

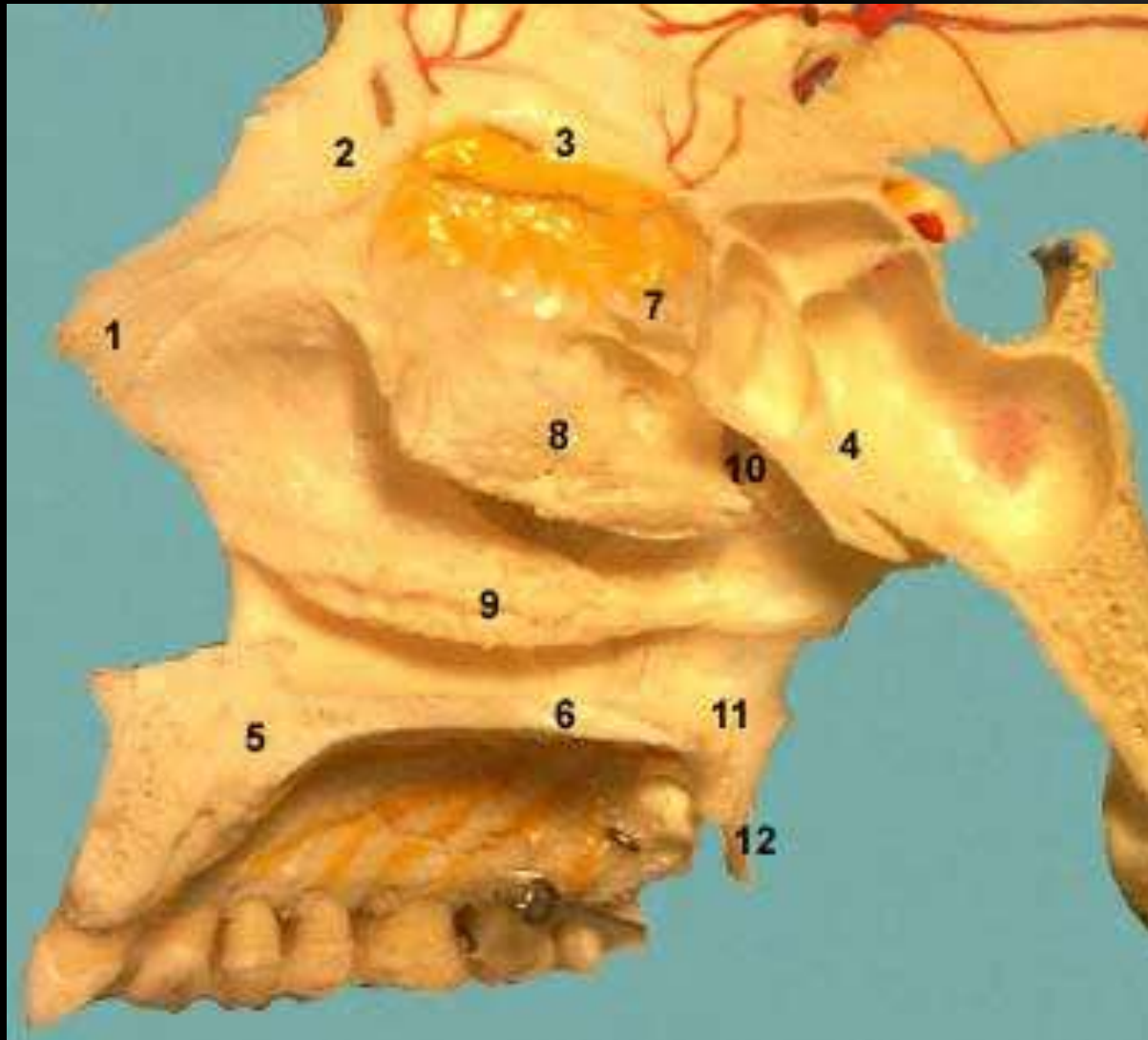


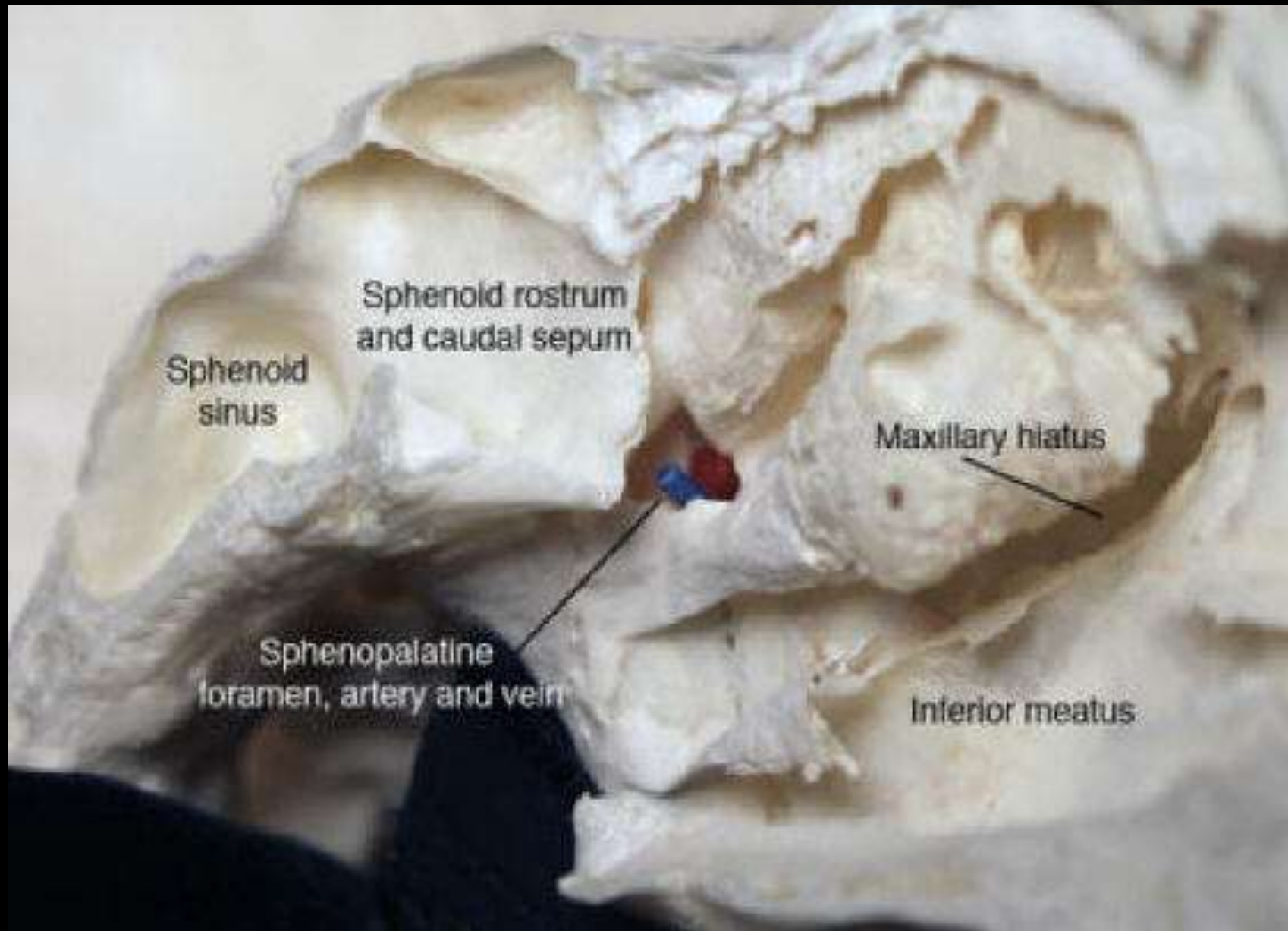
Juvenile Angiofibroma

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Sphenopalatine Foramen





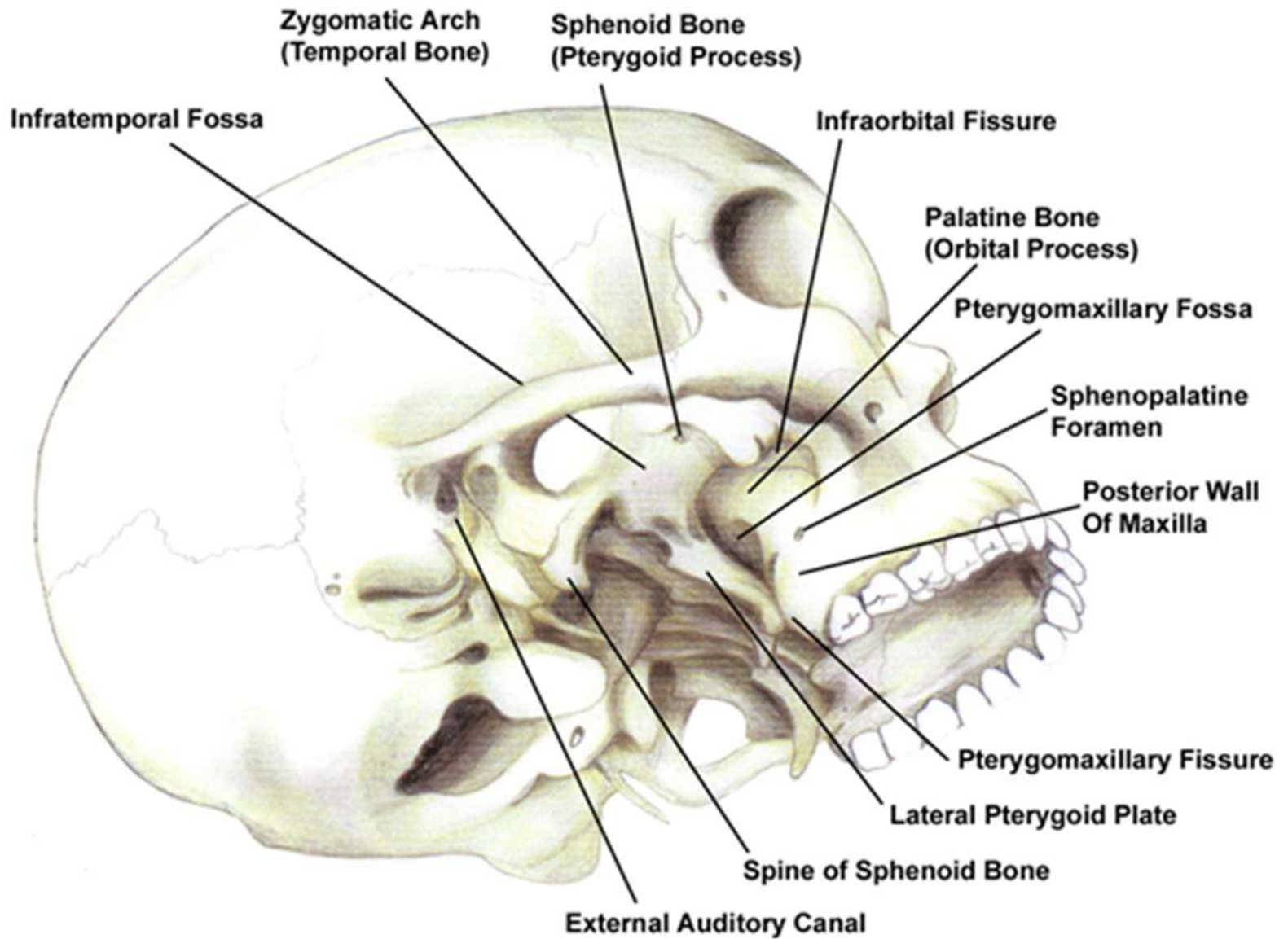
Sphenoid sinus

Sphenoid rostrum and caudal septum

Maxillary hiatus

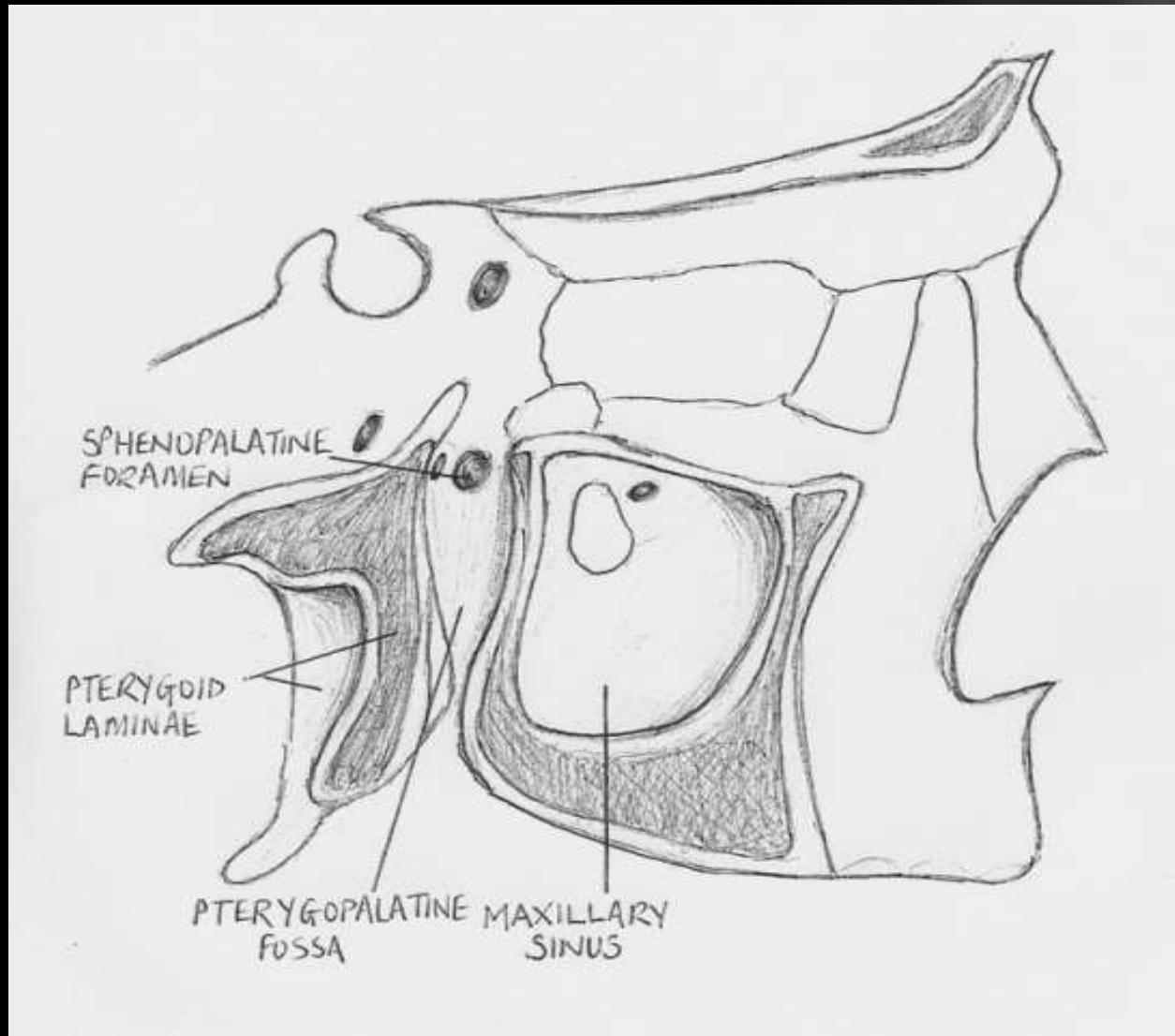
