Scholars Journal of Medical Case Reports
Abbreviated Key Title: Sch J Med Case Rep
ISSN 2347-9507 (Print) | ISSN 2347-6559 (Online)
Journal homepage: https://saspublishers.com

OPEN ACCESS

Radiology

Sacrococcygeal Chordoma: A Clinical and Imaging Review

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DOI: https://doi.org/10.36347/sjmcr.2024.v12i09.014 | **Received:** 27.07.2024 | **Accepted:** 02.09.2024 | **Published:** 05.09.2024

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Abstract **Case Report**

Sacrococcygeal chordoma is a rare, locally aggressive malignant tumor originating from remnants of the notochord, and it represents the most common primary malignancy of the sacrum. This review provides an in-depth discussion of the clinical presentation, pathological features, imaging characteristics, and management of sacrococcygeal chordomas. These tumors typically present with local pain, neurological deficits, and a palpable mass, and they are known for their high recurrence rate and challenging surgical management. Imaging, especially MRI, is crucial for accurate diagnosis and surgical planning, given its ability to delineate the extent of the tumor and its relationship with surrounding structures. Complete surgical excision remains the cornerstone of treatment, with prognosis largely depending on the ability to achieve negative surgical margins. We present a case of a 45-year-old male who presented with bowel and bladder dysfunction and a painful, progressively enlarging mass over the sacrum. MRI revealed a large, well-defined soft tissue mass involving the distal sacrum and coccyx, extending into the retrorectal space. The patient underwent successful en bloc surgical resection, and histopathological examination confirmed the diagnosis of chondroid chordoma.

Keywords: Sacrococcygeal chordoma, notochord remnants, MRI, surgical resection, chondroid chordoma, sacral tumor, clinical presentation, imaging characteristics, malignant tumor, recurrence.

INTRODUCTION/BACKGROUND

Chordoma is a rare malignant neoplasm that arises from notochord remnants. Chordoma is the second most common primary malignancy in the spine (behind myeloma) and is the most common primary malignancy of the sacrum [1]. Sacrococcygeal is the most common location, accounting for approximately 30-50% 2,3 of all chordomas and commonly involving the fourth and fifth sacral segments 2 and the clivus or the spheno occipital region in 30–35% cases [2].

Local invasiveness and destructiveness are characteristic features of the disease. Complete surgical excision is the main therapeutic modality able to effect a cure. The importance of radiation has gradually increased overtime. Imaging techniques in particular MRI, play a crucial role in surgical planning [4].

CASE REPORT

A 45-year-old male presenting with bowel and bladder dysfunction, along with a progressively enlarging and painful swelling in the lower back, specifically over the sacrum, which had been present for over a year. On physical examination, a firm, immobile swelling measuring 6 cm × 8 cm was identified. Further examination revealed an indentation on the posterior rectal wall.

MRI findings showed a large, well-defined soft tissue mass involving the distal sacrum and coccyx, with extension into the retrorectal space and dorsal soft tissues. The mass is hypointense T1, and hyperintense on T2 sequences (Figure 1A, 1B & 1C) with linear hypointense septa, and post-contrast T1-weighted images (Figure 1D) revealed heterogeneous enhancement, which was suggestive of a sacrococcygeal chordoma.

