

Case Report

A case report on a rare primary bone tumour: sacral chordoma presenting as sacral swelling with sphincter dysfunction

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Received: 01 November 2024

Accepted: 05 December 2024

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ABSTRACT

Chordoma is a rare, locally aggressive malignant tumour arising from remnants of the notochord, primarily affecting the axial skeleton, seen twice often in males above 40 years. This case report presents a 50-year-old male with primary sacral chordoma, emphasizing the need for early evaluation, treatment modalities and focusing on the importance of sacral nerve preservation during surgery for better functional outcome. Purpose of the study was to demonstrate the significance of early evaluation and diagnosis, emphasizing the importance of at least unilateral sacral nerve root preservation for optimizing postoperative bowel and bladder function. A 50-year-old male presented with sacral swelling, saddle anaesthesia, lower backache, urinary incontinence, and constipation. Examination revealed a firm, immobile sacral mass with reduced anal sphincter tone. Magnetic resonance imaging (MRI) suggested the presence of a sacral chordoma, leading to the planning of surgical resection. Intraoperatively, frozen section analysis confirmed the diagnosis of chordoma. Intraoperatively, the lesion was found to extend predominantly toward the left side, necessitating careful dissection and preservation of the sacral nerve roots on the right side. Post-operatively, the patient experienced improved bowel and bladder function, along with significant pain relief. The patient showed substantial postoperative improvement, with resolution of urinary and bowel dysfunction. Follow-up imaging revealed no tumour recurrence. Preservation of sacral nerve roots was crucial to the patient's recovery, improving overall quality of life. This case highlights the need to consider chordoma in the differential diagnosis of sacral masses and emphasizes the importance of sacral nerve root preservation during resection to optimize functional outcomes in patients with this rare tumour.

Keywords: Sacral chordoma, Nerve root preservation, Surgical management, Functional outcomes

INTRODUCTION

Chordomas are rare, slow-growing, malignant tumours derived from remnants of the notochord. These tumours account for approximately 1-4% of primary bone malignancies, with over 50% occurring in the sacrococcygeal region.^{1,2} Other common locations include the skull base and cervical spine. Chordomas are thought to arise from embryonic notochordal remnants and are considered congenital in origin.³ Sacral chordomas can reach a large size before compressing nearby structures,

leading to neurological symptoms. As the tumour enlarges, it often impinges on the sacral nerve roots, leading to sphincter dysfunction. The S2-S4 nerve roots are primarily responsible for bowel, bladder, and sexual function. Compression or injury to these nerve roots can result in urinary incontinence, constipation, and decreased anal sphincter tone. Preservation of these nerve roots during surgical resection is essential to maintain sphincter control and prevent permanent neurological deficits.⁴ This case report presents a primary sacral chordoma in a 50-year-old male, emphasizing the importance of recognizing such tumours early and highlighting the role of preserving

sacral nerve roots during surgery to optimize postoperative functional outcomes.

CASE REPORT

A 50-year-old male presented with complaints of urinary incontinence for 7 months, characterized by involuntary leakage, particularly during movement or coughing. The patient also reported dull aching pain in the lower back, localized to the sacral region, that had persisted for 6 months. Additionally, he had progressive constipation for past 3 months with reduced bowel movement frequency and had taken multiple conservative treatment with no much improvement. The patient denied any leg weakness or numbness but reported decreased sensation in the perineal region and inner thigh.

On examination, a 5×6 cm swelling was noted in the sacrococcygeal region (Figure 1a). The swelling was firm, non-tender, immobile, and fixed to the underlying bone. The surface was irregular with well-defined edges, and the overlying skin was pinchable. Neurological examination revealed positive saddle anaesthesia, no motor deficits, and his gait was normal. Per rectum examination revealed reduced anal sphincter tone, with a firm, irregular, fixed mass palpable in the posterior rectal wall, 4 cm from the anal verge.

Magnetic resonance imaging of the pelvis and sacrum revealed a large, well-defined lobulated mass involving the sacrococcygeal region (Figure 1b). The mass extended into the sacral canal, compressing the sacral nerves, particularly at the S2-S4 levels. No metastasis was noted.

