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CASE REPORT

When Chordoma Isn't Chordoma: Lessons from a Misleading Diagnosis

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Abstract:

Primary bone malignancies are exceptionally rare, and sacral tumors often pose significant diagnostic challenges because of their insidious presentation and overlapping radiological features. Chordoma is the most common malignant tumor of the sacrum, while solitary plasmacytoma of bone (SPB) is much rarer and even less frequently considered in Asian populations. We report the case of a 50-year-old man who presented with a one-year history of progressive low back pain, without neurological deficit or systemic features. Initial imaging demonstrated a destructive lytic lesion of the sacrum with an associated soft tissue mass, radiologically suggestive of chordoma. Fine-needle aspiration cytology was inconclusive and yielded conflicting impressions, including Ewing's sarcoma and chordoma. Definitive biopsy, however, revealed sheets of plasmacytoid cells with Russell bodies and amyloid deposits, establishing the diagnosis of plasmacytoma. This case highlights the diagnostic dilemma posed by solitary sacral lesions and emphasizes the importance of histopathological confirmation, as management of plasmacytoma differs substantially from that of chordoma. Recognition of this rare entity is crucial to avoid unnecessary surgical morbidity, since radiotherapy remains the cornerstone of treatment for sacral plasmacytoma.

Keyword: Chordoma, Plasmacytoma, Primary bone malignancies, Solitary sacral lesions

Introduction

Primary bone malignancies are rare, accounting for only about 0.2% of all cancers.[1] Among these, chordoma represents a small subset, comprising approximately 2–4% of primary bone tumors,[2] with an annual incidence of 0.8 cases per million population.[3] Nearly half of all chordomas arise in the sacral or coccygeal region.[4] An even rarer entity of axial skeleton malignancies is solitary plasmacytoma of bone (SPB), with an estimated incidence of 3.5 per million population.[5] While data on Western populations are available, reports from Asian cohorts are limited; one Taiwanese study noted that SPB accounted for 6.2% of all plasma cell neoplasms.[6] Both chordoma and SPB are indolent but destructive tumors of the sacrum. They usually progress insidiously, producing vague symptoms that can delay recognition until significant local destruction or neurological compromise has occurred.[7,8] This overlap in clinical and radiological features makes distinction between the two particularly challenging. Here, we report a rare case of a sacral lesion that was initially suspected to be chordoma but was ultimately confirmed to be plasmacytoma, highlighting the importance of careful diagnostic evaluation in such complex presentations.

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Case history

A 50-years male presented to the Department of Orthopaedics at Bangabandhu Sheikh Mujib Medical University (BSMMU), Dhaka, in February 2023, with a history of persistent low back pain. The discomfort had been dull, aching, and localized to the sacral region, gradually increasing in intensity over the preceding year. He reported increasing difficulty in carrying out his daily activities, such as prolonged standing, bending, or lifting, and noted occasional disturbance of sleep due to nocturnal discomfort.

Clinical examination revealed localized pain elicited during the straight leg raising test; without radiation. However, motor and sensory functions of the lower limbs remained intact, reflexes remained intact and there was no disturbance of bowel or bladder control. There were no clinical signs of radiculopathy, and systemic evaluation did not reveal features of anemia, hypercalcemia, or renal dysfunction.

The insidious onset of pain without neurological deficit initiated the first diagnostic confusion.The patient's hematological profile revealed mild anemia (Hb 8.6 g/dL) and a raised ESR (40 mm in the first hour), while serum calcium, urea, creatinine, and eGFR levels remained within normal limits. The albumin-to-globulin ratio was preserved, and protein electrophoresis demonstrated a polyclonal gammopathy. Serum immunoglobulin levels (IgA, IgM, IgG) were normal, and β2-microglobulin was 2 mg/dL. To exclude multiple myeloma, a skeletal survey was performed, which did not reveal any additional punched-out lytic lesions. Bone marrow examination was normal, and no paraproteinemia or end-organ damage was evident.

Plain radiography (X-ray, Fig. 1) showed a destructive lytic lesion involving the sacrum. The lesion appeared as an area of bone rarefaction with poorly defined margins and cortical thinning, suggestive of an aggressive process. There was evidence of trabecular bone destruction with absence of sclerosis, consistent with a malignant pathology. The lesion expanded the sacral contour but did not show calcification or periosteal reaction, features that may help differentiate from other primary bone tumors.

