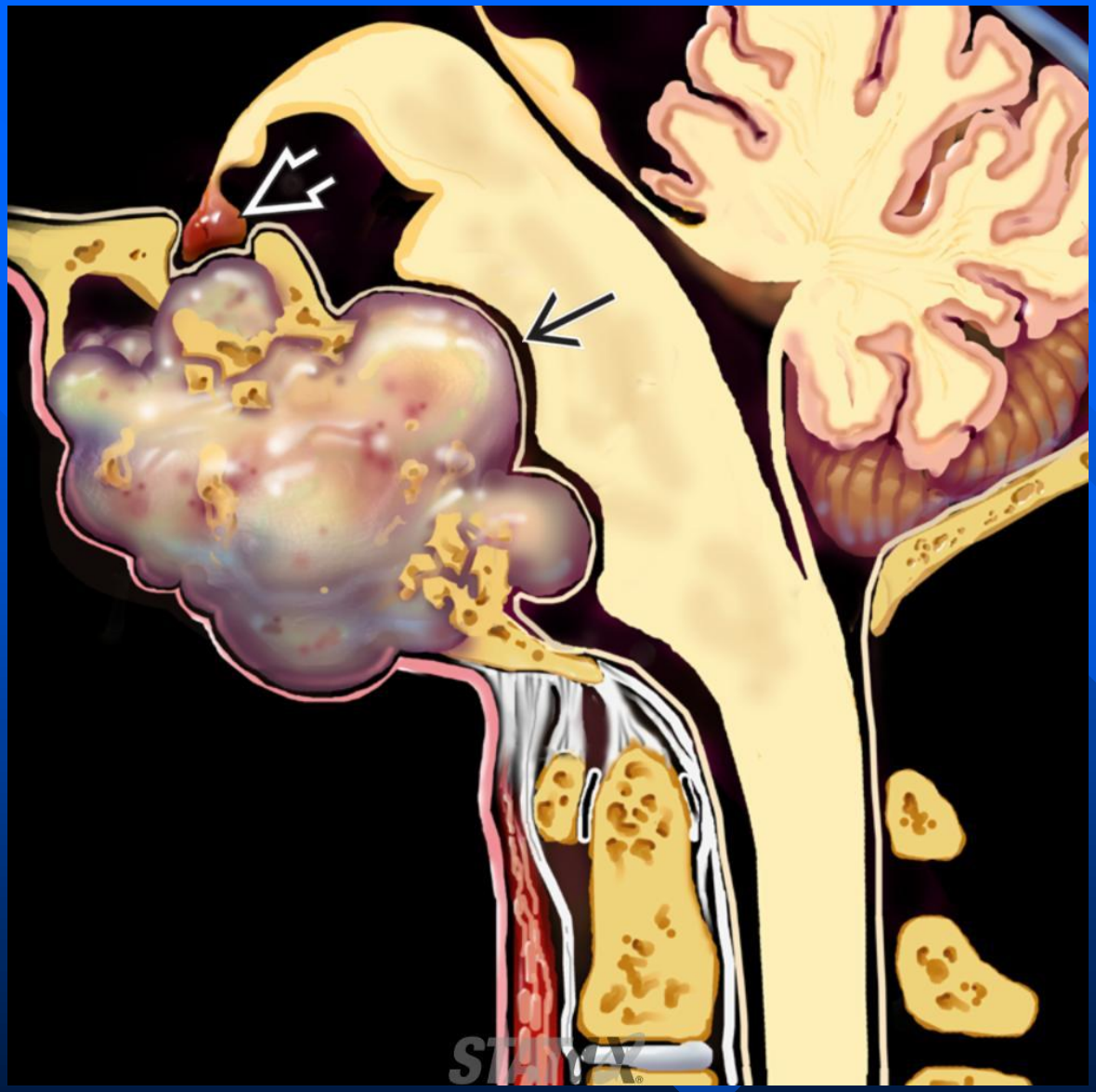
Chordoma

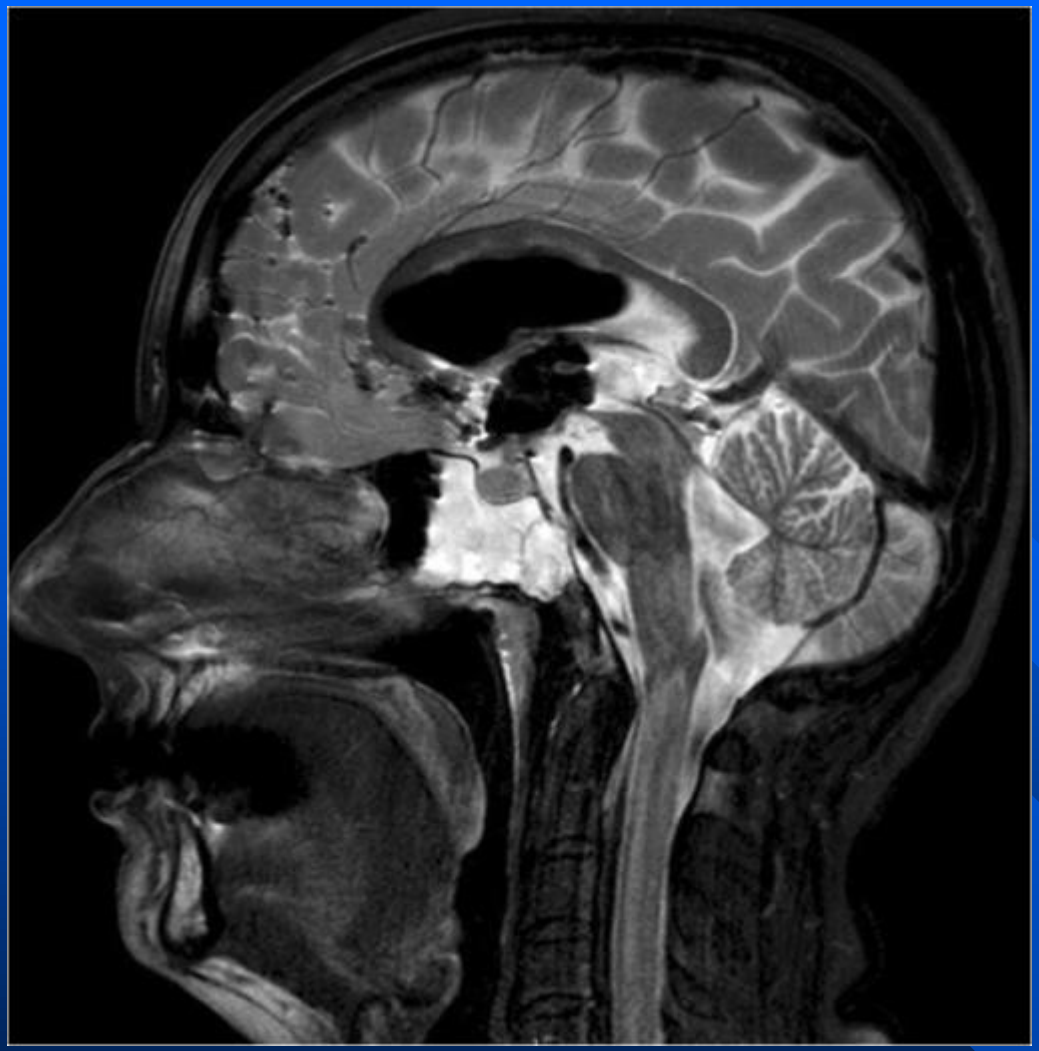
- Rare, locally aggressive tumor of clivus arising from cranial end of primitive notochord remnant
- Imaging
- Location: Clivus; sphenooccipital synchondrosis
  - Can occur anywhere along primitive notochord
- CT findings
  - Midline, expansile, multilobulated, well-circumscribed mass
  - Lytic bone destruction with intratumoral Ca^{++}
  - Variable enhancement
- MR findings
  - T1: Intermediate to low signal ≈ brain
  - T2: Classically ↑ ↑ signal
  - T1WI C+: Moderate to marked enhancement
  - DWI: Mean ADC value 1474 ± 117 x 10^{-6} mm^2/s, generally less than chondrosarcoma

