Metastatic sacral chordoma

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ABSTRACT

Sacral chordomas (SC) are rare primary malignant bone tumors of the vertebral column, with an incidence between 0.000005-0.000027%. This study aims to describe a case of metastatic SC. A 42-year-old man without comorbid conditions, arrived at the referral center, presenting with a sacral lesion. MRI showed a tumor measuring 9.3 cm that was suggestive of myeloma or chordoma. A biopsy with histopathology study was performed, confirming the diagnosis of SC. The patient underwent surgical tumor excision. Six months after surgery, the tumor recurred with metastatic vertebral column implants, soft tissues of the chest wall, liver, and pleural space, and the patient developed paraplegia. There was no indication of adjuvant radiotherapy and/or chemotherapy. There was also no possibility that the Unified Health System would approve imatinib. At about 28 months of monthly clinical follow-up, the patient died. The case presented showed unsuccessful SC surgery, which is associated with a worse prognosis. The patient had systemic tumor dissemination and paraplegia a few months after surgery, dying at 28 months of follow-up.

Keywords: Chordoma, Sacrum, Tumor metastasis, Case reports.

INTRODUCTION

Initially described in 1857 by Virchow, chordomas are the most common primary malignant bone tumors of the spinal column. Chordomas originate from remnants of the notochord, and its incidence is 0.18-0.84 cases/million people per year, with sacral involvement (SC) in around 29-32% of cases[1-3].

The treatment of choice for chordomas is surgical excision. There are currently no efficient adjuvant therapeutic strategies in these cases since chordomas are resistant to the available chemotherapy and radiotherapy regimens[3,4].

This study aims to report a case of sacral chordoma with metastases to the vertebral column, soft tissues of the chest wall, liver, and pleural space.

CASE REPORT

A 41-year-old man, without any comorbid conditions, was transferred to the referral center for diagnostic investigation of a sacral lesion, complaining of pain and discomfort in the region for about two months. Magnetic resonance imaging (MRI) was performed and detected a large lobulated expansive formation measuring 9.3 cm with characteristics of myeloma or chordoma, involving the sacrum with S2-S5 dissemination, invasion of the canal and sacral foramens and a large anterior exophytic component that occupied the presacral space. Other small similar iliac lesions at the sacroiliac joint were detected.

Biopsy by curettage and histopathological analysis of the lesion with hematoxylin-eosin (H&E) staining were performed. Results indicated a moderately differentiated chordoma (G2), with a mitotic index of 1 mitosis/10 high-power microscope fields, necrosis, and negative angiolymphatic invasion.

Tumor marginal resection, performed via the posterior route, was unsuccessful. Histopathological and immunohistochemical studies of the surgical specimen were not performed since these tumors are not responsive to the currently available adjuvant radiotherapy and/

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https://doi.org/10.11606/issn.2176-7262.rmrp.2022.195609

or chemotherapy. At about six months after surgery, there was a tumor recurrence with chest wall lesions (Figures 1A and 1B), and the patient developed paraplegia. CT scan of the column showed multiple metastatic implants in the region.