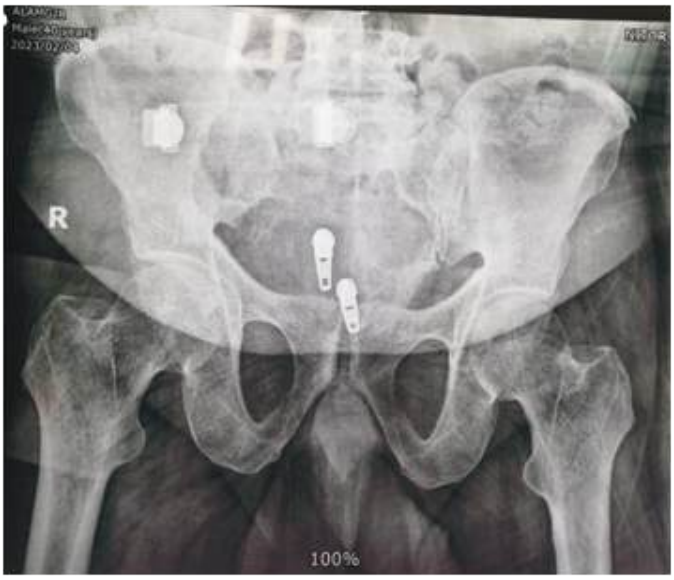

Figure-1: Lytic lesion of sacrum on plain x-ray

Magnetic resonance imaging (MRI, Fig. 2) revealed a large, lobulated soft tissue mass arising from the sacrum, measuring approximately 11 × 9 × 7.6 cm. The lesion caused destruction of all sacral vertebrae and extended into the presacral space. On T1-weighted imaging, the lesion was predominantly hypointense, while on T2-weighted imaging, it demonstrated a hyperintense signal. Post-contrast sequences showed strong, heterogeneous enhancement, reflecting its vascular nature and cellular density. The tumor encroached upon adjacent pelvic soft tissues, although without definitive invasion of bowel or bladder structures. These imaging features were strongly suggestive of a malignant sacral tumor, with chordoma initially favored in the differential diagnosis.

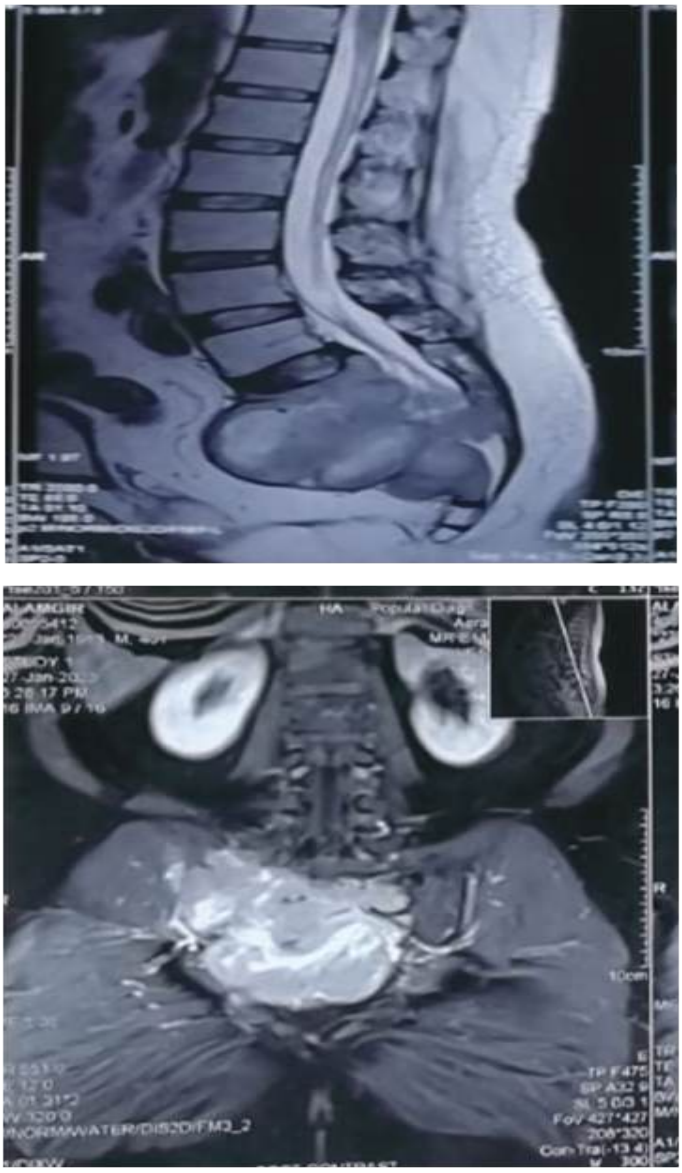

Figure-2: MRI showing large lobulated soft tissue lesion with bony destruction of all sacral vertebrae

CT-guided fine-needle aspiration cytology (FNAC) report was inconclusive. One cytology report suggested malignant cells consistent with Ewing's sarcoma, while another indicated features compatible with chordoma. However, subsequent biopsy from surgical tissue samples provided a definitive diagnosis. Histopathology (Fig. 3) revealed sheets of plasmacytoid cells with eccentrically placed nuclei and abundant cytoplasm. Foci of Russell bodies and amorphous amyloid deposition were also noted.

