Sacral chordoma:case report and literature review

Summary

Sacral chordoma is a rare malignant tumor originating from embryonic remnants of the notochord, primarily affecting adults aged 50 to 70. Diagnosis involves MRI and biopsy, with complete surgical resection as the main treatment. Due to its aggressive nature and recurrence tendency, long-term follow-up is essential. Ongoing research aims to improve targeted therapies and immunotherapies.

Case: A 66-year-old patient, known to be hypertensive under treatment, was referred for management of sacralgia with a sacral mass and constipation evolving for 4 months. The clinical examination showed no deficit with hypoesthesia in the saddle territory S3 and a fixed mole consistency mass in relation to the deep plane, not very sensitive to palpation, without a motor deficit in the lower limb. Histological examination of a macrobiopsy of this swelling concluded that it was a chordoma. CT and magnetic resonance imaging (MRI) showed a tumor with multiple branches toward the scrotal region. A palliative lumpectomy was performed, and radiotherapy was indicated for this patient.

This case report highlights the importance of Interdisciplinary collaboration is essential to managing complex conditions such as sacral chordomas, ensuring the delivery of the highest quality care, and optimizing patient outcomes.

Introduction

Sacral chordoma is a rare and slow-growing malignant tumor that originates from the embryonic remnants of the notochord. Representing approximately 1 to 4% of all primary bone cancers, sacral chordomas are primarily located in the sacrum, the base of the skull, and along the spine (1). Despite their rarity, they pose a significant challenge due to their anatomical location and relative resistance to conventional treatments (2).

Epidemiologically, sacral chordomas primarily affect adults, with a peak incidence between 50 and 70 years, and show no significant gender predilection (3). Initial symptoms are often insidious, including chronic pelvic pain, neurological dysfunctions, and gastrointestinal disturbances, which can delay diagnosis (4).

The diagnosis of sacral chordoma relies on a combination of imaging techniques, notably magnetic resonance imaging (MRI) and computed tomography (CT), which allow visualization of the local extent of the tumor and its impact on surrounding structures (5). Histopathological confirmation through biopsy is essential to establishing a definitive diagnosis (6).

Regarding treatment, complete surgical resection remains the cornerstone, despite the technical challenges posed by the complex anatomy of the sacrum and the proximity of vital structures (7). Adjuvant radiotherapy, particularly proton therapy and stereotactic radiotherapy, can enhance local control of residual or inoperable tumors (8). However, systemic treatment options are limited, and ongoing research is needed to develop effective targeted therapies and immunotherapies (9).

Due to their aggressive nature and propensity for local recurrence, sacral chordomas require rigorous and long-term follow-up to detect recurrences early and adjust therapeutic strategies accordingly (10).

Case Presentation:

This is a 66-year-old patient, known to be hypertensive under treatment, referred for management of sacralgia with a sacral mass and constipation evolving for 4 months, in whom the clinical examination showed no deficit with hypoesthesia in the saddle territory S3 and a fixed mole consistency mass in relation to the deep plane not very sensitive to palpation, without fistulization to the skin, signs of inflammation, or motor deficit in the lower limb.

CT scan (CT) and magnetic resonance imaging (MRI) confirm that it is a tissue mass 18 cm in diameter, polylobed, well limited, in the gaze of the coccyx, pushing the bulb forward and the rectal canal without signs of invasion of these. This mass has a non-greasy consistency, improving after intravenous injection of contrast product.

The immunohistochemical study showed

Anti-AE1/AE3 antibodies: ++ Anti-ENA antibodies: ++ Anti-BACHYRURIA antibodies:++

The biopsy of the mass and the anatomopathological and immunohistochemical study made it possible to make the diagnosis of sacrococcygeal chordoma.

Total-body positron emission tomography-computed tomography (PET-CT) revealed localized disease without metastasis.

The tumor was surgically resectable with the help of visceral specialists. (Figure 1-2))

Therapeutic intervention and follow-up: radiotherapy was carried out on the tumor at a total dose of 66 Gy in 33 fractions per conformal radiotherapy.

