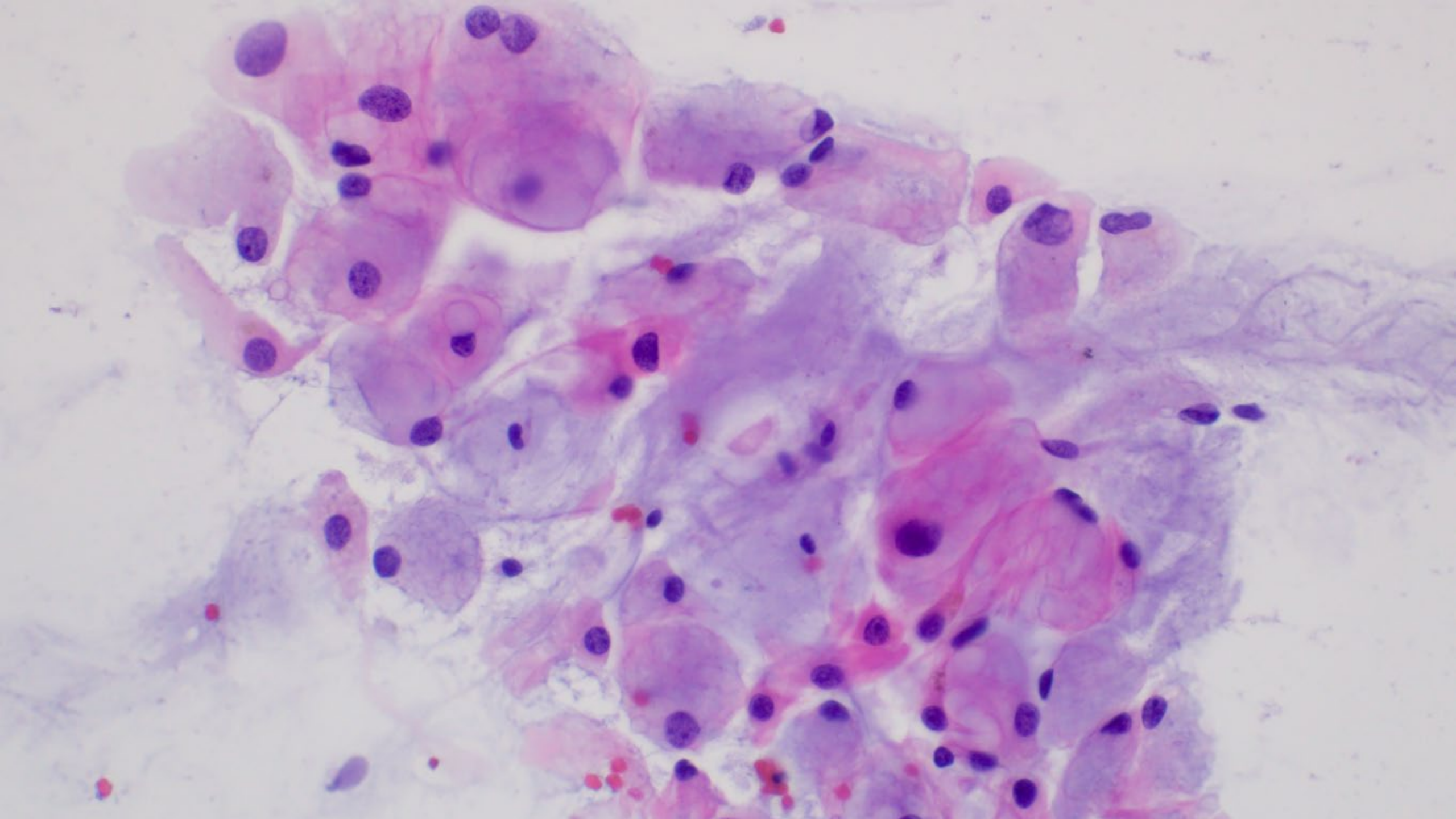
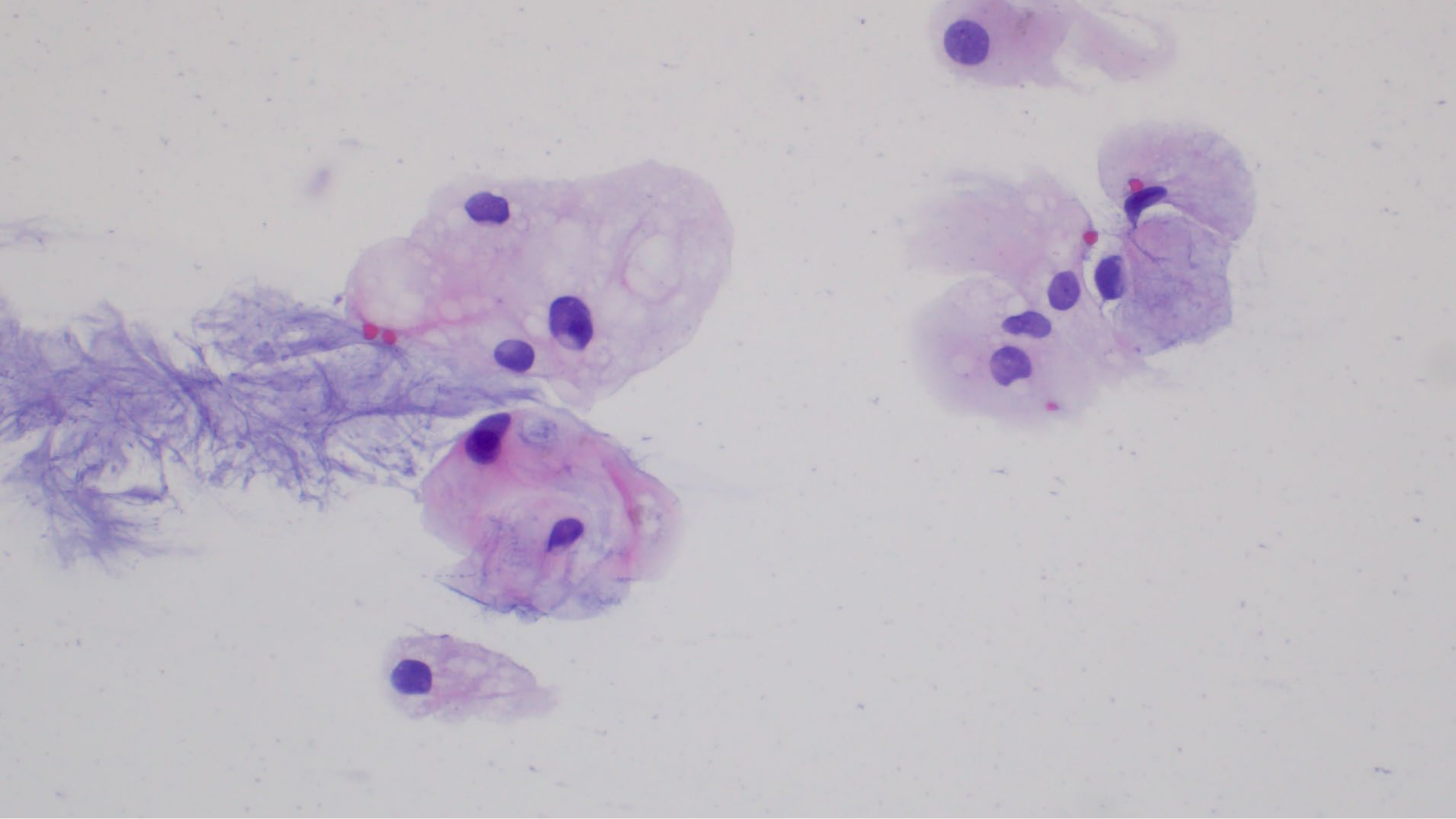
A 14-year-old female presented for evaluation of 3 days of periumbilical pain.  MRI revealed a well-circumscribed ventral/right lateral epidural lesion at L4-L5 level extending into the right neural foramina.  A laminectomy was performed and a small portion of gelatinous-appearing, lobular tissue was submitted for intraoperative review.





