Juvenile angiofibroma

E Post

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

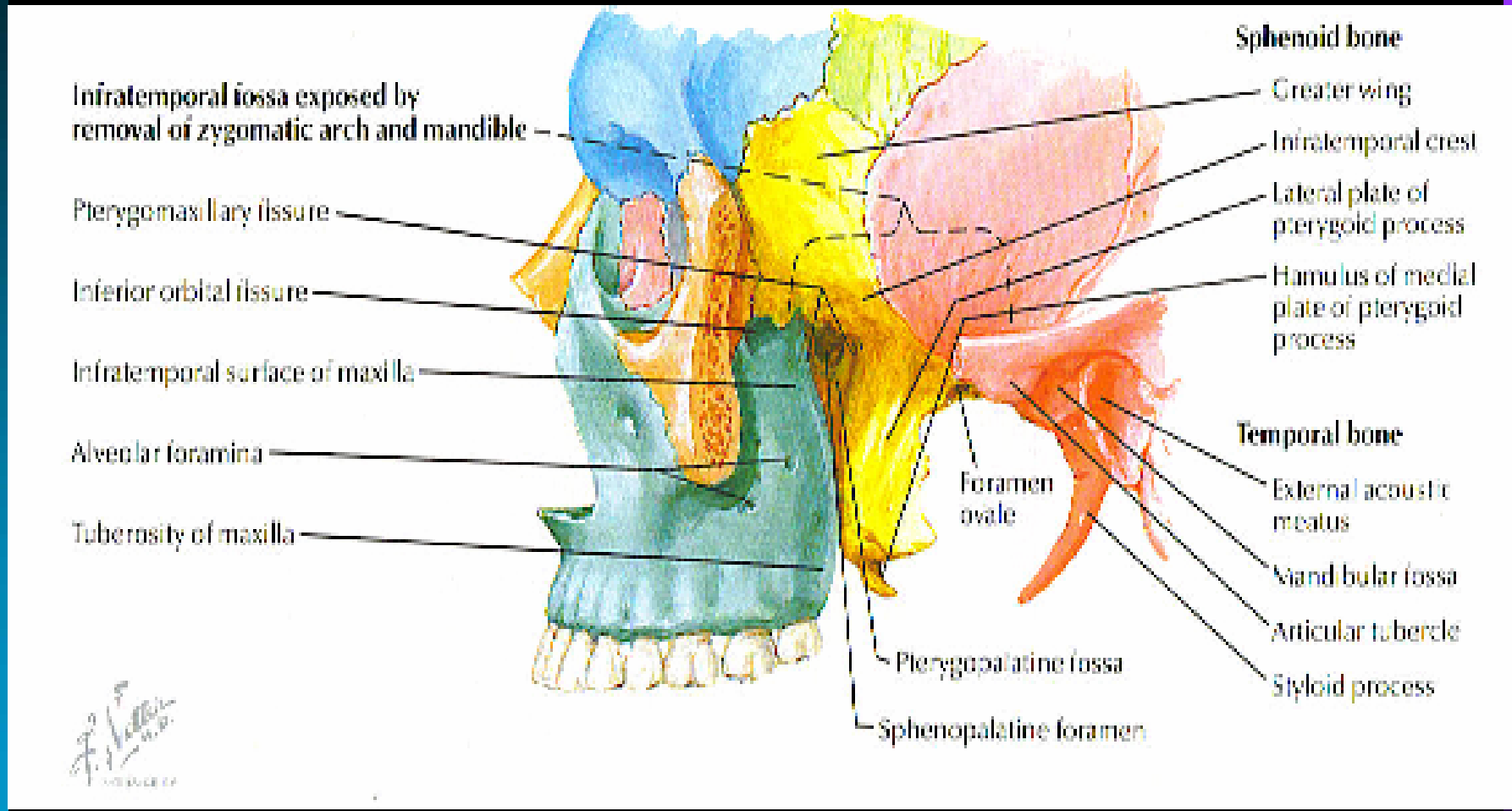
- Grows: Foramina + fissures; displace structures
  Large may erode bone
  Develop collateral blood supply

