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Tumors of the Skull

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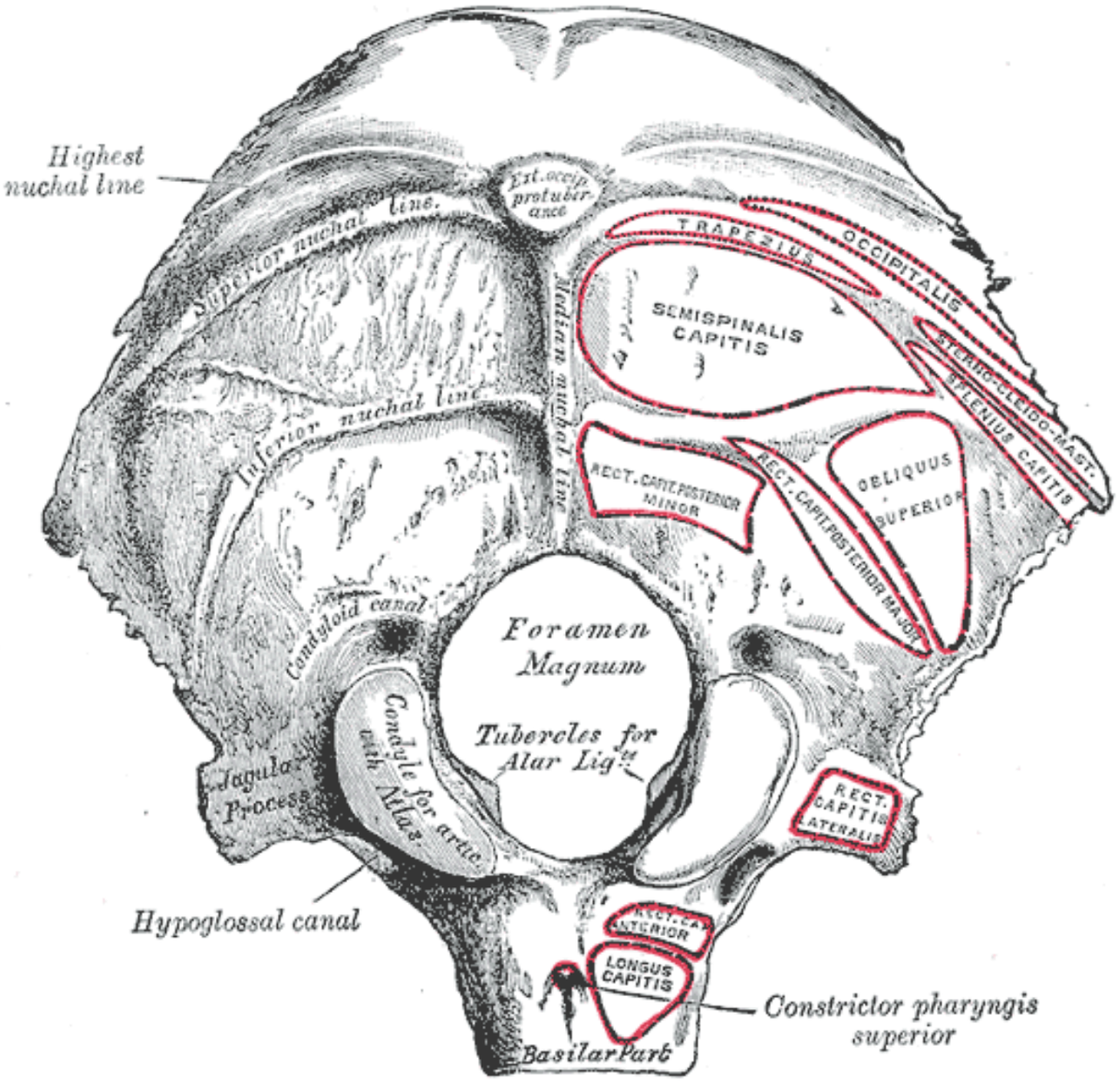
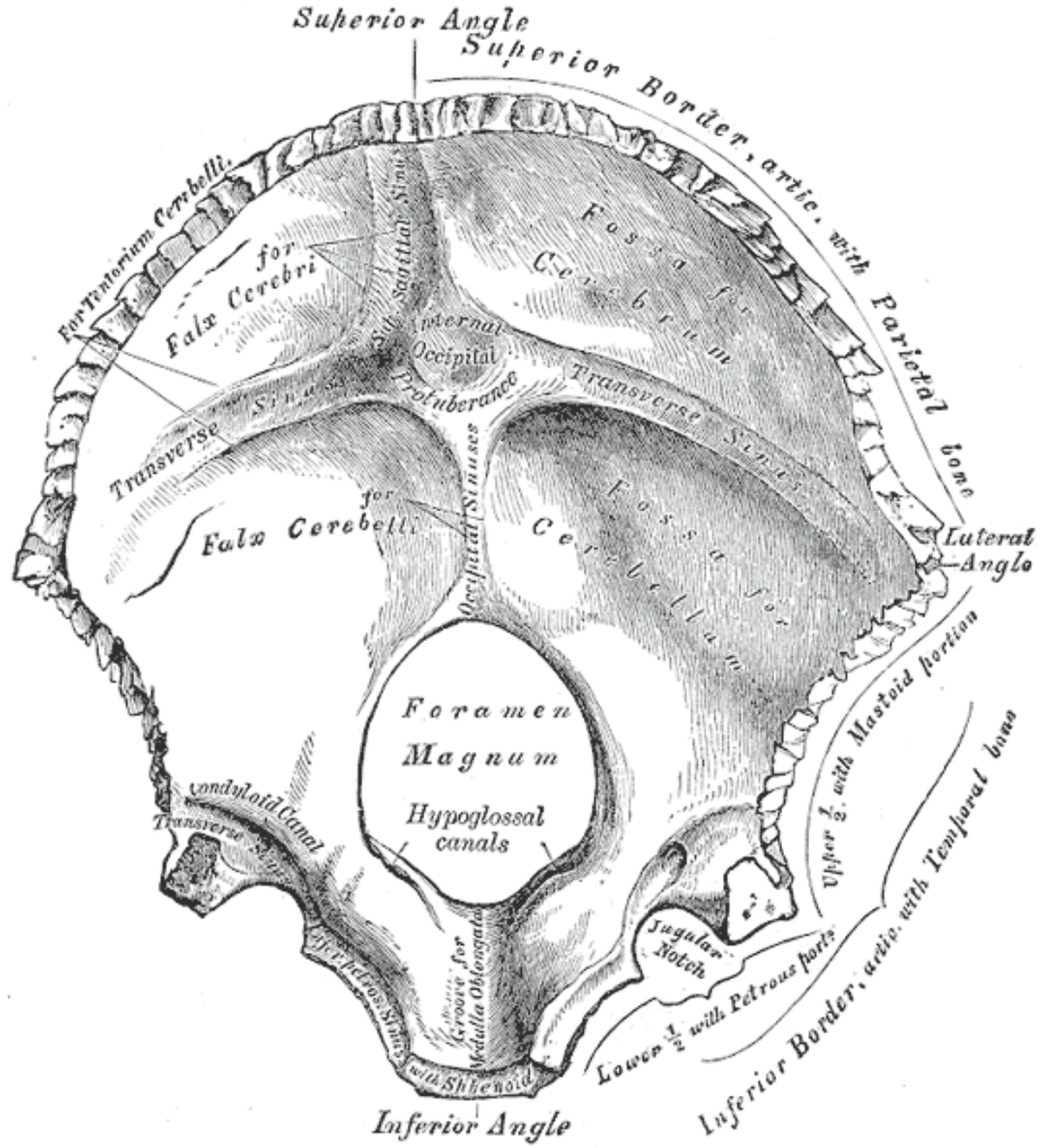
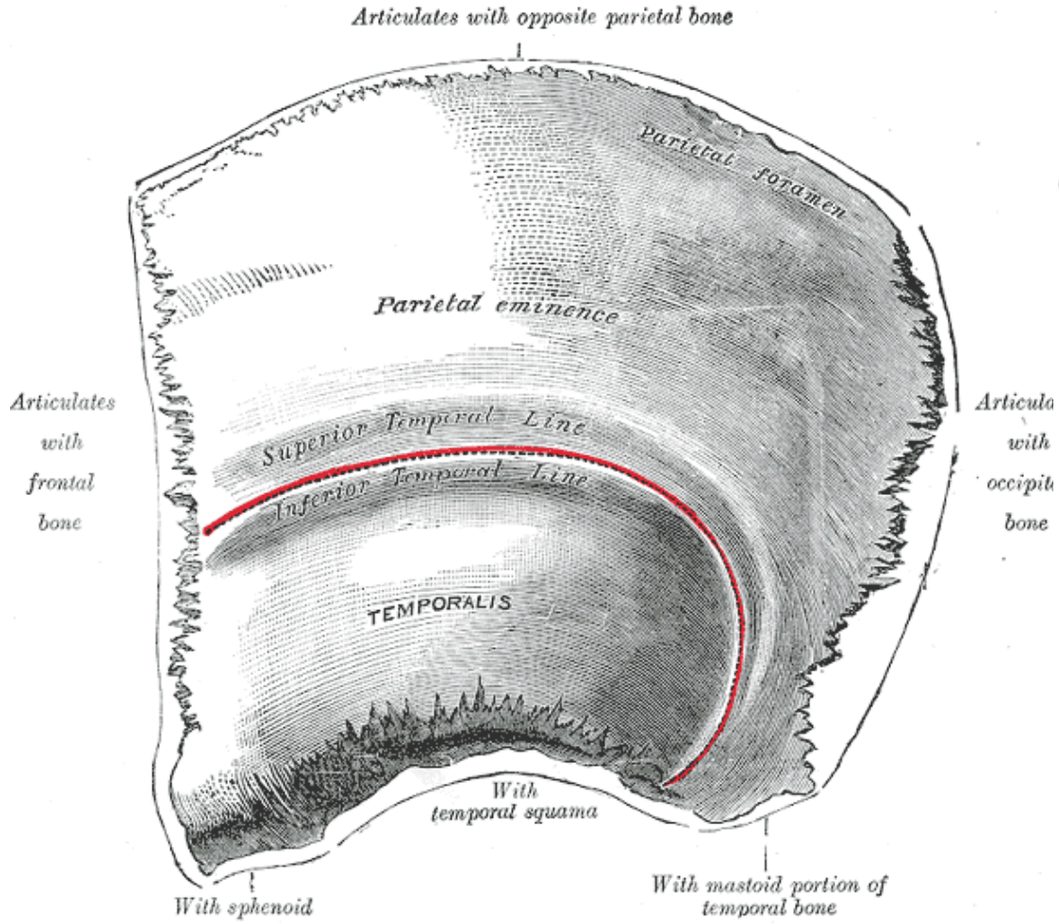
Outline

