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Sacral Chordoma: A Case Report and Literature Review

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Report

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ABSTRACT

Sacral chordoma is a type of cancer that starts from leftover tissues of the notochord and is mostly seen in individuals, between 50 to 70 years old. The diagnosis typically involves the use of MRI scans and a biopsy procedure to confirm it accurately; surgery is usually the approach for treatment. Given its behavior and tendency to come back after treatment (recurrence) continuous monitoring over a period is crucial. Researchers are actively working on enhancing treatments, like targeted therapies and immunotherapies through studies.

A 66 year old individual, with a history of blood pressure and receiving treatment was sent for evaluation due to lower back pain along with a lump in the back and ongoing constipation for the past four months. Upon examination by the doctor or healthcare provider (or appropriate professional title) no physical impairment was detected except for sensation in the S3 area and a solid mass within the tissue that felt somewhat firm when touched but not very tender; there was no loss of movement in the leg. After performing an analysis, on a small tissue sample taken from this growth mass (or lump) it was confirmed through examination that it is classified as a chordoma. CT scans and MRI images revealed a tumor extending into branches, near the area of a patients body.

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The patient underwent a lumpectomy procedure, with radiotherapy recommended for treatment. This report emphasizes the significance of teamwork, in handling conditions, like sacral chordomas to provide top notch care and improve patient results effectively.

Keywords: Sacral chordoma; sacral mass; anatomy; malignant tumor; pelvic pain.

1. INTRODUCTION

1.1 Background

Sacral chordoma is a rare and slow growing malignant tumor of notochordal remnants that form embryonic spinal structures. Sacral chordomas make up approximately 1-4% of all primary bone cancers, primarily affect the sacrum at the base of the skull and vertebral column [1]. Although they are rare, because of their elaborate anatomy and not being typically responsive to conventional therapies [1].

1.2 Epidemiology and Presentation

Sacral chordomas almost exclusively arise in adults, with a peak age group between 50 and70 years of life [2]. It is not known to be more common in males or females though the symptoms develop insidiously and prompt diagnosis is therefore delayed [3]. Initial symptoms often include chronic pelvic pain, neurologic findings and functional gastrointestinal issues* [4].

1.3 Diagnosis and Imaging

Imaging techniques and histopathology confirm the diagnosis of sacral chordoma. Both Magnetic Resonance Imaging (MRI) and Computed Tomography (CT) are very helpful tools for evaluating the reach of a tumor and its implications on adjacent structures [5]. Biopsy is necessary to collect biopsy specimens for pathological examination and rule out other differential diagnoses [6].

1.4 Treatment and Management

The ideal strategy of treatment for sacral chordomas is complete surgical resection. Nevertheless, the sacrum has complex anatomy with critical neighboring structures contributing to that often almost impossible to obtain a R0 resection [7]. Often adjuvant radiotherapy, including proton therapy or stereotactic radio-surgery, is implemented to target any residual disease or portions of tumor that could not be resected [8]. Effective systemic treatment options are few, and current research efforts are aimed at the development of targeted therapies and immunotherapies [9].

1.5 Prognosis and Follow-Up

Sacral chordomas have a poor prognosis with high rates of local recurrence. Follow-up is most important over the extensive term to attain an early notice of recurrences and review therapeutic strategies [10]. The outlook for patients with sacral chordoma can differ based on variables including the size of the tumor, whether or not it has spread

2. 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.

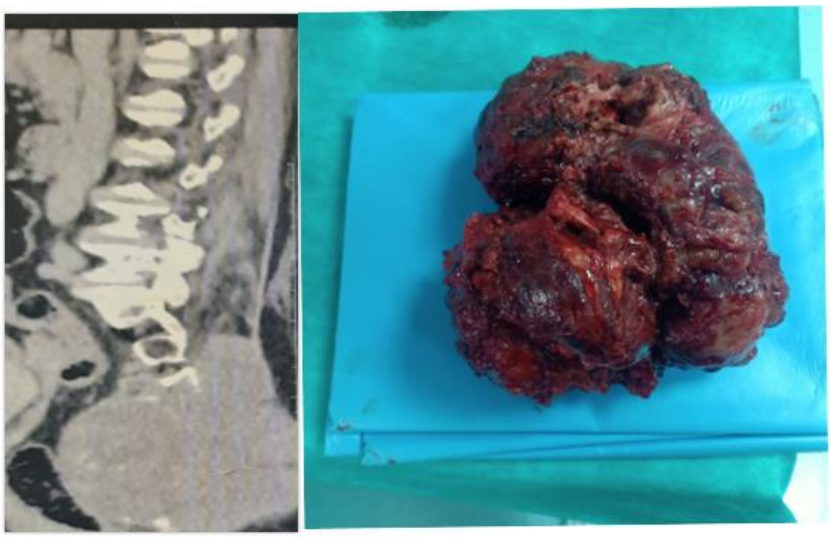

Fig. 1. Neoplastic invasion of the posterior rectal wall. The tumor is therefore resected en bloc while preserving the sacrum and rectum

