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En Bloc Resection of a Giant Sacrococcygeal Chordoma Presenting with Urinary Bladder Distension up to L4 Vertebrae: A Case Report

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Case Report

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Abstract

Chordoma is an uncommon primary bone malignancy that originates from residual notochordal tissue. We report the case of a 54-year-old male who presented with chronic lower lumbar pain radiating to the right lower limb, followed by progressive urinary retention. Physical examination revealed tenderness in the sacral region. MRI of the lumbosacral spine demonstrated a large, lobulated, expansile lytic mass measuring 6.3 × 8.0 × 7.1 cm (AP x TS x CC) involving the S3 to S5 vertebrae and coccyx. The mass resulted in marked urinary bladder distension, extending to the mid-abdominal region and up to the level of the L4 vertebra. Due to chordoma's poor response to chemotherapy and radiotherapy, en bloc resection remains the mainstay of treatment. The patient underwent a successful en bloc resection of the tumor, including the S3 to S5 vertebrae and coccyx, with preservation of the S1–S2 nerve roots to maintain bladder, bowel, and lower limb function. The surgery was challenging due to the tumor's proximity to the urinary bladder, rectum, and critical neurovascular structures. Postoperatively, the patient recovered without complications and with significant improvement in pain and mobility. Final histopathology confirmed an R0 resection. This case highlights the value of early advanced imaging in patients with atypical lower back symptoms and the importance of optimally timed and executed surgical intervention.

Introduction

Chordomas are rare primary malignant bone tumors arising from notochordal remnants, predominantly affecting the axial skeleton. The distribution includes 40%–50% in the sacrum, 35%–40% in the clivus or skull base, and 15%–20% in the mobile vertebral bodies. They comprise 1%–4% of primary malignant bone tumors, with an annual incidence of less than 0.1 per 100,000 individuals (1). They typically present between the ages of 40 and 70, with a 2:1 male-to-female predominance (2, 3).

Sacrococcygeal chordomas often cause insidious lower back pain exacerbated by sitting, which is frequently unresponsive to non-steroidal anti-inflammatory drugs. As the tumor enlarges, sacral nerve root compression may lead to lower limb weakness, urinary and bowel incontinence, or impotence in males. Due to this indolent growth pattern, the disease is often diagnosed at a locally advanced stage. Diagnosis relies on histopathology, revealing characteristic physaliphorous cells on a background of myxoid stroma, supported by magnetic resonance imaging (MRI) and computed tomography (CT). As slow-growing tumors with limited response to conventional chemotherapy and radiotherapy, chordomas are primarily managed with surgical resection, with en bloc excision offering the best chance for cure (4). We present a case of a surgically treated sacrococcygeal chordoma notable for causing massive urinary bladder distension, highlighting the complexities of managing this rare malignancy.

Case Report

A 54-year-old male presented to the emergency department with a 5-month history of progressively worsening urinary retention and a 4-month history of lower back pain. He reported straining to urinate, a

weak stream, frequent nocturia (3–4 times/night), and a persistent sensation of incomplete bladder emptying. The lower back pain was gradual in onset, progressive, and aching in nature, radiating to the right posterior leg and associated with numbness. The pain was exacerbated by sitting and unrelieved by over-the-counter analgesics. There was no history of trauma, fever, or weight loss.

On examination, the patient was alert with a GCS of 15/15. Neurological assessment revealed full motor strength (5/5) in both lower limbs with intact sensation. Deep tendon reflexes were normal. Physical examination of the lumbosacral region showed tenderness over the sacrococcygeal area. On deep palpation, a firm, non-mobile mass was appreciated over the midline sacrum, without overlying skin changes.

Routine laboratory investigations, including complete blood count and renal function tests, were within normal range. An ultrasound of the kidneys, ureters, and bladder (KUB) revealed a post-void residual volume of 90 ml, indicating incomplete bladder emptying. The prostate was mildly enlarged at 38 grams, consistent with benign prostatic hyperplasia, which was considered a contributing but not primary factor.

Multiplanar, Multisequential MRI of the sacrococcygeal region with and without contrast was performed as shown in Figs. 1, 2, and 3.

The MRI revealed a large, lobulated, heterogeneous mass measuring 6.3 × 8.0 × 7.1 cm (AP × TS × CC), involving the S3-S5 vertebrae and the coccyx. The mass was isointense on T1-weighted images, heterogeneously hyperintense on T2/STIR sequences, and demonstrated avid heterogeneous post-contrast enhancement. It extended anteriorly into the presacral space, displacing the rectum, and posteriorly abutted the gluteal muscles. There was infiltration of the right neural foramina at S3-S4 with possible epidural extension and spinal canal stenosis. Critically, the mass caused significant anterior displacement and compression of the bladder outlet, resulting in massive urinary bladder distension that reached the level of the L4 vertebra. The imaging findings were highly suggestive of a sacrococcygeal chordoma.