Chordomas almost always arise within the axial skeleton, particularly in the skull base and the sacrococcygeal region, but anatomical distribution varies depending on age and histopathological type.

Conventional chordoma is divided into lobules by fibrous septa. Tumor cells are large with clear to eosinophilic cytoplasm characterized by vacuolated or bubbly cytoplasm (physaliphorous cells) arranged in cords or ribbons separated by a myxoid matrix.

Chondroid chordoma is a subtype of conventional chordoma containing extracellular matrix mimicking hyaline cartilage.

Dedifferentiated chordomas are biphasic, composed of conventional chordoma juxtaposed to high-grade sarcoma. Brachyury (nuclear) and cytokeratin expression are preserved in the conventional component but lost in the high-grade sarcomatous component.

Poorly differentiated chordomas are epithelioid and solid, with focal rhabdoid morphology and without physaliphorous cells, and they are characterized by a loss of SMARCB1 (INI1) expression but retained brachyury expression.

***From: Brat, D. J. (2021). Who classification of tumours. International Agency for Research on Cancer (IARC).***

| Characteristic | Conventional chordoma | Chondroid chordoma | Dedifferentiated chordoma | Poorly differentiated chordoma, SMARCB1-deficient |
|---|---|---|---|---|
| **Age at diagnosis** | Adults (96%)<br>Median: 55 years | Adults (86%)<br>Median: 45 years | Adults (96%)<br>Median: 61 years | Children (86%)<br>Median: 7 years |
| **M:F ratio** | 1.7 | 1.1 | 1.8 | 0.7 |
| **Prior irradiation** | No | No | Yes (25%) | No |
| **Localization** | Sacrococcygeal region (55%) | Skull base (73%) | Sacrococcygeal region (60%) | Skull base (64%) |
| **Histopathology** | Classic | Chondroid | • Conventional juxtaposed with sarcomatous (91%)<br>• Chondroid juxtaposed with sarcomatous (2%)<br>• Conventional chordoma transformed into pure sarcomatous tumor (7%) | Epithelioid<br>No physaliphorous cells |
| **Immunohistochemical profile** | SMARCB1 (INI1) preserved<br>Brachyury+<br>Pancytokeratin+<br>EMA+<br>S100+ | SMARCB1 (INI1) preserved<br>Brachyury+<br>Pancytokeratin+<br>EMA+<br>S100+ | SMARCB1 (INI1) preserved<br>Brachyury+/−[a]<br>Pancytokeratin−<br>EMA−<br>S100−/+ | Loss of SMARCB1 (INI1)<br>Brachyury+<br>Pancytokeratin+<br>EMA+<br>S100+/− |