Sphenopalatine foramen, artery and vein

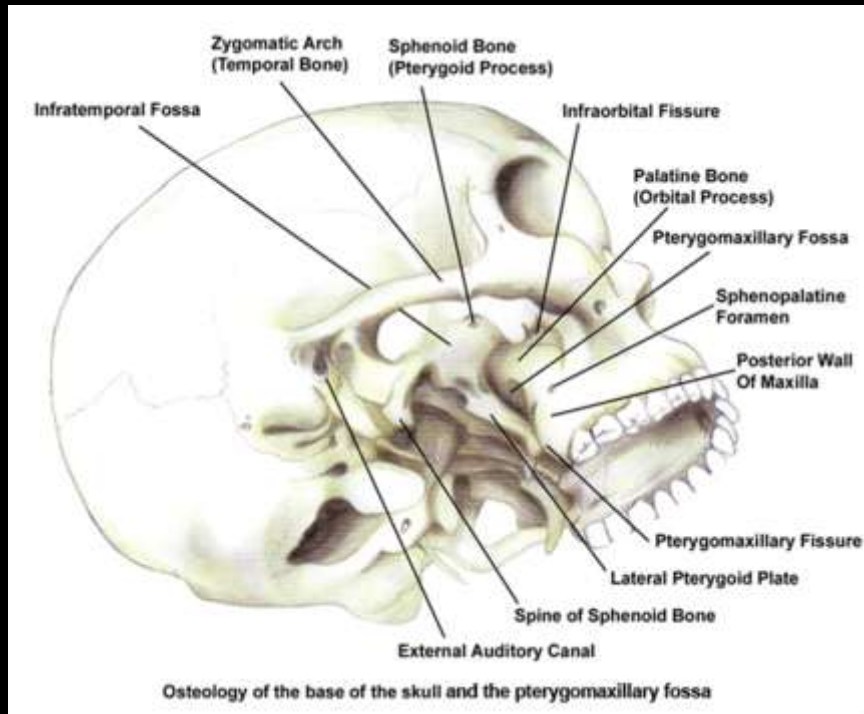
Inferior meatus



Osteology of the base of the skull and the pterygomaxillary fossa

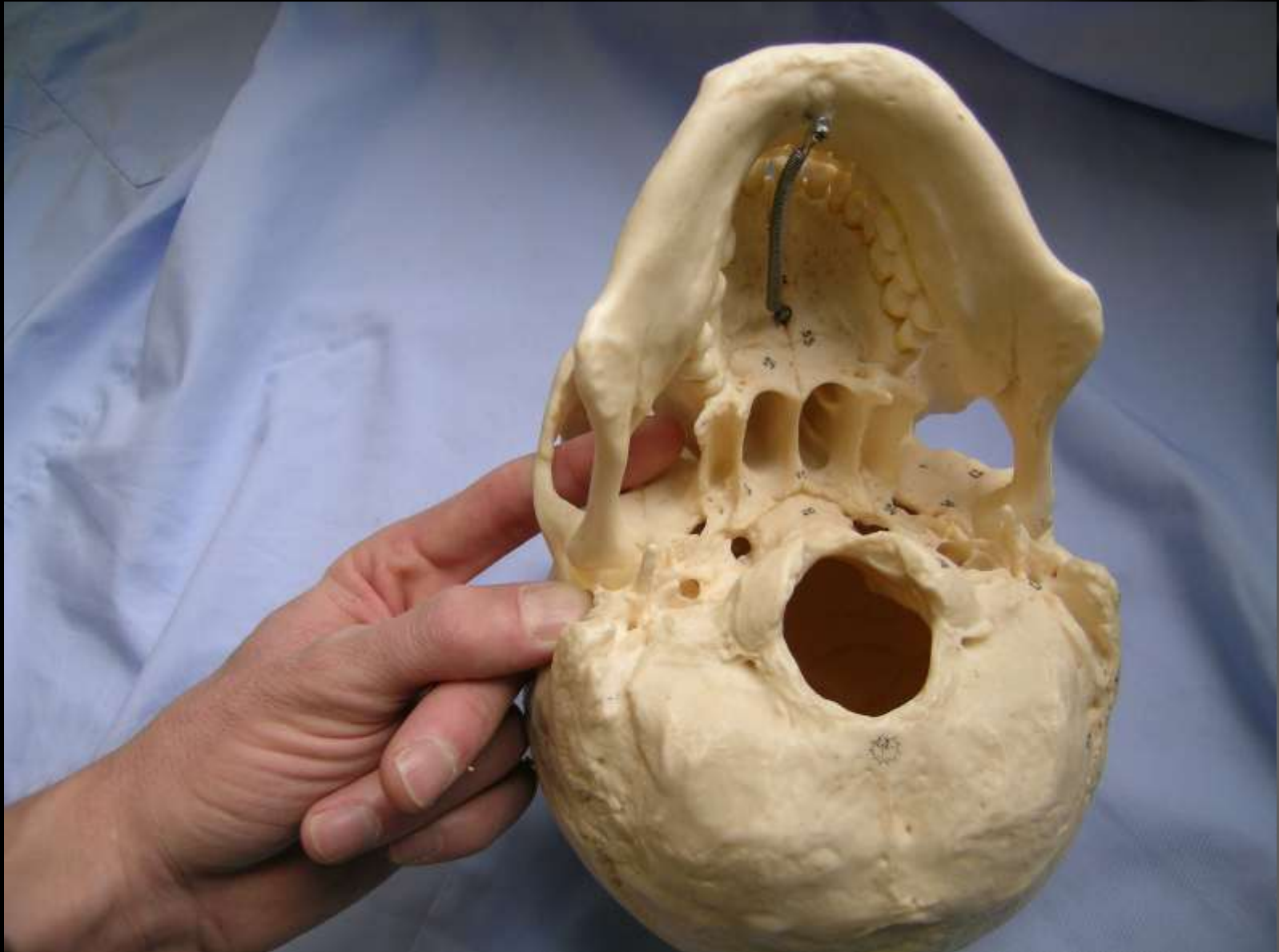
Pterygopalatine Fossa





- Laterally with the infratemporal fossa through the pterygomaxillary fissure
- Anteriorly to the orbit via the infraorbital fissure
- Posteriorly to the middle cranial fossa through the foramen rotundum and the pterygoid canal
- Medially to the inferior portion of the sphenoid recess through the sphenopalatine foramen
- To the oral cavity via the greater and lesser palatine foramina.

