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Case report

SACRAL CHORDOMA

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Abstract

Chordoma is an ultra-rare bone sarcoma of notochordal origin (incidence 1/1 000 000/year). Complete resection with negative margins remains the treatment of choice. We report a 78-year-old male with low back pain and a sacrococcygeal mass. Magnetic resonance imaging (MRI) showed a 75 mm lobulated tumor involving distal sacrum and adjacent soft tissues without visceral invasion. After multidisciplinary evaluation, surgery was indicated and a posterior oncological resection with sacral osteotomy (S4-S5) was performed. Pathology confirmed chordoma with negative margins. Sacral chordoma poses diagnostic and therapeutic challenges due to its infiltrative growth pattern and proximity to pelvic structures. Complete in bloc resection remains the cornerstone of treatment. This case highlights the importance of multidisciplinary management of a rare disease.

Key words: chordoma, sacrectomy, oncological resection, sacral tumor

Resumen

Cordoma sacro

El cordoma es un sarcoma óseo ultra-raro originado en vestigios de la notocorda (incidencia 1/1 000 000/año). El tratamiento de elección es la resección completa con márgenes negativos. Presentamos el caso de un hombre de 78 años con dolor lumbar y masa sacrococcígea. La resonancia magnética evidenció un tumor de 75 mm con compromiso distal del sacro y de partes blandas adyacentes, sin invasión rectal ni visceral. Tras evaluación multidisciplinaria se indicó cirugía realizándose resección oncológica mediante abordaje posterior y osteotomía sacra a nivel S4–S5. La anatomía patológica confirmó un cordoma con márgenes negativos. La resección en bloque con márgenes negativos continúa siendo la estrategia terapéutica principal. El caso presentado ilustra la importancia del enfoque multidisciplinario en esta enfermedad poco frecuente.

Palabras clave: cordoma, sacrectomía, resección oncológica, tumor sacro

Chordomas are considered ultra-rare sarcomas (1/1 000 000/year), accounting for 1-4% of skeletal tumors[1]. They arise from notochordal cells and occur in the axial skeleton, mainly in the sacrococcygeal region (50%), skull base (35%), and less frequently in the cervical, thoracic, or lumbar spine[2]. These tumors are locally aggressive and can metastasize to lungs, liver or bones. Sacral chordomas are often diagnosed late due to nonspecific symptoms such as gluteal pain or perineal discomfort. In localized disease, complete resection with negative

margins offers the best local control and survival rates, though recurrence (mainly locoregional) remains common[3].

Clinical case

A 78-year-old male presented with low back pain and a sacrococcygeal mass for one year. On clinical examination, a firm, non-mobile 15×10cm intergluteal mass adherent to deep planes was detected. There were no lymphadenopathy or neurological deficits, and sphincter function was preserved. MRI with gadolinium showed a lobulated, hyperintense T2/intermediate T1 tumor (60×65×75mm) involving the coccyx, subcutaneous tissue, and medial gluteal muscles without rectal invasion (Fig. 1). Image-guided biopsy confirmed the diagnosis of chordoma. Staging computed tomography (CT) showed no metastasis (T2N0M0). After multidisciplinary evaluation, upfront surgical resection was indicated.

A posterior approach was chosen. In prone position and iliac crests elevation, a vertical incision was made over the sacrococcygeal tumor, including the previous biopsy tract (Fig. 2). The gluteal muscles were detached from their distal sacral insertions, and the sacrotuberous and sacrospinous ligaments were divided. The midline raphe was sectioned, and dissection of the mesorectum was performed. Then, a sacral osteotomy was carried out at the S4-S5 level and the tumor was excised en bloc without tumoral rupture. After excision, a polypropylene mesh was placed to reinforce the sacral defect, and the wound was closed primarily. Recovery was uneventful, and the patient was discharged on day 4.

Histopathological examination (Fig. 2) revealed a tumor with an infiltrative growth pattern. At low magnification, the tumor showed a lobular architecture, with fibrous septa separating the lobules. Immunohistochemical (IHC) analysis demonstrated positive staining for cytokeratin, epithelial membrane antigen (EMA), S-100 protein, and brachyury, supporting the diagnosis of chordoma. Following multidisciplinary discussion, surveillance without adjuvant treatment was recommended. At 17-month follow-up, the patient remains disease-free on clinical and radiological evaluation.

Written informed consent was obtained from the patient for the publication of this case report and related images.