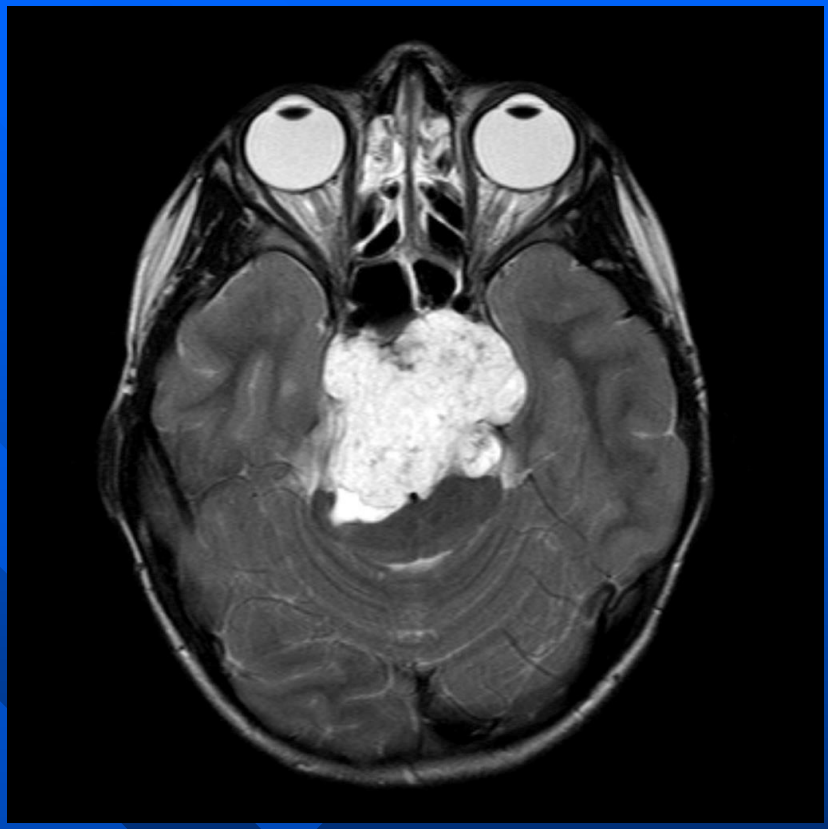
DDX:

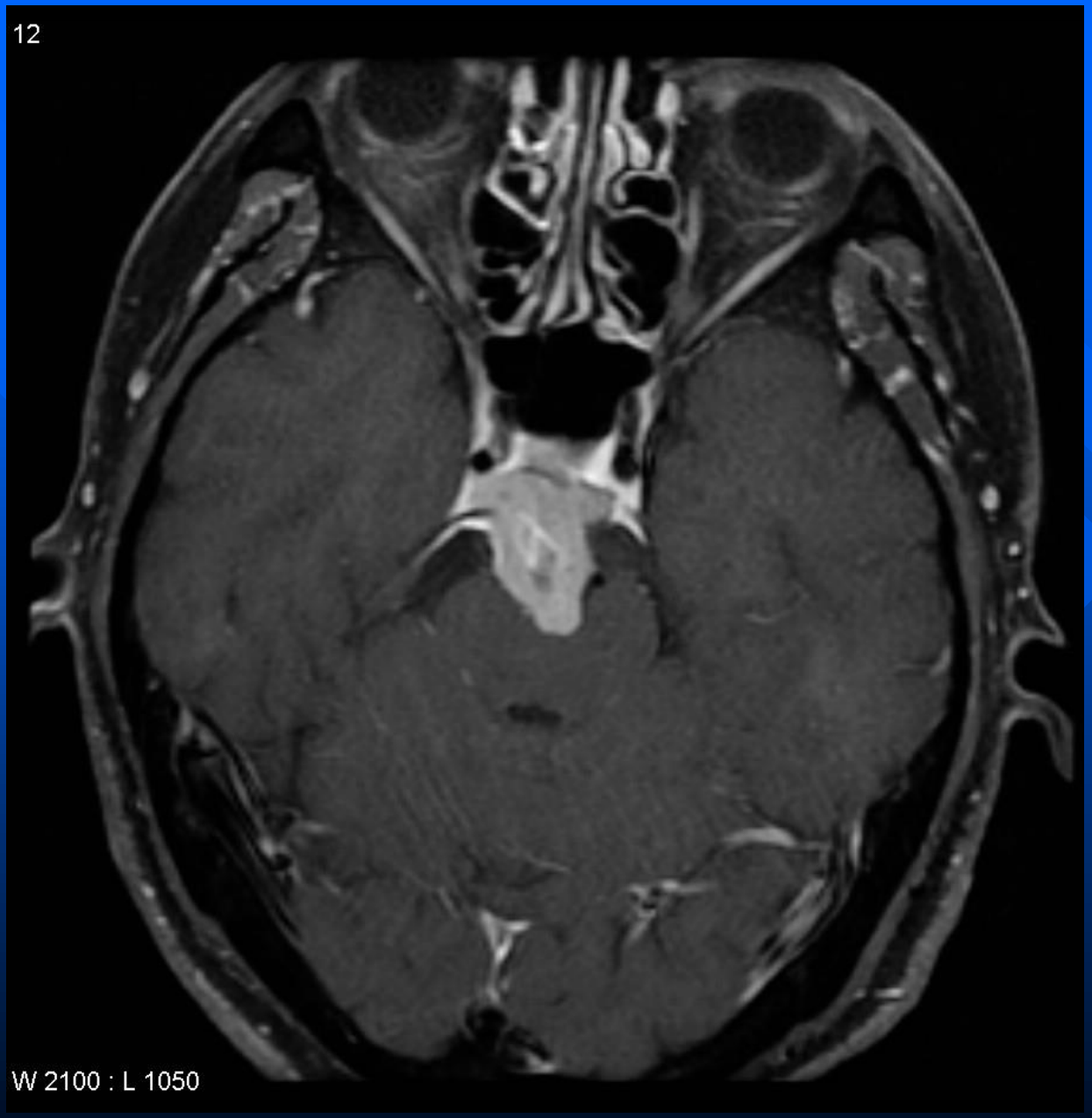
- Invasive Pituitary Macroadenoma
  - Originates in sella & involves pituitary gland
  - Extends into sphenoid sinus, not prepontine cistern
- Ecchordosis Physaliphora
  - Rare, benign notochord remnant lesion
  - Nonenhancing, T2-hyperintense mass posterior to clivus
- Skull Base Chondrosarcoma
  - Arises off midline at petrooccipital fissure
  - Similar T1 & T2 characteristics to CCh
  - Chondroid calcifications more common
  - ADC values 2051 ± 261 x 10^{-6} mm^2/s, much higher than chordoma
  - IDH1 mutations common but not seen in chordoma
- Skull Base Plasmacytoma
  - Can be midline destructive mass of clivus
  - T2 signal usually intermediate to low
- Skull Base Metastasis
  - Destructive lesion; extraosseous component < CCh
  - Known primary neoplasm
- Skull Base Meningioma
  - Sclerosis/hyperostosis of adjacent bone
  - Homogeneous enhancement with dural tails
  - Commonly causes narrowing of encased vessels



