Chordoma

- Low- to intermediate-grade malignant tumor that recapitulates notochord.
- Slowly growing, often with nonspecific symptoms
  - Eventually develops symptoms related to location of spread into adjacent soft tissues
- 1-4% of primary malignant bone tumors
- Of sacral tumors, 40% are chordomas
- High recurrence rate (80%) with marginal resection
- 5-year survival rate: 50-84% in different studies
- Recent study shows that lesion is highly malignant over long term
- May contain dedifferentiated foci; poorer prognosis

Chordoma

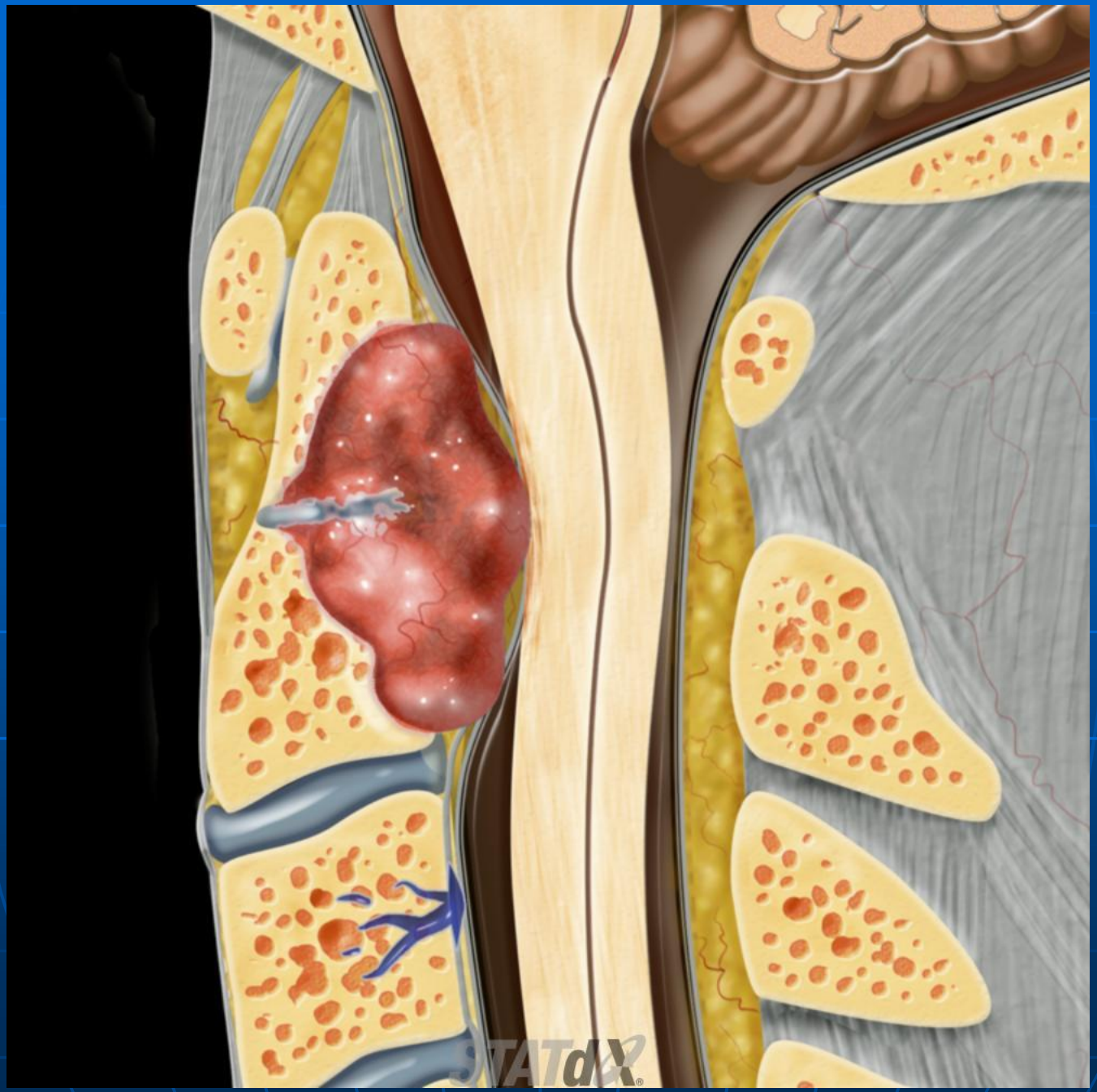
- **Demographics**
- **Age**
  - Generally > 30 years; extremely rare < 20 years
    - Younger patients tend to have sphenooccipital involvement more frequently than other regions
  - Most commonly presents in 6th decade (30%)
  - Skull and spine lesions tend to present 10 years earlier than sacral lesions
  - Chondroid chordomas generally occur in patients < 40 years old
- **Gender**
  - Male > female (1.8:1)
  - Female > male for chondroid chordoma variety