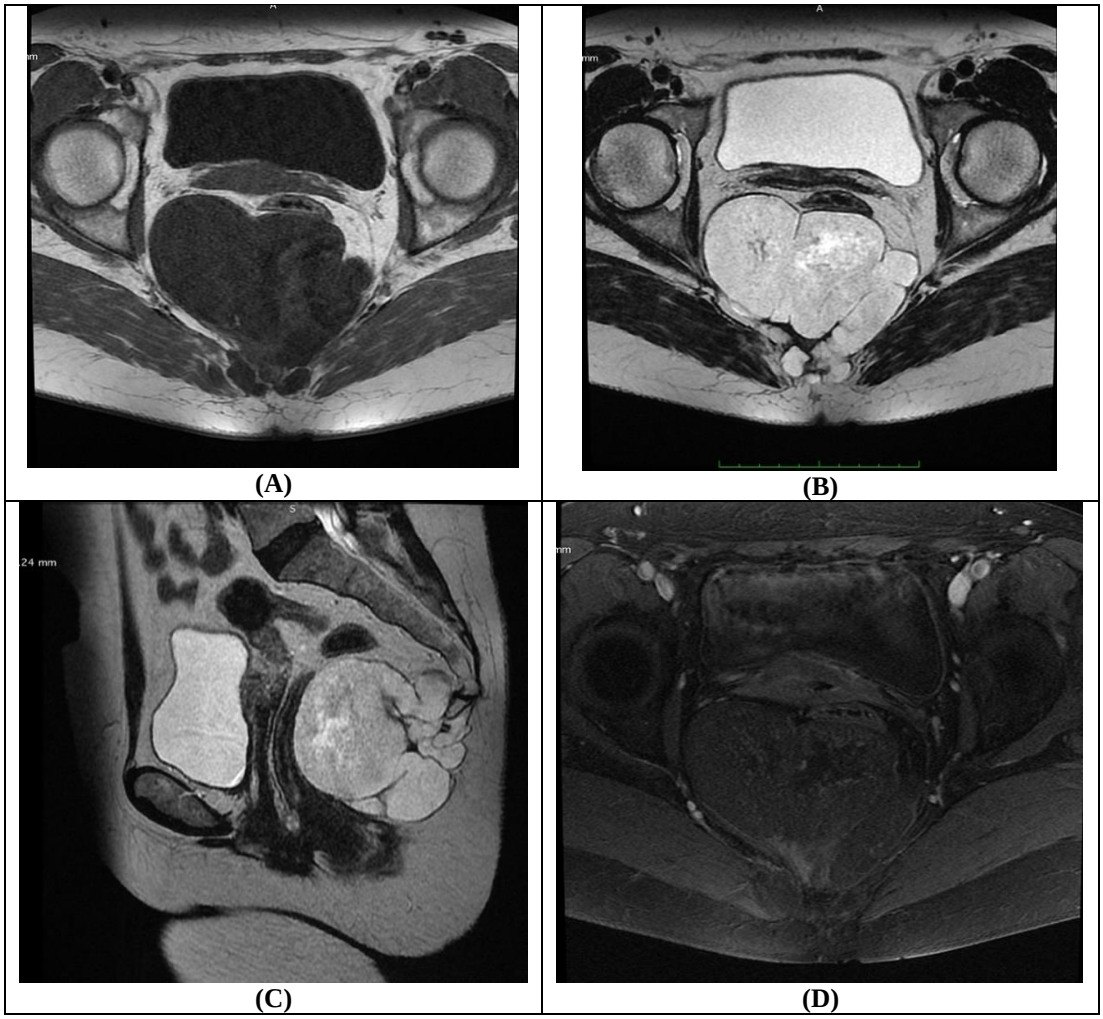

Figure 1: Magnetic resonance imaging (MRI) of the pelvis was performed without and with contrast; (A) axial T1, axial (B) and sagittal T2 (C) and post-contrast T1-weighted images (D) demonstrate a well-defined soft tissue mass involving the distal sacrum and coccyx, with extension into the retrorectal space and dorsal soft tissues. The mass is hypointense T1 (A), and hyperintense on T2 sequences (B, C) with linear hypointense septa, revealed heterogeneous enhancement

En bloc surgical resection with negative margins was performed. Pathology revealed an irregular grey-brown to grey-black specimen with soft tissue and bony bits, with features suggestive of a chondroid chordoma.

DISCUSSION

Tumours that occur in the retrorectal space comprise an uncommon and mixed group. They are estimated at one in every 40,000 hospital admissions. Retrorectal tumours maybe classified as congenital, neurogenic, osseous or miscellaneous [5].

Chordoma is a rare malignant neoplasm that arises from notochord remnants. Chordoma is the second most common primary malignancy in the spine (behind myeloma) and is the most common primary malignancy of the sacrum [4]. Greater than 50% of chordomas arise in the sacrococcygeal area, and more than 30% arise at the base of the skull; the remainder are dispersed throughout the rest of the spine. Peak incidence for sacrococcygeal chordomas occurs in the fifth to seventh decades, whereas the peak for sphenooccipital lesions is the fourth to sixth decades. Most series show a marked male predominance (2:1), especially for sacrococcygeal tumors [3, 16].

