Skull Base and Vault: Osseous and Cartilaginous Lesions

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Learning objectives
- Review skull anatomy
- Create algorithm for skull bony lesions based on lesion patterns
- Review differential for diffuse calvarium thickening

Anatomy

Anterior Cranial Fossa
- Middle Cranial Fossa
- Posterior Cranial Fossa

Primary bone lesions
- Primary neoplasm of the calvarium account for 0.8% of all bone tumors
- Benign tumors are more common than primary malignancy

Calvarium (Vault) lesions
- Lytic Lesions:
  - Solitary
    - Osteosarcoma/Osteoid, Hemangioendothelioma: Sclerotic rim
    - GI: well defined, button sequenam
    - Mets
  - Multiple
    - GI: Geographic pattern
    - Multiple Myeloma: Punch hole
    - Paget's disease: Osteosclerosis circumscripta
    - Mets
- Sclerotic Lesions:
  - Focal
    - Fibrous dysplasia: Ground glass with regular inner table
    - Intraosseous Meningioma: Ground glass with irregular inner table
  - Diffuse
    - Paget's disease: Cotton-wool
    - PTH: Salt and pepper
Solitary lytic lesion with sclerotic rim

Dermoid/Epidermoid

- Result of persistence of ectodermal elements at the sites of closure (fissures, neural tube, and diverticulations of cerebral hemispheres)
- Contain ectoderm and skin
- Common locations – midline/frontotemporal– parietal location
- May be associated with external skin-cysts or deep sinus tract
- CT – fat/fluid attenuation
- MRI – Hyperintense on T1WI, variable signal on T2WI; no enhancement, fat suppression

Midline calvarial dermoid

Intraosseous Hemangioma

- Benign lesions
- Occur in all age groups – F:M = 3:1
- Common in the frontal and parietal bones
- X-ray – Coarse honeycomb or sunburst trabecular pattern
- CT – Outer table is involved with preservation of the inner table
- Inhomogeneous contrast enhancement
Langerhans Cell Histiocytosis

- AKA: Eosinophilic Granuloma
- Monostotic involvement is more common than multifocal
- 50% have skull involvement
- Well defined sclerotic lesion w/o sclerotic borders
- Unequal involvement of inner and outer tables= “beveled edge”
- May or may not have soft tissue mass or dural invasion

Eosinophilic granuloma

- Button sequestrum (Bone sequestrum)

Multiple Myeloma

- MR is most sensitive for evaluation of marrow involvement (STIR)

Osteosarcoma

- Head and neck: mandible and maxilla more common than calvarium
- Lesion can have radiographic features of a benign lesion
Osteoma

- Benign, slow growing, osteoma
- Gardner syndrome
  - autosomal dominant
  - familial colorectal polyposis
- Radiodense lesions, similar to normal cortex

Intraosseous Meningioma

- Expands the inner and outer tables of the calvarium
- There may be no dural component
- May extend into the scalp
Fibrous Dysplasia

- Commonly affects adolescents and young adults
- Monostotic and polyostotic forms
- Replacement of the medullary cavity with fibrocellular tissue resulting in expansion of the diploic space
- Outer table tends to bulge more than the inner table
- Variable enhancement
- Radionuclide bone scan can be used to exclude polyostotic disease

Fibrous Dysplasia

- Three radiographic patterns:
  - Predominately sclerotic (38%)
  - Predominately lytic (22%)
  - Mixed (40%)
- Involves both inner and outer tables
- Often has “ground glass” appearance
- Asymmetric involvement of the cranium – 10% of monostotic and 50% of polyostotic cases

Metastatic disease

Skull metastasis

- Breast, lung, or prostate malignancy
- Skull may be the only site of bony metastasis in up to 11.6% of patients
- DWI: ↑ sensitivity
GH-secreting pituitary adenoma

Liver produces insulin-like growth factor 1 (IGF-1)

Causes abnormal growth of soft-tissues and bones

Also note wide mandibular angle, frontal bossing

AD Osteopetrosis (Albers-Schonberg Disease)

- Failure of osteoclasts to resorb bone due to lack carbonic anhydrase
- Multiple forms associated with multiple genes (more than 10)
- May be autosomal recessive, dominant, or X-linked recessive
- Cranial nerve symptoms due to foraminal compression

Acromegaly

Skull base metastatic disease
Hyperparathyroidism

- Excessive parathyroid hormone
- Salt and pepper sign
- Primary
  - Parathyroid adenoma, hyperplasia or carcinoma
- Secondary
  - Renal failure
- Tertiary
  - Longstanding secondary HPT

Hyperostosis frontalis interna

- First described by Morgagni more than 200 years ago.
- Headaches, obesity, virilism, and hypertrichosis (Morgagni's syndrome)
- Diabetics

Paget Disease

- Chronic progressive disease with initial destruction of bone followed by a reparative process
- Usually affects older patients
- Usually polystotic (can affect any bone)
- Three stages of disease in the skull:
  - Vascular – enlargement of skull
  - Advancing sclerosis/leading edge – thickening of the cortex
  - Complete diffuse sclerosis – loss of distinction between the diploic space and calvarium

Paget Disease

- X-ray patterns - osteolytic, osteosclerotic, or mixed
- "Cotton ball" – round areas of sclerosis with surrounding demineralization
- Symmetric involvement of the cranium
- CT/MR – diploic widening and thickened cortex
Paget Sarcoma

- 1% of patients with Paget’s disease will develop osteosarcoma
- More common with polyostotic Paget’s - worse prognosis
- Common sites - femur, humerus, pelvis, skull and tibia.
- X-ray: ill-defined, primarily osteolytic lesion of the medullary cavity, with aggressive cortical destruction and extension into the soft tissues

Anemia

- Skull alterations can occur due to overactivity of the red marrow in response to anemia
- Marrow hyperplasia widens the diploic space and thins the outer table

- Thalassemia major
- Iron deficiency anemia
- Sickle cell disease
- Spherocytosis

Thalassemia
Skull base Lesions

- Chordoma
- Chondrosarcoma
- Fibrous dysplasia
- Multiple Myeloma/Plasmacytoma
- Meningioma
- Ecchordosis Physaliphora

These fibro-osseous lesions have no particular predilection for site of presentation and can present in either an intra-axial or extra-axial location within the CNS. CAPNONs are thought to arise as a result of local compression and or irritation of adjacent tissues. They are slow growing, non-neoplastic lesions with no propensity for metastasis.

Ecchordosis Physaliphora

- Notochordal remnant appearing at the dorsal wall of the clivus
- 2% of autopsies
Chondrosarcoma

- Off-midline centered on petro-occipital fissure
- Sxs: CN VI palsy and headache
- Chondroid matrix Ca++ in 50%; bone destruction in >50%
- T1-usually isointense; T2-hyperintense; avid enhancement
- Association with Ollier’s (enchondromatosis) and Maffucci (enchondromatosis with ST hemangiomas in hands/feet) syndromes
- Main DDx=chordoma… DWI may differentiate
- Chondrosarcoma mean ADC=2051 +/-261
- Classic chordoma mean ADC=1174 +/- 117

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Chondrosarcoma

- Origin from remnants of embryonal cartilage or from metaplasia of meningeal fibroblasts

Chordoma

- Histologically benign but very aggressive
- Lesion originates from distal clivus and is fairly midline
- Iso on T1, hyperintense on T2
- Moderate to marked, typically heterogeneous, enhancement (contains mucoid material, hemorrhage, occ Ca++)

PEARL: Thumb printing on brainstem is only seen with this lesion and is pathognomonic
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