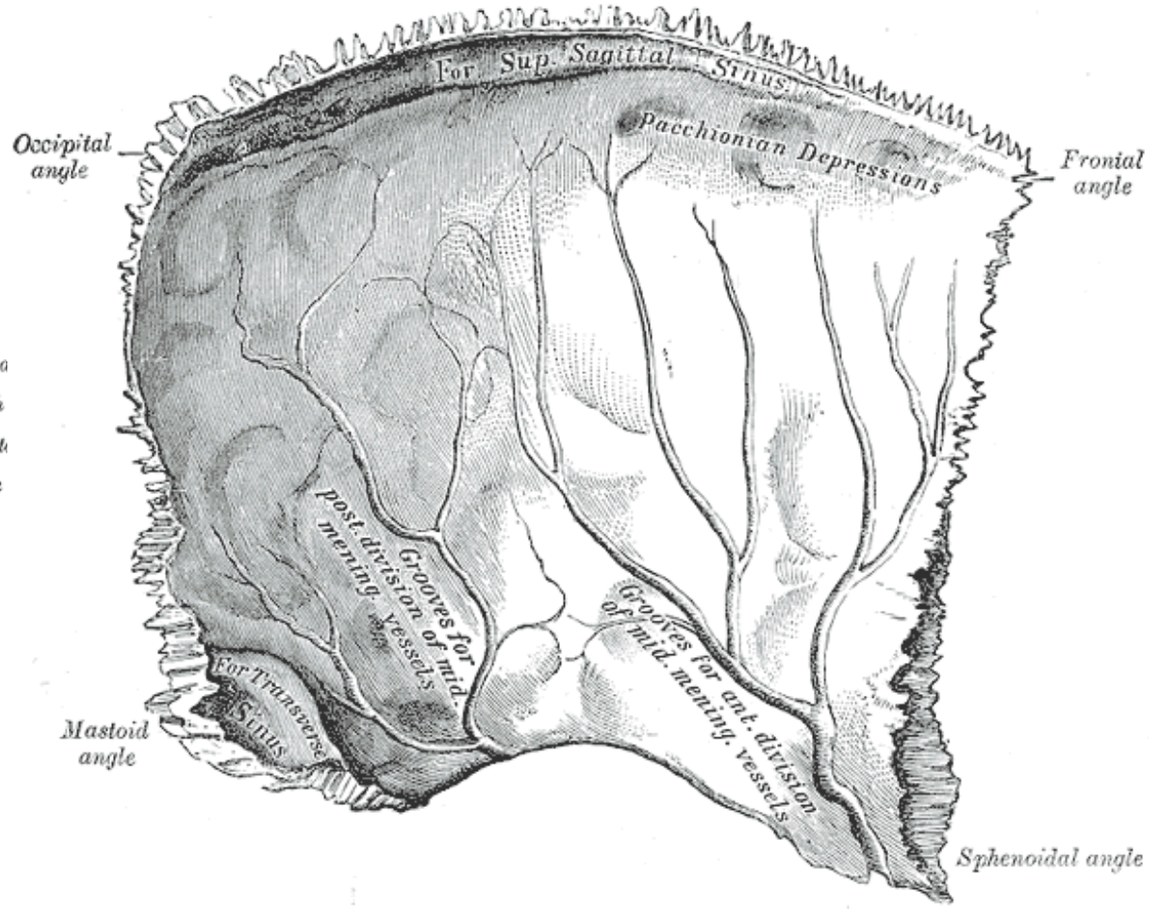
- Anatomy of the skull
- Non-neoplastic and Neoplastic lesions of the skull
- Esthesioneuroblastoma
- Chondroma and chondrosarcoma