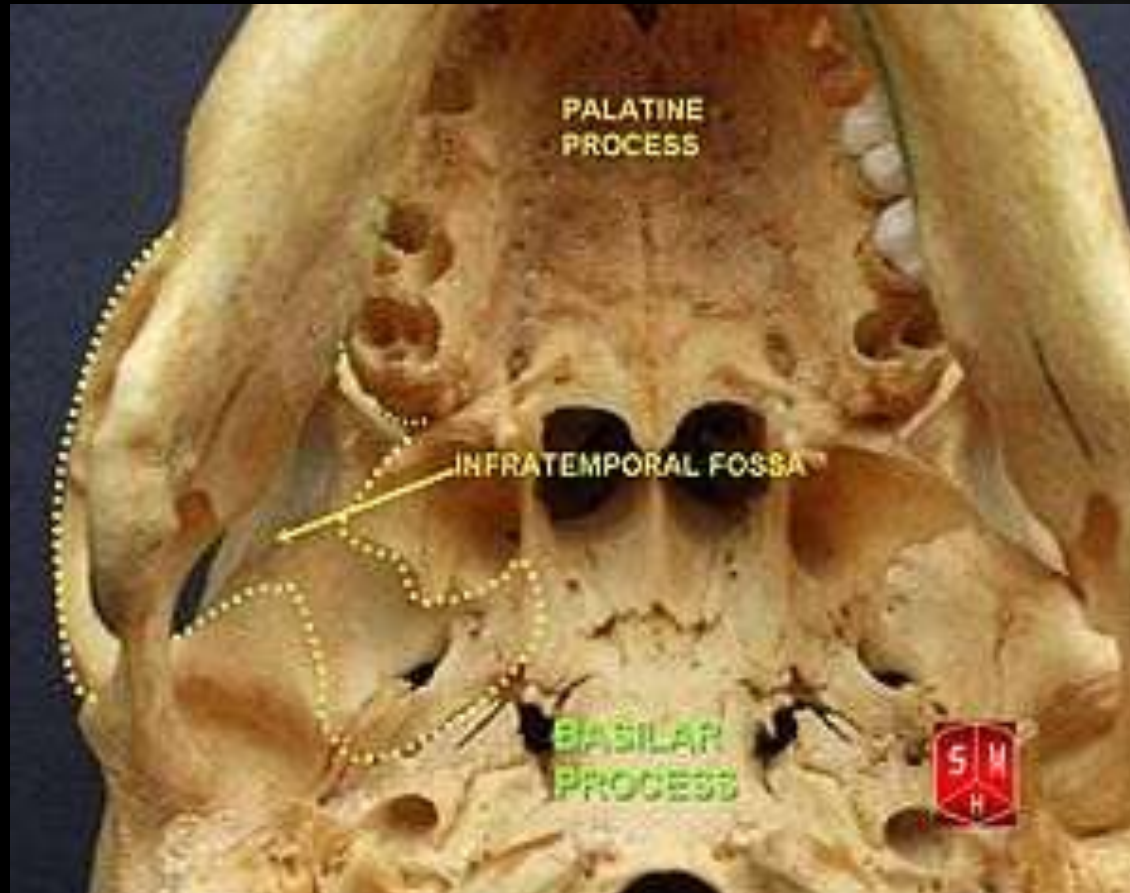
Infratemporal Fossa



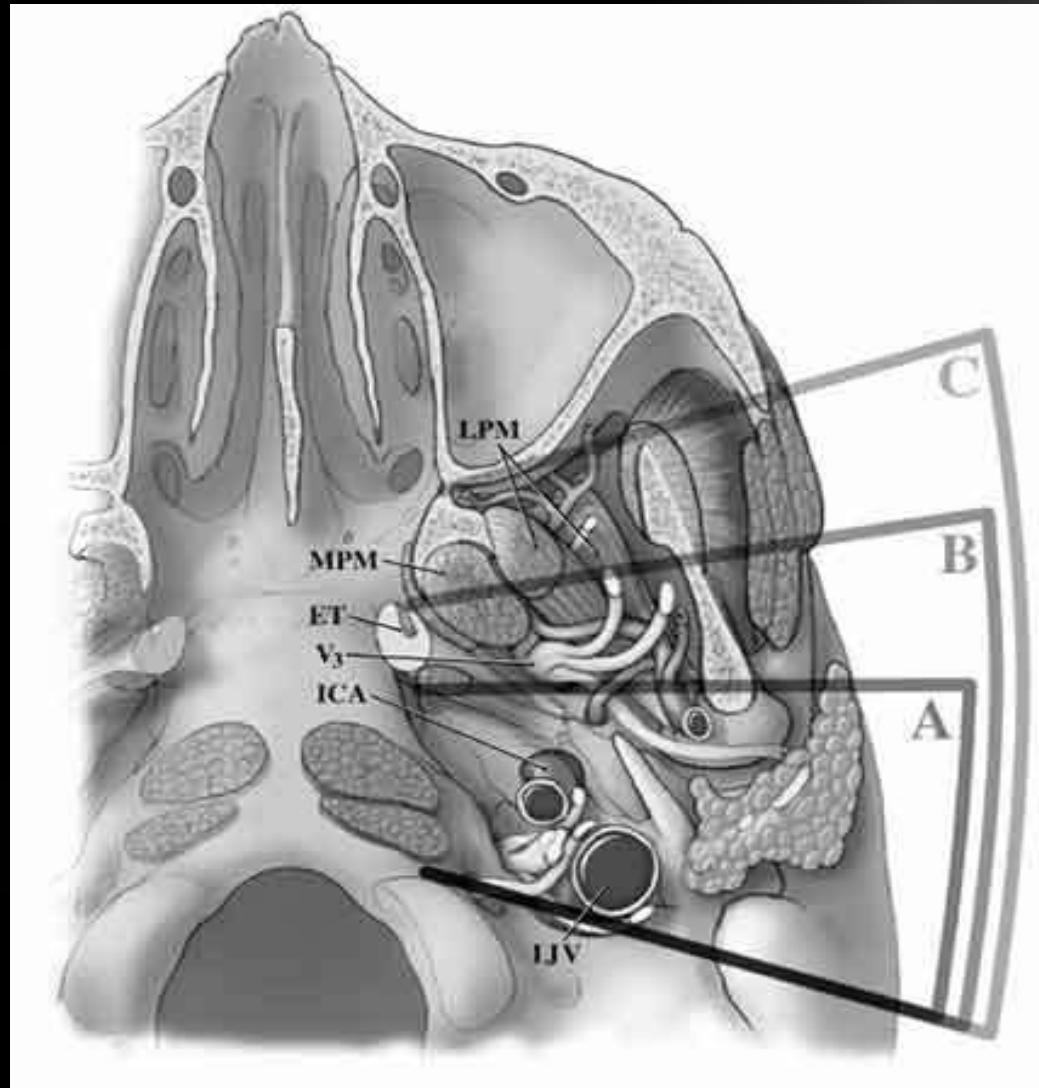
Infratemporal Fossa



Infratemporal Fossa



Infratemporal Fossa



Juvenile nasopharyngeal angiofibroma (JNA)

- ❖ It is a rare, benign, vascular neoplasm that accounts for less than 0.5% of all head and neck tumours.
- ❖ JNAs occur almost exclusively in the nasopharynx of adolescent males.
- ❖ The site of origin of JNA remains controversial.

Age:

- ❖ Onset most commonly is in the second decade; range is 7-21 years.
- ❖ Mean age at diagnosis is 14 years
- ❖ May regress in late teens but may persist into adulthood
- ❖ Rare in patients older than 25 years.

