevation
Midface Degloving with Maxillary Osteotomies

- Le Fort I osteotomies
Surgical Approaches

- Endoscopic transnasal
- Transpalatal
- Denker approach
- Facial translocation
- Medial maxillectomy
- Infratemporal fossa with or without craniotomy
Maxillectomy

- Maxillary osteotomies
- Sagittal osteotomy
Maxillectomy
Alternative Approaches to Nasal Cavities and Paranasal Sinuses

- Lateral Rhinotomry
- Weber-Ferguson incision
- Weber-Ferguson with Lynch extension
- Weber-Ferguson with lateral subciliary extension
- Weber-Ferguson with subciliary extension and supraciliary extension
Surgical Approaches

- Endoscopic transnasal
- Transpalatal
- Denker approach
- Facial translocation
- Medial maxillectomy
- Infratemporal fossa with or without craniotomy
Infratemporal Fossa with or without Craniotomy
Choosing the Surgical Approach

- Retrospective chart review of surgical intervention - 37 patients
- Staged using CT scan and/or MRI
- Follow-up CT scan or MRI: 3 months, 6 months x 3 years, yearly
- Recurrence rate = 27%

Surgical Planning

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

- Larger tumors (IIIA, IIIB)
  - Lateral rhinotomy
  - Midfacial degloving
  - Extensive resection with higher morbidity
  - Limited resection with higher recurrence

Changing Technique

- Retrospective chart review of surgical intervention - 30 patients

- Marked shift towards endonasal procedures while tumor stages remained the same

- Endonasal approach contraindicated in Stage IV and some Stage III cases
  - May be used in conjunction with other approach in these cases

Surgical Approach

## Surgical Technique

<table>
<thead>
<tr>
<th>Approach (65 pts)</th>
<th>Endoscopic</th>
<th>Open</th>
</tr>
</thead>
<tbody>
<tr>
<td>EBL</td>
<td>225 ml</td>
<td>1250 ml</td>
</tr>
<tr>
<td>Complications</td>
<td>1</td>
<td>30</td>
</tr>
<tr>
<td>Length of Stay</td>
<td>2 days</td>
<td>5 days</td>
</tr>
<tr>
<td>Recurrence Rate</td>
<td>0 %</td>
<td>24 %</td>
</tr>
</tbody>
</table>

Surgical Technique

- Retrospective study of 24 patients using Radkowski staging scale
- 10 patients IA through IIA had transpalatal approach
  - Before 1999
- 9 patients IA through IIIA had transnasal endoscopic approach
  - After 1999
- 5 patients IIA through IIIA had lateral rhinotomy or degloving approach
- Recurrence in 1 case with 12-56 month follow-up range
  - Transpalatal approach

Surgical Technique

- Transnasal endoscopic approach can replace transpalatal approach
  - Less morbidity
- Patients with IIA through IIIA previously treated with lateral rhinotomy may be treated with transnasal endoscopic approach
- Tumors extending to infratemporal fossa require lateral rhinotomy and degloving for optimal exposure
  - Greater morbidity

Surgical Technique

- Surgical limitations of endoscopic resection evaluated in literature review
- Extremely limited IIIA and IIIB may be approached endoscopically
- Preoperative embolization recommended
- Unlikely that limits on endoscopic resection of JNA have been reached

Gamma Knife Surgery

- 2 case reports used as booster treatment for residual tumor after surgery
  - No change in tumor size of one patient, regression in other patient

- 1 case report used as primary treatment modality successfully


External Beam Radiation

- Retrospective review of efficacy of radiation as primary treatment modality for JNA
- 15 patients received 3000-3500 cGy
- Recurrence rate of 15%
- External beam radiation is effective mode of treatment of advanced JNA

External Beam Radiation

- Retrospective review of efficacy of radiation as primary treatment modality for JNA
- 27 patients received 3000-5500 cGy
- Recurrence rate of 15% 2-5 years post-treatment
- External beam radiation is effective mode of treatment of advanced JNA

External Beam Radiation

- Long-term sequelae of concern
  - Growth retardation, panhypopituitarism, temporal lobe necrosis, cataracts, radiation keratopathy

- Retrospective review reported 2 cases out of 55 patients developing secondary malignancies
  - Thyroid carcinoma 13 years after receiving 3500cGy
  - Basal cell carcinoma of skin 14 years after receiving 3500cGy initially, then 3000cGy for recurrence

Chemotherapy

- Chemotherapy alternative therapy

- 1 unresectable tumor had chemotherapy for palliation
  - Adriamycin and decarbazine
  - Extensive regression of tumor
  - Possible alternative to radiation?

Shick et al. HNO. 1996.
Hormonal Therapy

- Estrogen, progesterone, and androgen receptors have been identified with varying frequencies in JNAs
  - Some JNAs lack these receptors

- Limited utility
  - Delays surgery
  - Feminizing side effects
  - Cardiovascular complications
Hormonal Therapy

- Efficacy of treatment with flutamide evaluated in 7 patients
- Before and after measurement comparison made using CT scan
  - No statistically significant difference in size
  - No difference in blood loss
- No advantage with treatment

Surveillance

- Frequent physical examinations
- CT Scan / MRI
Recurrence Rates

- **Post-operative**
  - Stage I and II = 7%
  - Stage III = 39.5%

- **Tumor stage – extracranial vs. intracranial tumor**
  - Extracranial = 5%
  - Intracranial = 50%


Conclusions

- Rare, benign, vascular tumor found almost exclusively in young males
- Surgery is the gold standard with a trend towards endoscopic approaches
- Frequent follow-up after treatment is necessary
Questions
Bibliography