Location

- Sacrum (60%);
  - predominantly in lower sacral elements
- Sphenooccipital/nasal (25%)
  - 33% are of chondroid chordoma variety
- Cervical spine (10%)
- Thoracolumbar spine (5%)
- Usually midline in origin

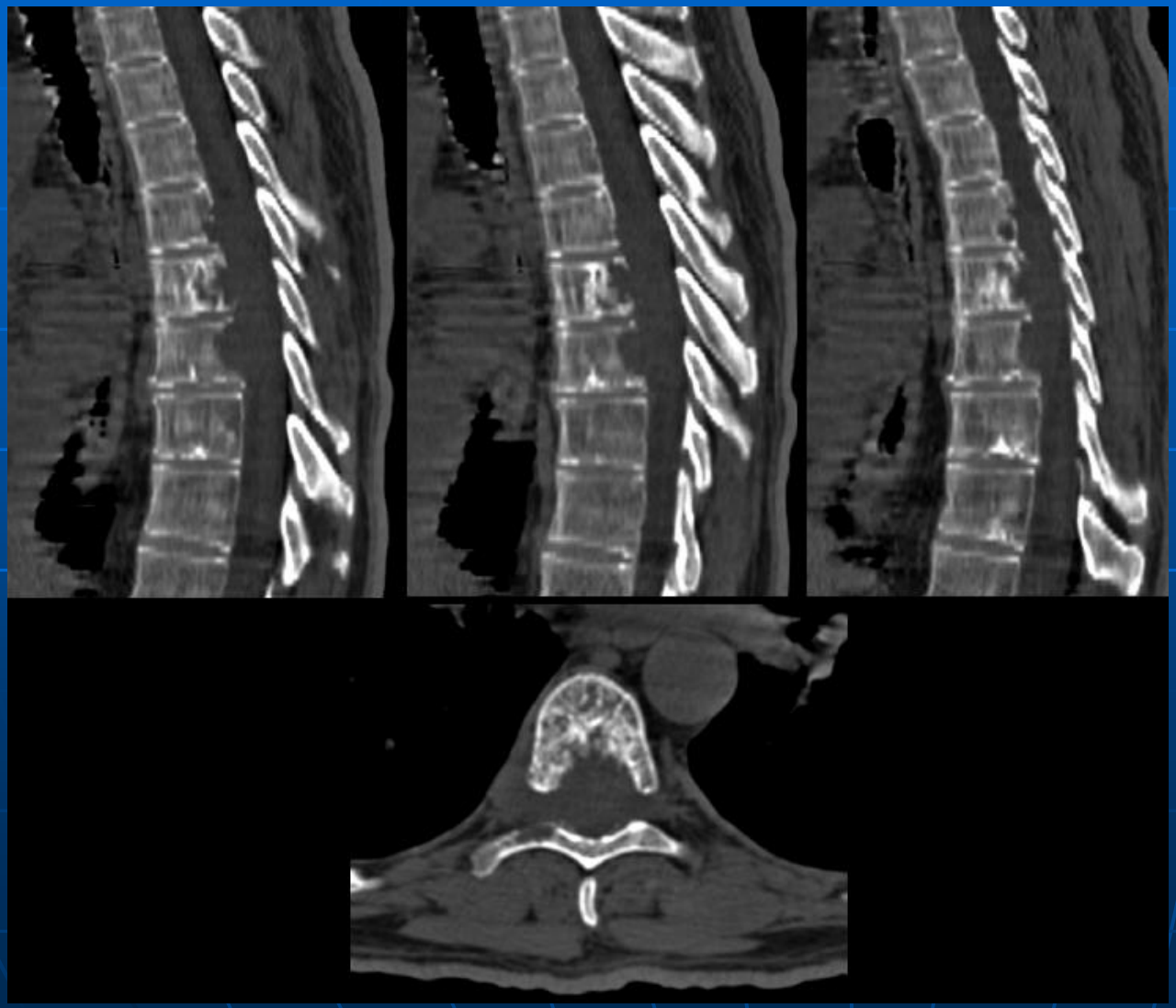
Imaging

- Location: Sacrum (60%)
  - Sphenooccipital/nasal (25%)
  - Cervical spine (10%)
  - Thoracolumbar spine (5%)
- Radiographic appearance: Lytic destructive lesion
  - Internal calcification may be present (50-70%)
  - Calcification prominent in chondroid chordoma (located virtually only in clivus)
  - Bony debris often carried into soft tissue mass; may mimic matrix