Figure 1 |A y B: Contrast-enhanced CT scans demonstrating a tumor in the sacrococcygeal region, measuring 66 × 65 mm (white arrow). C y D: Axial contrast-enhanced MRI showing a lobulated sacrococcygeal mass with T2 hyperintensity and intermediate T1 signal, involving the coccyx and adjacent soft tissues (white arrow)

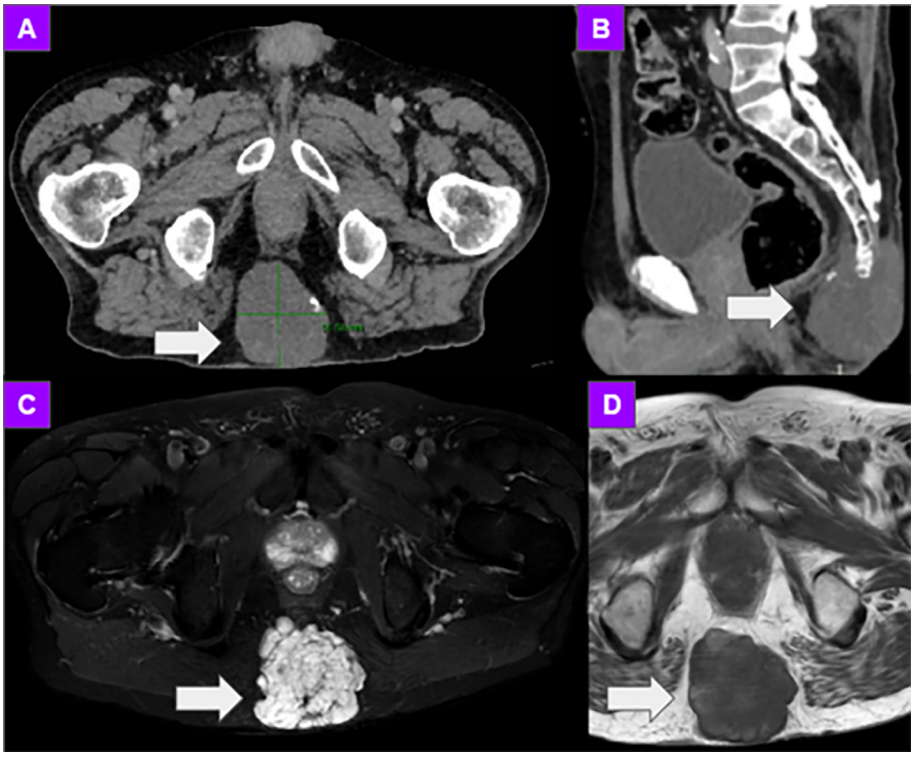
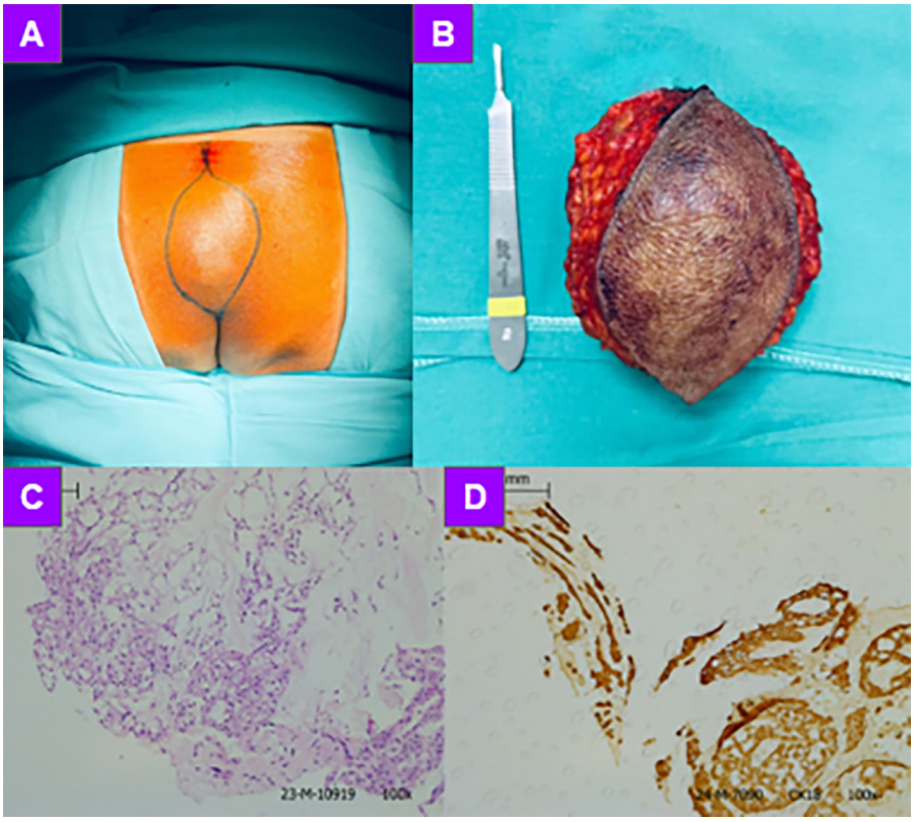


Figure 2 |A: Preoperative skin marking indicating the planned surgical margins. B: In bloc resected specimen of sacral chordoma. C: Histopathological section showing lobulated architecture with physaliphorous cells in a myxoid matrix (hematoxylin and eosin staining). D: Brachyury-positive tumor cells confirming chordoma (IHC).



