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# Intracranial Chordoma

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# Agenda

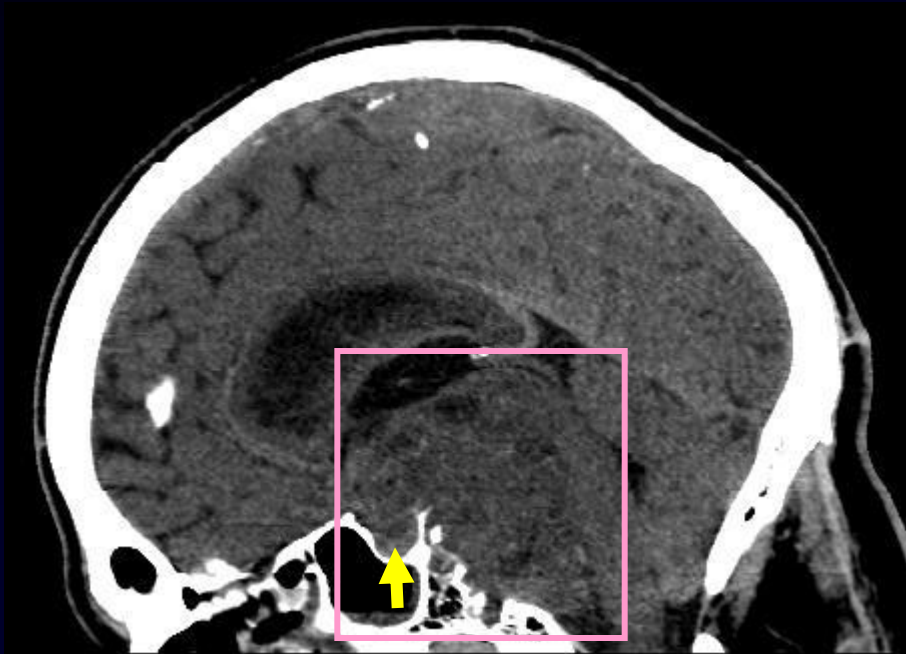
- Our Patient
- Basics of Chordoma
- Radiological Features
- Differential Diagnoses
- Take Home Messages