- CT: 90% show calcification
  - Though lesion is locally aggressive, time course is slow enough that it may not appear aggressive
- MR T1WI: Isointense to hypointense; inhomogeneous if dense calcification present
- MR fluid-sensitive sequences: Inhomogeneous but generally very high signal
  - May appear lobulated
  - Significant regions of low signal if dense calcification present



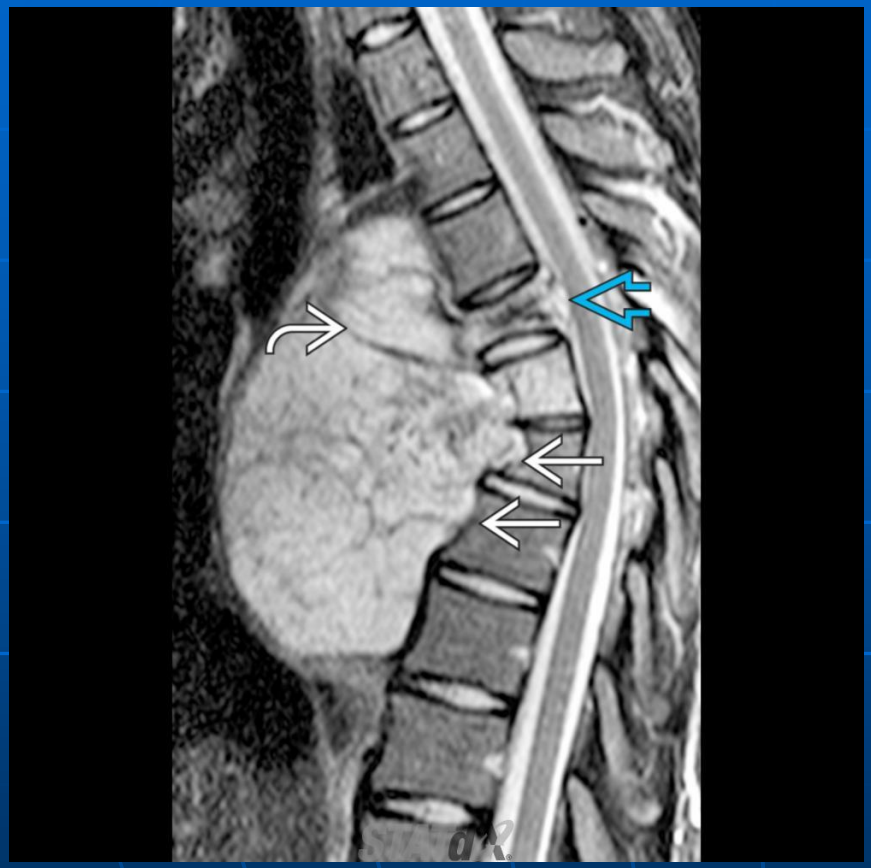
Sagittal graphic shows chordoma arising in the posterior aspect of the C2 vertebral body, extending posteriorly and causing compression of the spinal cord.

