

Sacral Chordoma with Thoracic Metastases: A Case Study

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Introduction

- Chordomas are rare locally aggressive neoplasms of bone originating from primitive notochord cells.¹
- These tumors most often occur in the sacral region of the spinal cord but can also occur in the cervical or skull base regions.
- Treatment of these tumors often consist of surgical en bloc resection, followed by radiation. Due to the nature of the pathology, complete maximal safe resection is recommended. Maximally safe resection can be difficult based on size, and involvement in surrounding structures.¹
- Local recurrence rate is 30-75%, and overall survival rate for five years is 70-86%.²
- The risk of metastasis is reported as 30%, typically occurring in the lungs, bone, soft tissue, and liver. Once metastasized, the survival rate can be less than 12%.³
- Despite these factors, there remains no current available treatment for metastatic chordoma patients.
- Pathologists' Assistants (PAs) play a vital role in evaluating the tumor, determining extent of tumor involvement with surrounding structures, and determining the margin status, which is highly indicative of risk of metastasis and recurrence.

Case Presentation

- 72-year-old male presented with worsening back pain following a fall with injury to his tailbone. He also reported urinary incontinence and changes in bowel movements.
- Imaging reported a large sacral mass coming off the S5 segment and coccyx, extending posteriorly.
- He then underwent a CT-guided biopsy that demonstrated the tumor to be consistent with chordoma.
- To achieve an en bloc resection, he underwent a low-sacral amputation with sacrifice of the S4-S5 nerve roots.
- Pathology was consistent with chordoma, grade 2, with negative surgical margins.
- Six months following surgery, he began experiencing shortness of breath and underwent imaging which demonstrated lesions within the lung which were biopsied and proven to be metastatic chordoma.
- The patient clinically declined and ultimately passed 11 months following resection.
- A full body autopsy was conducted for research under the Nervous System Tumor Bank (NSTB) at Northwestern University.

Pathologic Findings



Figure 1: Thoracic Cavity, image taken at time of autopsy, demonstrating multiple soft, pedunculated and flat, tan fleshy nodules extending from the chest wall of the rib cage, ranging in size from 1.0- 10.0 cm in greatest dimension



Figure 2: Left Lung (841g, expected: 325-480g), image taken at time of autopsy demonstrating multiple flat and pedunculated lesions ranging in size from 1.0- 7.0 cm in greatest dimension occasionally invading lung parenchyma.