An image-guided core needle biopsy was performed. Histopathology revealed fragments of a neoplastic lesion composed of atypical cells forming cords and sheets within a myxoid stroma. The tumor cells exhibited characteristic physaliphorous features with clear-to-eosinophilic, vacuolated cytoplasm. Immunohistochemical staining showed that the tumor cells were positive for pan-cytokeratin, S100, and brachyury, confirming the diagnosis of chordoma.

Following a multidisciplinary tumor board discussion, the patient was scheduled for surgical excision. Intraoperatively, the patient was placed in a prone position. A midline incision was made over the sacrum, and the paraspinal muscles were retracted to expose the posterior sacral elements. A gelatinous, lobulated mass involving the S3 to S5 vertebrae and coccyx was identified. An en bloc resection of the tumor was performed, which included the complete excision of the S3 to S5 vertebrae and coccyx. The S1 and S2 nerve roots were meticulously preserved. The surgical cavity was irrigated, hemostasis was achieved, and a closed suction drain was placed before layered closure.

Final histopathological analysis of the resected specimen confirmed the diagnosis of chordoma with tumor-free margins (R0 resection).

Ethics and consent

This study was conducted in accordance with the 1964 Declaration of Helsinki. Formal ethics committee approval was not required for this single case report per institutional policy. Informed written consent for publication of this case report and accompanying images was obtained from the patient

Discussion

This case report illustrates the successful en bloc resection of a giant sacrococcygeal chordoma that presented with the unusual finding of massive urinary bladder distension up to the L4 vertebral level. The patient's uneventful recovery with no new neurological deficits and complete resolution of preoperative symptoms underscores the value of meticulous surgical planning and advanced preoperative imaging in preserving critical neurovascular structures while achieving an R0 resection.

Sacral chordomas pose significant therapeutic challenges due to their high local recurrence rates (43%–85%) and risk of distant metastasis (5%–40%) within 10 years, with inadequate margins being the strongest predictor of recurrence (5). The en bloc approach in this case, which achieved tumor-free margins, minimizes the risk of intraoperative tumor seeding—a complication associated with intratumoral resections that carry significantly higher recurrence rates (60%–64%) compared to marginal resections (25%–28%) (1).

The patient's presentation with advanced urinary retention, rather than the more commonly reported incontinence, highlights the tumor's strategic location for extrinsic compression of the bladder neck and outlet. This case reinforces the need for a high index of suspicion for malignancy in patients with non-specific lower back pain and unexplained urinary symptoms, particularly when pain is mechanical and unresponsive to conventional therapy. Advanced imaging, especially MRI, is indispensable for characterizing the tumor's extent, planning the surgical approach, and anticipating involvement of adjacent viscera and neurovascular structures (4).

The histopathological diagnosis was confirmed by the presence of physaliphorous cells and a supportive immunohistochemical profile, including positivity for brachyury, which is a highly sensitive and specific marker for chordoma (6). Achieving an R0 resection was the primary goal, as adjuvant therapies, including conventional radiotherapy and chemotherapy, play a limited role in the curative management of chordoma (7). While proton beam therapy and targeted molecular agents (e.g., imatinib) offer options for unresectable or recurrent disease, surgical excision remains the cornerstone of treatment for resectable tumors (7).

Despite the successful short-term outcome, this report has limitations. The follow-up period is insufficient to evaluate long-term oncological control, given chordoma's indolent nature and propensity for late recurrence. Furthermore, the feasibility of en bloc resection in this case may not be generalizable to all patients, particularly those with tumors encasing bilateral nerve roots, invading pelvic viscera, or with significant medical comorbidities. This case did not involve molecular profiling, which is increasingly relevant for guiding targeted therapies in the event of future recurrence.

In conclusion, this case demonstrates that en bloc resection of a giant sacrococcygeal chordoma is feasible and can provide excellent short-term functional outcomes and symptom relief. It highlights the critical importance of a multidisciplinary approach, meticulous preoperative planning with advanced imaging, and the pursuit of negative surgical margins.