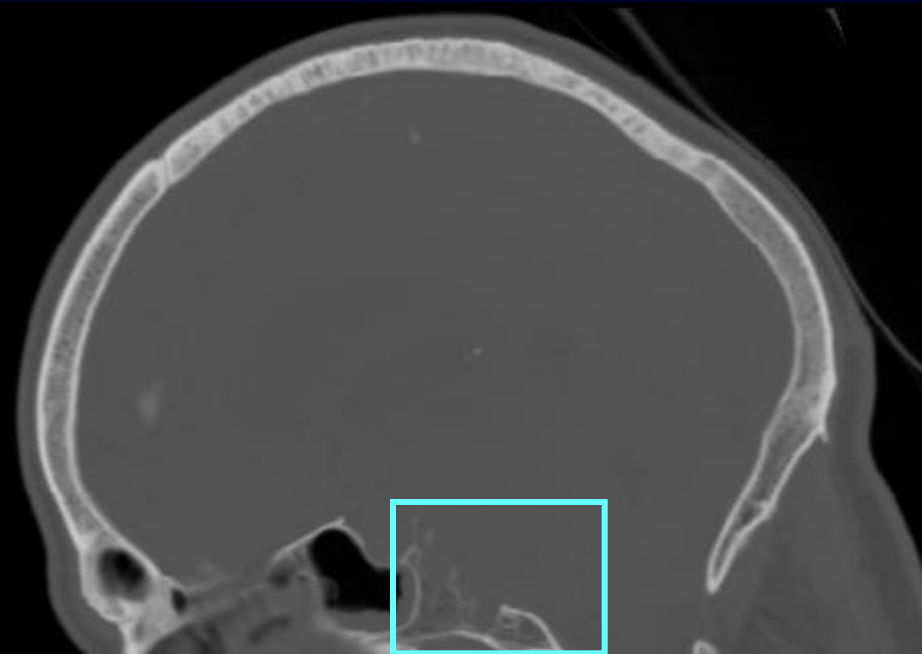
# Our Patient: Clinical Presentation

- ❑ 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



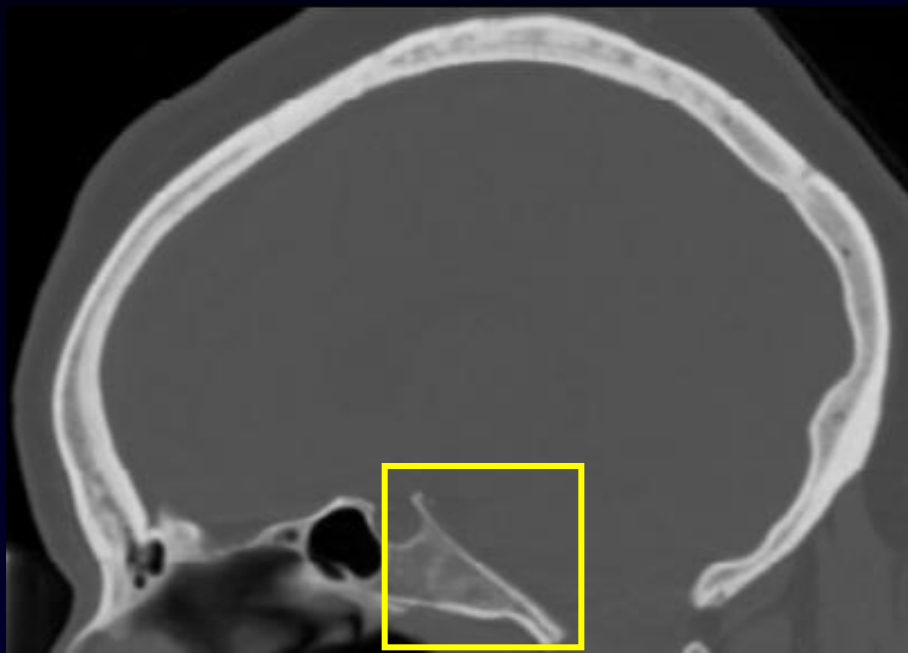
Sagittal C(-) CT Soft Tissue Window. PACS, BIDMC



Sagittal C(-) CT Bone Window. PACS, BIDMC

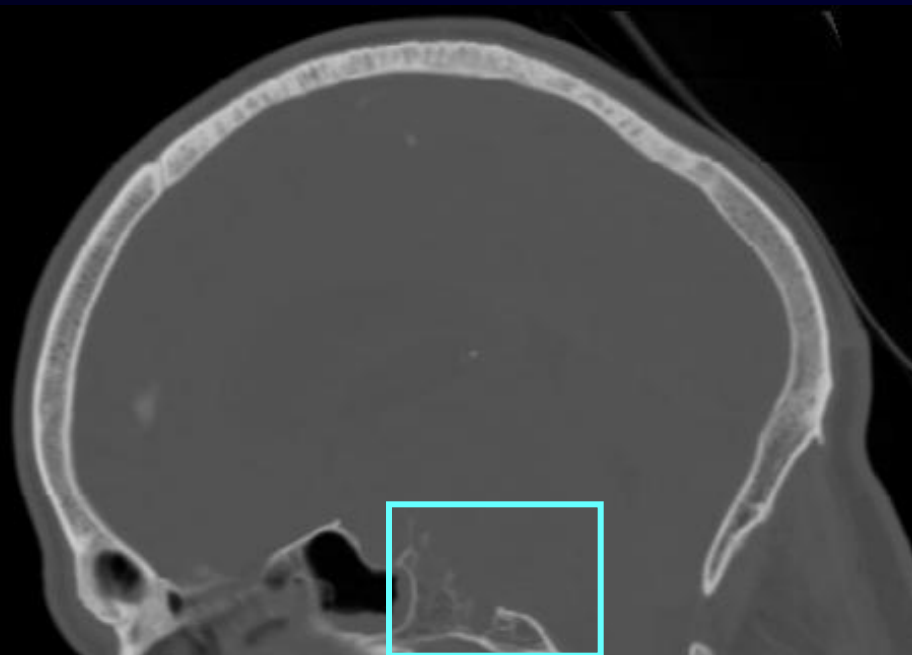
- Large, poorly delineated mass appearing to be centered anterior to, and intimately associated with, the brainstem
- Slightly expanded in the sella turcica
- Bone destruction of the clivus