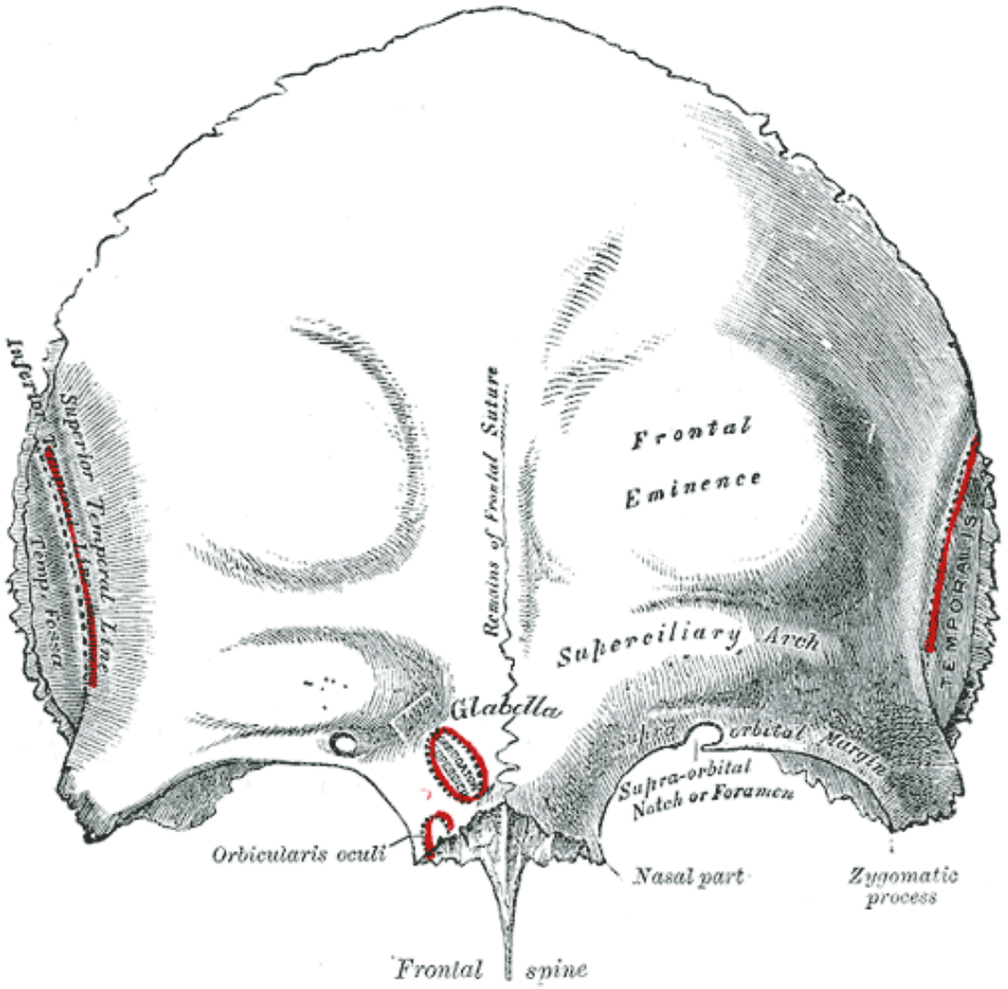
Anatomy of the Skull

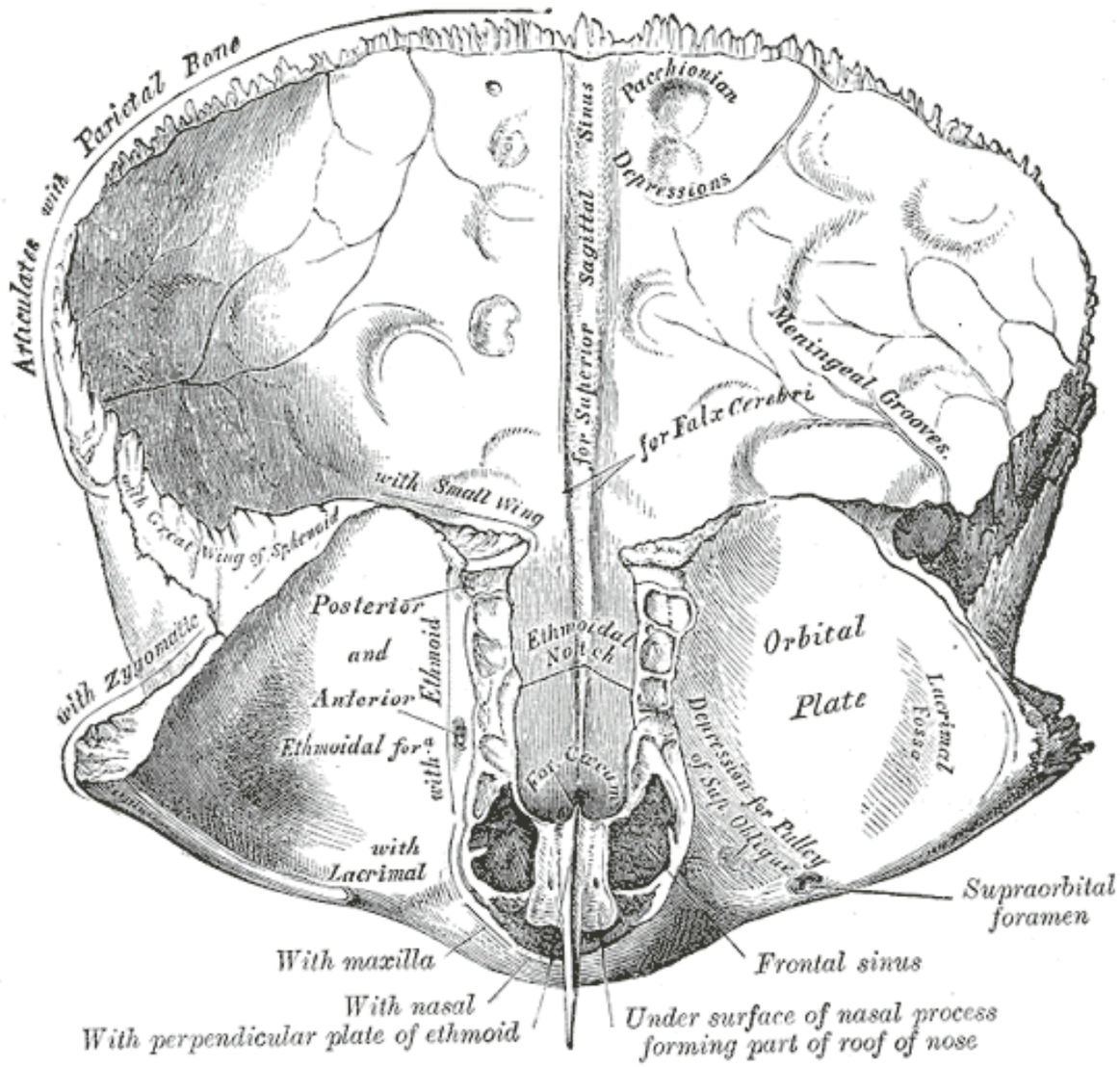

Highest nuchal line
Superior nuchal line
Inferior nuchal line
Ext. occip. protuber-ance
Median nuchal line
TRAPEZIUS
OCCIPITALIS
SEMISPINALIS CAPITIS
STERNO-CLEIDO-MAST.
SPLENIUS CAPITIS
RECT. CAPIT. POSTERIOR MINOR
RECT. CAPIT. POSTERIOR MAJOR
OBLIQUUS SUPERIOR
Condyloid canal
Foramen Magnum
Tubercles for Alar Lig.ts
Condyle for artic. with Atlas
Jugular Process
RECT. CAPITIS LATERALIS
Hypoglossal canal
RECT. CA. ANTERIOR
LONGUS CAPITIS
Constrictor pharyngis superior
Basilar Part

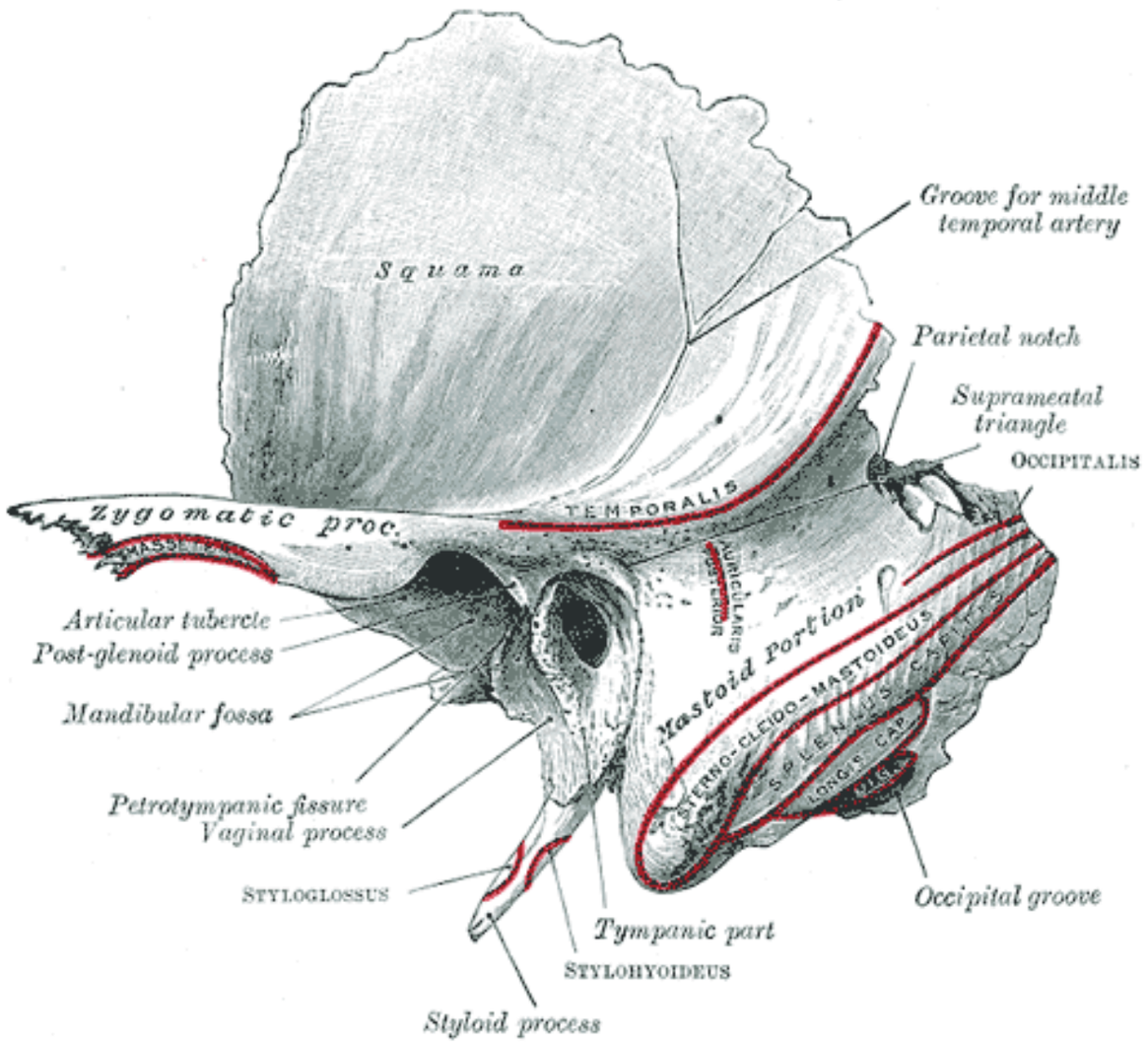

Superior Angle
Superior Border, artic. with Parietal bone
For Tentorium Cerebelli
for Falx Cerebri
Sagittal Sinus
Fossa for Cerebrum
Internal Occipital Protuberance
Transverse Sinus
Transverse
for Falx Cerebelli
Occipital Sinuses
Fossa for Cerebellum
Lateral Angle
Foramen Magnum
Hypoglossal canals
Condyloid Canal
Transverse Sinus
Groove for Arteria Occipitalis
Jugular Notch
Upper ½ with Mastoid portion
Lower ½ with Petrous port.
Inferior Border, artic. with Temporal bone
with Sphenoid
Inferior Angle

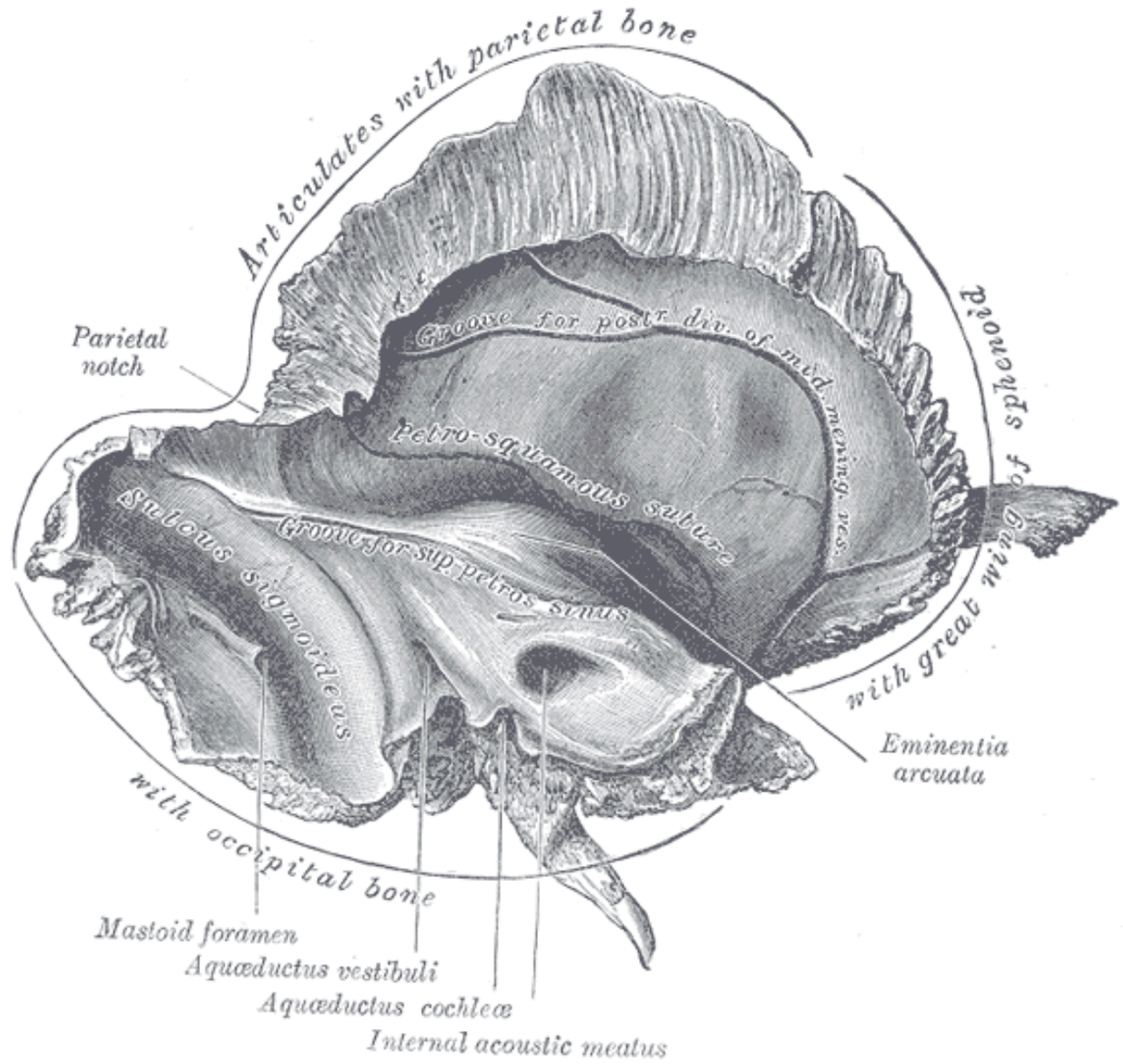

Articulates with opposite parietal bone
Parietal foramen
Parietal eminence
Articulates with frontal bone
Superior Temporal Line
Inferior Temporal Line
TEMPORALIS
Articula with occipit bone
With temporal squama
With sphenoid
With mastoid portion of temporal bone