Clinical Presentation

Clinical signs and symptoms may vary depending on the location, the size of the tumour and the extent of neural invasion. The most common presenting complaint for patients with sacrococcygeal tumors is low back pain. Bowel and bladder disturbance and sciatic pain are also common with sacral tumors. A palpable mass frequently is present on rectal examination [7].

Pathology

Macroscopically, sacrococcygeal chordomas are usually well demarcated by a pseudocapsule. Most of them involve the bone and the surrounding soft tissue and skeletal muscle almost always. The cut surface of the tumour is characteristically soft, gelatinous, mucoid, and

haemorrhagic [6]. Histological features include a mixture of epithelioid and physaliferous cells. Chordomas are classified into classical or conventional, chondroid and de-differentiated types. Most common is chondroid type and de-differentiated type has the worst prognosis amongst them. True malignant forms of chordomas occasionally have areas of typical chordoma as well as undifferentiated areas, most often fibrosarcoma. The prognosis in such cases is poor [8, 17, 18].

Imaging:

Radiographically, chordomas appear as destructive lesions. They virtually always arise from the midline. Sacrococcygeal lesions often are missed on the initial radiographic examination because of overlying bowel gas. They are usually seen more easily on a lateral view of the sacrum. Likewise, radioisotope accumulation in the bladder can obscure a sacral tumor on a bone scan. More than 50% of chordomas exhibit radiographically detectable calcification [7].

MRI and CT scan have complementary roles in tumor evaluation. CT evaluation is needed to assess the degree of bone involvement and to detect patterns of calcification within the lesion. MRI provides excellent anatomical delineation of adjacent structures and is able to characterize the signal of the lesion usually allowing for a confident preoperative diagnosis.

On CT, a centrally located, well-circumscribed, destructive lytic lesion is observed, sometimes with marginal sclerosis. This lesion is associated with an expansile soft-tissue mass. The soft-tissue mass is often disproportionately large relative to the bony destruction, with irregular intratumoral calcifications thought to represent sequestra of normal bone rather than dystrophic calcifications. Moderate to marked enhancement is commonly seen [7].

On MRI, a chordoma typically shows a lobulated sacral mass in intermediate to low signal intensity on T1-weighted images, with small foci of hyperintensity that may indicate intratumoral hemorrhage or a mucus pool. On T2-weighted images, most chordomas exhibit very high signal intensity. Post-contrast T1-weighted images reveal heterogeneous enhancement with a characteristic honeycomb appearance, corresponding to low T1 signal areas within the tumor. Greater enhancement has been associated with a poorer prognosis. SWI/GE sequences may show variable intralesional hemorrhage, suggested by the presence of blooming artifacts. Diffusion-weighted imaging (DWI) and apparent diffusion coefficient (ADC) values vary, with conventional chordomas showing an ADC of approximately $1474 \pm 117 \times 10^{-6}$ mm^2/s, while dedifferentiated chordomas exhibit lower ADC values around $875 \pm 100 \times 10^{-6}$ mm^2/s [9, 10].

Treatment and prognosis

The primary treatment is surgical resection with wide margins, even if this creates a neurologic deficit, because progressive growth of the tumor would create a neurologic deficit anyway and possibly metastatic disease. Resection that preserves the S3 nerve roots bilaterally results in relatively normal bowel and bladder function, whereas resection above this level results in incremental loss of bowel and bladder function. Resection of bilateral S2 nerve roots results in complete loss of control of bowel and bladder function. If wide margins cannot be obtained or if tumor contamination occurs intraoperatively, radiation may be beneficial [11, 12]. Radiation may also be beneficial for patients in whom resection is not feasible, although a cure is rarely, if ever, achieved in these patients [13, 14]. Chemotherapy is of no proven benefit. Likewise, distant metastases are treated surgically [12].

Prognosis depends on the resection of the tumour in surgery and postoperative treatment. Although metastases also may occur, most patients who succumb to the disease do so because of local recurrences. 5-year survival rate is 51% and 10-year survival rate is 35% [15].

Declaration of Interest: All authors declare no conflicts of interest.

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