Conclusion

En bloc resection successfully treated a rare case of giant sacrococcygeal chordoma presenting with massive bladder distension, achieving an R0 resection and preserving neurological function. At 12-month follow-up, the patient demonstrated an excellent clinical outcome with complete resolution of preoperative urinary retention and back pain. Neurological examination was unremarkable, with no motor, sensory, or sphincter deficits. Postoperative imaging showed no evidence of residual or recurrent tumor. This case highlights the importance of maintaining a high index of suspicion for chordoma in patients with persistent lower back pain associated with urinary symptoms.

Declarations

Conflicts of Interest Disclosure

All authors have no conflict of interest.

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Figures

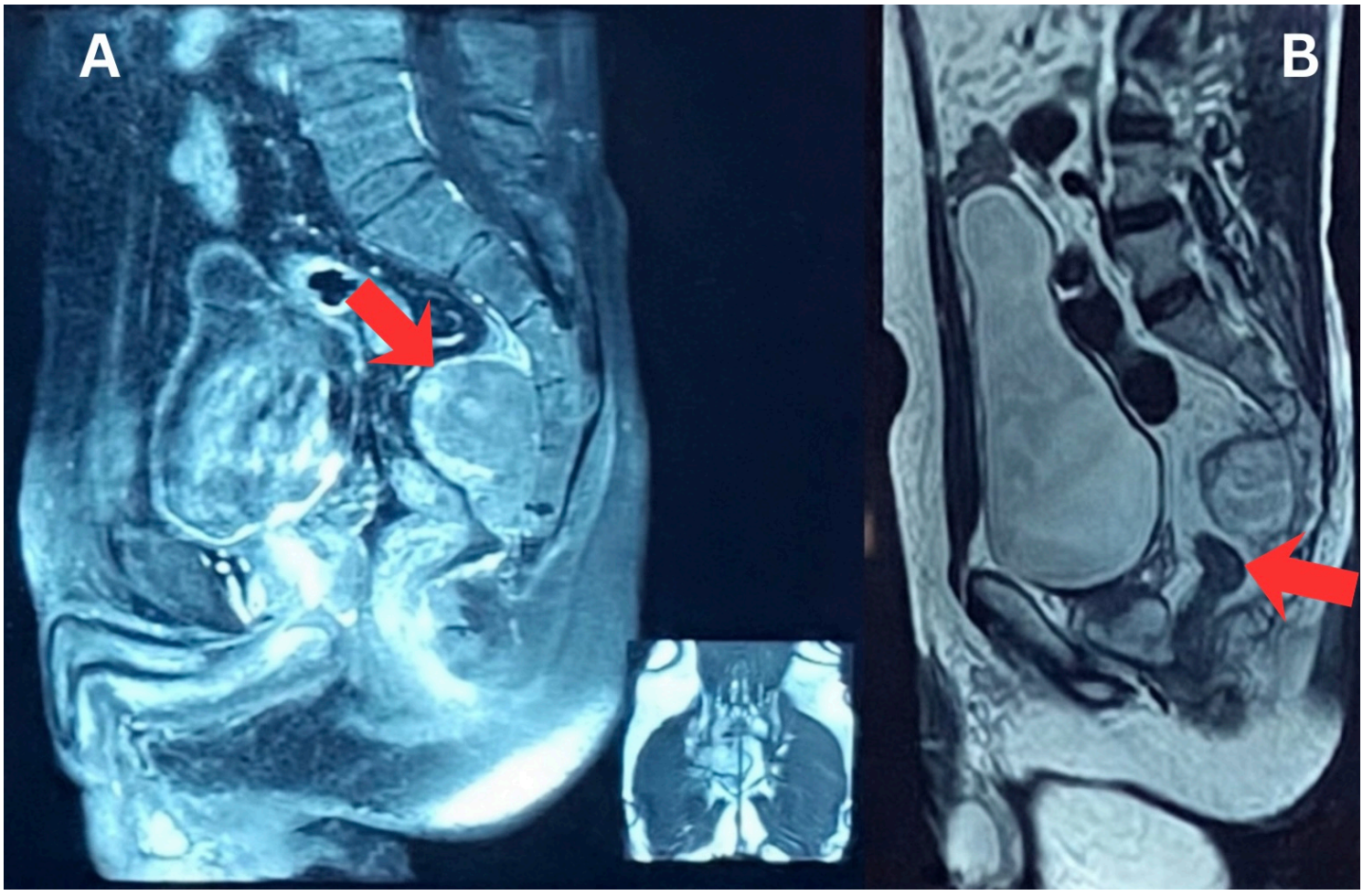


Figure 1

Preoperative Sagittal MRI of the Sacrococcygeal Chordoma.