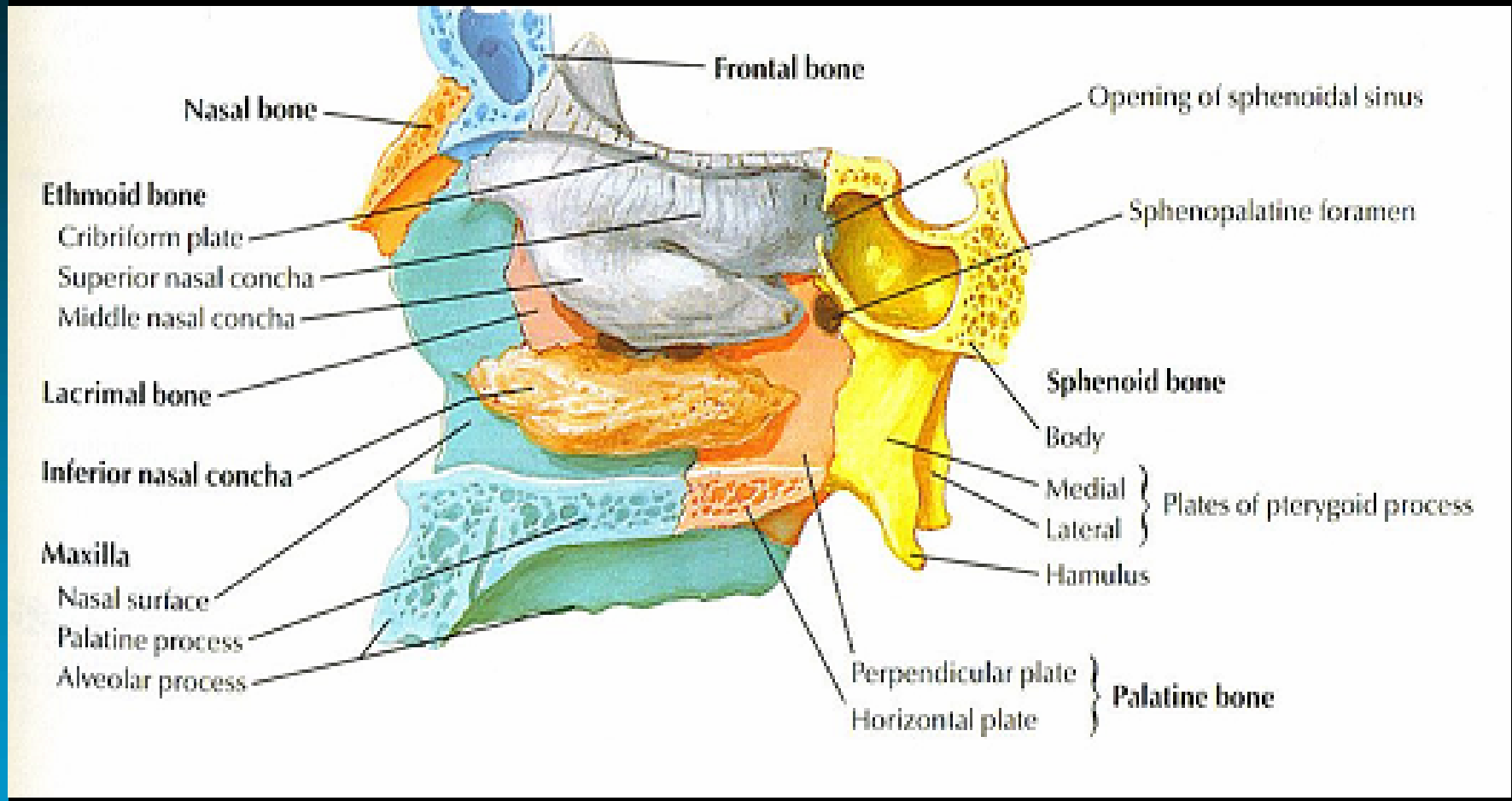
Tumour spread

- Lateral: Sphenopalatine foramenl – pterygomaxillary fissure – infratemporal fossa – cheek
- 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
Foramen ovale
Pterygopalatine fossa
Sphenopalatine foramen


Frontal bone
Nasal bone
Opening of sphenoidal sinus
Ethmoid bone
Cribriform plate
Superior nasal concha
Middle nasal concha
Sphenopalatine foramen
Lacrimal bone
Sphenoid bone
Body
Inferior nasal concha
Medial
Lateral
Plates of pterygoid process
Maxilla
Hamulus
Nasal surface
Palatine process
Alveolar process
Perpendicular plate
Horizontal plate
Palatine bone

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

I	Tumour limited to nasopharyngeal cavity; bone destruction negligible or limited to sphenopalatine foramen
II	Invading pterygopalatine fossa or maxilla, ethmoid, or sphenoid sinus with bone destruction
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