# Our Patient: Comparison With Normal Clivus



Sagittal C(-) CT Bone Window. PACS, BIDMC

Normal head CT  
Normal clivus



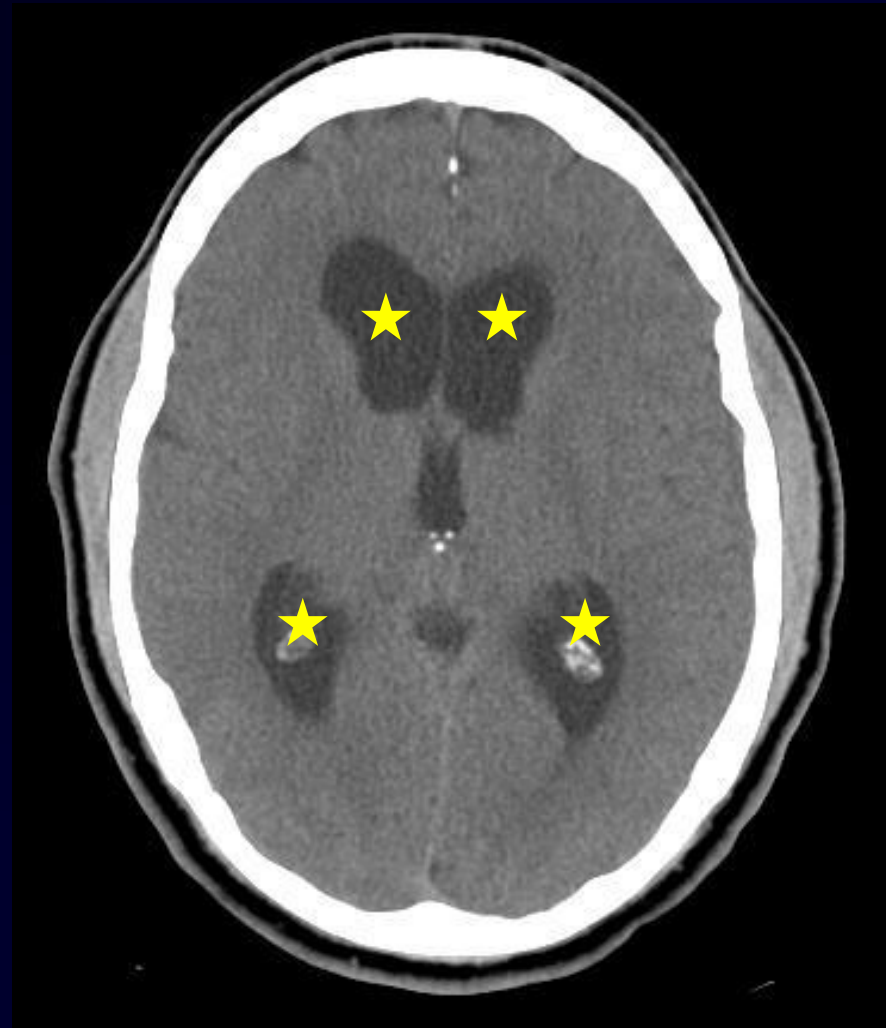
Sagittal C(-) CT Bone Window. PACS, BIDMC