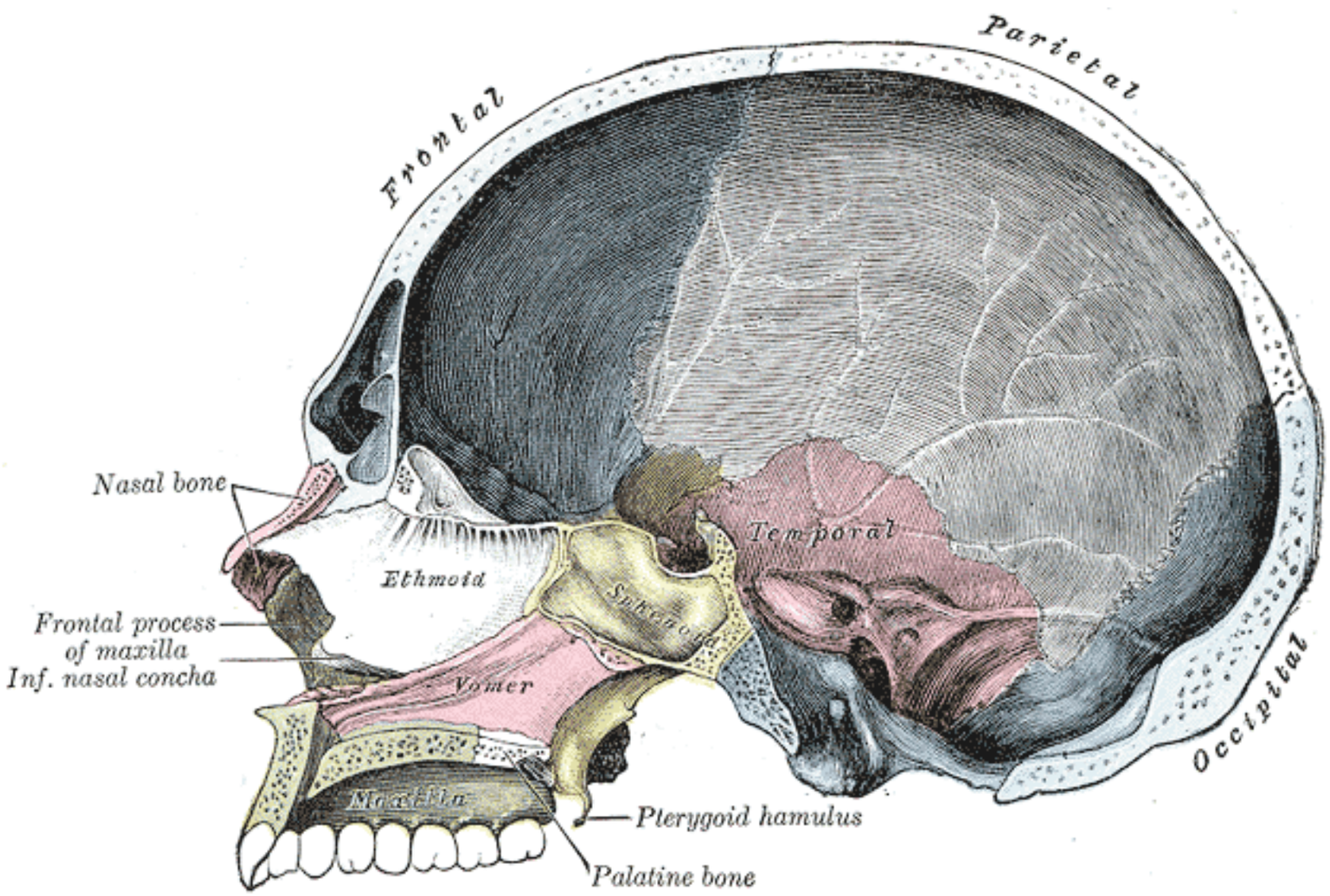

For Sup. Sagittal Sinus
Occipital angle
Pacchionian Depressions
Frontal angle
Grooves for post. division of mid. mening. vessels
For Transverse Sinus
Grooves for ant. division of mid. mening. vessels
Mastoid angle
Sphenoidal angle

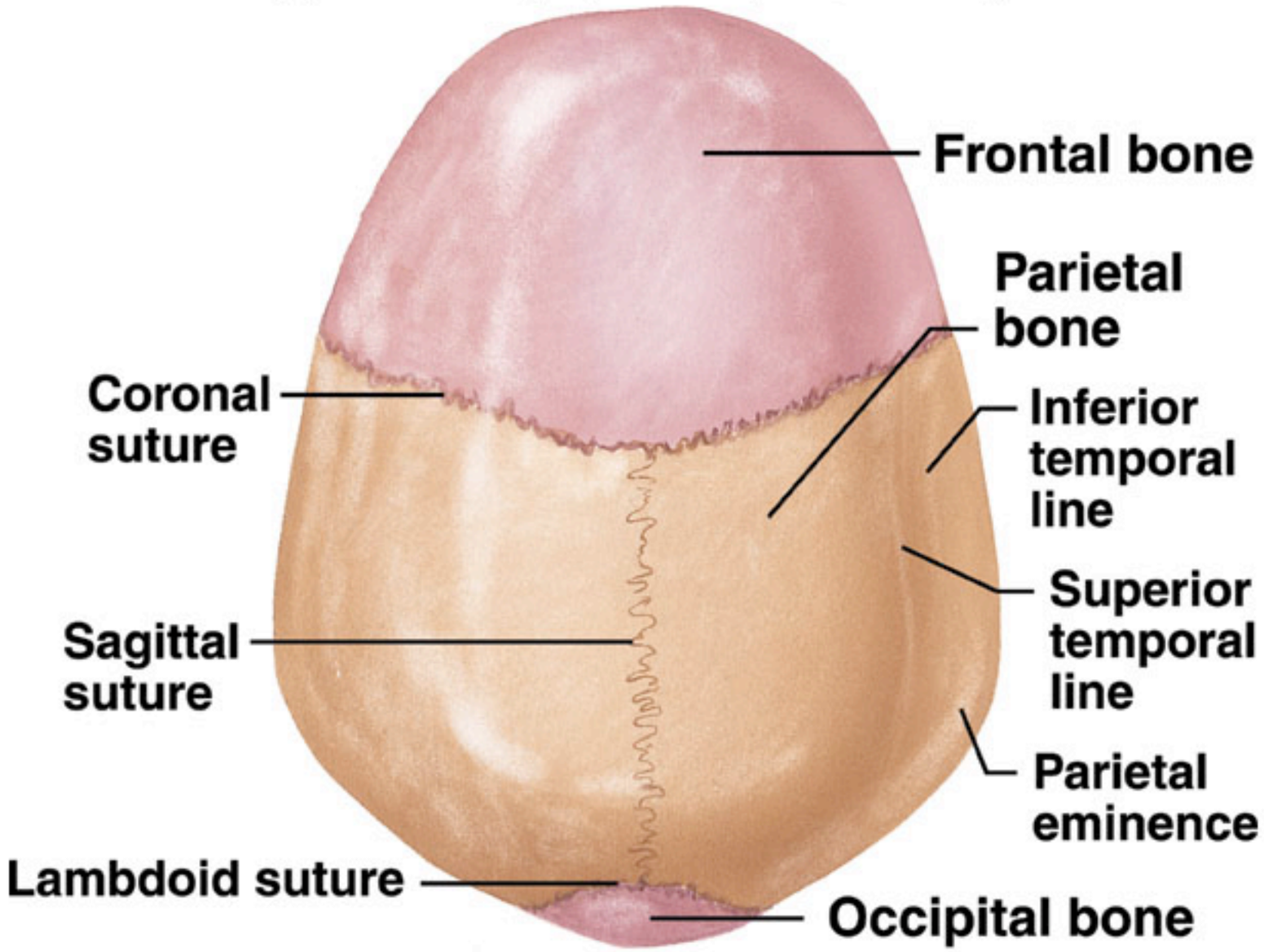

Inferior Temporal Line
Superior Temporal Line
Temporal Fossa
Remains of Frontal Suture
Frontal Eminence
Superciliary Arch
Glabella
Supra-orbital Notch or Foramen
Supra-orbital Margin
TEMPORALIS
Orbicularis oculi
Nasal part
Zygomatic process
Frontal spine

