Intracranial Chordoma

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Agenda

- Our Patient
- Basics of Chordoma
- Radiological Features
- Differential Diagnoses
- Take Home Messages
Our Patient: Clinical Presentation

- Man with acute onset of headache one week ago at dinner
- Pressure sensation both posterior and lateral on his skull
- The headache wakes him up from sleep
- He rates the pain a 6/10
- No visual problems, fevers, chills, or night sweats
- Vitals: T 99.0; BP 144/101; P 77; RR 18; O2 sat 99%
- Neurological exam unremarkable
Our Patient: CT Findings Sagittal

- Large, poorly delineated mass appearing to be centered anterior to, and intimately associated with, the brainstem
- Slightly expanded in the sella turcica
- Bone destruction of the clivus
Our Patient: Comparison With Normal Clivus

Normal head CT
Normal clivus

Index patient’s head CT
Bone destruction
Our Patient: CT Findings Axial

- Moderate hydrocephalus
Our Patient: MRI Findings Axial T1W

- Large heterogeneously enhanced mass centered within the prepontine cistern arising from, or invading into, the clivus
Our Patient: MRI Findings Mass Effect

- Significant mass effect with associated moderate hydrocephalus
Our Patient: MRI Findings FLAIR

In case of an invasion of the brainstem, a parenchymal edema would have been demonstrated in the FLAIR images. In our case there was no evidence of a parenchymal edema.
Chordoma: Basics

- Relatively rare malignant tumor
  - 1% of intracranial tumors
  - 4% of all primary bone tumors
- Originate from embryonic remnants of the primitive notochord
- Most common in the 4th to 7th decade of life
- 2:1 male predilection
- Generally slow growing
  - Subtle clinical presentation
- Extremely high recurrence rate
- Intimate relation to critical structures
- Distant metastasis is rare
- Extensive surgical removal and postoperative fractionated proton beam radiation therapy are the most effective treatments

In the past high mortality rates
Chordoma: Location

- Any site along the course of the embryonic notochord
  - Cranial 32%
  - Spinal 33%
  - Sacral 29%
- Other locations 4%
Chordoma: Intracranial Location

- Most often originates from the clivus
  - Especially from the sphenoid-occipital synchondrosis

- Occasionally unilaterally arising from the petrous apex

- Other sites of origin
  - Sellar area
  - Sphenoid sinus
  - Rarely nasopharynx, maxilla, paranasal sinuses, or intradural region
Chordoma: Anatomy Of The Clivus: Sagittal

- **Clivus**
- **Sphenop-occipital synchondrosis**
- **Sphenoid sinus**

Chordoma: Anatomy Of The Clivus: Axial

Chordoma: Appropriate Imaging

- MRI best modality for radiologic evaluation
  - Detection of intracranial chordomas: MRI equal to CT
  - Delineation of lesion extent: MRI is superior to CT
- Pretreatment evaluation: Both CT and MR imaging are usually required
  - Planning of radical resection
  - Planning of irradiation to demarcate the tumor margins, neighboring cranial nerves, and vital vascular structures
- Postsurgical follow-up and detection of recurrence: MRI
Chordoma: Histopathologic Features

- Gelatinous, multilobulated tumor

- Majority of the lesions are 2-5 cm in size

- Typical chordoma
  - Cells set in a matrix of mucopolysaccharides with a characteristic physaliphour appearance
  - Areas of necrosis, recent and old hemorrhage, and entrapped bone trabeculea
MRI Features: Planes

- **Sagittal images**
  - Most valuable in defining the posterior margin of the tumor
  - Showing the relation between the tumor and brainstem
  - Depicting nasopharyngeal extension of the tumor
  - Disclosing transdural transgression
    ⇒ Surgical planning

- **Coronal images**
  - Extension into the cavernous sinus
  - Depicting the position of the optic chiasm and tract
MRI Features: T1-Weighted Images

- Intermediate to low signal intensity
  - Easily recognized within the high signal intensity of the fat of the clivus

- Small foci of hyperintensity
  - Hemorrhage
    - Can be confirmed with gradient-echo imaging (dark areas)
  - Mucus pool
MRI Features: T2-Weighted Images

- **High signal intensity**
  - High fluid content

- Intratumoral areas of heterogeneous hypointensity
  - Calcification
  - Hemorrhage
  - Highly proteinaceous mucus pool

- Low-signal-intensity septations

⇒ Used to differentiate from adjacent neural structures
MRI Features: Contrast Enhancement

- Moderate to marked enhancement
- Occasionally slight or even absent
  - Necrosis
  - Large amount of mucinous material
- Sometimes “honeycomb” appearance
  - Intratumoral areas of low signal intensity

Axial C(+) MRI T1W. PACS, BIDMC
MRI Features: Fat Suppression

- Differentiation of enhanced tumor margins from adjacent fatty bone marrow
- Small intraclival chordomas can be better demarcated
CT Features: Classic Appearance

- Centrally located
- Well-circumscribed
- Expansile soft-tissue mass that arises from the clivus
- associated extensive lytic bone destruction
- Usually hyperattenuating relative to the adjacent neural axis
- Irregular intratumoral calcifications
  - Sequestra from bone destruction
- Solitary or multiple low-attenuation areas sometimes seen within the soft-tissue mass
  - Myxoid and gelatinous material
CT Features: Contrast Enhancement

- Unenhanced and contrast-enhanced images
  - Moderate to marked contrast enhancement

- Limited capacity to show soft-tissue structures in the posterior fossa
  - Beam-hardening artifacts
Differential Diagnosis: Based On Images

Common
- Chondrosarcoma
- Clival meningioma
- Nasopharyngeal malignancies
- Rhabdomyosarcoma (pediatric patients)

Rare
- Metastasis
- Aggressive pituitary adenoma
- Langerhans Cell Histiocytosis
- Dermoid
- Epidermoid cysts
- Trigeminal neuroma
- Fibrous dysplasia
Differential Diagnosis: Chondrosarcoma

- Most often confused with intracranial chordomas

- **Majority arises along the petro-occipital fissure**
  - Chordomas typically have a midline skull base location

- Similar signal intensity on T1W and T2W to chordoma

- **Linear, globular, or arclike calcifications possible**
  - Uncommon for chordoma
Differential Diagnosis: Clival Meningioma

- Dural attachment
- No appearance of a destructive bone lesion
  - Causes bone sclerosis!
- Homogeneous enhancement
- Characteristic angiographic appearance
Differential Diagnosis: Nasopharyngeal Malignancies

- Usually extend more anteriorly
- Have associated head and neck lymphadenopathy
Differential Diagnosis: Metastasis

- Relatively infrequent in the skull base in the absence of a primary neoplasm

- Extraosseous tumor component of metastases is usually small
Index Patient: 3-years follow-up

- S/p transphenoidal resection, vp shunt and radiation
- Last MRI noted a small area of T2 signal abnormality in the lateral cavernous sinus which potentially represented a small amount of hemorrhage within the old tumor and surgical bed
  - Suspicious for recurrence
Take Home Messages

- 4th to 7th decade
- Most often originates from clivus
- MRI for radiologic evaluation
  - T1w intermediate to low signal intensity
  - T2w high signal intensity
- Septations
  - T2w low-signal-intensity
- Moderate to marked enhancement
- Areas of necrosis, hemorrhage, and bone trabeculae (CT)
- Extremely high recurrence rate
- Extensive surgical removal and fractionated proton beam irradiation are the most effective treatments
References


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