Sagittal graphic shows an expansile, destructive mass originating from clivus, "thumbing" pons (black solid arrow) & elevating the pituitary gland (white open arrow). Note bone fragments floating in chordoma.



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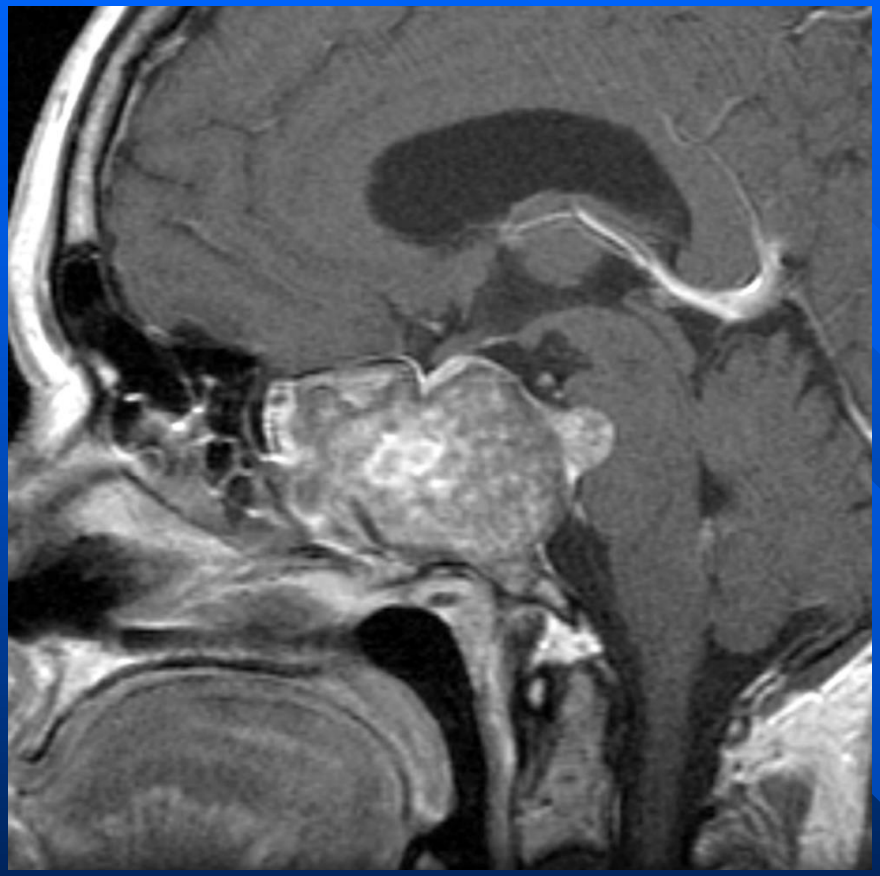
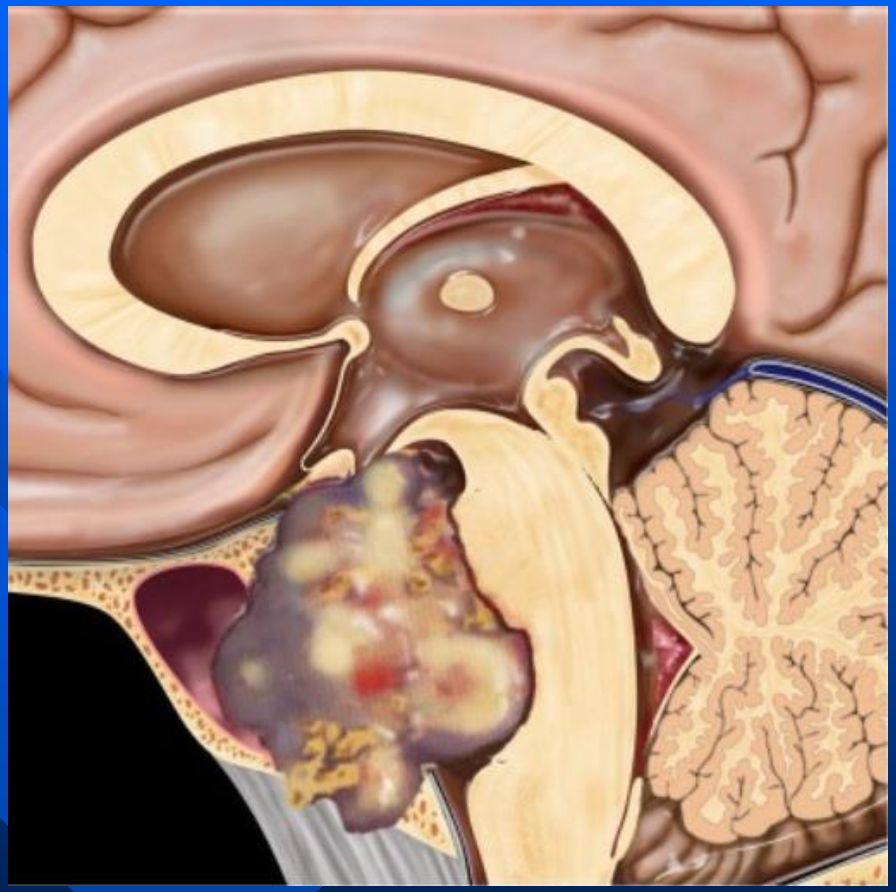
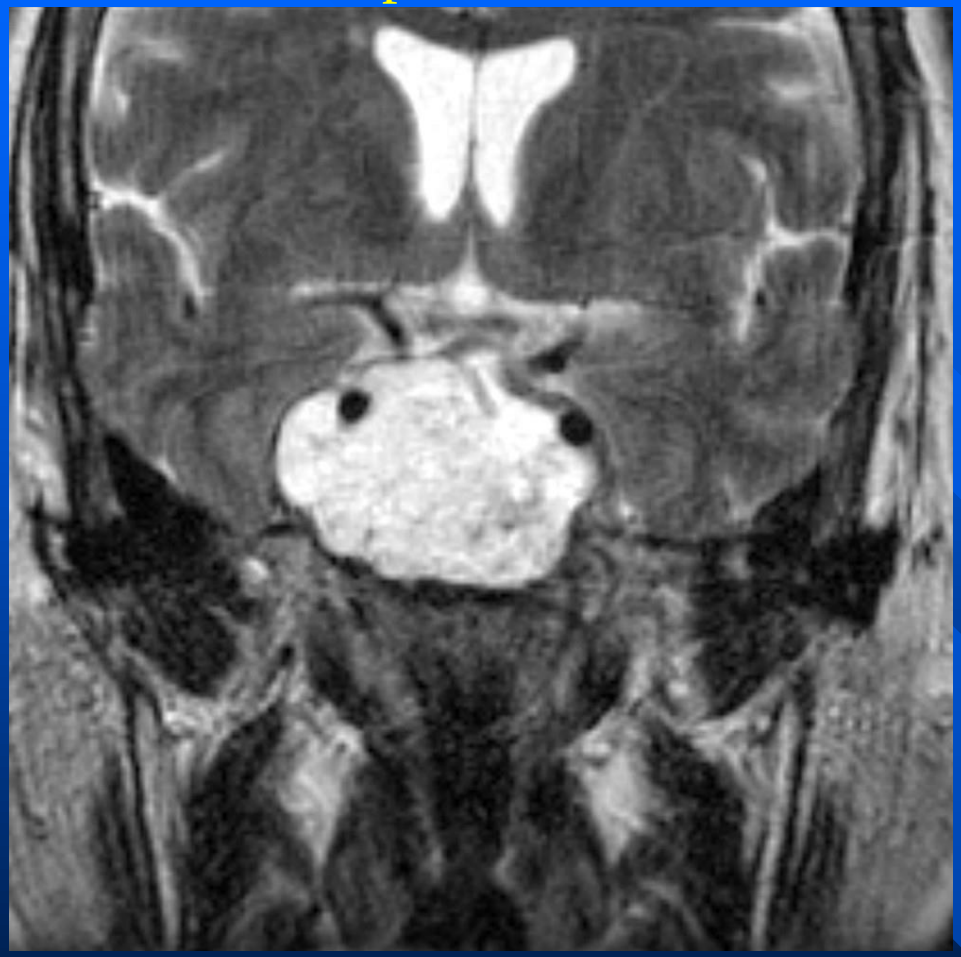
W 2100 : L 1050

