Layout

- Pathology
- Presentation
- Diagnosis
- Special investigations
- Classification
- Management
- Preoperative embolisation
Pathology

- Benign “tumor”
- Highly vascular; **vascular malformation:**
  - Immunohistochemical and electron microscopic profile
- Comparative genomic hybridisation show genetic imbalances
- Adolescent males
- Originates: posterior nasal + Nasopharyngeal area
- Biologically aggressive / expansive growth
- Histology:
  - Fibrous connective tissue interspersed with endothelium-lined spaces
Incidence / Pathogenesis

1: 5,000 to 1: 50,000
Onset second decade
Theories:

1. Fibroblastic: abN growth of connective tissue
2. Hormone dependent tumour: E-A imbalance androgen receptors
3. Harmartomatous origin: similar choana tissue
### Clinical presentation

#### Symptoms:
- Nasal obstruction 16/20
- Nasal bleeding 15/20
- Double vision 4/20
- Hearing loss 3/20

#### Signs:
- Intranasal tumor (reddish purple) 18/20
- Tubal dysfunction 6/20
- Exophthalmos 4/20
- Cranial nerve palsy: II 2/20
  - III - IV ------
  - VI 2/20
Origin

- Posterolateral wall of the nasal cavity + adjoining superolateral nasopharyngeal wall.
- Always involve sphenopalatine foramen
- Broad base

- Grows: Foramina + fissures; displace structures
  - Large may erode bone
  - Develop collateral blood supply
Tumour spread

- Inferior orbital fissure – apex of orbit – superior orbital fissure
- Pressure erosison of pterygoid plate + greater sphenoid wing – dura / middle cranial fossa
- Medial: fill nasopharynx, distort septum / turbinates
  Erode posterior ethmoidal + sphenoidal sinusses – direct extension into orbit, cavernous sinus, parasellar region

Sao Paulo, Rhinology Dec 2003: 33 pt; no relation between pt age and invasion route. Growth in all directions. Most lateral and superior. 3 sites invaded through > 1 route: pterygoid fossa, middle cranial fossa, maxillary sinus
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
- Hamulus of medial plate of pterygoid process

Temporal bone:
- External acoustic meatus
- Mandibular fossa
- Articular tubercle
- Styloid process

Pterygopalatine fossa
Sphenopalatine foramen
Diagnosis

- “Male teenager with epistaxis / nasal obstruction and nasopharyngeal mass”
- Radiology: CT - contrast
  Angiogram – IADSA (intra-arterial digital subtraction angiography)
- Differential: polypoid nasopharyngeal CA
  nasopharyngeal Ca
- ? Biopsy – hemorrhage
Radiological diagnosis

1. Nasopharyngeal soft tissue mass
2. Enlargement of superior orbital fissure (proptosis)
3. Distortion of nasal septum, erosion / opacification of paranasal sinuses
4. Widening of pterygopalatine fissure

**Classic sign** (not pathognomonic)
- ant. Bowing of maxillary antrum post wall
- post bowing of pterygoid plate
CT scan
Classification:

- Andrew et al.
- Chandler
- Radkowski
- Sessions
## Classification: Andrews et al

<table>
<thead>
<tr>
<th>Stage</th>
<th>Description</th>
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<tbody>
<tr>
<td>I</td>
<td>Tumour limited to nasopharyngeal cavity; bone destruction negligible or limited to sphenopalatine foramen</td>
</tr>
<tr>
<td>II</td>
<td>Invading pterygopalatine fossa or maxilla, ethmoid, or sphenoid sinus with bone destruction</td>
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</tbody>
</table>
| III   | Invading infratemporal fossa or orbital region  
|       | **A** without intracranial involvement  
|       | **B** with " " extradural |
| IV    | Intracranial intradural  
|       | **A** without infiltration of cavernous sinus/ pituitary fossa / optic chiasma  
|       | **B** with " " " " " |
Angiography