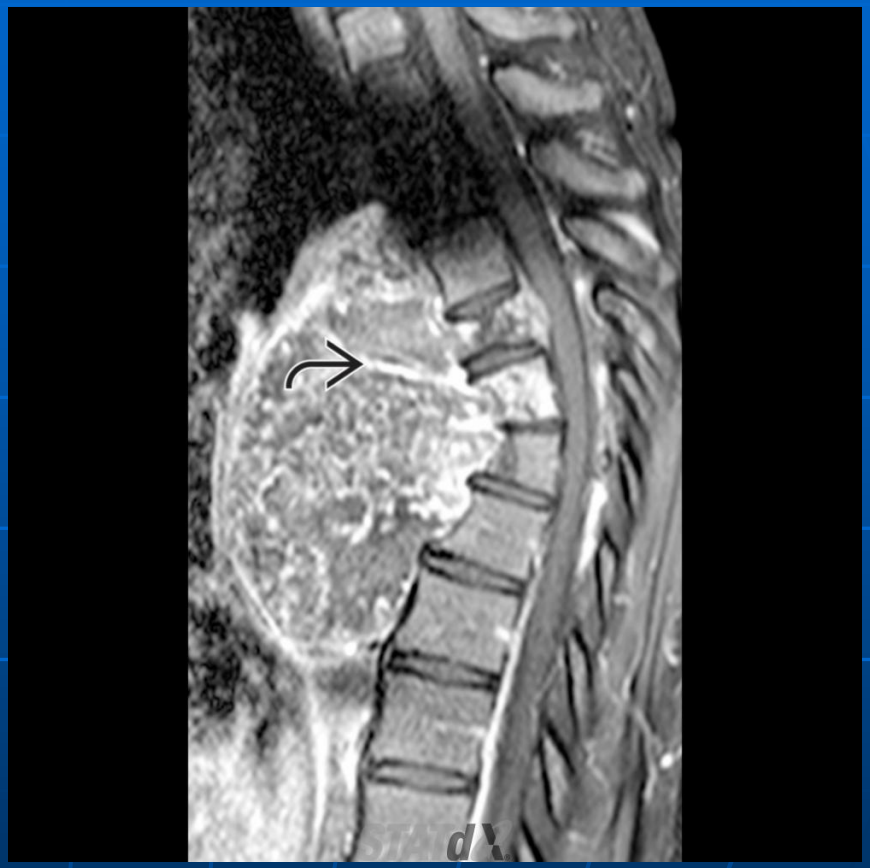
Chordoma



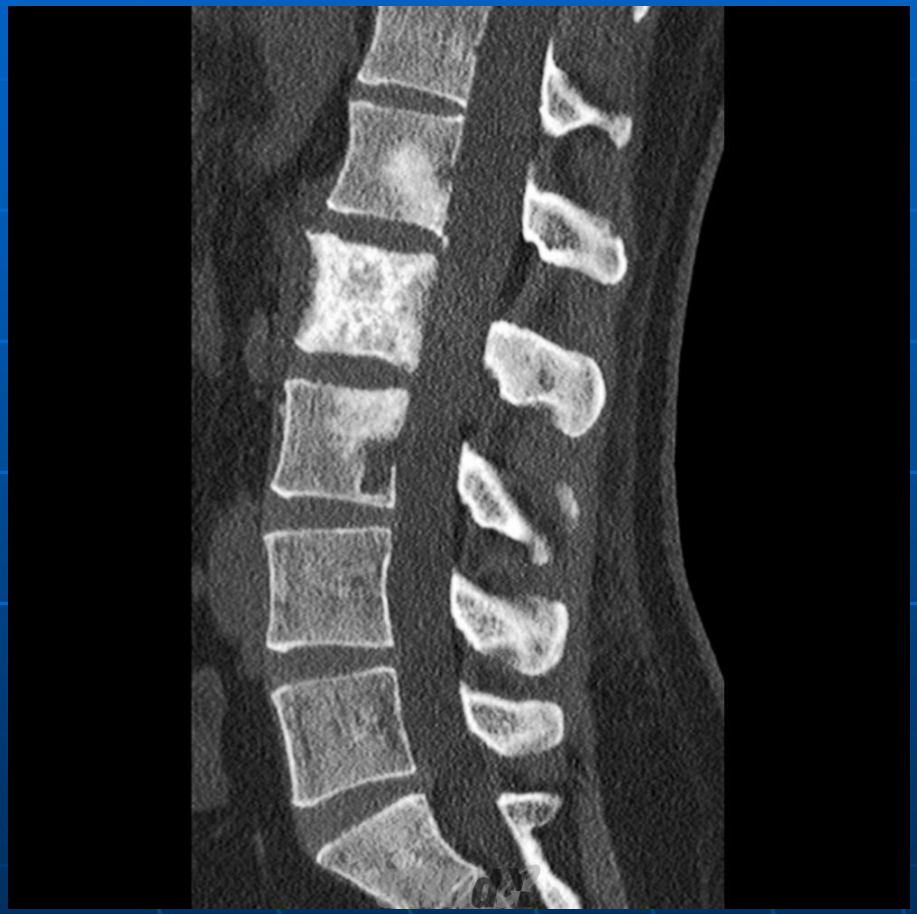
Chordoma and Lymphoma are tumors that can cross the disc space