Index patient's head CT  
Bone destruction



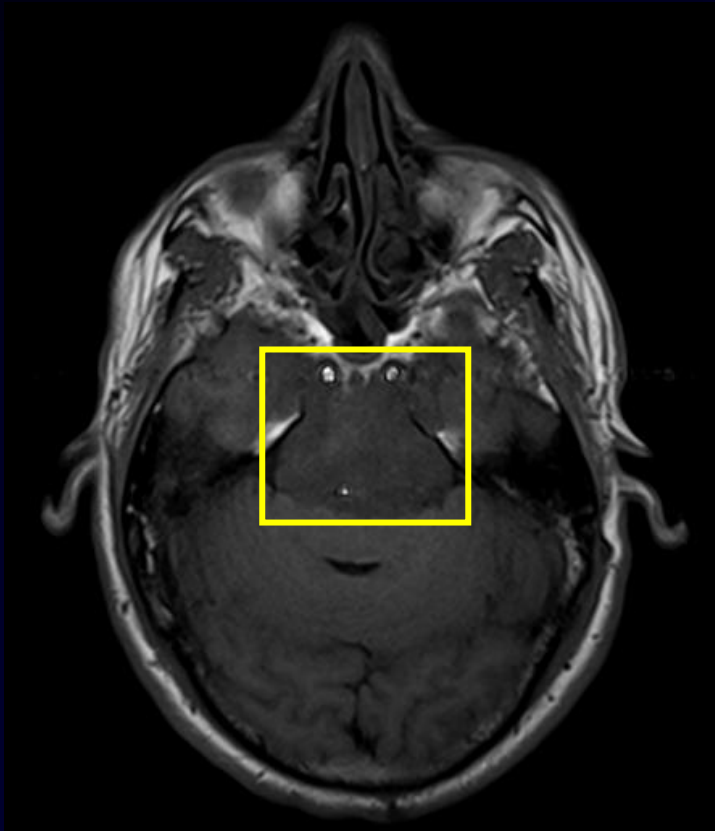
# Our Patient: CT Findings Axial

- Moderate hydrocephalus

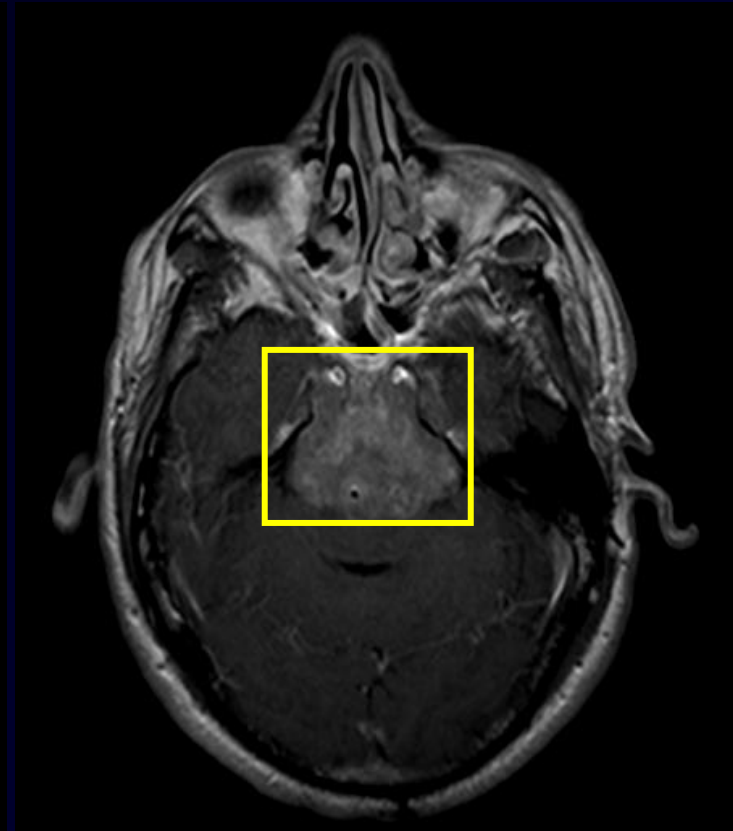




# Our Patient: MRI Findings Axial T1W



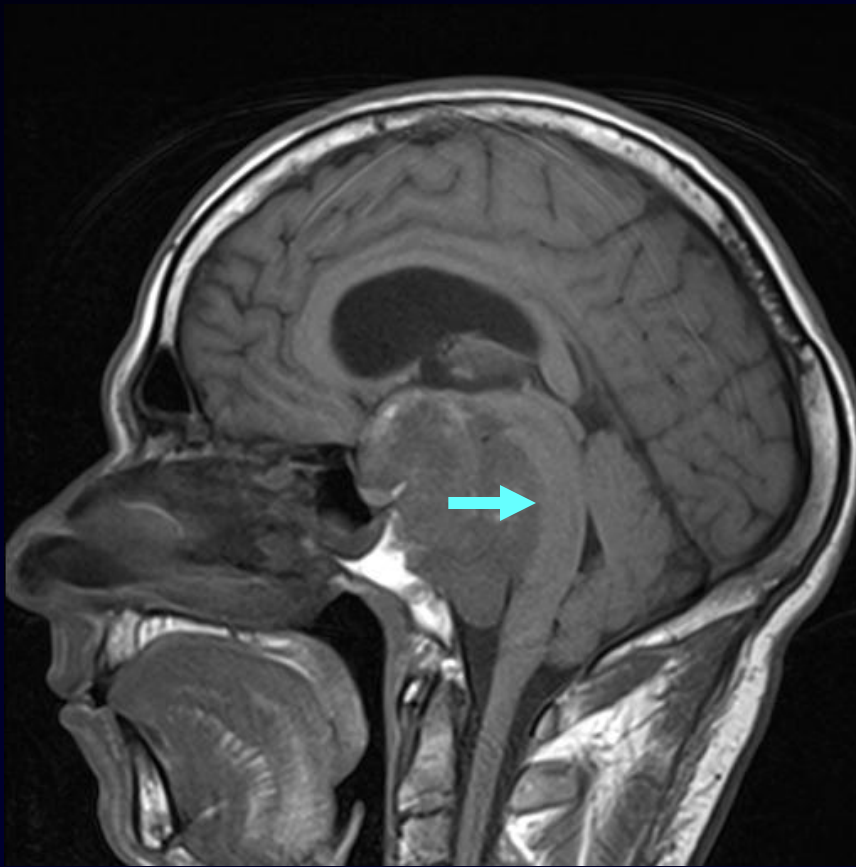
Axial C(-) MRI T1W. PACS, BIDMC



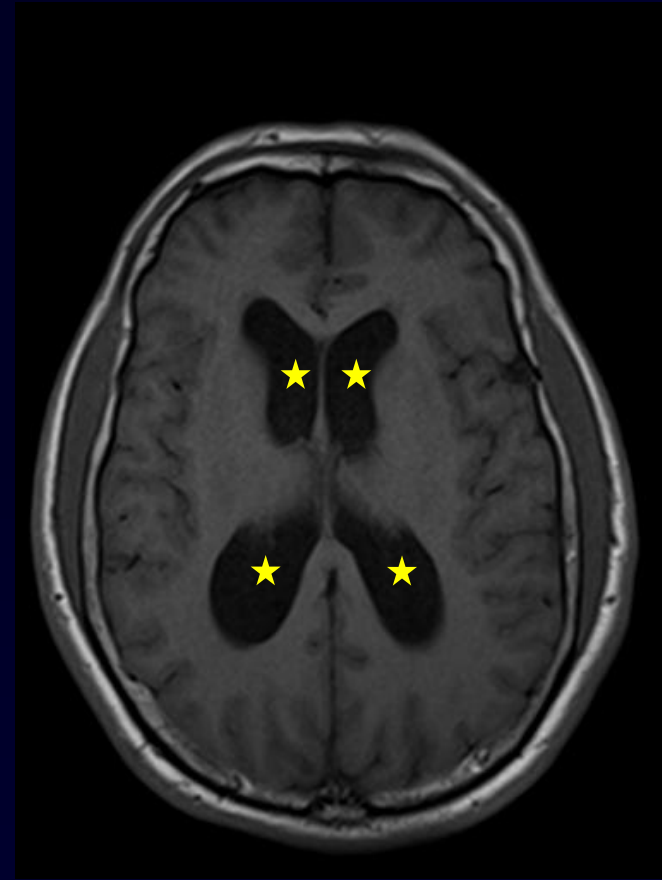
Axial C(+) MRI T1W. PACS, BIDMC

- **Large heterogeneously enhanced mass** centered within the prepontine cistern arising from, or invading into, the clivus