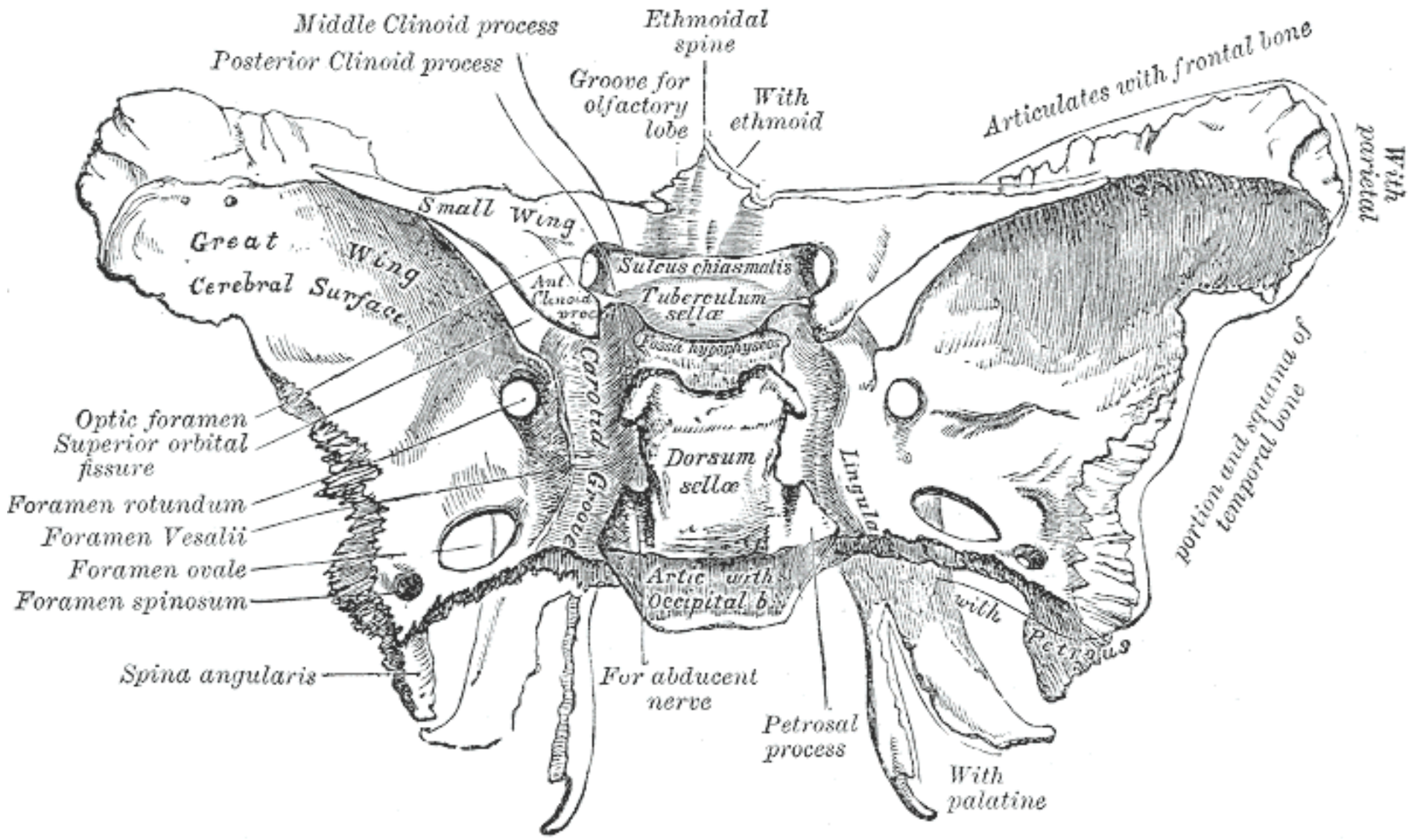

Articulates with Parietal Bone
Pacchionian Depressions
for Superior Sagittal Sinus
Meningeal Grooves.
for Falx Cerebri
with Great Wing of Sphenoid
with Small Wing
with Zygomatic
Posterior and Anterior Ethmoidal for.a
with Ethmoid
Ethmoidal Notch
for. Cæcum
Orbital Plate
Lacrimal Fossa
Depression for Pulley of Sup. Oblique
with Lacrimal
Supraorbital foramen
With maxilla
Frontal sinus
With nasal
With perpendicular plate of ethmoid
Under surface of nasal process forming part of roof of nose

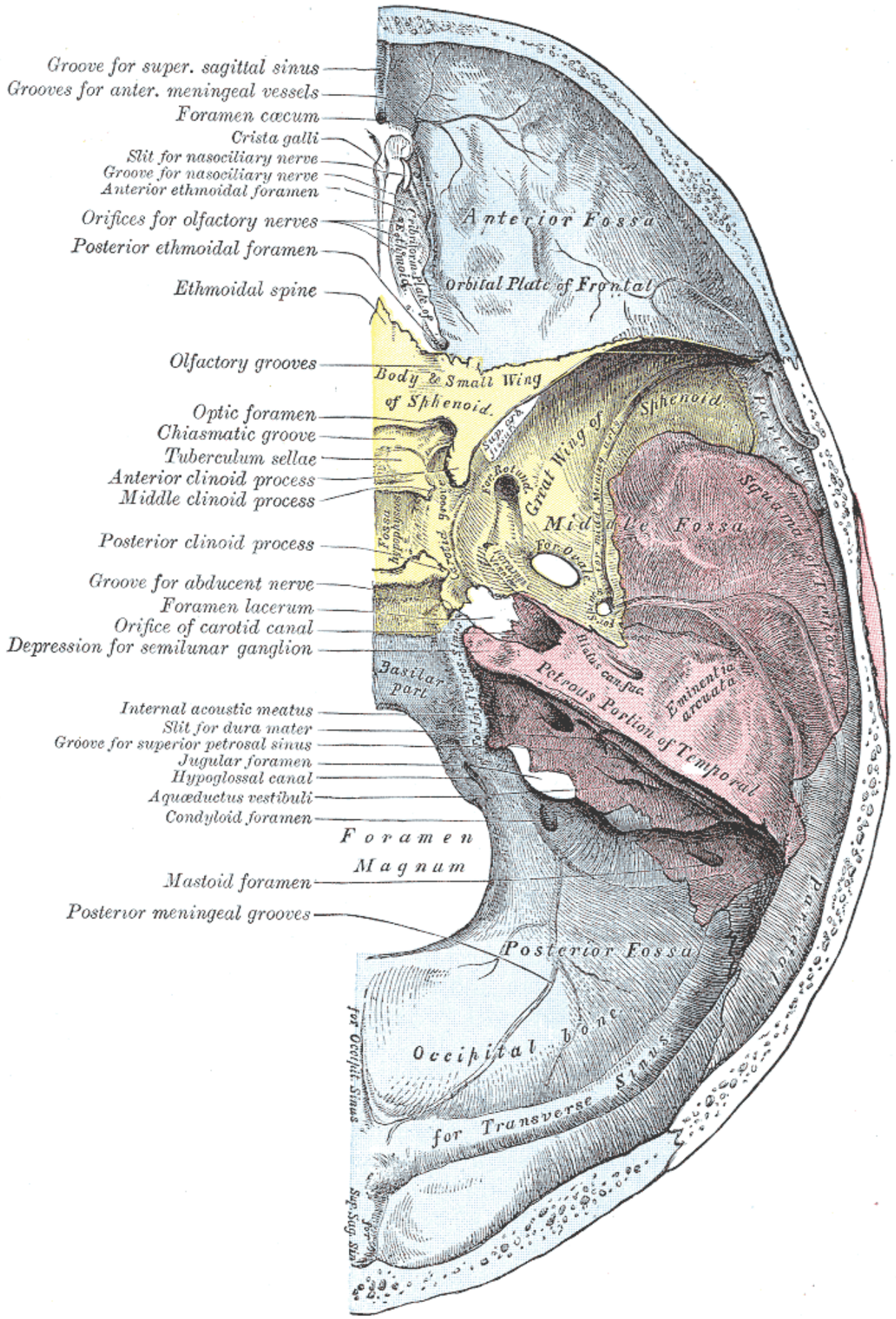

Squama
Groove for middle temporal artery
Parietal notch
Suprameatal triangle
Occipitalis
Zygomatic proc.
Temporalis
Articular tubercle
Post-glenoid process
Mandibular fossa
Petrotympanic fissure
Vaginal process
Styloglossus
Mastoid Portion
Sterno-cleido-mastoideus
Splenius capitis
Longissimus cap.
Occipital groove
Tympanic part
Stylohyoideus
Styloid process


Articulates with parietal bone
Parietal notch
Groove for post. div. of mid. menin. vs.
Petro-squamous suture
Groove for sup. petros. sinus
Sulcus sigmoideus
with great wing of sphenoid
Eminentia arcuata
with occipital bone
Mastoid foramen
Aquæductus vestibuli
Aquæductus cochleæ
Internal acoustic meatus


Frontal
Parietal
Nasal bone
Temporal
Ethmoid
Sphenoid
Frontal process
of maxilla
Inf. nasal concha
Vomer
Occipital
Maxilla
Pterygoid hamulus
Palatine bone




Middle Clinoid process
Posterior Clinoid process
Ethmoidal spine
Groove for olfactory lobe
With ethmoid
Articulates with frontal bone
With parietal
Small Wing
Great Wing
Cerebral Surface
Ant. Clinoid proc.
Sulcus chiasmatis
Tuberculum sellæ
Fossa hypophyseos
Optic foramen
Superior orbital fissure
Foramen rotundum
Foramen Vesalii
Foramen ovale
Foramen spinosum
Spina angularis
Carotid Groove
Dorsum sellæ
Lingula
Artic. with Occipital b.
For abducent nerve
Petrosal process
With palatine
with Petrous
portion and squamous of temporal bone


Groove for super. sagittal sinus
Grooves for anter. meningeal vessels
Foramen cæcum
Crista galli
Slit for nasociliary nerve
Groove for nasociliary nerve
Anterior ethmoidal foramen
Orifices for olfactory nerves
Posterior ethmoidal foramen
Ethmoidal spine
Olfactory grooves
Optic foramen
Chiasmatic groove
Tuberculum sellae
Anterior clinoid process
Middle clinoid process
Posterior clinoid process
Groove for abducent nerve
Foramen lacerum
Orifice of carotid canal
Depression for semilunar ganglion
Internal acoustic meatus
Slit for dura mater
Groove for superior petrosal sinus
Jugular foramen
Hypoglossal canal
Aquæductus vestibuli
Condyloid foramen
Foramen Magnum
Mastoid foramen
Posterior meningeal grooves
Anterior Fossa
Orbital Plate of Frontal
Body & Small Wing of Sphenoid
Great Wing of Sphenoid
Parietal
Squama of Temporal
Middle Fossa
Petrous Portion of Temporal
Basilar part
Posterior Fossa
Occipital bone
for Transverse Sinus
Parietal

NEOPLASMS OF THE SKULL

Approach...

Primary Neoplasms

Benign

Osteoma
Hemangioma, lymphangioma
Embryonic skull neoplasms
Chondroma
Giant cell tumors
Aneurysmal bone cyst
Lipoma

Malignant

Osteogenic sarcoma
Fibrosarcoma
Chondrosarcoma
Chordoma

Approach...

Secondary Neoplasms

Malignant Disease

carcinoma (lung, breast, kidney, thyroid, prostate)

multiple myeloma

lymphoma

Ewings Sarcoma

Neuroblasmtoma

Direct Extension

esthesioneuroblastoma

paraganglioma

meningioma

Approach...