Etiopathogenesis

Many theories have been propounded but none is entirely convincing

Hormonal theory has been suggested due to the lesion's occurrence in adolescent males

Other theories include:

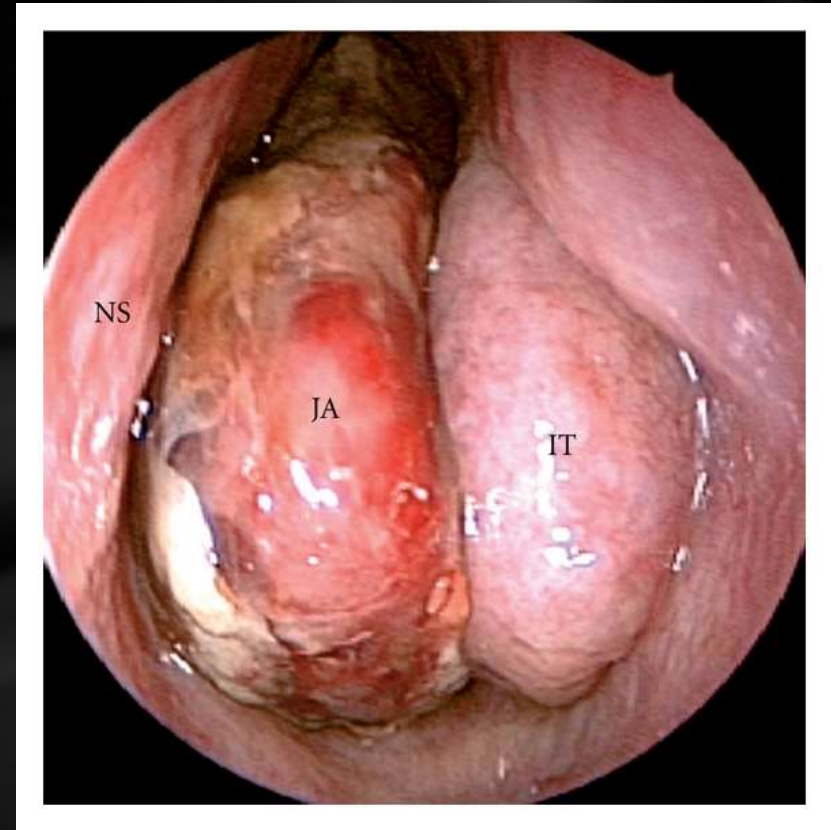
- ❖ Desmoplastic response of the nasopharyngeal periosteum or
- ❖ Embryonic fibrocartilage between the basiocciput and the basisphenoid
- ❖ Hamartomas
- ❖ Nest cells [undiff. epitheloid]
- ❖ Vestiges of atrophied stapedia artery

Site of Origin

- ❖ 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.
- ❖ Bone of the vidian canal

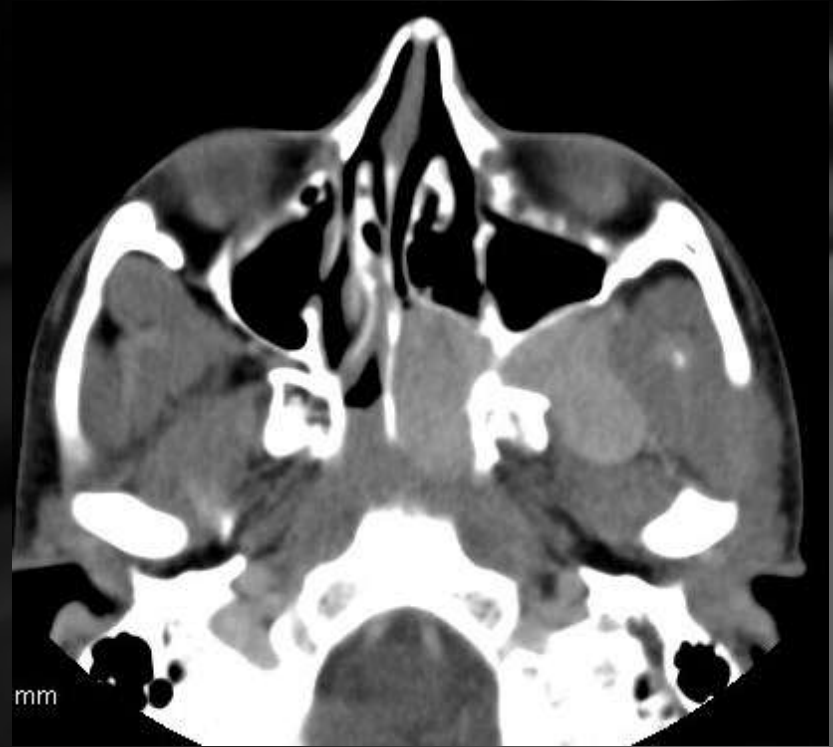
Gross Examination

- ❖ Usually sessile, lobulated, rubbery, and red-pink to tan-gray in appearance.
- ❖ In rare cases, the tumor is polypoid or pedunculated
- ❖ Usually is encapsulated and composed of vascular tissue and fibrous stroma with coarse or fine collagen fibers.



Spread

- ❖ 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.

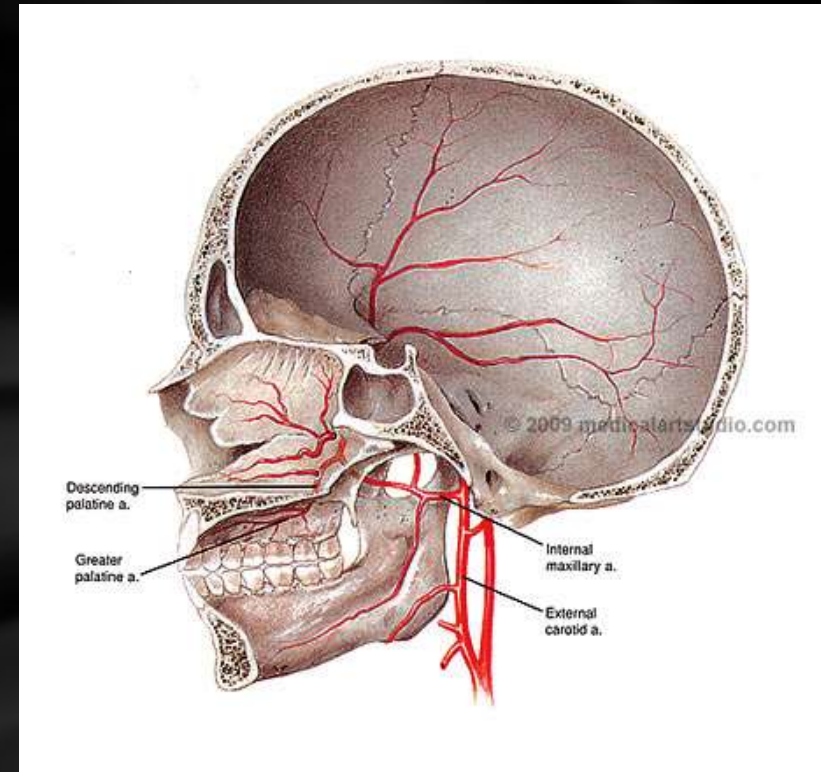


The blood supply to these benign tumours is most commonly from the internal maxillary artery.

May also be supplied by the:

- External carotid artery
- Internal carotid artery
- Common carotid artery
- Ascending pharyngeal artery