After 3 years of evolution, the patient is still alive with no pain, and the tumor looks clinically and radiologically stable.

Discussion

Sacral chordoma is a rare, slow-growing, but locally aggressive malignant tumor originating from remnants of the notochord. Despite surgical resection being the primary treatment modality, achieving complete tumor removal is often challenging due to the tumor's proximity to critical structures such as nerves, blood vessels, and the pelvic organs. This incomplete resection leads to a high rate of local recurrence, making adjuvant therapies essential in managing this malignancy (1, 2, 3, 4, 5).

** Shaping surgical margins**

Surgical margin status is the most important prognostic factor in sacral chordoma patients undergoing surgery . En- bloc tumor-sacrum resection is the cornerstone of treatment of both primary and recurrent localized disease. Regardless of theapproach used, the final goal is to achieve a wide local excision with negative microscopic margins[8,11,12,].

Several retrospective analyses demonstrated the negative prognostic impact of positive microscopic margins on the outcome of sacral chordoma. However, even in patients where resection is microscopically complete, loco-regional relapses are seen in >50% of cases. Recurrences may occur late and only a minority of patients are disease-free at 15 years[8,11,12,].

Radiotherapy has emerged as a crucial adjunct in the treatment of sacral chordoma. Traditional radiotherapy, however, is often limited by the tolerance of surrounding healthy tissues to radiation. High doses required to control chordoma can lead to significant morbidity, making advanced radiotherapy techniques more suitable for this purpose (6).

Proton Therapy and Stereotactic Radiotherapy

Proton therapy offers a distinct advantage due to its ability to deliver high-dose radiation with precision, minimizing exposure to surrounding healthy tissues. Studies have shown that proton therapy, when used in conjunction with surgical resection, can improve local control rates and potentially enhance overall survival (8, 10). Proton beams have a distinct physical advantage known as the Bragg peak, which allows maximum energy deposition directly in the tumor with minimal exit dose, thereby sparing adjacent normal tissues from high-dose radiation (10).

Stereotactic radiotherapy (SRT) is another advanced technique that delivers high doses of radiation in a highly focused manner over fewer sessions. This precision reduces damage to surrounding tissues and is particularly beneficial for chordomas that are difficult to resect completely. The use of

SRT has shown promising results in improving local control and reducing recurrence rates in patients with sacral chordoma (8).

Efficacy and Safety

The combination of surgery and radiotherapy, particularly with advanced techniques like proton therapy and SRT, has been shown to provide better outcomes in terms of local control and progression-free survival. Fuchs et al. (5) and Park et al. (10) highlight the efficacy of high-dose proton/photon beam therapy in managing residual disease post-surgery. This multimodal approach mitigates the high recurrence rates traditionally associated with chordomas, which are inherently resistant to conventional radiotherapy and chemotherapy (7, 9).

Future Directions

Despite these advancements, challenges remain in optimizing the timing, dosage, and combination of radiotherapy with other treatment modalities. Ongoing research is focusing on enhancing the radiosensitivity of chordoma cells and combining radiotherapy with novel systemic therapies, such as targeted therapies and immunotherapies. Further clinical trials are essential to establish standardized protocols and improve the therapeutic outcomes for patients with sacral chordoma (1, 4).