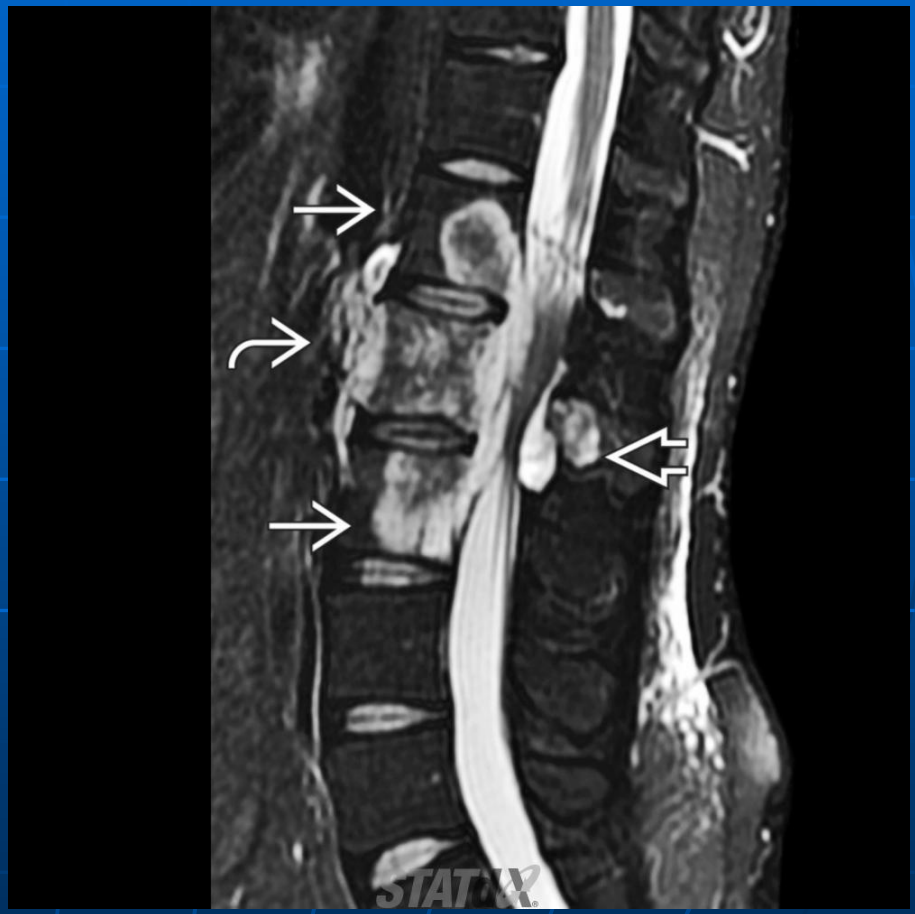
T2 - hyperintense





Sagittal T2WI FS MR shows a large posterior mediastinal mass with involvement of 4 adjacent thoracic vertebral bodies. The mass is hyperintense with fine septations (white curved arrow). There is a pathologic compression fracture of the top vertebra, with ventral epidural tumor (cyan open arrow). Inferiorly, there are scalloped anterior margins (white solid arrow).