Discussion

Sacral chordomas are slow-growing but locally aggressive tumors, usually diagnosed late because of nonspecific symptoms such as low back pain, perineal heaviness, or a painless mass[3,4]. Advances in imaging techniques, including MRI and 3D CT reconstruction, have improved preoperative planning and assessment of tumor extent. Positron emission tomography (PET) is not routinely required, as chordoma cells have slow metabolic activity and may not be detected on PET imaging[5]. The initial assessment consists of preoperative imaging and imaging-guided percutaneous core biopsy (14-16 gauge). In patients with localized disease the treatment of choice is en bloc resection with negative margins, which offers the best chance of local control and long-term survival[6]. The proximity of these tumors to pelvic organs and neural structures increases surgical complexity and the risk of morbidity. The level of sacral resection determines postoperative outcomes: lower resections may cause minor sensory deficits, while higher ones can lead to sphincter or motor dysfunction[7]. Therefore, only 40-55% of cases are amenable to wide resection[8]. Hsieh et al. (2009) reported a series of 20 patients showing that complete surgical resection with negative margins improves disease-free survival with acceptable perioperative morbidity[9]. Despite complete surgical resection, locoregional recurrence is frequently observed. The rate of locoregional recurrence may be as high as 70% depending on type of resection margins (R0-R1)[10]. In a recent report of Waldsperger et al. 31(14%) out of 214 patients treated with surgery alone had recurrent disease with a tendency to locoregional and multifocal spread, mainly in osseous and muscular structures[8].

The surgical approach-posterior or combined abdominoperineal-depends on tumor extension and the need for visceral or vascular resection[11]. When complete removal is not feasible, or margins are positive, adjuvant radiotherapy may be considered to enhance local control[12]. The presence of micro-skip metastasis was associated with a higher rate of local recurrence. The maximum tumor diameter and the presence of histologic vascular invasion were associated with poorer overall survival[13]. Recent molecular studies suggest that alterations in genes such as *CLDN9* and *brachyury* may be involved in tumor pathogenesis, potentially serving as targets for future biological therapies[14,15].

Although evidence is mostly derived from small, retrospective series, current data support en bloc resection with negative margins as the cornerstone of treatment, achieving improved disease-free survival with acceptable morbidity and mortality. Long-term follow-up is essential, as late recurrences are common. Continued research into molecular pathways may offer novel therapeutic options for this rare and challenging disease.

Conflict of interest: None to declare

References

1. Stacchiotti S, Frezza AM, Blay JY, et al. Ultra-rare sarcomas: a consensus paper from the Connective Tissue Oncology Society community of experts on the incidence threshold and the list of entities. *Cancer* 2021; 127: 2934-42.
2. Court C, Briand S, Mir O, et al. Management of chordoma of the sacrum and mobile spine. *Orthop Traumatol Surg Res* 2022; 108: 103169.
3. Radaelli S, Fossati P, Stacchiotti S, et al. The sacral chordoma margin. *Eur J Surg Oncol* 2020; 46: 1415-22.
4. Varga P, Bors I, Lazary A. Sacral tumors and management. *Orthop Clin North Am* 2009; 40: 105-23.
5. New diagnosis. Chordoma Foundation. In: https://www.chordomafoundation.org/treatment-guidelines/new-diagnosis; accessed October 2025.
6. Fourney D, Rhines L, Hentschel S, et al. En bloc resection of primary sacral tumors: classification of surgical approaches and outcome. *J Neurosurg Spine* 2005; 3: 111-22.
7. Zuckerman S, Lee S, Chang G, et al. Outcomes of surgery for sacral chordoma and impact of complications: a report of 50 consecutive patients with long-term follow-up. *Global Spine J* 2021; 11: 740-50.
8. Waldsperger H, Lehner B, Geisbuesch A, et al. Recurrence patterns after resection of sacral chordoma: toward an optimized postoperative target volume definition. *Cancers* 2025; 17: 2521.
9. Hsieh P, Xu R, Sciubba D, et al. Long-term clinical outcomes following en bloc resections for sacral chordomas and chondrosarcomas: a series of twenty consecutive patients. *Spine* 2009; 34: 2233-9.
10. Ruggieri P, Angelini A, Ussia G, Montalti M, Mercuri M. Surgical margins and local control in resection of sacral chordomas. *Clin Orthop Relat Res* 2010; 468: 2939-47.
11. Denaro L, Berton A, Ciuffreda M, et al. Surgical management of chordoma: a systematic review. *J Spinal Cord Med* 2020; 43: 797-812.
12. Dial B, Kerr D, Lazarides A, et al. The role of radiotherapy for chordoma patients managed with surgery: analysis of the National Cancer Database. *Spine* 2020; 45: 742-51.
13. Akiyama T, Ogura K, Gokita T, et al. Analysis of the infiltrative features of chordoma: the relationship between micro-skip metastasis and postoperative outcomes. *Ann Surg Oncol* 2018; 25: 912–9.
14. Xu Z, Zhang L, Wen L, et al. Clinical and molecular features of sacrum chordoma in Chinese patients. *Ann Transl Med* 2022; 10: 61.
15. Chen S, Ulloa R, Soffer J, et al. Chordoma: a comprehensive systematic review of clinical trials. *Cancers* 2023; 15: 5800.