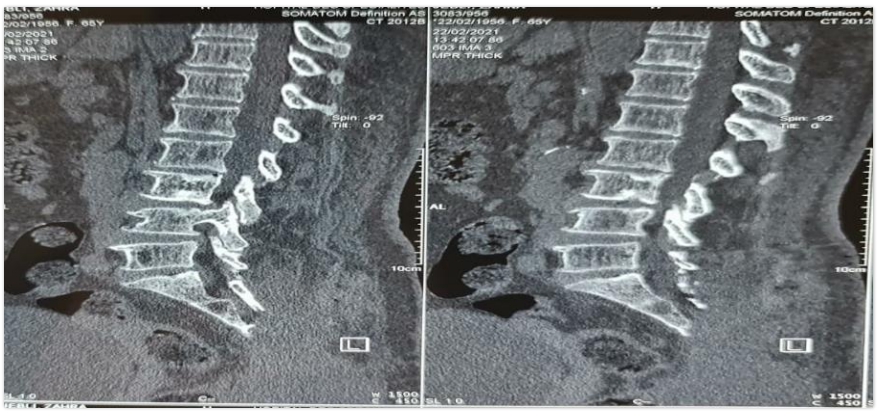

Fig. 2. CT scan of the control

The tumor was surgically resectable with the help of visceral specialists. (Figs. 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.

3. DISCUSSION

Sacral chordoma is a rare and highly malignant tumor, which has few effective therapeutic options due to its complex structure and non respondence to traditional treatments. Surgical resection is the foundation of management although complete tumor excision is frequently prevented by close association with critical structures. This often results in increased rates of local recurrence and the necessity for adjuvant therapies [1-5].

The most important prognostic factor in surgically treated sacral chordoma patients is the quality of resection. En bloc resection of the tumor with negative microscopic margins is ideal; however, this can be difficult to accomplish. To this day, local recurrence rates continue to be significant even after optimal microscopically complete resection leading the way for other therapeutic strategies [8, 11-12].

Recent advances in radiotherapy, such as proton therapy and stereotactic radiotherapy (SRT), have become important adjuncts to surgery.

These minimal doses target a very small volume with minimal radiation delivered to the surrounding normal tissues. Randomized clinical trials have shown proton therapy and SRT to be effective in increasing rates of local control [8–10], leading to a possible benefit on overall survival, especially when used in combination with surgical resection.

Surgery combined with modern radiotherapy techniques is promising in terms of decreasing the high local recurrence rates for sacral chordoma. Nonetheless, challenges remain in treatment optimization. Current research is aimed at improving the radiosensitivity of chordoma cells, and developing new systemic therapies, such as targeted therapies or immunotherapies. Future clinical trials are needed to define standardized protocols and optimize outcomes for this difficult-to-treat malignancy.

4. CONCLUSION

While it is uncommon, sacral chordoma is an aggressive malignancy that poses complex anatomical and therapeutic challenges due to its natural resistance against current interventions. Although surgical resection has been the majorstay of cure, complete resection is difficult to achieve given that these lesions are often in close proximity to important structures. This often results in high local recurrence rates, highlighting the need for adjuvant therapies.

For the surgical patients with sacral chordoma, the most important prognostic factor is degree of resection. An ideal scenario would include achieving complete en-bloc tumor-sacrum resection with free microscopic margins, however, this could be technically very demanding. Despite microscopically complete resection, local recurrence rates still high and further therapeutic efforts are needed.

Combination of Radiotherapy with Surgery has become a cornerstone in the era of modern radiation techniques, especially for such advanced techniques as Proton Therapy and Stereotactic Radiotherapy. These allow one to deliver radiation even more precisely with little damage to surrounding normal tissue. Several studies have shown that local control can be achieved by using proton therapy or SRT and suggests an improvement in overall survival, when combined to surgical resection.

The combination of surgery and high-precision radiotherapy techniques with stereotaxy has demonstrated increasing evidence to improve the very poor local control rates in sacral chordoma. Nevertheless, the burden of optimizing treatment strategies are still there. Current areas of research in the field are seeking to improve radiosensitivity of chordoma cells or exploring alternative systemic therapies including targeted and immunotherapies. More clinical trials are needed; the only way to determine standardization is through further rigorous investigations and collaboration. This adverse malignancy will continue to cause challenge for at least 40% of potential subjects with this disease in their lifespan.

DISCLAIMER (ARTIFICIAL INTELLIGENCE)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc) and text-to-image generators have been used during writing or editing of this manuscript.

CONSENT

As per international standards or university standards, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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