# Our Patient: MRI Findings Mass Effect



Sagittal C(-) MRI T1W. PACS, BIDMC



Axial C(-) MRI T1W. PACS, BIDMC

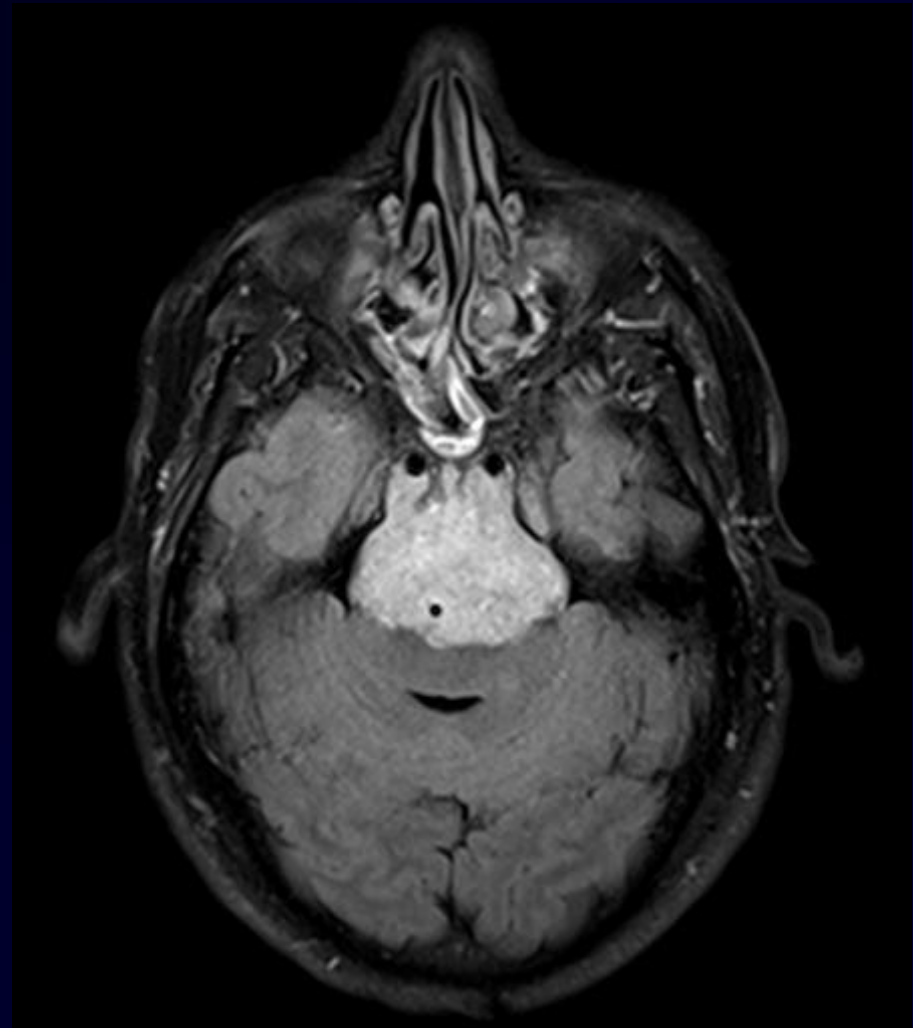
- Significant mass effect with associated moderate hydrocephalus