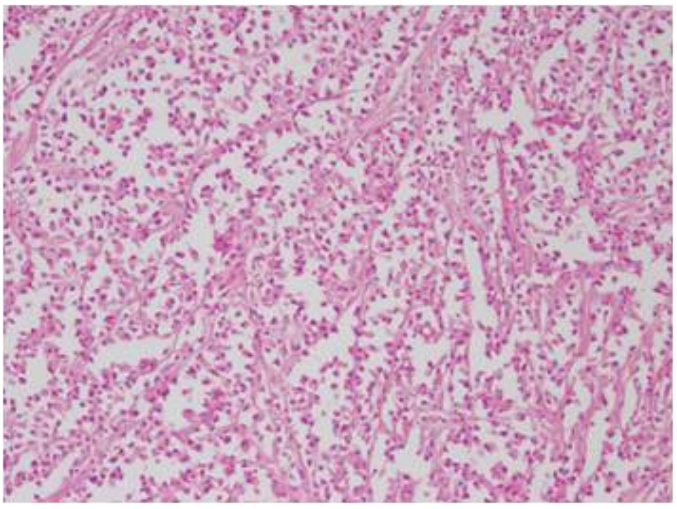

Figure-3: Plasmacytoid cells arranged in sheets, having eccentrically placed prominent nuclei with abundant cytoplasm with foci of Russell body

Discussion

Solitary plasmacytoma of bone (SPB), also referred to as osseous plasmacytoma, represents one of the two recognized forms of solitary plasmacytoma, the other being extramedullary plasmacytoma. SPB arises most commonly in the axial skeleton, particularly the vertebral bodies and skull.[6,9] It demonstrates a male predominance, with a male-to-female ratio of approximately 2:1, and the median age at diagnosis is around 55 years.[10] Local recurrence is relatively uncommon, occurring in fewer than 5% of cases, whereas dissemination to multiple myeloma has been reported in 35–70% of patients.[11]

The clinical manifestations of SPB are largely determined by the site of involvement. Pain at the lesion site is the most frequent presenting symptom, reflecting progressive bone destruction by infiltrating plasma cells. Patients may also present with a palpable painful mass, pathological fractures, or features of nerve root or spinal cord compression, depending on the anatomical location. Long bone involvement frequently predisposes patients to fractures, whereas vertebral or sacral lesions may cause neurological deficits due to compression.[11] The case we reported only presented with pain without a palpable mass, pathological fracture or features of nerve involvement.

Radiographically, SPB typically appears as a solitary expansile lytic lesion with cortical thinning and destruction, often exhibiting a "bubbly" or trabeculated pattern.[11,12] On MRI, plasmacytomas generally demonstrate homogeneous low signal intensity on T1-weighted sequences and high signal intensity on T2-weighted sequences.[13] Contrast-enhanced MRI is particularly valuable, as it shows marked homogeneous enhancement of the mass and provides superior delineation of soft tissue involvement compared to CT. Lesions in the pelvis or sacrum often display these imaging characteristics, and MRI can be especially useful in identifying subtle extensions into adjacent tissues.[14]

The diagnosis of SPB requires stringent criteria: (a) histopathological confirmation of a plasma cell tumor, (b) bone marrow plasma cell infiltration less than 10% of nucleated cells, and (c) normal skeletal survey findings apart from the primary lesion.[15,16] Additional features supporting the diagnosis include the absence of systemic events such as anemia, hypercalcemia, renal impairment, or widespread osteolytic lesions.[11] The presence of Bence-Jones proteinuria or a monoclonal band on serum protein electrophoresis does not necessarily exclude SPB, provided other criteria are met.[17,18] Our reported case met the cardinal features of SPB diagnosis; outcasting the shadow of doubt at the end.

Taken together, the rarity of SPB, especially in Asian populations, combined with its nonspecific clinical and radiological presentation, makes it a challenging diagnosis. In the present case, the sacral lesion mimicked chordoma radiologically and was initially suspected to be a different malignancy based on FNAC, before definitive histopathology confirmed plasmacytoma. This highlights the importance of a comprehensive diagnostic approach, including histopathological confirmation, biochemical workup, and imaging correlation, in differentiating SPB from other destructive sacral tumors.

Conclusion

Nonspecific clinical and radiological features of SPB often leads to diagnostic confusion with other primary sacral tumors. Failure to consider SPB as a potential differential diagnosis may expose patients to unnecessary or high-risk interventions, as these tumors

are highly vascular and although incisional biopsy was essential, it can cause profuse bleeding. Moreover, unlike chordoma, the standard treatment for SPB is local radiotherapy rather than extensive surgical resection. Diagnostic dilemma unnecessarily burdens the patient with an invasive intervention.

Author contributions

All authors participated equally in preparing, drafting and cross-checking of the study materials

Consent

Informed written consent was taken from the patient's family member to publish this case report and any accompanying images in accordance with the journal's patient consent policy.

Conflicts of Interest

The authors declare no conflicts of interest

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Data availability statement

All medical reports and corresponding images are available from the corresponding author upon reasonable request.

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