· **(A) Sagittal T2-weighted MRI:** Demonstrates a large, well-circumscribed, heterogeneously hyperintense mass in the sacrococcygeal region (red arrow). The lesion shows extensive osteolytic destruction of the S3-S5 vertebrae and coccyx. (Figure 1a)

· **(B) Sagittal T1-weighted MRI:** Clearly illustrates the significant anterior displacement of the rectum (red arrow) and the massive secondary distension of the urinary bladder. The bladder is seen extending superiorly toward the level of the L4 vertebral body due to extrinsic outlet compression by the tumor. (Figure 1b)

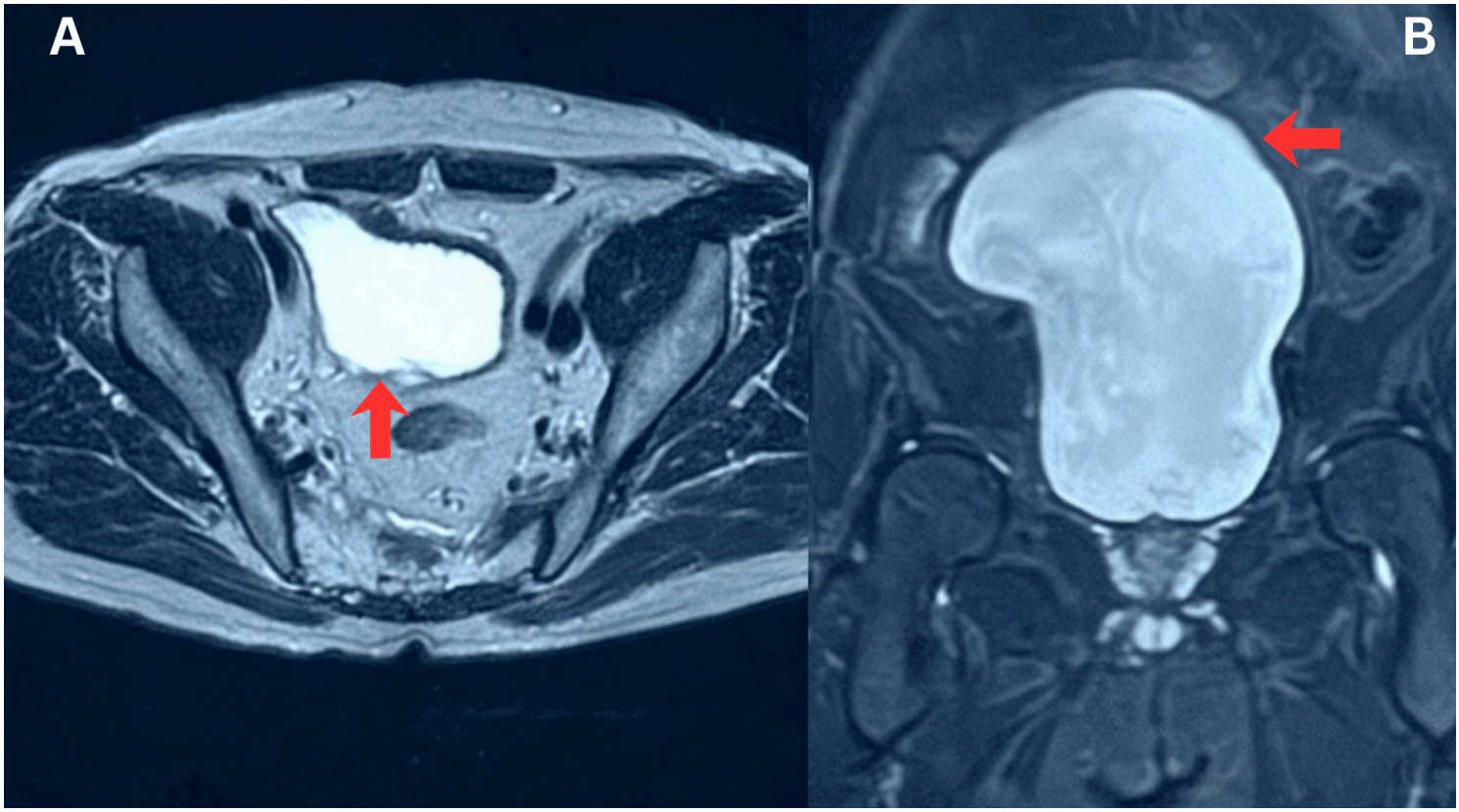


Figure 2

Cross-sectional Imaging of the Sacrococcygeal Chordoma.