Reactive Proliferative and Preneoplastic Lesions

fibrous dysplasia
Paget's disease
Langerhans cell histiocytosis
hyperostosis frontalis interna
sinus pericranii
cephalohematoma
mucocele
leptomeningeal cyst

Benign Primary Neoplasms

Osteoma

- benign slow growing osteoblastic tumor
- common primary skull neoplasm
- more common in females 3:1, any age
- patient may complain of a slow growing painless mass
- may manifest as sinusitis
- common locations:

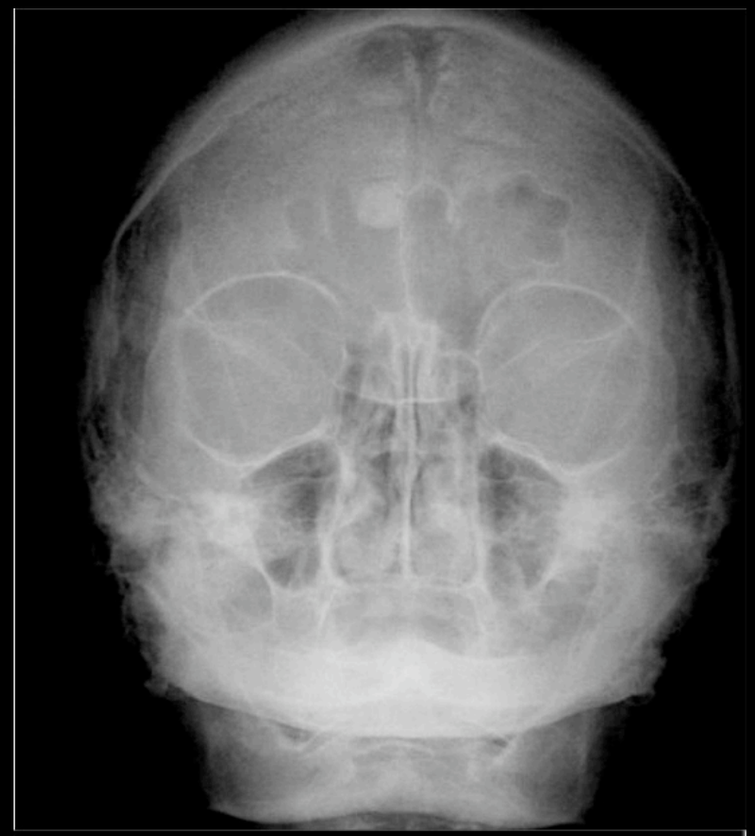
  frontal sinus > ethmoid > maxillary > sphenoid

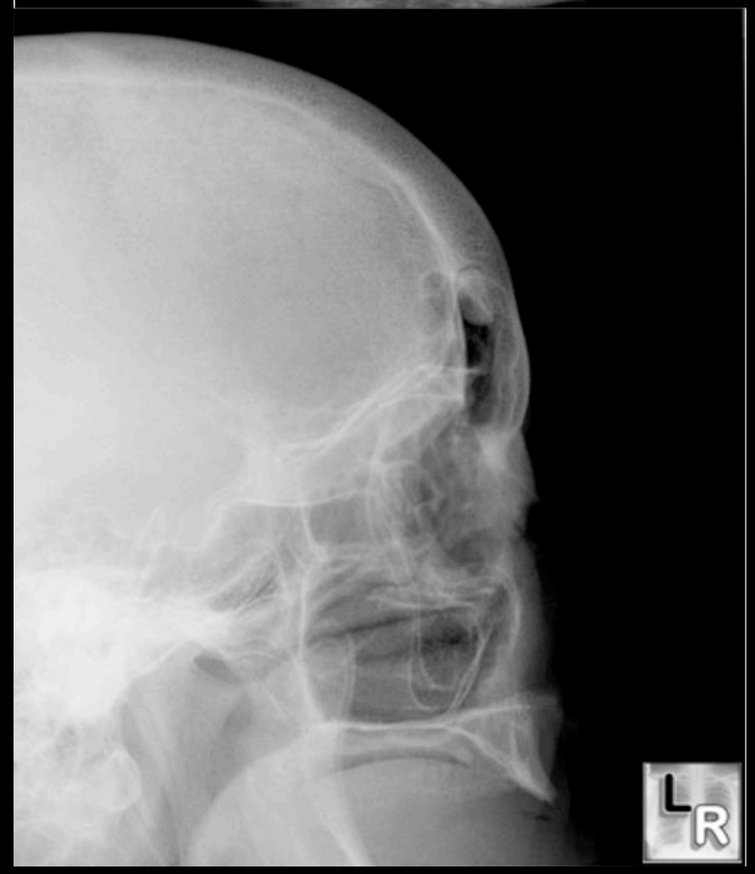
Osteoma

- may also be found in calvaria and mandible

- multiple osteomas may imply Gardner's syndrome

- in paranasal area, may arise from sinus walls





Osteoma - Histopathology

- compact - resembles cortical bone
- cancellous - includes compact bone, trabeculae, fat and fibrous tissue
- fibrous - mature lamellar bone

Osteoma - Treatment

- depends on size and location
- a small lesion involving outer table may easily be drilled leaving inner table intact
- larger lesions involving inner table may need craniotomy and cranioplasty

Hemangiomas and Lymphangiomas

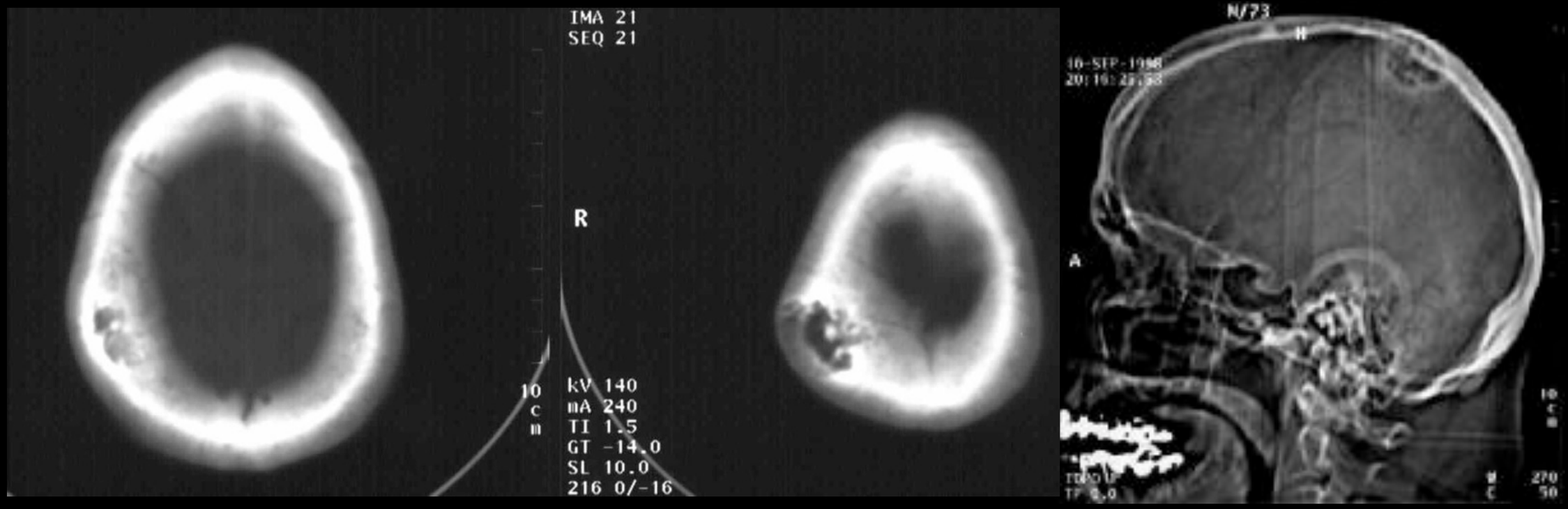
- second most common benign neoplasm of skull
- most common at 4th decade of life
- parietal and frontal bones most commonly involved followed by mandible
- some patients may present with headache
- soft tissue of scalp or dura rarely involved
- usually unifocal

Hemangiomas and Lymphangiomas

- Classification:
    - cavernous - slowly enlarging blood filled sinusoidal cells most commonly found at the calvaria
    - capillary - multiple tufts of capillaries
    - venous - consist of thick walled venous channels

Note - there are no feeding or draining vessels associated with these lesions!

Hemangiomas and Lymphangiomas



skull x-ray will show lucency and classically a honeycomb appearance

Hemangiomas and Lymphangiomas

Treatment:

- en bloc removal of tumor
- ? radiation therapy

Giant Cell Tumors

- aka osteoclastomas
- mostly involve long bones, mandible or maxilla
- involvement of skull is rare
- usually occur as painful masses that occur in temporal relation to a trauma
- variable pathology: soft and gelatinous to firm and rubbery...cell of origin unknown
- treatment: radical resection
    - recurrence treated with repeat surgery
    - radiotherapy for inaccessible lesions

Malignant Neoplasms

Osteogenic Sarcoma

- most common primary malignancy of bone HOWEVER skull involvement is rare
- may present as rapidly enlarging mass +/- pain...may grown internally or externally
- various histopathologic types, but in general present as nonspecific lytic lesion with permeation/thickening of surrounding bone
- treatment is surgery + radioation + chemotherapy
- may get metastases to lung