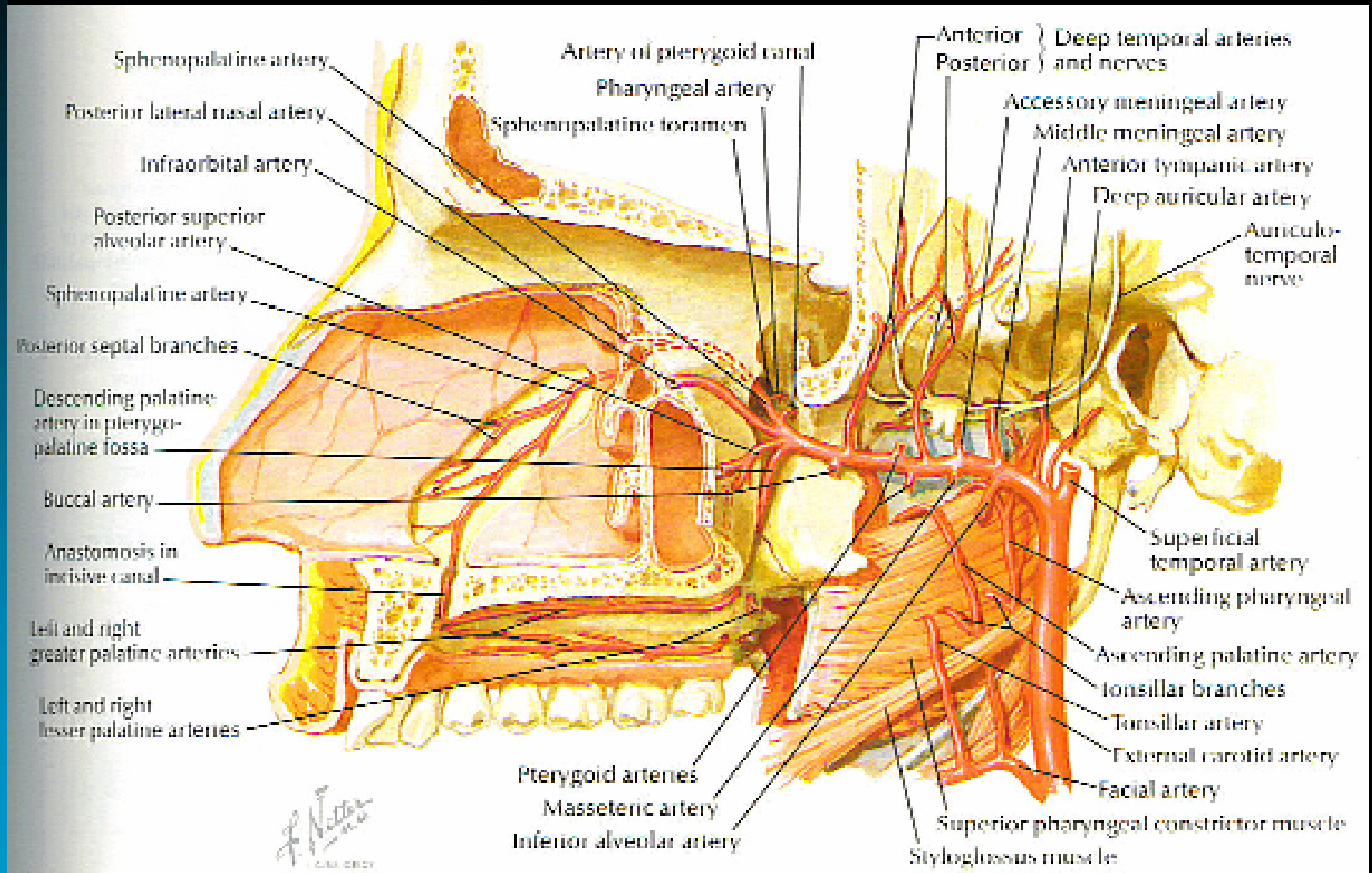
  Superficial temporal / External facial arteries –

  - 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 in pterygo-palatine fossa
Buccal artery
Anastomosis in incisive canal
Left and right greater palatine arteries
Left and right lesser palatine arteries
Artery of pterygoid canal
Pharyngeal artery
Sphenopalatine foramen
Pterygoid arteries
Masseteric artery
Inferior alveolar artery
Anterior
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

IADSA

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 JA: 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 Trendellenburg, 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