Blood Supply

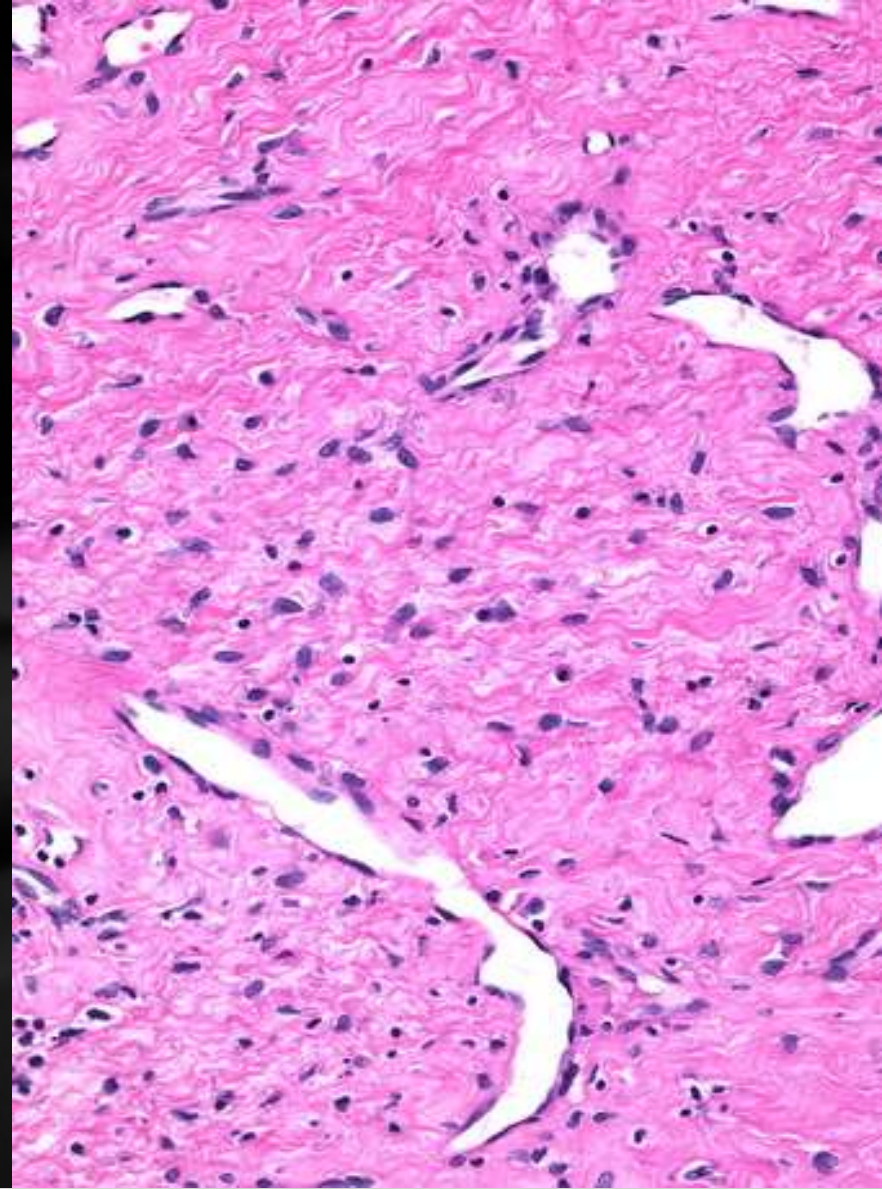


Structure

- ❖ Histologically, JNAs originate from myofibroblasts.
- ❖ The tumour 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.

Histopathology

The tumour is composed of a variable admixture of blood vessels and fibrous tissue



Presentation

- Unilateral nasal obstruction.
- **Epistaxis**
- Nasopharyngeal mass in adolescent males with an average age of onset of 15 years of age

Not so rare presentations

- ❖ Conductive hearing loss
- ❖ Dacrocystitis
- ❖ Rhinolalia
- ❖ Hard and soft palate deformity
- ❖ Hyposmia or anosmia

Advanced lesions may cause

- Facial swelling
- Proptosis
- Cranial neuropathy
- Massive hemorrhage



Investigations

Plain x-ray:

- ❖ View of the sinuses may demonstrate nasopharyngeal polyp.
- ❖ Bowing of the posterior wall of the maxillary sinus and maxillary sinus opacification is very suggestive of JNA.
- ❖ Newer radiographic modalities have surpassed plain films in usefulness



Holman-Miller sign

- ❖ CT Scan is excellent for evaluation of bone detail and will enhance with contrast.
- ❖ The characteristic anterior bowing of the posterior maxillary wall due to the presence of a mass in the pterygomaxillary space known as the Holman-Miller sign is a finding noted on CT Scan.