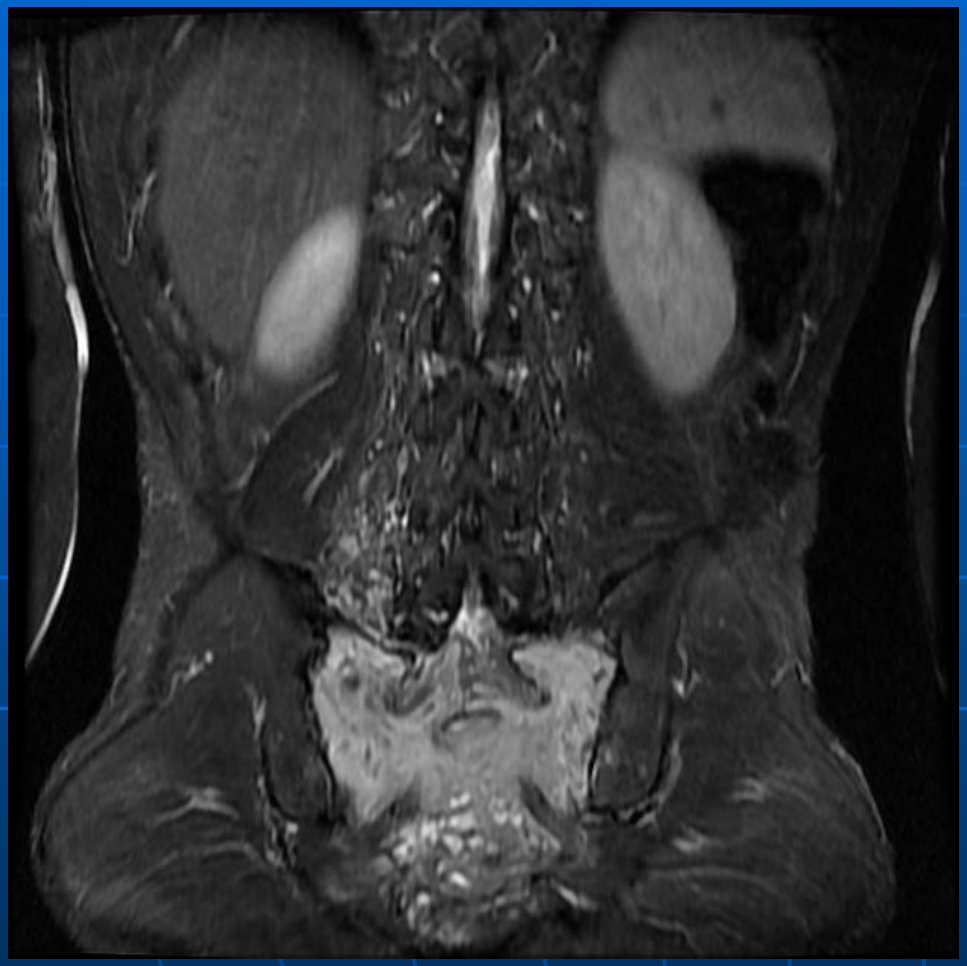


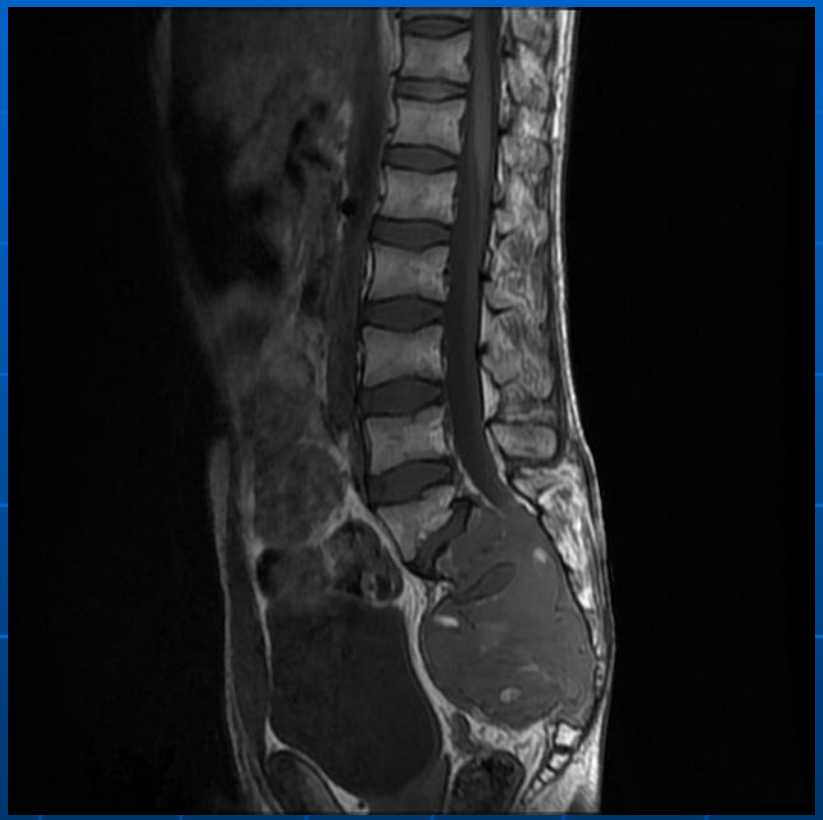


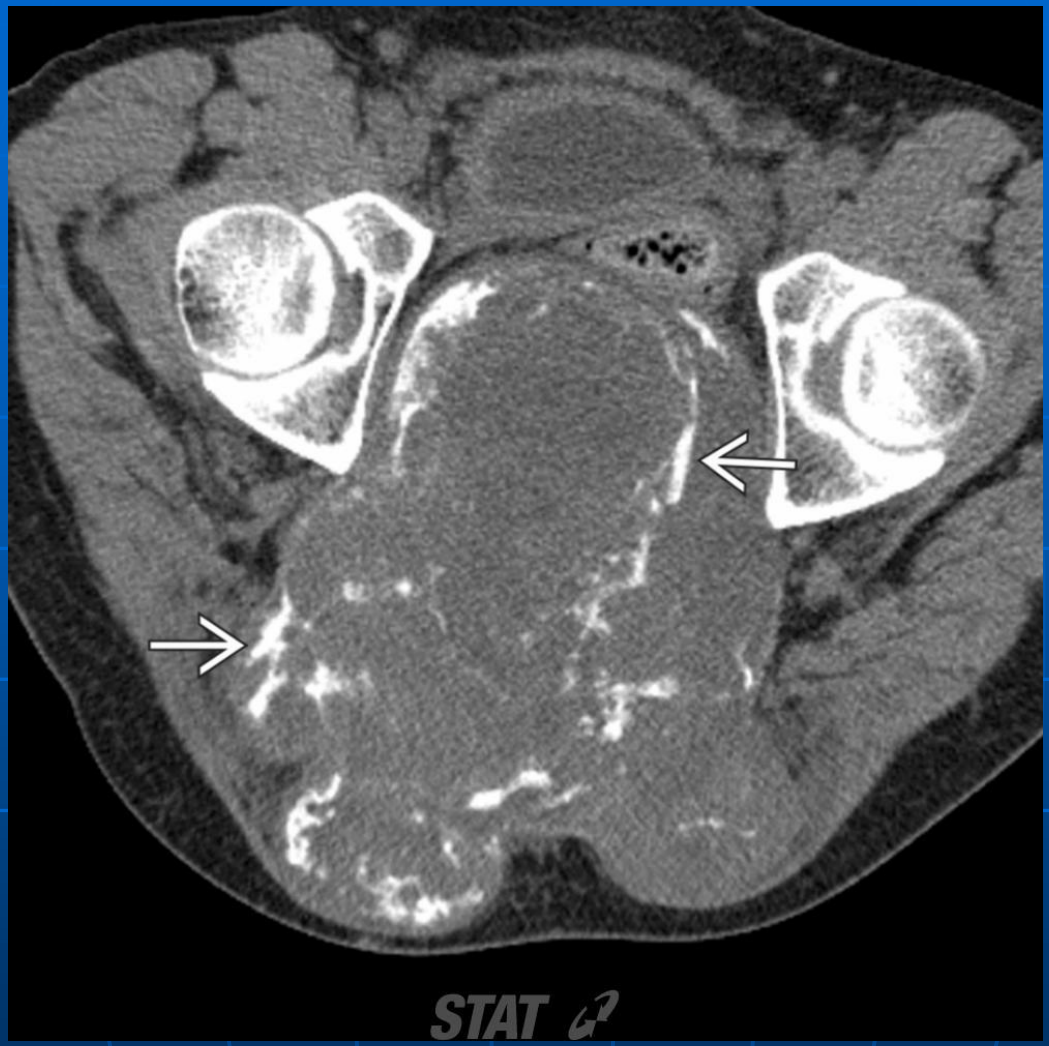
Sagittal NECT shows predominately osteosclerotic involvement of the L1-L3 bodies in this large lumbar chordoma.