· **(A) Axial T2-weighted MRI:** Demonstrates a heterogeneous, lobulated mass occupying the sacral and presacral space (red arrow). The mass involves the sacral vertebrae and shows extension into the right neural foramen. (Figure2a)

· **(B) Coronal T2-weighted MRI:** Clearly illustrates the massive superior distension of the urinary bladder (red arrow), extending into the mid-abdominal cavity. The tumor's relationship to the pelvic floor and lateral sidewalls is well-demarcated. (Figure 2b)

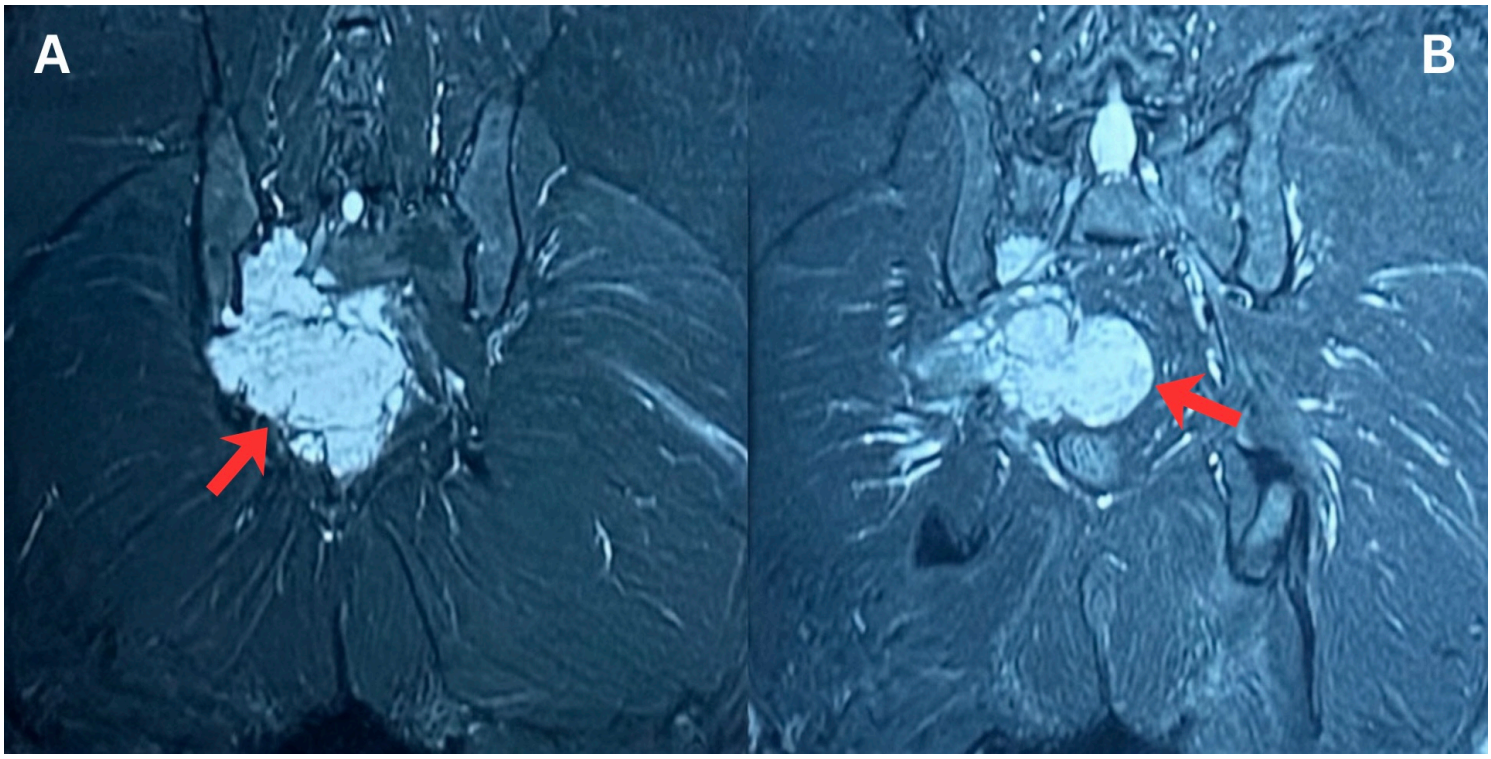


Figure 3

Coronal MRI of the Sacrococcygeal Mass.

(A & B) Coronal STIR/T2-weighted MRI: Sequential slices showing a large, hyperintense, lobulated mass (red arrows) involving the lower sacral segments and extending toward the pelvic sidewalls. The tumor's relationship to the piriformis muscle and pelvic floor is clearly demarcated. (Figure 3)