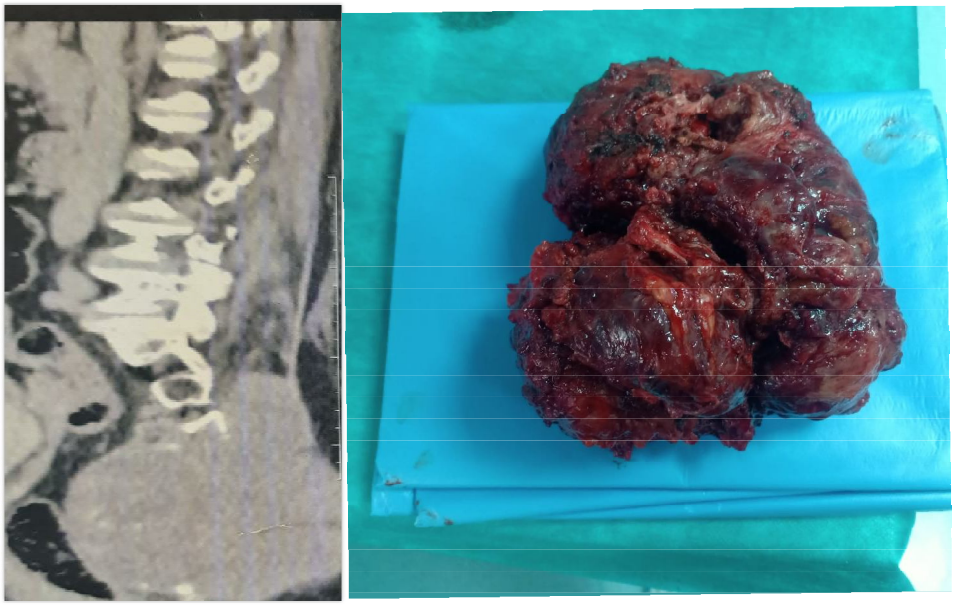

Fig. 1. Neoplastic invasion of the posterior rectal wall. The tumor is therefore resected en bloc while preserving the sacrum and rectum.

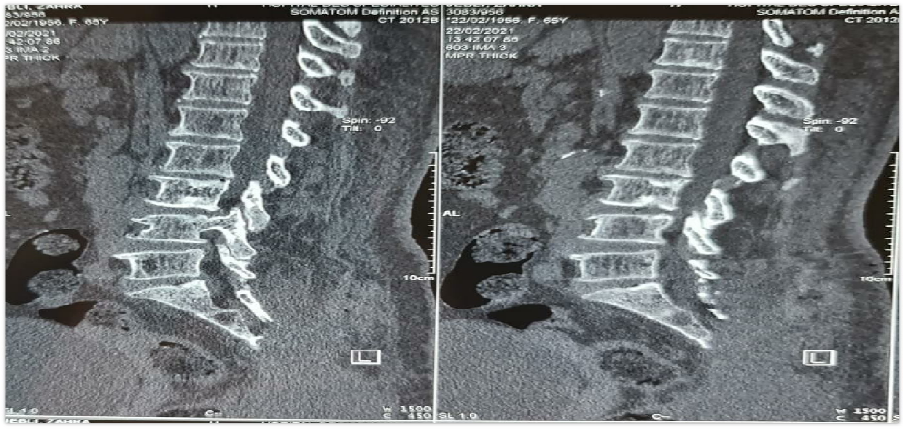

Figure 2 CT scan of the control

CONCLUSION :

Chordoma is the most common primary malignant tumor of the sacrum, but it remains a rare condition. The clinical approach to sacral chordoma is always very delicate due to clinical polymorphism, so it is often unknown and diagnosed late in relation to its location and the mass effect it exerts on neighboring organs. The paraclinical approach consists of performing a CT-MRI ANAPATH. The preferred treatment is the widest possible surgical resection (ensuring local control of the tumor, decompressing the nervous structures, and improving quality of life). Their evolution is often favorable, but this requires aggressive treatment. first surgical, followed by radiotherapy, particularly advanced techniques like proton therapy and SRT, plays a vital role in

themultidisciplinary management of sacral chordoma. These modalities offer significant benefits in terms of local tumor control and minimizing damage to surrounding tissues, ultimately improvingpatient outcomes and quality of life .the obligation of regular follow-up to detect local recurrence even 20 years after resection.Local recurrence may require re-intervention; if the size of the recurrent tumor is large or in the face of an operative contraindication, radiotherapy and/or chemotherapy remain the only recourse.During its evolution, The prognosis of sacral chordomas depends mainly on the quality of the excision.

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Fig. 1. Neoplastic invasion of the posterior rectal wall. The tumor is therefore resected en-bloc with the sacrum and the rectum.

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