# 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%



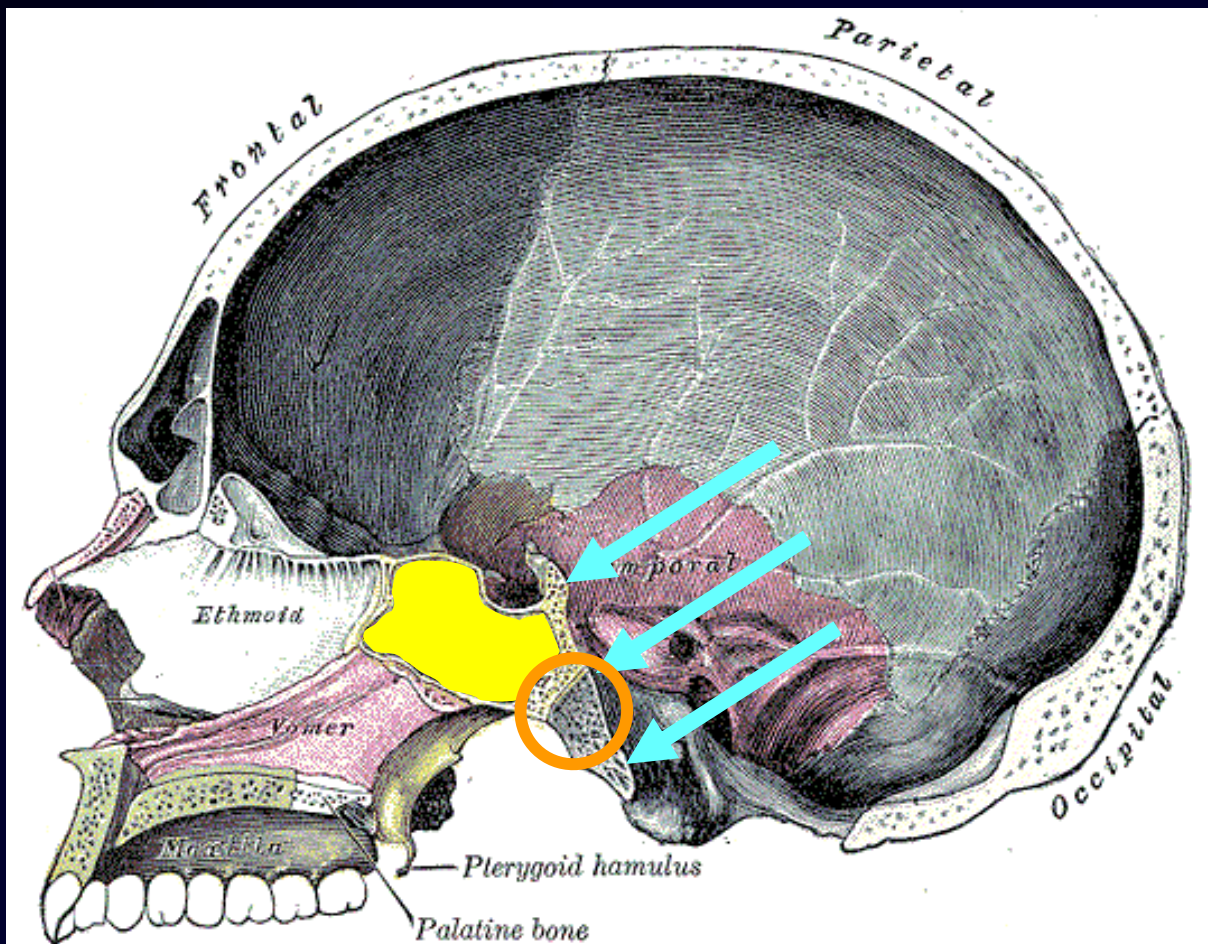
# Chordoma: Intracranial Location

- Most often originates from the **clivus**
  - Especially from the spheno-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



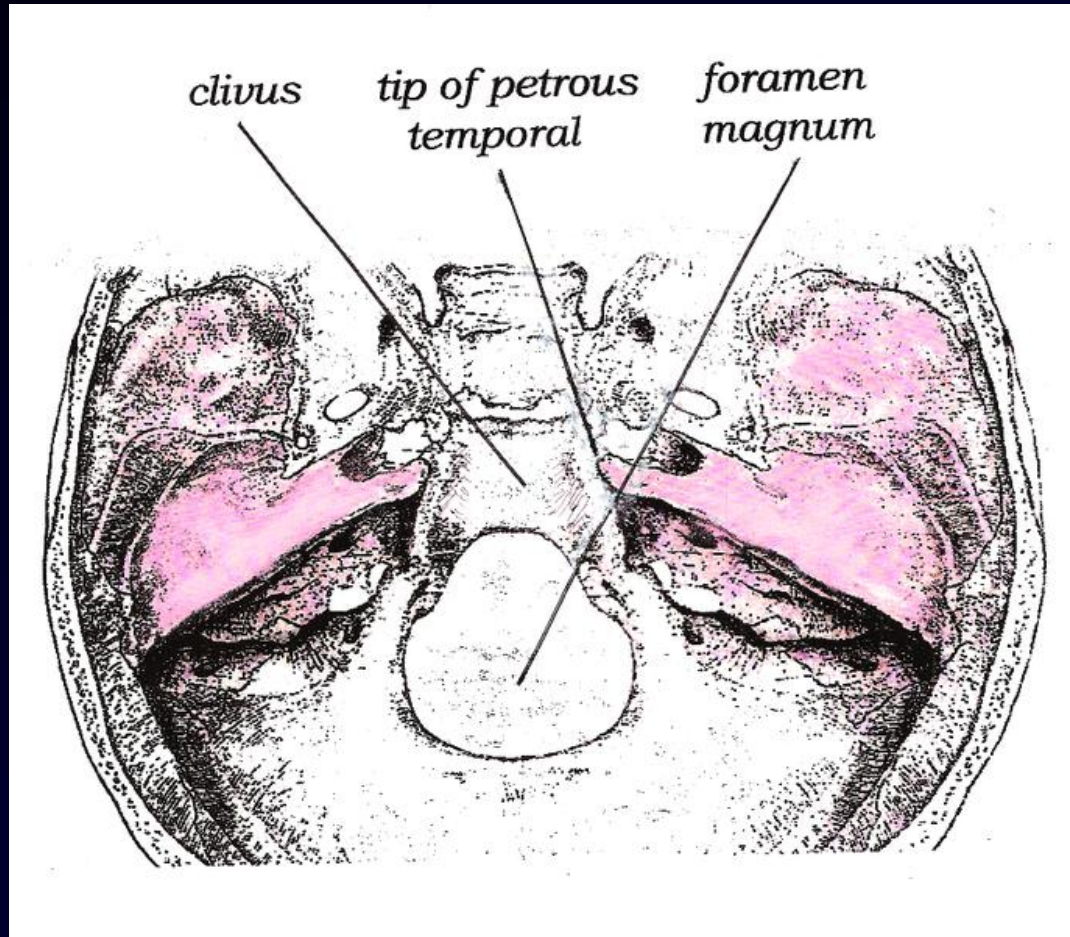
# Chordoma: Anatomy Of The Clivus: Sagittal