Chordoma

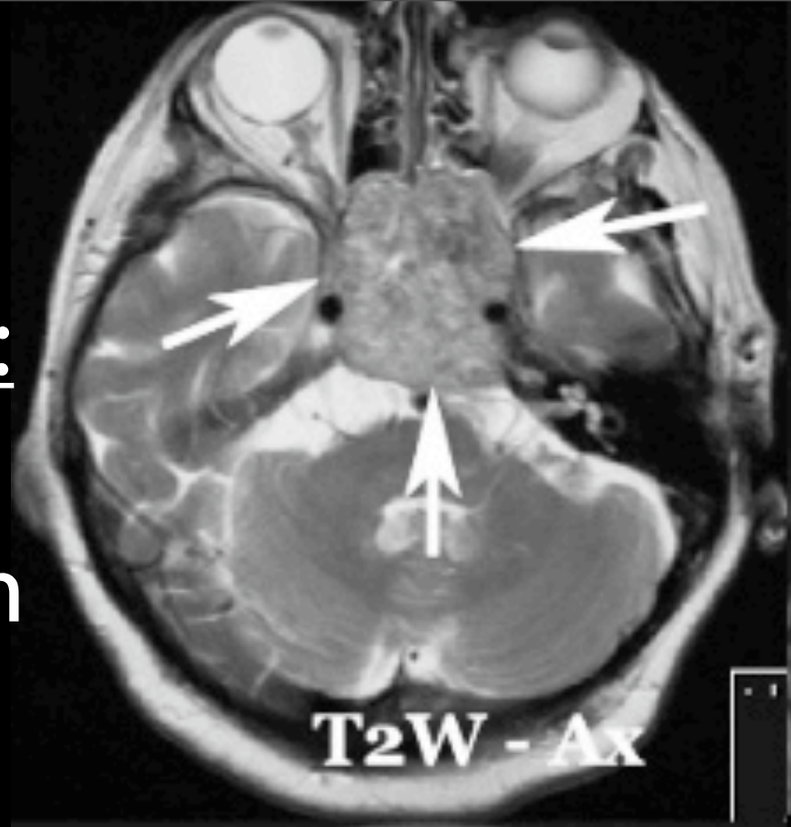
- uncommon
- arise from notochordal remnants
- most often found in the sacrococcygeal and clival regions
- 30-40% of chordomas are cranial in location mostly situation in the midline and involving clivus
- found in people ages 20-40yo

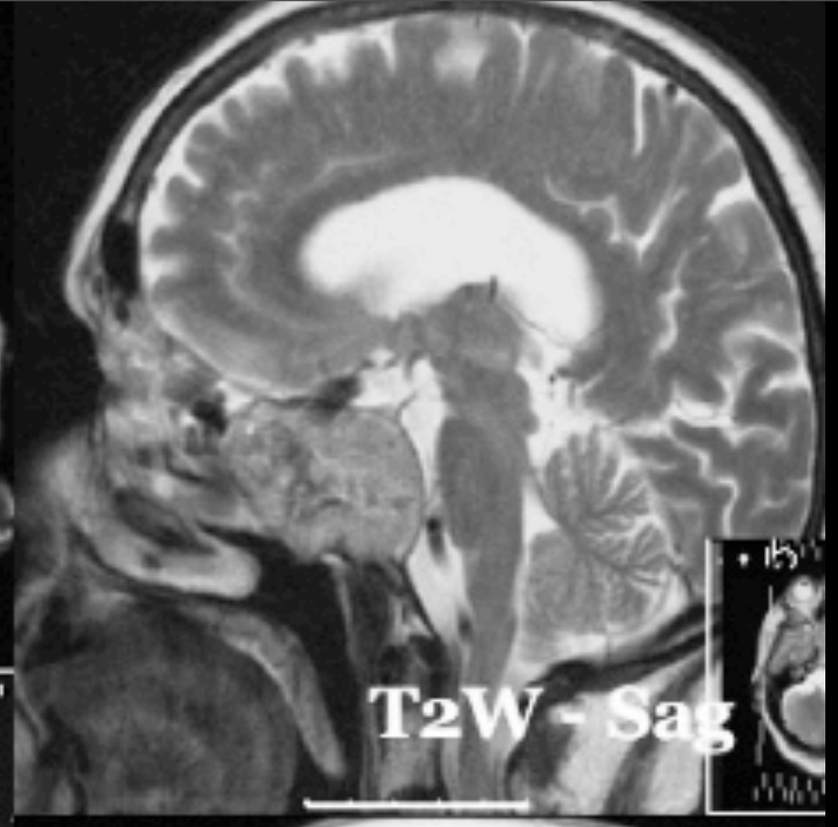
Chordoma

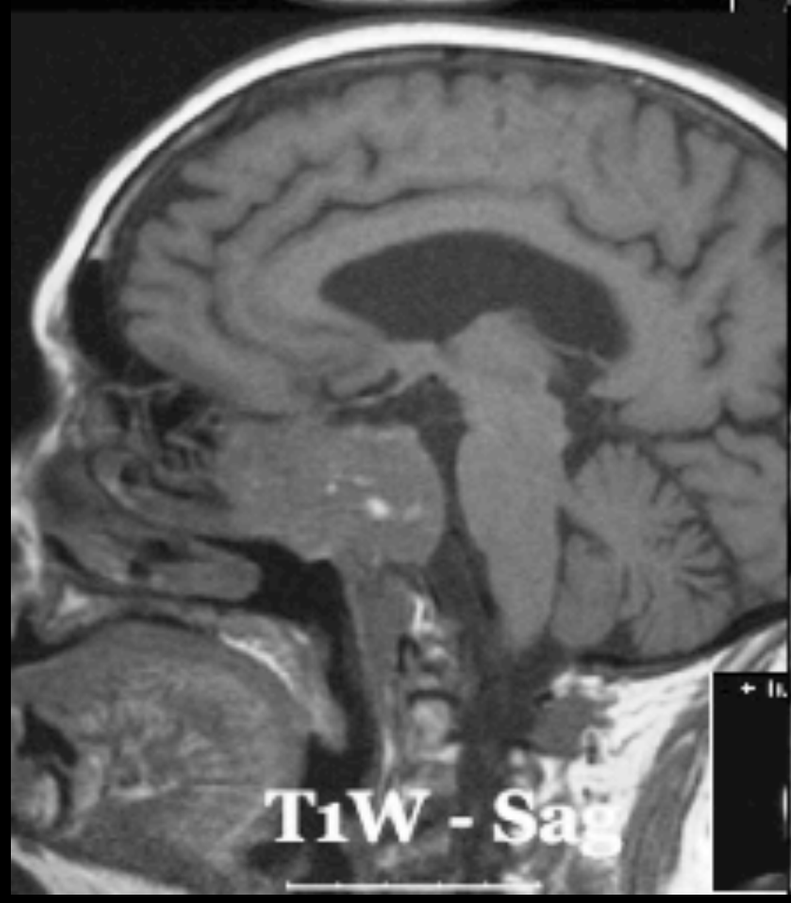
- few histologic features of malignancy
  HOWEVER very aggressive locally

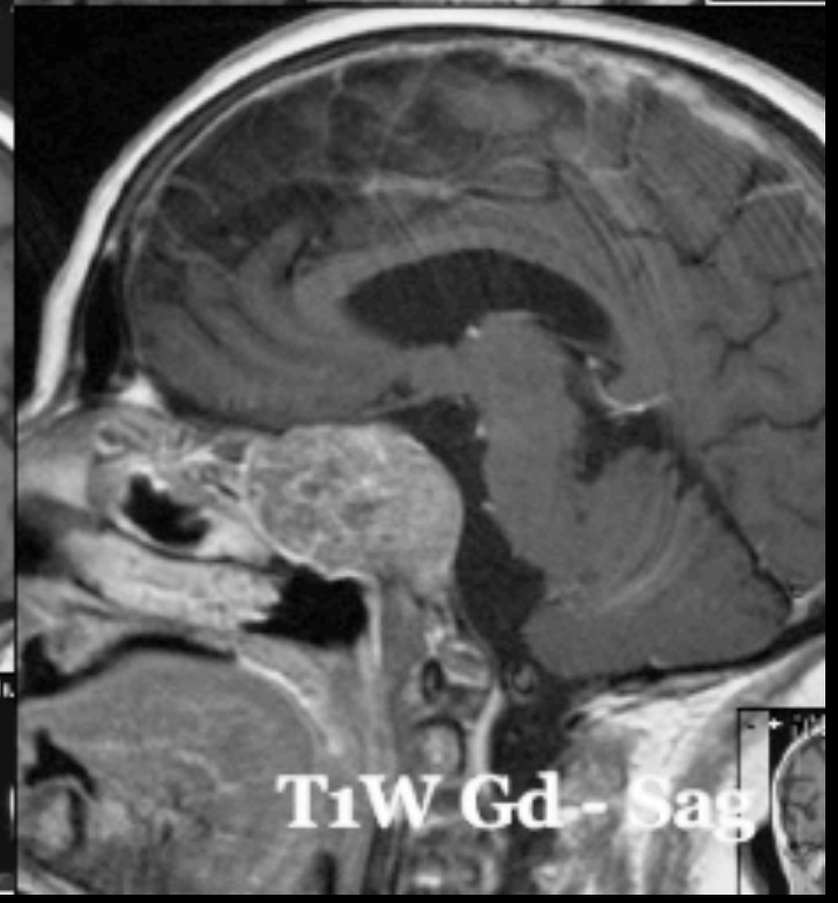
Radiological features:

- poorly enhancing on CT
- punctate calcification
- destroys bone
- usually high signal on T2

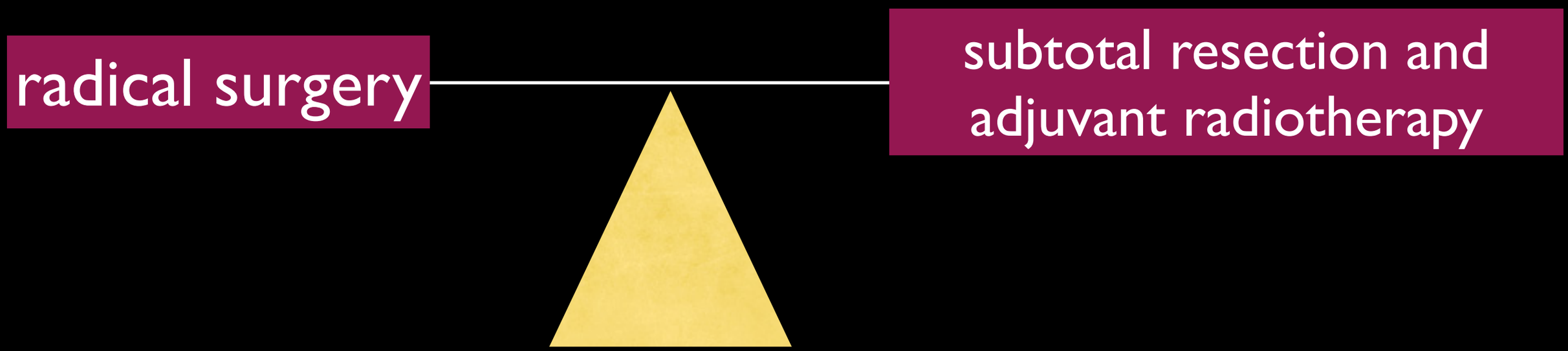








Chordoma: Treatment



CHONDROMA &
CHONDROSARCOMA

Clinical Features

- Comprise 0.15% of all intracranial tumours, and 6% of skull base tumours
- Mean age at presentation is 37 y.o., M = F
- Arise from persistent islands of embryonal cartilage that occur near the cranial base synchondroses (petrosphenoclival junctional area)
- Often occur in the para-median position