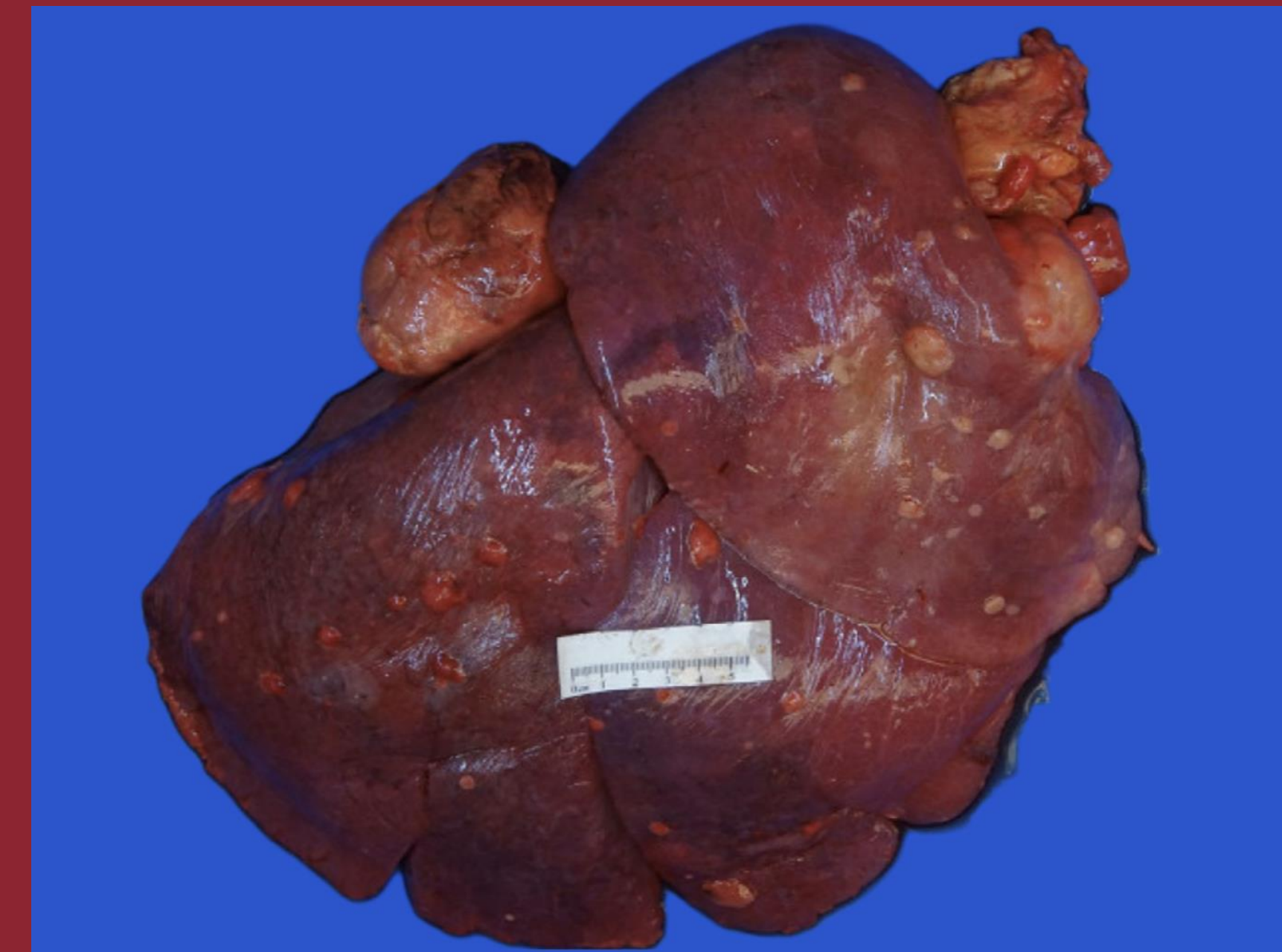


Figure 3: Right Lung (1348g, expected: 360-570g), image taken at time of autopsy demonstrating multiple flat and pedunculated lesions ranging in size from 1.0- 8.0 cm in greatest dimension occasionally invading lung parenchyma.



Figure 4: Diaphragm, image taken at time of autopsy demonstrating multiple soft, tan, fleshy nodules ranging in size from 1.0- 6.0 cm on the thoracic surface of the diaphragm. Larger masses demonstrated necrosis and hemorrhage in the centers.

Final Autopsy Diagnosis:

Sacral Chordoma, metastatic to thoracic cavity, bilaterally

Involving the right and left lung, chest wall and rib cage, and diaphragm

Microscopically, lesions are composed of nodules of atypical spindle cells with bubbly cytoplasm in a fibromyxoid background, consistent with the patient's history of metastatic chordoma

Discussion

- Chordomas are rare, slow growing malignant bone tumors and account for less than 5% of all primary malignant bone tumors in adults.¹
- Complete surgical excision remains the gold standard of treatment. Incomplete resection are associated with increased rates of recurrence and metastatic disease.
- Histologic diagnosis can be challenging; therefore, immunohistochemical staining is useful for distinguishing chordomas from other bone and soft tissue tumors. Brachyury positivity is sensitive and specific and can be used to help distinguish chordoma from other cartilaginous tumors.¹
- Patients with multifocal or unresectable metastatic disease have limited treatment options available, highlighting the need for continued investigation into novel treatment strategies.
- As a PA, these cases present unique grossing challenges due to the complexity of multiorgan involvement and extensive surgical resections. Thorough documentation, accurate measurements, and strategic tissue sampling of margins are essential for diagnosis and staging.⁴

Conclusion

- Metastatic chordoma is an uncommon manifestation of an already rare malignancy and may present significant diagnostic and management challenges.
- Recognition of characteristic gross, microscopic, and immunohistochemical features are essential for accurate diagnosis.
- This case highlights the aggressive nature of chordomas, and its ability to metastasize despite achieving negative margin status.
- Research autopsies can be useful for determining extent of tumor progression, and evaluation of treatment strategies.
- Biobanks are useful resources, which can provide data and material on rare diseases.

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