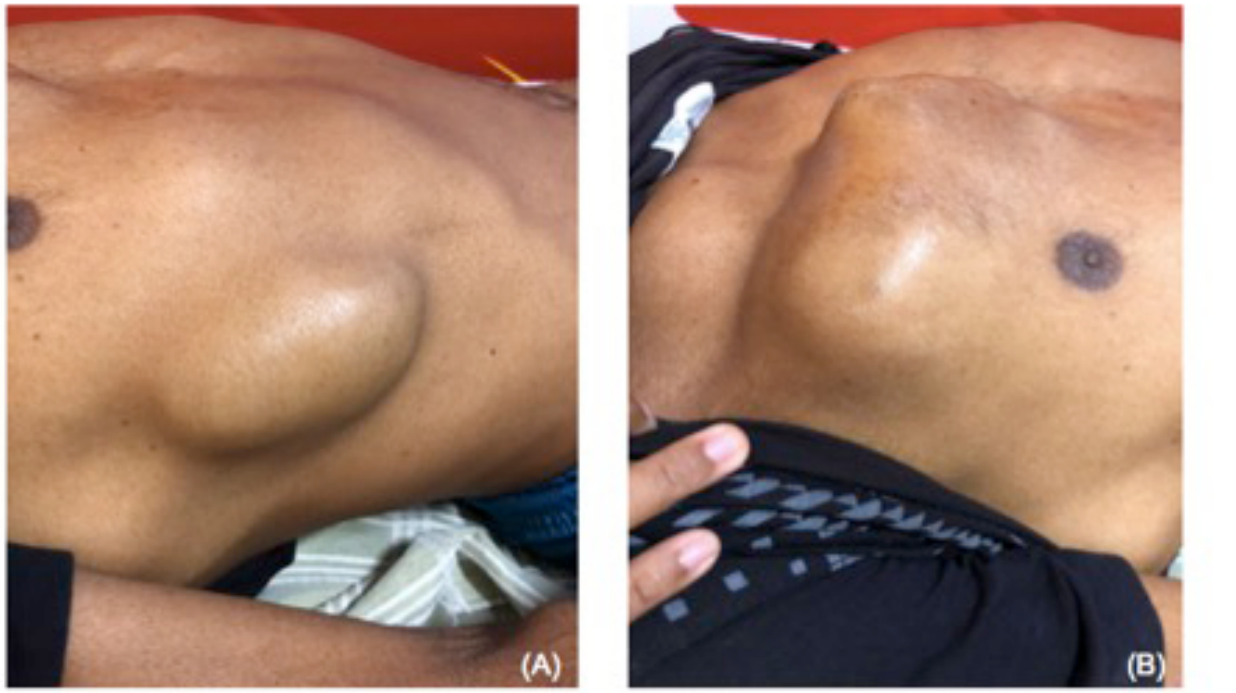

Figure 1: (A and B): Chest lesions that occurred in the patient.

CT scan (Figure 2A) showed expansive lesions with a large soft tissue component, involving several costal arches bilaterally, associated with extensive bone lysis, with some lesions projecting into the pleural space, as well as soft tissues of the chest wall, the largest measuring 8.4 cm in the mid-third of the right hemithorax and 9.2 cm in the anterosuperior region anterosuperior of the same hemithorax. Compressive fracture at T8 with a large mass of soft tissues in a region that presented paravertebral extension, and inside the vertebral canal. On MRI (Figure 2B), a large heterogeneous expansive formation with cystic and hemorrhagic areas, imprecise limits, and lobulated contours with heterogeneous post-contrast enhancement, measuring 28 cm in the posterior pelvic region, largest component at the right, characterized by extensive involvement of the sacrum, ilium and ischia-pubic rami at the right. There was a large component in the right gluteal region, spreading to the root of the thigh. At the right, the lesion occupied the obturator and ischiatic foramens, as well as the left ischiatic foramens, with extension to gluteal muscle planes on this side. Multiple focal lesions in the left iliac, proximal femoral region, and left ischium of a secondary aspect. A nodular lesion with post-contrast heterogeneous enhancement in the anterior abdominal wall muscle compatible with a secondary implant. Liver node measuring 2.3 cm in the VII segment that may represent a secondary lesion. Multiple bone lesions occurred in dorsal and lumbar vertebral bodies, a large component of paravertebral soft tissues, and inside

the spinal canal, with spinal canal stenosis and medullary compression at T8, L2, and L3. Other lesions occurred in the lower costal arches that were compatible with secondary implants.

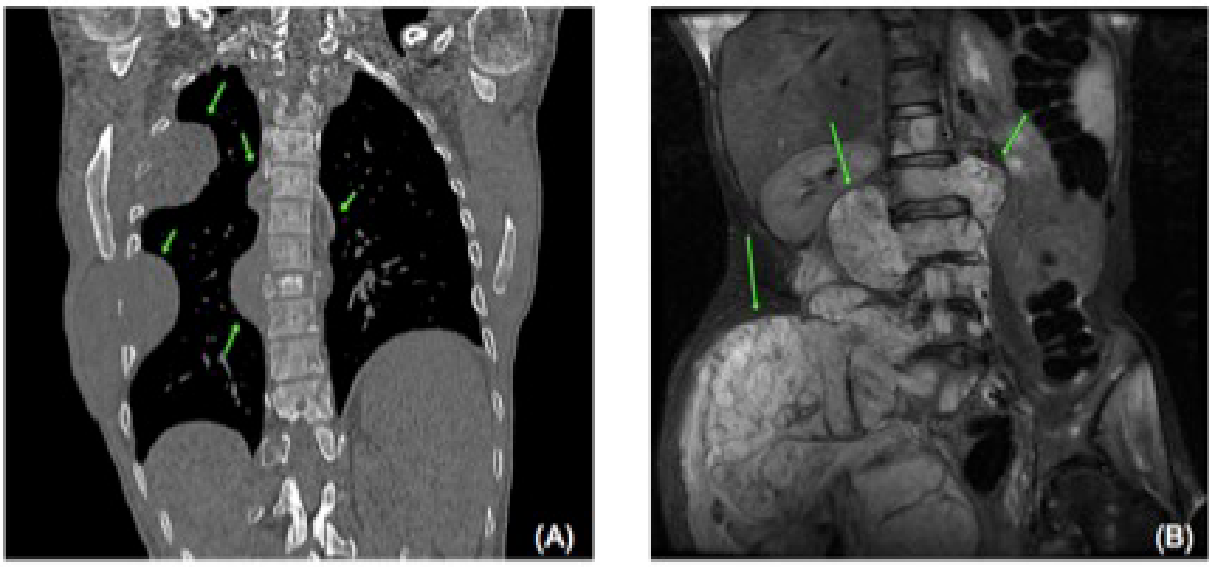

Figure 2: Patient metastases in the vertebral column, soft tissues of the chest wall, liver, and pleural space. (A): Computed tomography scan of the chest. **(B)**: Magnetic resonance imaging.