- Clivus
- Spheno-occipital synchondrosis
- Sphenoid sinus





# Chordoma: Anatomy Of The Clivus: Axial



<http://en.wikipedia.org/wiki/File:Clivus.png>



# 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 trabeculae**



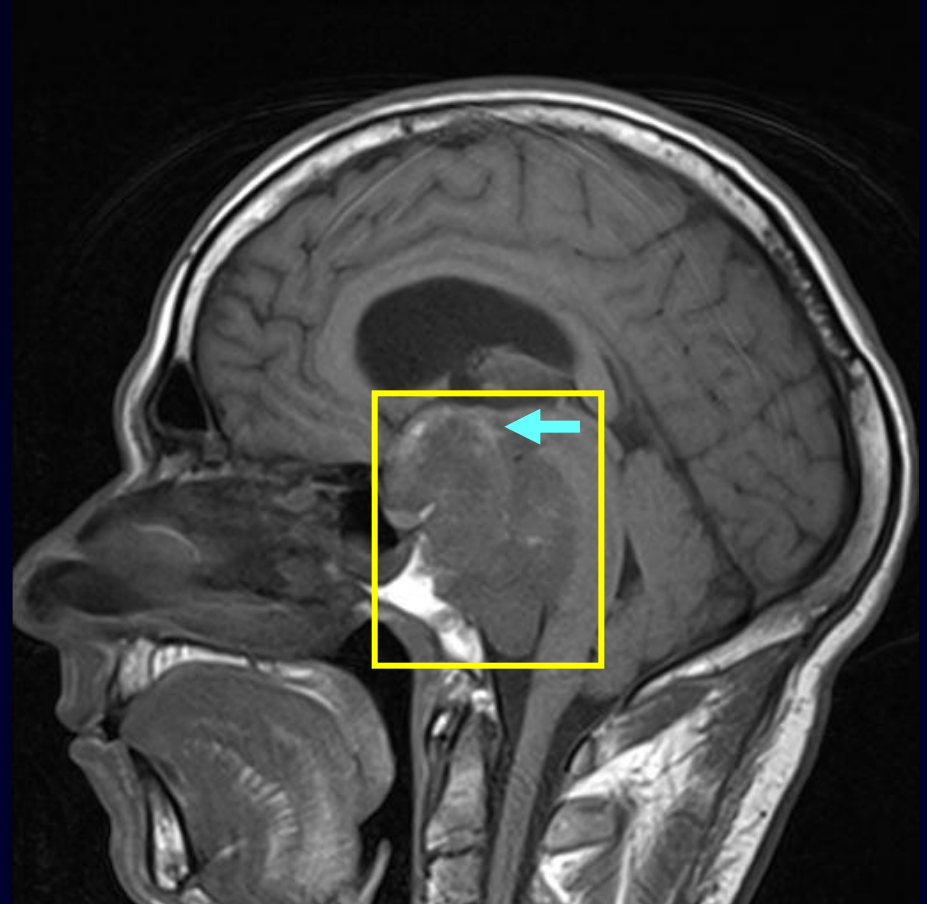


# MRI Features: Planes

- 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

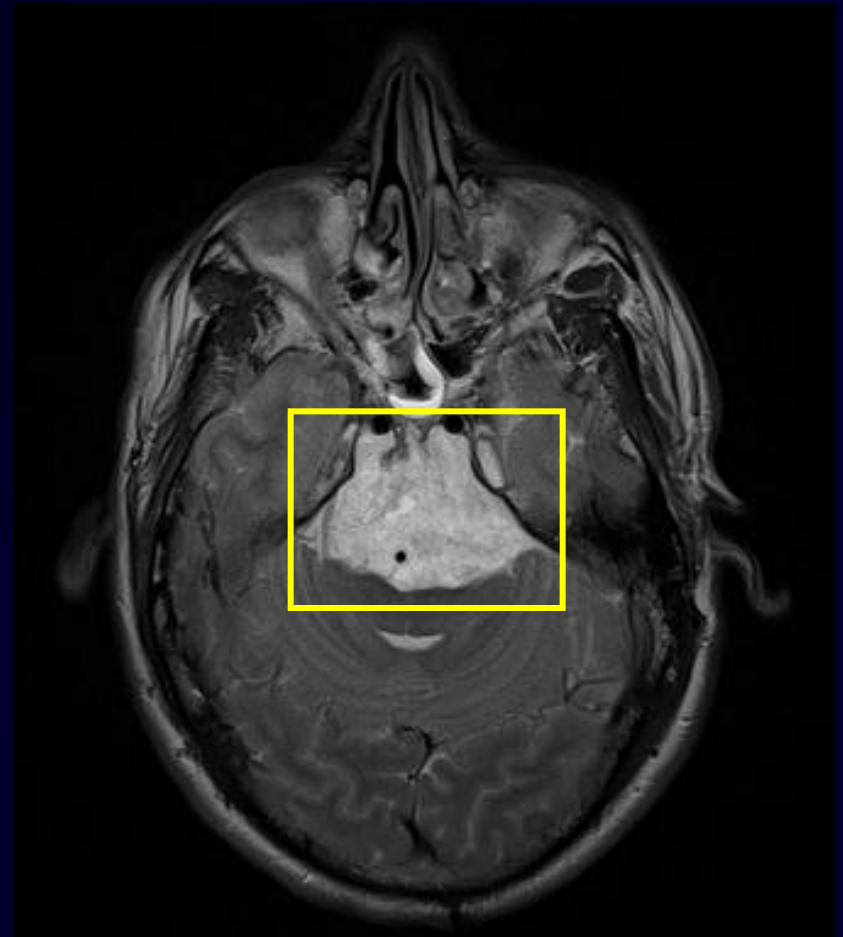
# MRI Features: T1-Weighted Images

- 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

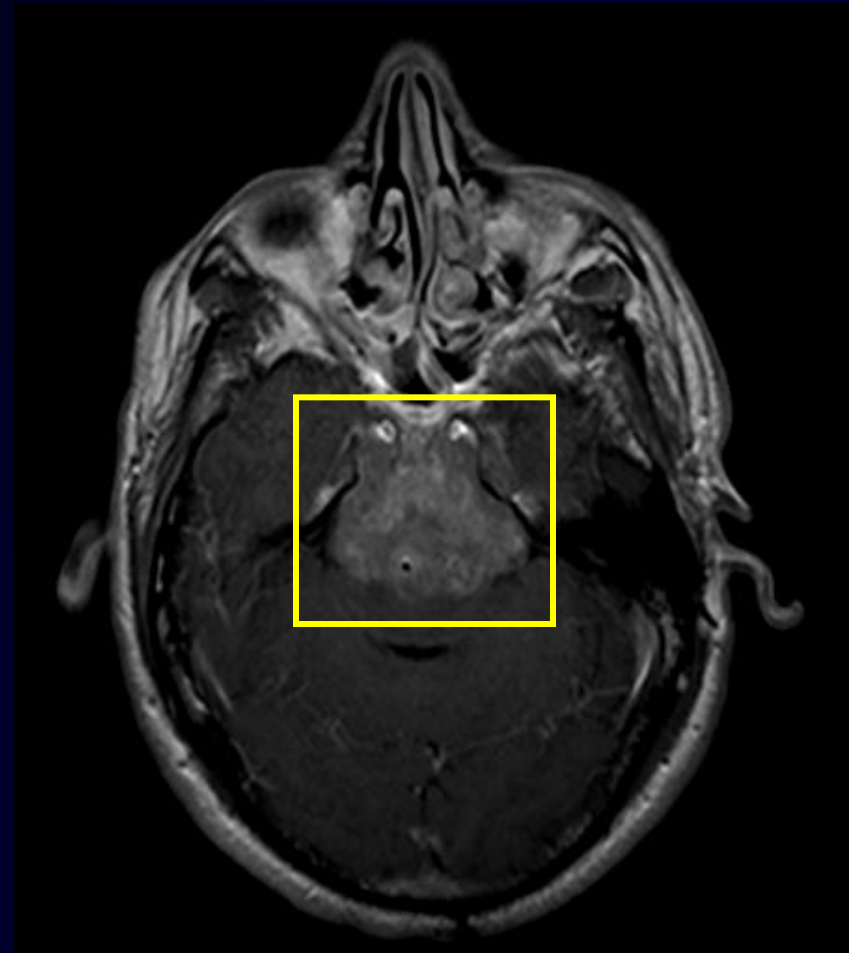
- **High signal intensity**
  - ⇒ High fluid content
- Intratumoral areas of heterogeneous hypointensity
  - ⇒ Calcification
  - ⇒ Hemorrhage
  - ⇒ Highly proteinaceous mucus pool
- Low-signal-intensity septations
- ⇒ Used to differentiate from adjacent neural structures



Axial C(-) MRI T2W. PACS, BIDMC

# 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





# MRI Features: Fat Suppression

- Differentiation of enhanced tumor margins from adjacent fatty bone marrow
- Small intracavitary 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 trabeculea (CT)
- Extremely high recurrence rate
- Extensive surgical removal and fractionated proton beam irradiation are the most effective treatments



# References

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- ❑ Erdem E, Angtuaco EC, Van Hemert R, Park JS, Al-Mefty O. Comprehensive review of intracranial chordoma. *Radiographics*. 2003; 23(4): 995-1009
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- ❑ <http://www.bartleby.com/107/Images/large/image194.gif>
- ❑ <http://en.wikipedia.org/wiki/File:Clivus.png>



# Acknowledgements

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