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MRI





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T1 FS +Gd axial

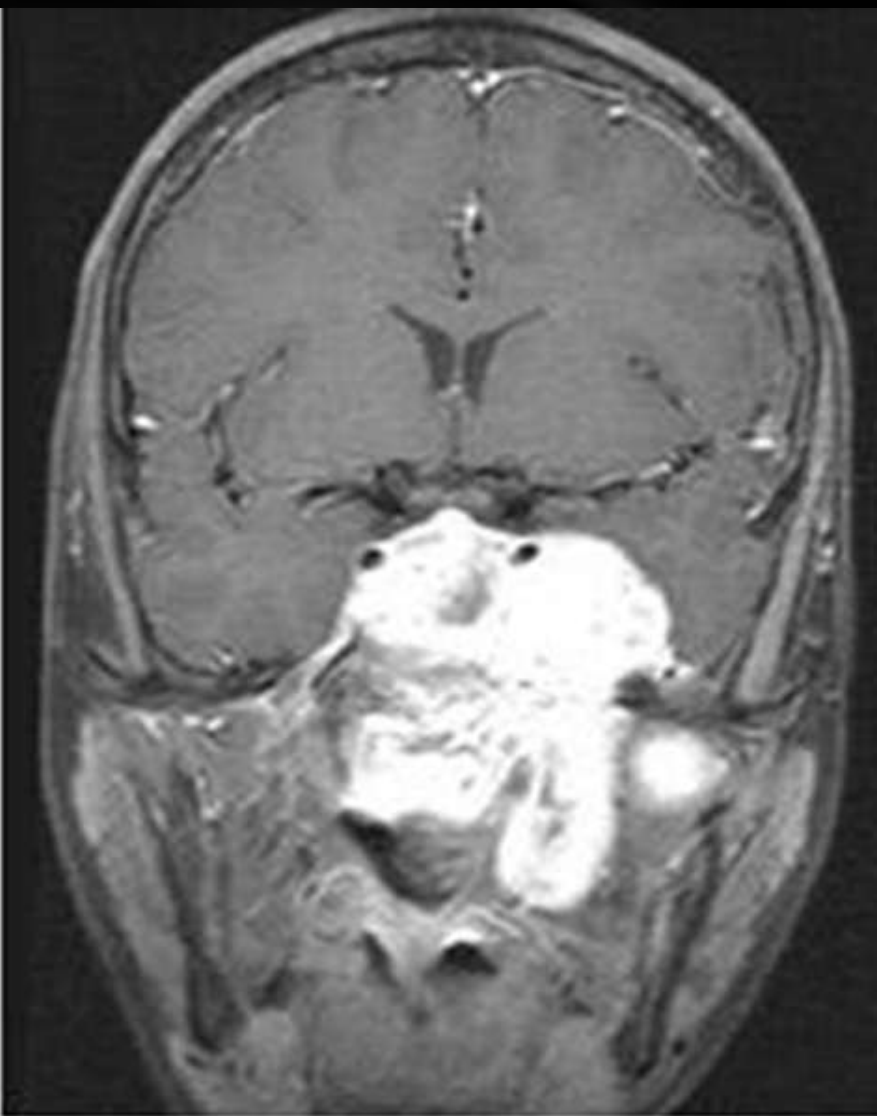
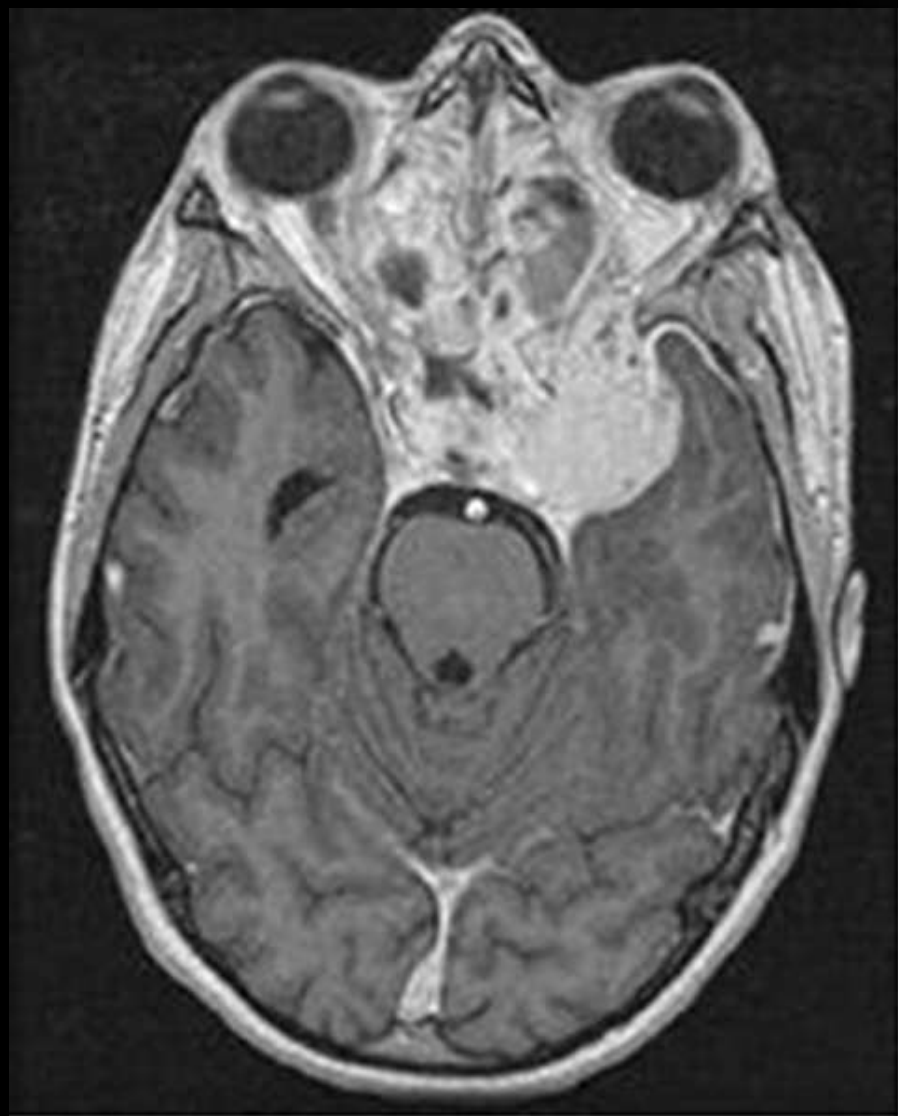
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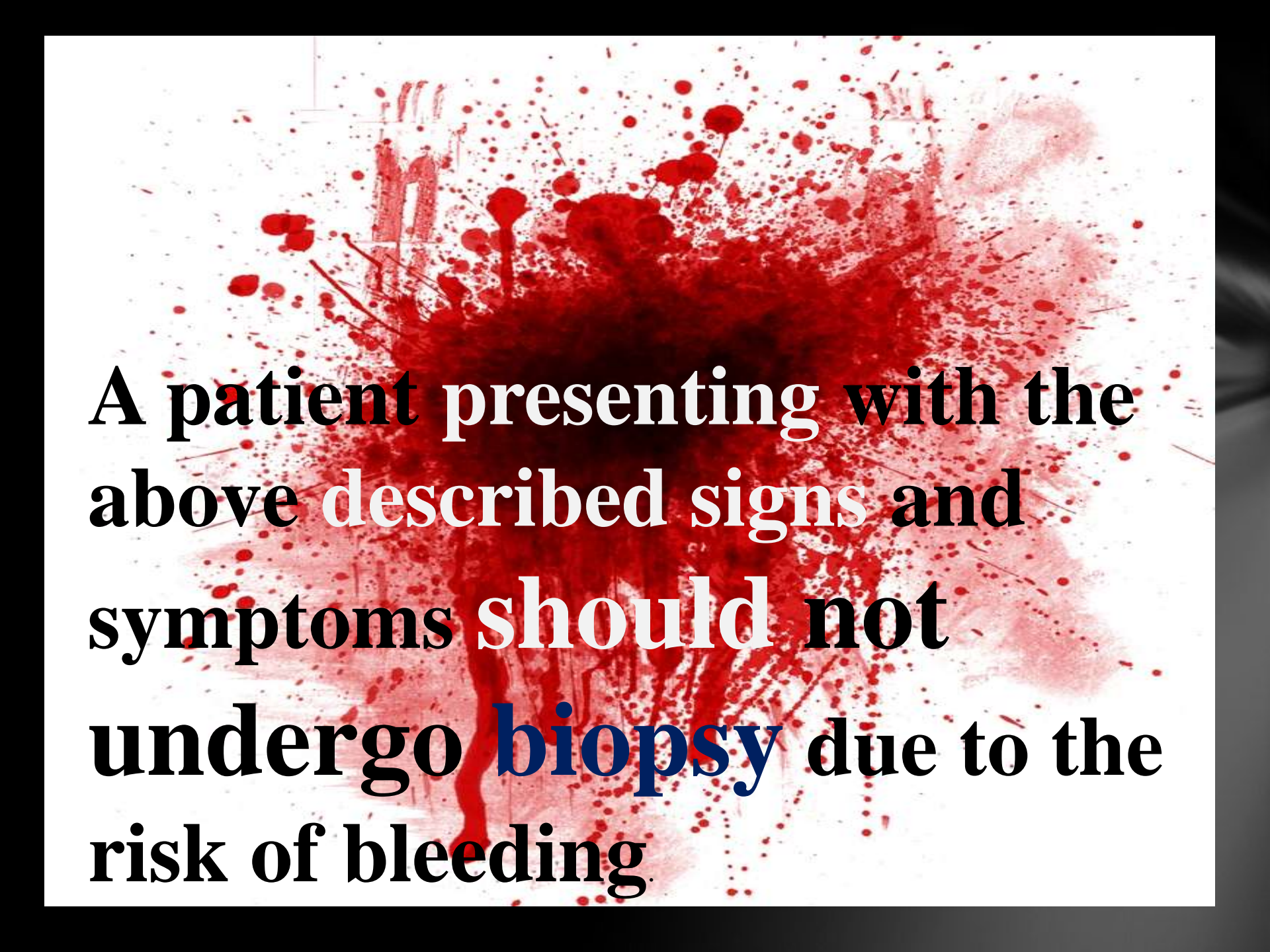
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Angiography



Angiography confirms the hyper vascularity of the lesion, which is supplied by a hypertrophic maxillary artery (arrow).



**A patient presenting with the
above described signs and
symptoms should not
undergo biopsy due to the
risk of bleeding.**

Staging: Classification according to Fisch

Stage I - Tumors limited to nasal cavity, nasopharynx with no bony destruction

Stage II - Tumors invading pterygomaxillary fossa, paranasal sinuses with bony destruction

Stage III - Tumors invading infratemporal fossa, orbit and/or parasellar region remaining lateral to cavernous sinus

Stage IV - Tumors invading cavernous sinus, optic chiasmal region, and/or pituitary fossa

Differentials

Other causes of nasal obstruction:

- ❖ Nasal polyps
- ❖ Antrochoanal polyp
- ❖ Teratoma
- ❖ Encephalocele
- ❖ Dermoids
- ❖ Inverting papilloma
- ❖ Rhabdomyosarcoma
- ❖ Squamous cell carcinoma

Treatment options for JNAs

- ❖ Surgery
- ❖ Radiation therapy
- ❖ Chemotherapy
- ❖ Hormone therapy
- ❖ Surgery is the gold standard of treatment