Sacrococcygeal

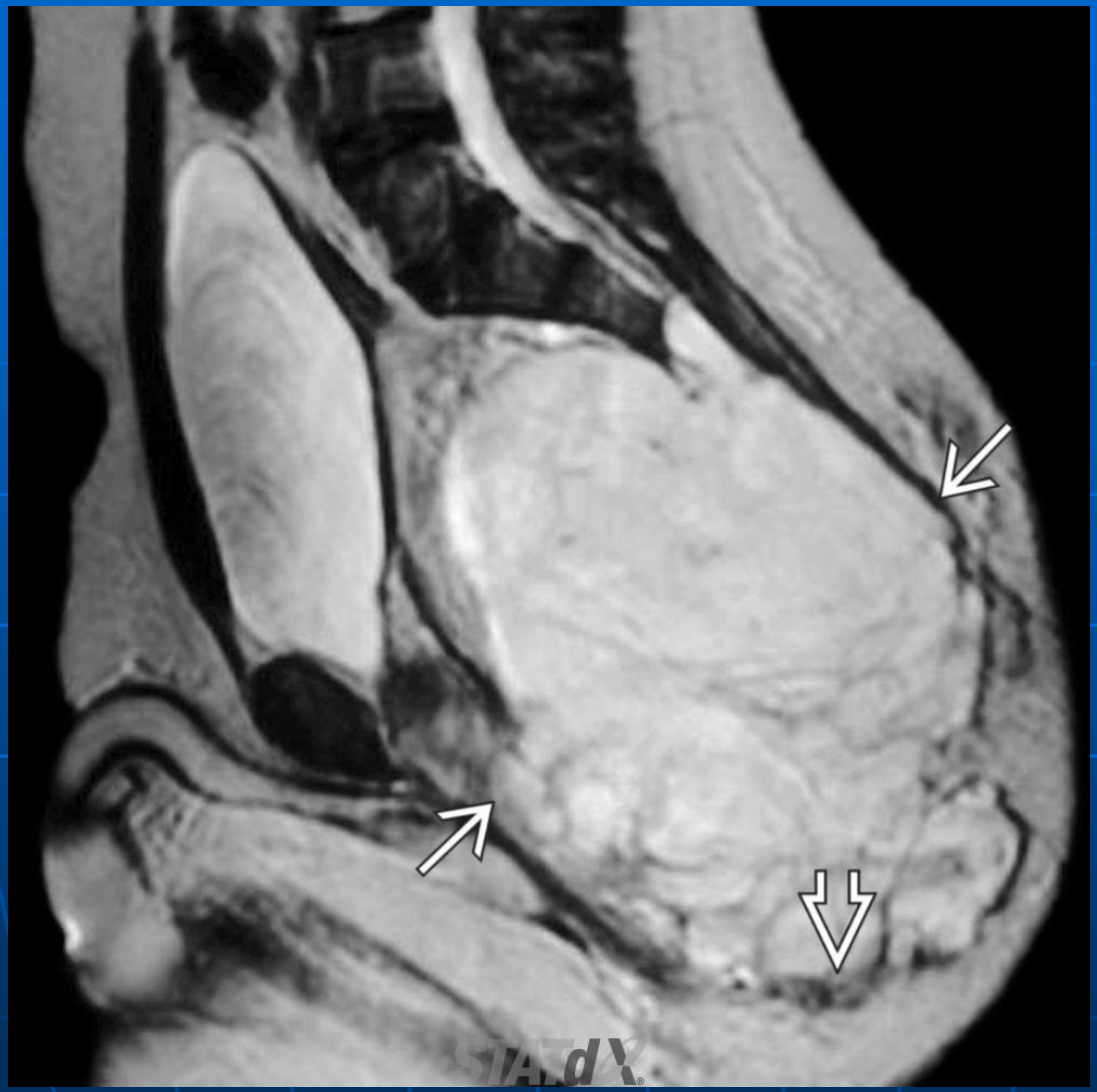
- Most common location, accounting for approximately 30-50% 2,3 of all chordomas and commonly involving the fourth and fifth sacral segments.
- In this location, a male predilection has been reported (M:F ratio of 2:1), and the tumor may be particularly large at presentation.
- Most common primary malignant sacral tumor.







- Axial CT in the same patient shows the large size of the sacral mass.
- Note that calcification (white solid arrow) does not have a chondroid appearance.
- The calcification is mostly distributed peripherally; this appearance is typical of chordoma. Such calcification is seen in 90% of chordomas by CT.



Sagittal T2WI MR shows the mass to be mildly lobulated and intensely high signal (white solid arrow), typical of chordoma. Note that the mass has replaced the osseous elements from which it arose. There are peripheral low signal calcifications (white open arrow), as is seen in up to 90% of chordomas. The bladder is displaced anteriorly and compressed.