Clinical Features

- Malignant, but slow growing
- Duration of symptoms at presentation – 23 to 26 months

<u>Symptoms</u>
headache (80%)
diplopia (74%)
dysphagia/hoarseness (38%)
facial numbness/pain (32%)

*** very difficult to distinguish chondroma from chondrosarcoma preoperatively

Chordoma

- arise at remnants of notochord - found only in nucleus pulposus of intervertebral discs or clival bone marrow
- usually arise extradurally at cranial base - only invade dura late in course
- rarely metastasize
- regional mass effect is the major issue

Chondrosarcoma

- three histologic subtypes:
    - classic
    - mesenchymal
    - dedifferentiated
- generally occur in paramedian position
- most common place of origin is petrosphenoclival junctional area

Imaging

- CT (enhanced): slightly hyperdense
- MRI: useful to assess relationshiop to adjacent structures
  - T1 images show isodense lesion
  - T2 images brightly hyperintense

Treatment Options

- Goal of treatment = Increase likelihood of recurrence-free survival
- Options include:
    - SURGERY as primary treatment modality

Treatment Options

- Goal of treatment = Increase likelihood of recurrence-free survival
- Options include:
    - SURGERY as primary treatment modality

followed by...

- radiation therapy
- stereotactic radiosurgery
- proton radiation therapy
- radiation therapy with heavy charged particles
- chemotherapy

Surgical Approaches

Anterior Approaches

- Extended subfrontal
- Transethmoidal
- Maxillotomy and extended maxillotomy
- Transsphenoidal and extended transsphenoidal
- Le Fort I approach and modifications
- Midface degloving approach
- Transpharyngeal-transpalatal
- ❖ Anterolateral Approaches

Anterolateral Approaches

- Frontotemporal ± orbital/orbitozygomatic osteotomy, transcavernous
- Subtemporal, transpetrous apex, and transcavernous
- Preauricular subtemporal-infratempora

Lateral and Posterolateral approaches

- petrosal approaches
- extreme lateral

Surgery - Complications

COMPLICATION	PERCENT
Cerebrospinal fluid leak	27
Surgery required	11
With meningitis	10
New cranial deficits	
VI	21
V	15
VIII	10
Death within 3 months	5

Radiation

- Because piecemeal resection is required for skull based surgery, gross total resection may not result in an oncologically complete resection
- Fractionated conventional external beam radiation is not currently recommended for chondrosarcomas as most of the published results are disappointing

Stereotactic Radiosurgery

TABLE 1. Univariate analysis of prognostic variables for imaging and clinical local control in 29 patients with cranial base chordomas and chondrosarcomas[a]

Variable	No.	4-yr freedom from local relapse by magnetic resonance imaging	
		%	*p*
Sex			
Female	19	75	0.58
Male	10	50	
Age (yr)			
<50	16	89	0.39
>50	13	53	
Histology			
Typical chordoma	19	55	0.24
Chondroid chordoma	6	100	
Chondrosarcoma	4	100	
Tumor volume, cm^3			
<10	9	100	0.15
>10	20	49	
Previous resection			
GTR	6	100	0.19
<GTR	23	59	
Radiosurgical dose, Gy			
<15	11	51	0.25
≥15	18	73	
EBRT			
Yes	19	57	0.63
No	10	90	

[a] EBRT, external-beam radiation therapy; GTR, gross total resection.

Krishnan *et al* (2005)

Outcomes

TABLE 7. Long-term outcomes in patients with cranial base chondrosarcomas[a]

Authors, yr	No. of patients	Mean age (yr)/sex	Therapy	Recurrence-free survival for primary tumor			Mean follow-up (mo)	Tumor-related deaths, no.
				3 yr	5 yr	10 yr		
Gay et al., 1995 (9)	14	41/12 M, 2/F	Radical surgery ± adjunctive therapy	85%	—	—	32	3 (21%)
Hug et al., 1999 (13)	25	44/16 M, 9/F	Proton beam radiation	87%	—	—	33	5 (20%)
Crockard et al., 2001 (7)	17	36/12 M, 5/F	Radical surgery	84%	—	—	60	4 (23%)
Present study	47	39/29 M, 18/F	Surgery ± adjunctive therapy	83%	52%	32%	86	4 (8.5%)

[a] M, male; F, female; —, no data available.

Recurrence-free survival according to tumor resection	3 yr	5 yr	10 yr
GTR	92%	78.3%	42.3%
STR + radiotherapy	78%	41%	13.8%
Overall recurrence-free survival			
Average total	83%	52%	32%

[a] GTR, gross total resection; STR, subtotal resection.

ESTHESIONEUROBLASTOMA

Histopathology

- similar to neuroblastomas of adrenal glands and sympathetic nervous system
- lobular architecture with sheets of dense neurofibrillary material
- occasional olfactory rosettes or pseudorosettes
- membrane bound neurosecretory granules

Clinical Presentation

- generally a very uncommon tumor type (fewer than 350 reported cases)
- presents with nasal obstruction and epistaxis
- hyposmia present (asymptomatic)
- age at presentation spans 3-78yrs
- no associated geographic, environmental or lifestyle factors

Clinical Presentation

- may get metastasis (17-48% of patients)
- metastasis most commonly found at cervical lymph nodes
- other sites include: liver, mediastinum, adrenal gland, ovary, spleen, parotid, spinal epidural space

Patient Evaluation

- one should involve an otolaryngologist and neuro-ophthalmology
- history/physical
- enhanced CT scan
- MRI if the tumor extends intracranially
- may need metastatic work up

Staging

Modified Kadish Staging of Esthesioneuroblastoma

Stage A	tumor confined to nasal cavity
Stage B	tumor extension to paranasal sinuses
Stage C	tumor beyond nasal cavity/sinuses and involves cribriform plate, base of skull and extends intracranially
Stage D	metastases

Treatment

- mainstay of treatment is SURGERY
- role of radiotherapy and chemotherapy is very controversial
- some local control established with adjuvant therapy alone
- combination of surgery, radiation and chemotherapy seem to provide best results

Radiotheraphy

- role for pre-operative radiotherapy?
    - some support this to reduce tumor bulk
        - reduce cell viability to reduce tumor dissemination during time of surgery
- 20% recurrence at 5yrs with surgery and radiotheraphy vs 50% recurrence with surgery alone

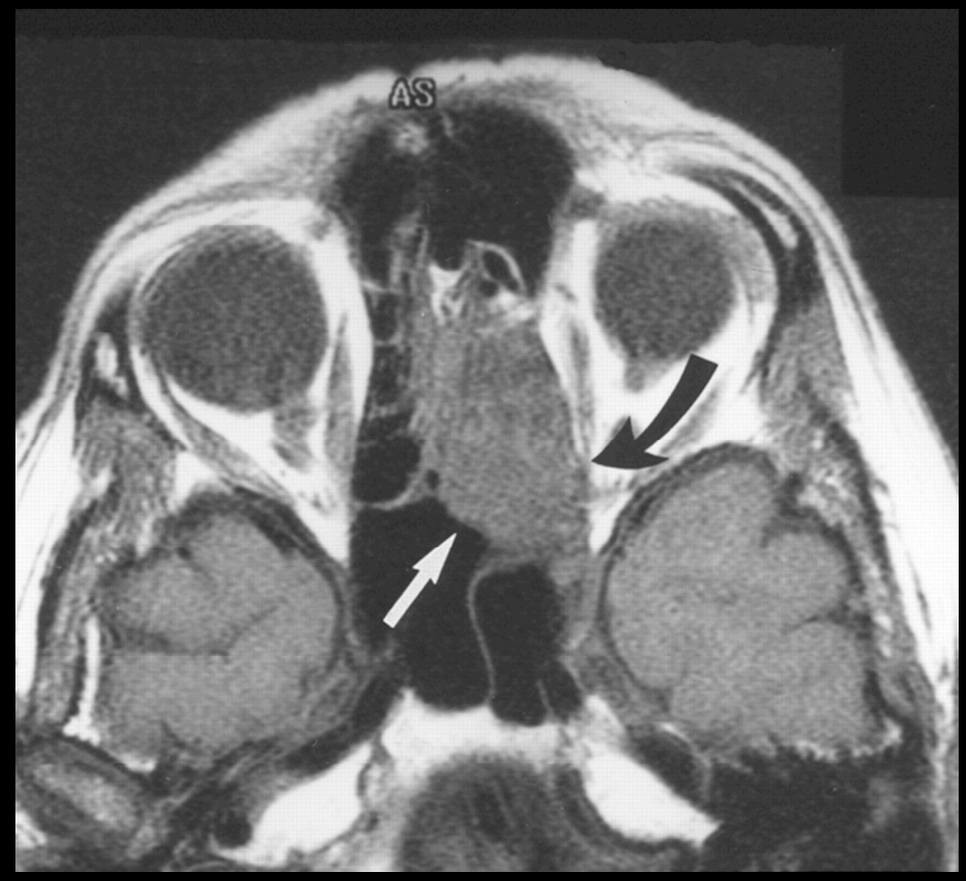
Fibrous Dysplasia

Clinical features

- developmental anomaly of mesenchymal precursors - unknown cause
- affects men and women often before age of 30
- monostotic
- commonly involves sphenoid, frontal, ethmoid and maxillary
- symptoms related to bony expansion causing local swelling, proptosis, visual loss and headache
- may undergo malignant degeneration to osteosarcoma (rare)

Fibrous Dysplasia

- three forms described:
    - pagetoid, sclerotic, cystic
- CT shows ground glass appearance of sclerotic form
- Cystic form seen as inhomogenous texture in expanded bone



...total removal recommended if cranial nerves are involved otherwise cosmetic reasons. Not chemo or radiosensitive...