- **Arterial phase**: rapid filling, dilate, homogenous blush
- **Subtraction**: tumour vascular supply
- **Major supply** = *ipsilateral internal maxillary artery*
- **Collateral** =
  - ascending pharyngeal artery
  - internal max. contralateral
  > palatine artery
  - superficial temporal / External facial arteries –
    - infratemporal fossa
  branches of internal carotid system
    - large / into CNS / ECA ligation
  bilateral ECA - cross midline
Sphenopalatine artery
Posterior lateral nasal artery
Infraorbital artery
Posterior superior alveolar artery
Sphenopalatine artery
Posterior septal branches
Descending palatine artery
Buccal artery
Anastomosis in incisive canal
Left and right greater palatine arteries
Left and right lesser palatine arteries
Pterygoid arteries
Masseteric artery
Inferior alveolar artery
Artery in pterygoid canal
Pharyngeal artery
Sphenopalatine foramen
Anterior deep temporal arteries
Posterior deep temporal arteries and nerves
Accessory meningeal artery
Middle meningeal artery
Anterior tympanic artery
Deep auricular artery
Auriculo-temporal nerve
Superficial temporal artery
Ascending pharyngeal artery
Ascending palatine artery
Tonsillar branches
Tonsillar artery
External carotid artery
Facial artery
Superior pharyngeal constrictor muscle
Styloglossus muscle
Angiogram
Treatment

- **SURGERY**
- **PREOPERATIVE EMBOLISATION**
- Radiotherapy if unresectable or residual
- **Past:** Oestrogens, androgens
  - Radiotherapy – Preoperative (30 Gy)
    - **SE:** skeletal growth, sarcoma
- Sclerotherapy
- Properative ECA ligation
Preoperative embolisation

- Reduce intraoperative hemorrhage, more complete excision, less recurrence, less complications
- Absorbable Gelfoam particles (1mm)
- TBH, Radiology dept: 24-48hr preop
  > 72 hr collateral supply
- Eur Arch ORL, China, Oct ‘97; 22 pt, bleed intraop (+)
  E. 677 ml, without (-) E. 1136 ml
Embolisation complications

- Accidental brain or ophthalmic artery (ICA branches)
- Numbness and mild pain ipsilateral
- Trismus

- Eur Arch ORL, China, Oct ’97; 11/11 no complications “safe and effective to reduce blood loss”
Surgery

- “challenge for the Otorhinolaryngologist”

- **Approach:**
  1. Transnasal-maxillary
  2. Transpalatal
  3. Transfacial/ Sublabial mid-facial degloving
  4. Infratemporal
  5. Lateral rhinotomy
Endoscopic surgery for J A: When + How

- Laryngoscope, May 2003, Philadelphia;
- Prospective, 6 years, 15 pt
  - I 2, II 9, IIIA 3, IIIB 1
  - Vascular supply unilateral in 11
- Preoperative embolisation
- Intraoperative blood loss 80 – 600 (372) ml
- Endoscopic and MRI followup
  - 1st year 4 monthly, after this every 6 /12s
  - 24 – 93 (50) months
  - Only 1 residual lesion on MRI
- Endoscopic good option for small – intermediate JNA.
Endonasal Endoscopic Excision

- For I – III A

- Must **reduce hemorrhage:**
  - Hypotensive, reverse Trendelenburg, topical vasoconstrictors, bipolar coagulation used for mucosa

- **Disadvantage:** need extra hand / assistant

- **Advantage:**
  1. Magnified, multiangled view of the mass
  2. No surgery to skin / osteotomies
     - midfacial growth N.
• Uncinectomy, Partial / total middle turbinectomy, anterior + post ethmoidectomy, wide middle antrostomy (expose post wall), remove post wall (as lateral as needed), gentle dissection, clip sphenopalatine (+ internal maxillary prn), Sphenoid involved – anterior wall removed, dissect subperiosteal plane – free from nasopharyngeal wall,

• Resect post septum with mass if adhere.

• If extend into cavernous sinus / infratemporal fossa – nasal-nasopharyngeal portion divided 1st and then use diode laser, remove transnasal / transoral

• Pterygoid process involved – skull base drilled; disclose tumour along Vidian nerve
Endoscopic surgery

- Int J Pedi ORL, Jan 2004, Egypt; endoscopic-assisted midfacial degloving approach for type III JNA. Good exposure and cosmoses. 2/15 recurrence. 1/15 CSF rhinorrhea intraop, Rx imm.

- Int J Pedi ORL, Nov ‘03, Turkey; endoscopic surgery, no preop embolisation. 12 pt, bled 1 – 1,5 l (IIIA). Suggest for small tumor (<III)

- J Otolaryngol. Aug ’03, China; 12 pt. Huge lobulated JNA with multiple sites: choice is transantral approach via midfacial degloving with one other approach PRN