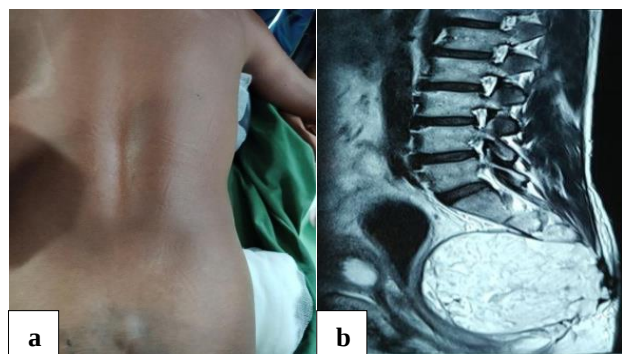


Figure 1: (a) Showing a swelling in the sacrococcygeal region, and (b) MRI showing the lesion in the sacral region.

The patient underwent En bloc resection of the tumour, with intraoperative frozen section confirming the diagnosis of chordoma Figure 2. The tumour extended more significantly toward the left side; hence care was taken to preserve the sacral nerve roots on the right. Postoperatively, the patient demonstrated considerable improvement in bowel and bladder function, along with substantial pain relief and resolution of saddle anaesthesia. At 3-month follow-up, there was no evidence of tumour

recurrence on magnetic resonance imaging (MRI) and sphincter control continued to improve progressively.



Figure 2: Intraoperative picture of sacral chordoma.

DISCUSSION

Chordomas, though rare, present significant diagnostic and therapeutic challenges due to their slow-growing nature and nonspecific symptoms. Early detection is crucial, as smaller tumours are more suitable for complete resection, improving prognosis and reducing recurrence risk.^{1,2} Delayed diagnosis allows for tumour growth, complicating surgery and increasing the risk of neurological deficits, such as bowel and bladder dysfunction.^{3,4}

Achieving negative surgical margins remains the cornerstone of chordoma treatment, as these tumours are resistant to chemotherapy and only minimally responsive to radiation.⁵ The balance between tumour removal and nerve preservation is critical. Sacrificing the sacral nerve roots often results in permanent bowel, bladder, and sexual dysfunction.^{6,7} In this case, unilateral nerve preservation contributed to the patient's improved functional outcomes. Studies show that even partial nerve preservation enhances postoperative quality of life.^{8,9}

Advances in imaging, intraoperative navigation, and nerve monitoring have improved outcomes in chordoma surgery, allowing for complete resection while minimizing neurological damage.^{10,11}

The prognosis for sacral chordoma depends largely on the ability to achieve complete resection with negative margins.^{12,13} Even with successful surgery, recurrence rates can be as high as 30-50%, and recurrent tumours are often more difficult to treat.^{14,15} Long-term follow-up is essential, with regular imaging to monitor for recurrence. While surgical resection offers the best chance for long-term survival, the morbidity associated with sacral chordoma surgery can be significant, especially in cases

requiring extensive resection. In some instances, adjuvant therapies, such as proton beam therapy or carbon ion radiotherapy, have been utilized to target residual tumour cells and reduce the risk of recurrence. However, these treatments are typically reserved for cases where complete resection is not possible or for recurrent disease. In this case, the patient benefited from an early intervention that allowed for partial nerve preservation and an overall good functional recovery. While long-term follow-up is still needed to monitor for recurrence, the immediate postoperative outcomes were favourable, with significant improvement in pain and sphincter function

CONCLUSION

This case of primary sacral chordoma, presenting with sacral swelling and sphincter dysfunction, highlights the importance of early detection in improving surgical outcomes. Early recognition and accurate diagnosis are crucial for achieving complete surgical resection with negative margins, which significantly reduces the risk of recurrence. Additionally, preserving sacral nerve roots during surgery plays a vital role in maintaining postoperative bowel and bladder function, as demonstrated in this case. The preservation of unilateral sacral nerve roots contributed to improved functional outcomes, emphasizing the need for careful surgical planning in sacral chordoma cases. Long-term follow-up is essential due to the high risk of recurrence in these patients.

Funding: No funding sources

Conflict of interest: None declared

Ethical approval: Not required

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Cite this article as: Latheef NA, James E, Paulose L, Karilly P, Karuthillam P. A case report on a rare primary bone tumour: sacral chordoma presenting as sacral swelling with sphincter dysfunction. *Int Surg J* 2025;12:110-2.