External beam radiation

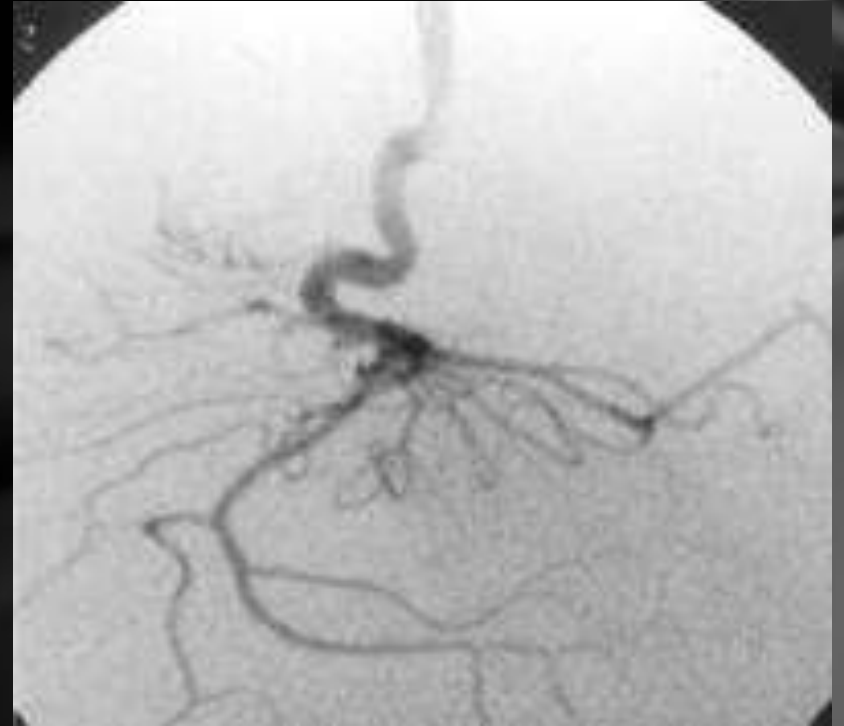
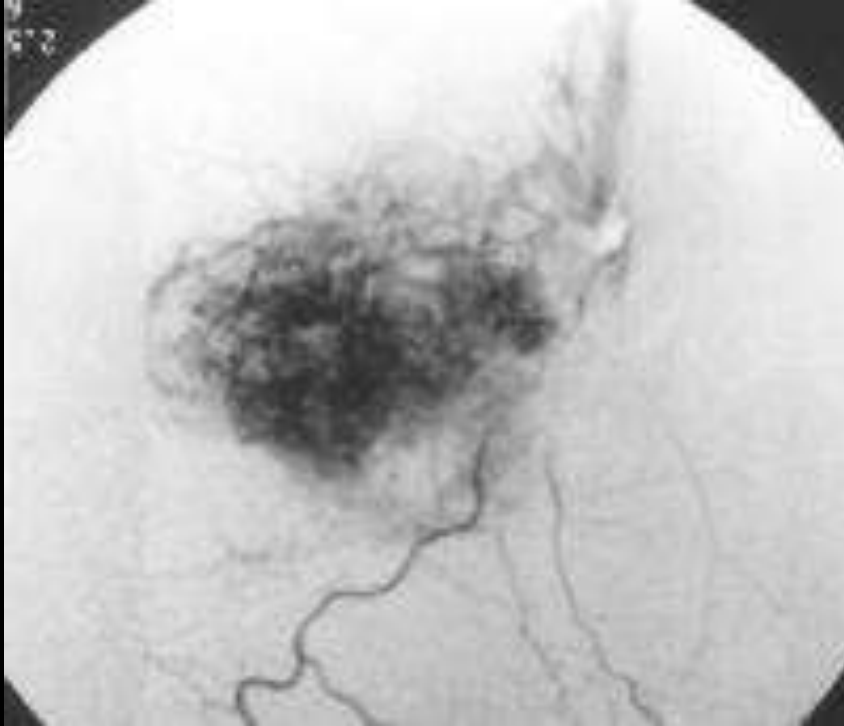
- ❖ Generally reserved for larger and/or unresectable tumors and tumors that are life threatening do to their location.
- ❖ Reason for limited use of radiation:
 - ❖ Carcinogenic side effects
 - ❖ Growth retardation
 - ❖ Temporal lobe radionecrosis
 - ❖ Panhypopituitarism
 - ❖ Cataracts
 - ❖ Radiation induced keratopathy

- ❖ Chemotherapy is used when previous surgery and radiation have failed.
- ❖ Hormone therapy has been proposed due to the androgen receptors associated with JNAs in an attempt to decrease tumour size and vascularity.
- ❖ Estrogen has been shown to decrease size and vascularity of the tumour, but has feminizing side effects.

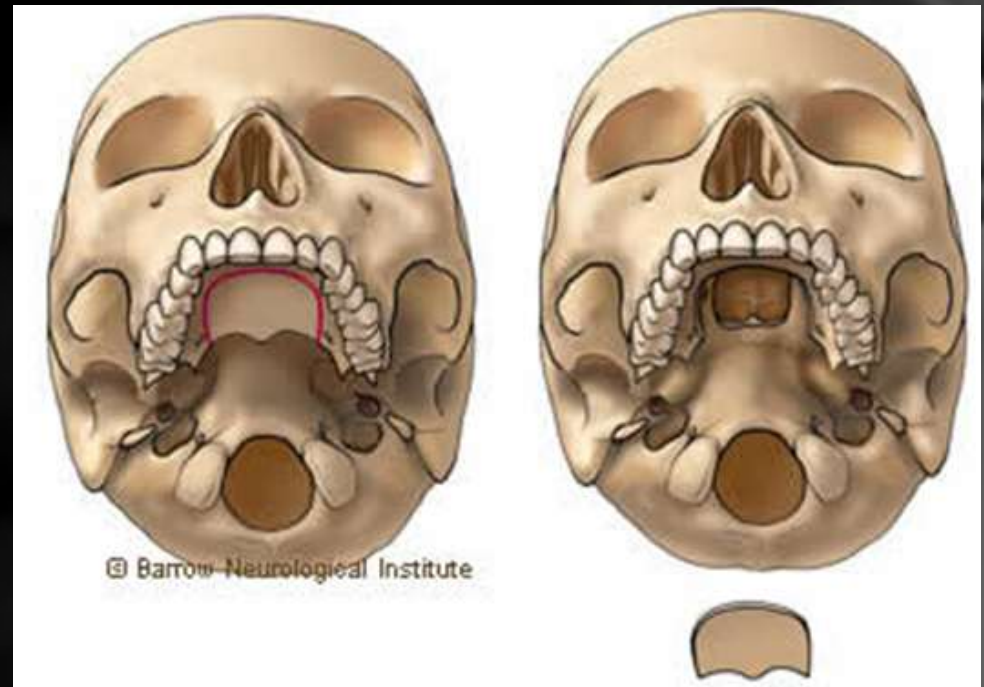
The treatment of choice in the vast majority of patients is surgical resection.

- ❖ Preoperative selective arterial embolization of feeding vessels from the external carotid artery has significantly decreased intraoperative blood loss and facilitated resection of larger tumors.
- ❖ Embolization is typically performed 24-72 hours prior to resection.
- ❖ Materials often used include gelfoam and polyvinyl alcohol foam.
- ❖ Gelfoam lasts approximately two weeks, while polyvinyl alcohol foam is more permanent.

Pre and Post Embolization



Lateral rhinotomy, transpalatal, transmaxillary, or sphenoethmoidal route is used for small tumors (Fisch stage I or II).



Endoscopic Excision

- ❖ It has been suggested that tumors involving the ethmoid, maxillary, or sphenoid sinus, the sphenopalatine foramen, nasopharynx, or pterygomaxillary fossa and have limited extension into the infratemporal fossa are amenable to endoscopic resection.
- ❖ JNAs that involve the orbit or middle cranial fossa are not ideal for endoscopic excision

Spontaneous Regression

- ❖ JNAs have the potential to regress which usually occurs when the patient is 20-25 years old.
- ❖ Complete regression does not occur in all patients.
- ❖ Spontaneous regression is valuable for residual tumor following treatment.
- ❖ Recurrence rates have been reported between 30 and 50%.