Classic Chordoma Of Clivus

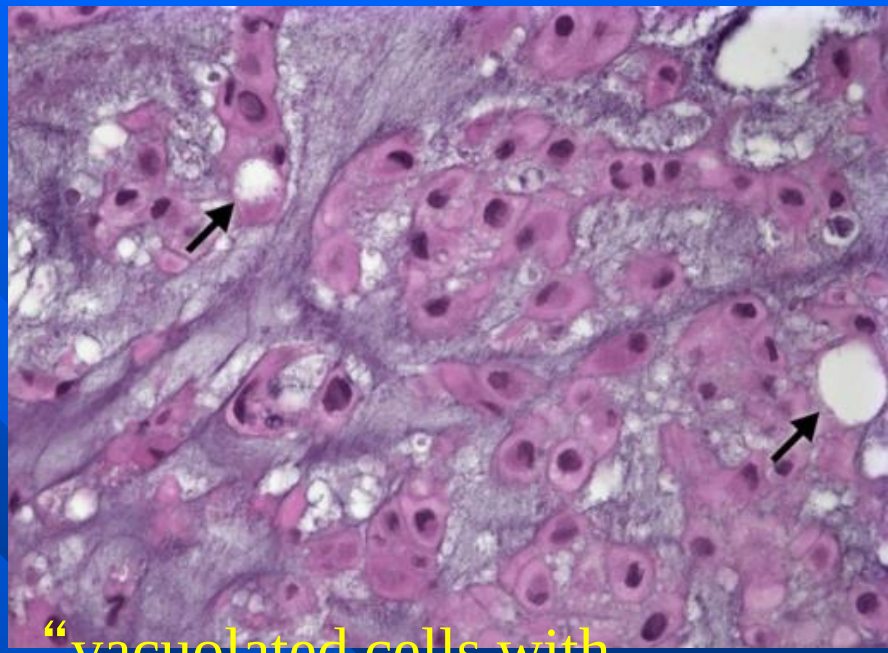




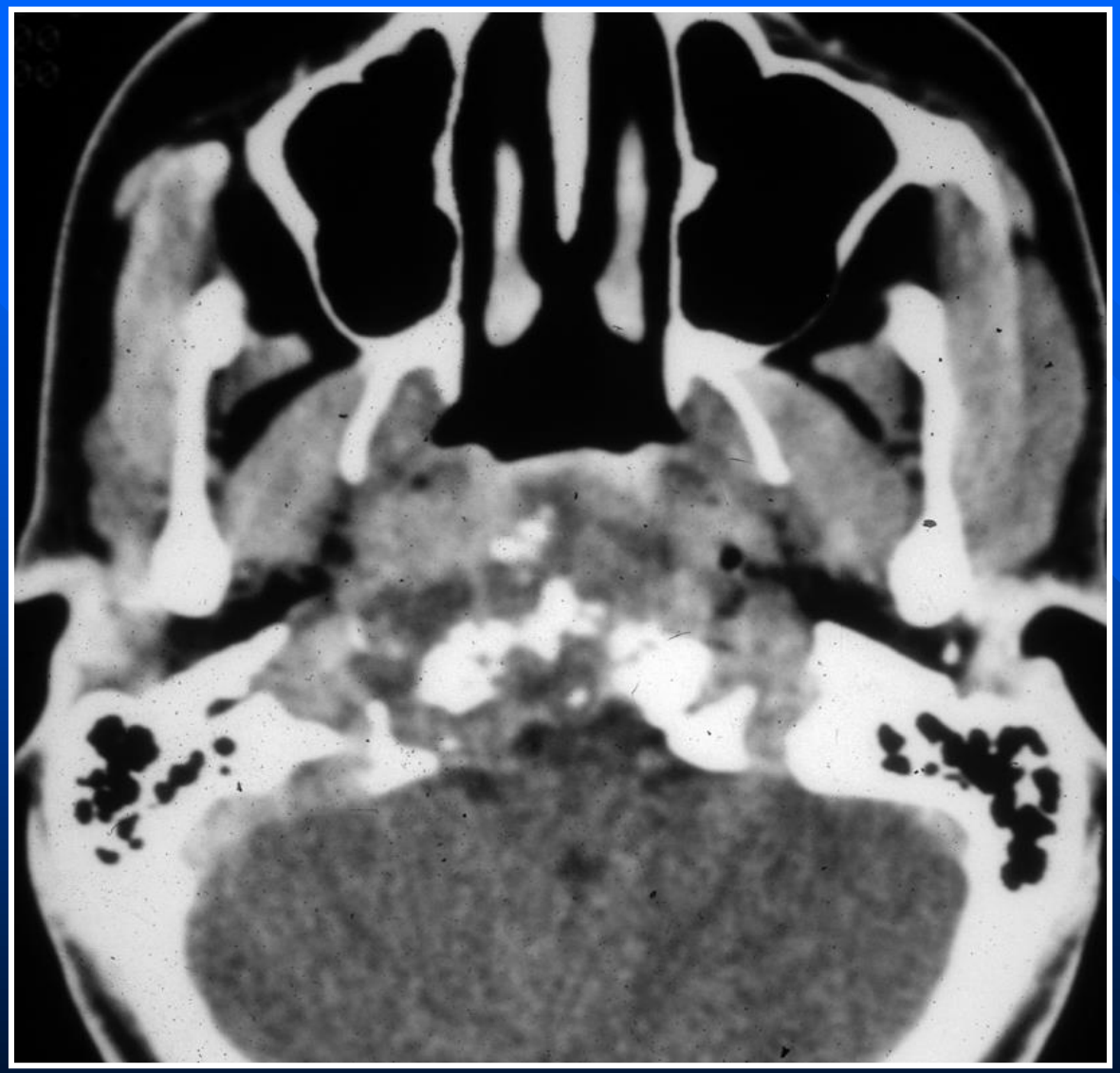
Example: Chordoma

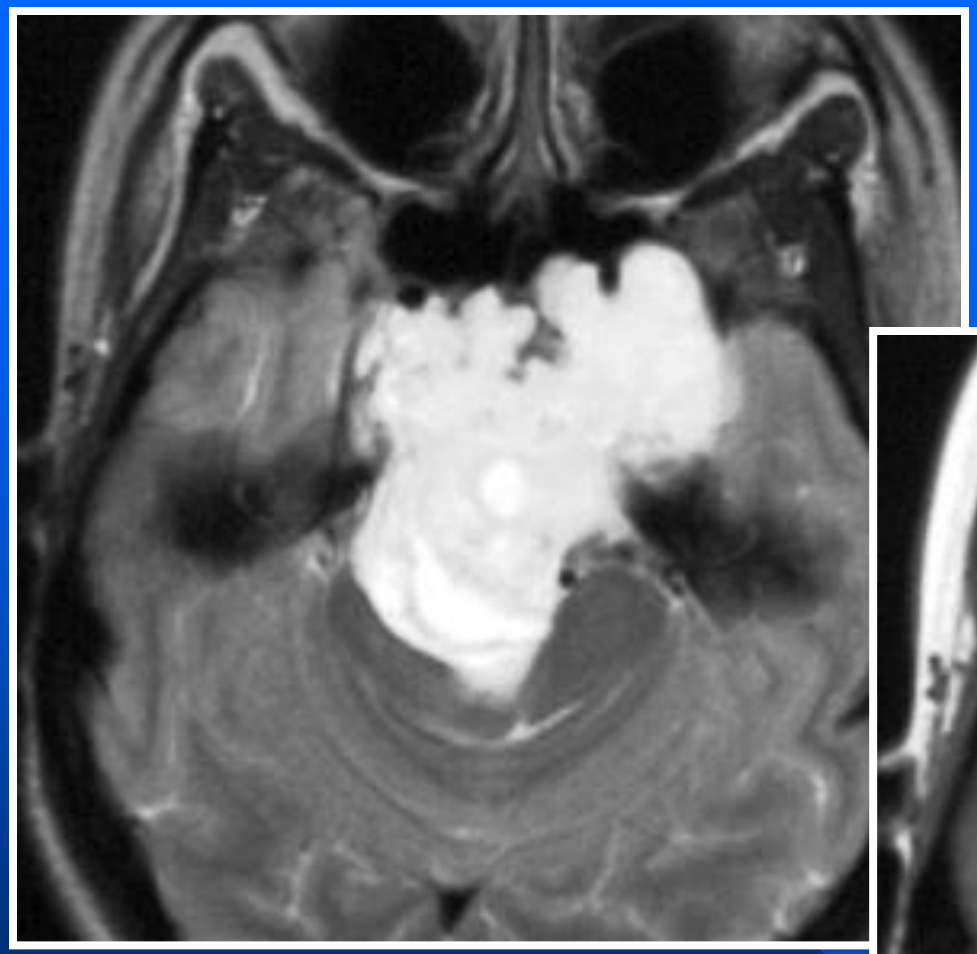


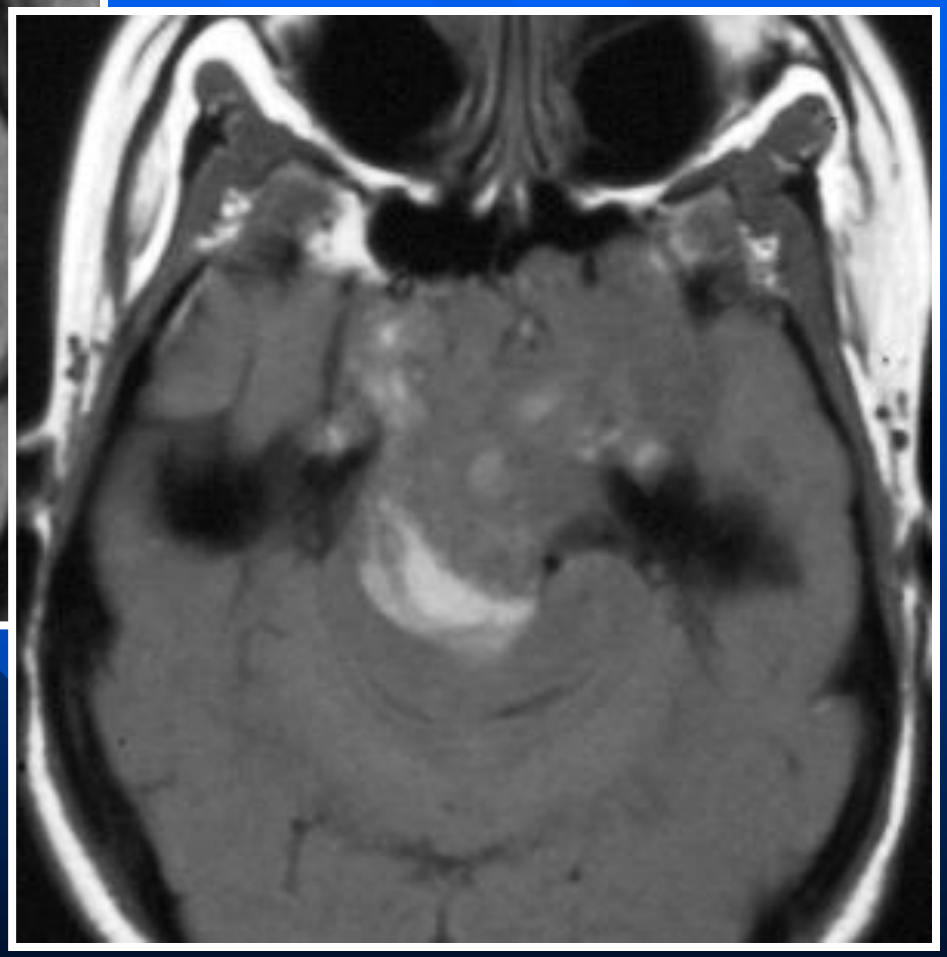
T2W



"vacuolated cells with intracytoplasmic mucus droplets (physaliphorous)"
Erdem, E. Radiographics 2003 23:995







Chordoma vs. Chondrosarcoma

- Both are rare skull base tumors
- Both are typically hyperintense on T2 and cause lytic destruction on CT
- Both are typically slow growing
- Both may present with cranial neuropathies
- At light microscopy, Chordoma were thought to have “chondroid” features (ex: chondroid Chordoma)

Chordoma vs. Chondrosarcoma

- At light microscopy, chordomas were thought to have “chondroid” features (ex: chondroid chordoma)
- These tumors are immunohistochemically distinct. Chordoma actually has no chondroid elements.
- Chordomas occur in midline: Clivus
- Chondrosarcomas typically Petro-occipital Fissure