The lack of therapeutic possibility for the patient was discussed with the family since the disease had extensive systemic dissemination, and there is no classical possibility of adjuvant therapy for this tumor. The use of targeted molecular therapy with imatinib would not be approved by the Unified Health System. The protocol was initiated for symptomatic control. At around 28 months of monthly clinical follow-up (8/31/2019 – 12/20/2021), the patient died.

This study was approved by the Research Ethics Committee of the State University of Piauí, Teresina (PI), Brazil (number: 5.209.669; CAAE: 52368921.5.0000.5209; approval date: Jan 24, 2022). The family member responsible for the patient signed the free informed consent term (FICT).

DISCUSSION

SC are rare tumors, with an incidence of about 0.000005-0.000027%. Therefore, it is essential to consider the possible differential diagnoses when faced with a suspicion of SC since several conditions can mimic this tumor. Chondrosarcoma is similar to SC radiologically and histologically, and it is the main differential diagnosis. Giant cell tumors and plasmacytomas can also have radiological similarities to SC. Furthermore, metastatic lesions in the sacrum derived from prostate, breast, thyroid, lung, and colon cancers, especially in the elderly, are not uncommon findings[2,3,5].

SC is a diagnostic hypothesis after the performance of a suggestive MRI exam. Definitive diagnosis is usually made by H&E stained histopathology after biopsy. Nevertheless, in more challenging diagnoses, immunohistochemistry may be used, in which chordomas are positive for epithelial membrane antigen (EMA) and pan cytokeratin (pan CK)[3,6]. In the current case, a definitive diagnosis of chordoma was obtained by H&E histopathology only, and immunohistochemistry was not required.

Sacral chordomas commonly occur between the fifth and sixth decades of life, with a recurrence rate of 40-50% after surgery. Chordomas do not respond to currently available radiotherapy and chemotherapy regimens, and there is still no possibility of classical adjuvant therapies in these modalities[3,4,6]. Nevertheless, in cases of recurrent or advanced chordomas, imatinib may be used as a first-line agent, based on its inhibition of platelet-derived growth factor receptor-beta, expressed in chordomas. Some studies have also shown that erlotinib may be used (epidermal growth receptor factor inhibitor) in cases of advanced chordomas that are not responsive to imatinib[7].

In general, chordomas are not very aggressive tumors. Distant metastases tend to occur, mainly to the lungs, soft tissues, lymph nodes, liver, and skin. Furthermore, it has been described that around 40-60% of patients with chordoma develop distant metastases. Negative margins during surgery remain the major marker of better prognosis, which is associated with a lower local recurrence rate and better survival. In patients with positive margins or inoperable tumors, palliative radiotherapy may be given. However, evidence suggests that its benefits are very limited. In a series of 25 patients with chordoma, Ferraresi et al. showed an overall survival of 76.7% and 59.7%, respectively, and disease-free survival of 78.3% in five years[8].

In the case presented in this study, the tumor recurred six months after surgery, with extensive posterior metastatic involvement of the vertebral column, soft tissues of the chest wall, liver, and pleural space, resulting in paraplegia of the patient. This case demonstrates the typical progression of SC when surgery is unsuccessful since these tumors have a worse outcome under these conditions, and classical adjuvant therapeutic strategies cannot be used. This case report is of interest since it addresses a rare tumor with little therapeutic possibilities, adding data to the literature that may be useful to evaluate prognostic factors and survival related to this disease.

CONCLUSION

The case report shows the progression of sacral chordoma with unsuccessful surgical resection. Since chordomas do not respond to the currently available adjuvant radiotherapy and chemotherapy regimens, the prognosis is usually worse in these cases. The patient had systemic tumor dissemination and paraplegia a few months after surgery and died 28 months after his referral to the oncology clinic.

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Authors' contributions:
REARC, ARLV: Study concept, data curation, formal analysis, project management, and writing (original draft, review and editing). ESCN, CECS: Study concept, data curation, formal analysis, project management, and writing (review and editing). All authors have read and approved the final draft.

Acknowledgements
We thank the Macroregional Hospital Dr. Everardo Aragão in Caxias for the institutional support.

Conflicts of interest
We deny having any conflicts of interest in the performance and/or publication of the study.

Funding
This study did not receive any external funding, either from the public or private sectors or nonprofit organizations.

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Editor:
Prof. Dr Felipe Villela Gomes

Received: mar 11, 2022
Approved: may 30, 2022
