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Radiotherapy

Radiosurgical Management of Sacral Chordoma: About a Case

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Abstract **Case Report**

Chordoma is a locally aggressive malignant tumour originating from ectopic notochordal cells. The sacrum is a frequent anatomical site of chordoma and represents approximately 50% of all chordomas. Due to the richness in nerve structures, surgery for sacral chordoma is often incomplete, radiotherapy is generally indicated as adjuvant treatment. We report in this work the case of a 52-year-old man, who presented a sacral chordoma treated with surgery and radiation therapy on residual disease. We shed light on the place of combined treatment, combining surgery and radiotherapy in the treatment of sacred chordomas.

Keywords: Sacral chordoma, surgery, radiation therapy.

INTRODUCTION

Although the most common malignant tumor of the sacrum is chordoma, this entity remains relatively very rare [1]. Sacral chordomas is a slow growing, locally invasive tumor, frequently recur, and are relatively radioresistant [2]. Sacral chordomas are often diagnosed at a late stage, making excisional surgery very difficult because of to the richness of the nervous elements in the sacrum [3]. Radiation therapy is used as an adjuvant treatment to control residual disease after surgery [4].

We report in this work the case of a 52-year-old man, who presented a sacral chordoma treated with surgery and radiation therapy on residual disease. With a follow-up of 5 years, the patient presents no symptoms with a radiological stability of the residual disease after surgery.

We shed light on the place of combined treatment, combining surgery and radiotherapy in the treatment of sacred chordomas.

CASE REPORT

A 60-years-old male, with unremarkable past medical history, presented with three years of lumbosacral spine pain mimicking sciatica with disabling constipation, without urinary or erectile dysfunction. Clinical assessment was normal, without any signs of physical or neurological deficits.

A pelvic magnetic resonance imaging (MRI) revealed a sacral mass of polylobed aspect, measuring 85 × 110 × 62 mm, lysing the anterior cortex of the sacrum from S1 to S5, and pushing the rectum forward (Fig 1). A scan-guided biopsy confirmed the diagnosis of sacral chordoma. Anterior abdominal excision surgery was performed, removing the endopelvically developing part of the tumor and leaving the sacral part in place. Postoperative pelvic MRI showed a residual sacral tumor of 61 × 42 × 32 mm (Fig 2).

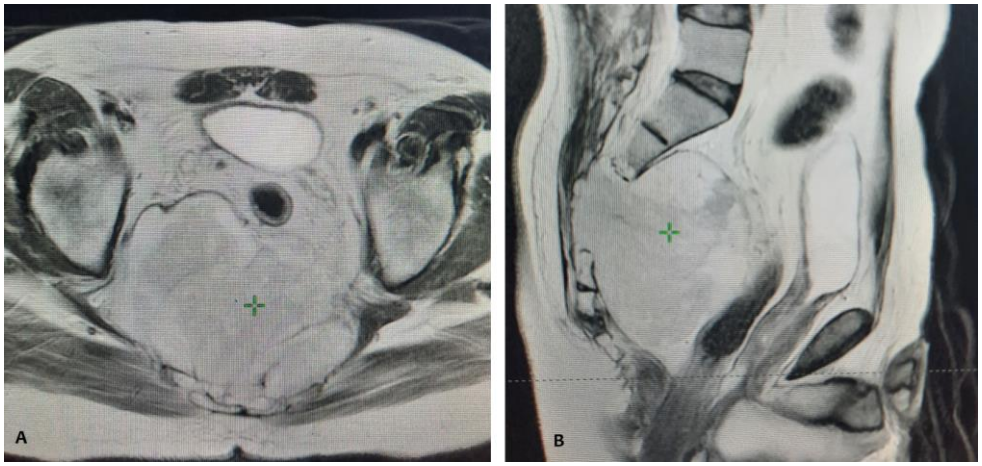

Figure 1: Axial slice (A) and sagittal slice (B) T2 weight pelvic MRI, showing sacral mass of polylobed aspect, lysing the anterior cortex of the sacrum and pushing the rectum forward

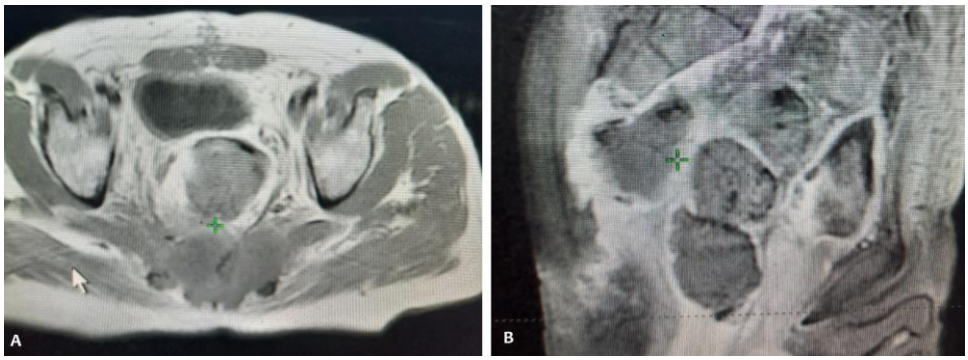

Figure 2: Axial slice (A) and sagittal slice (B) T1 Fat-Saturation weight pelvic MRI, showing residual sacral tumor of 61 × 42 × 32 mm

After discussion in a multi-disciplinary concretion meeting, it was decided to perform radiation therapy on the residual sacral mass.

A CT-simulation was performed, the patient was positioned supine, with arms on the chest. For delineation of target volume and organs at risk, a CT-MRI image registration was made. Two clinical target volumes (CTV) were delineated; CTV1 defined as gross tumor volume (GTV) before surgery and CTV2 defined as residual GTV after surgery. CTVs were expanded by 10 mm to generate two planning target volume (PTV); PTV1 and PTV2 (Fig 3).

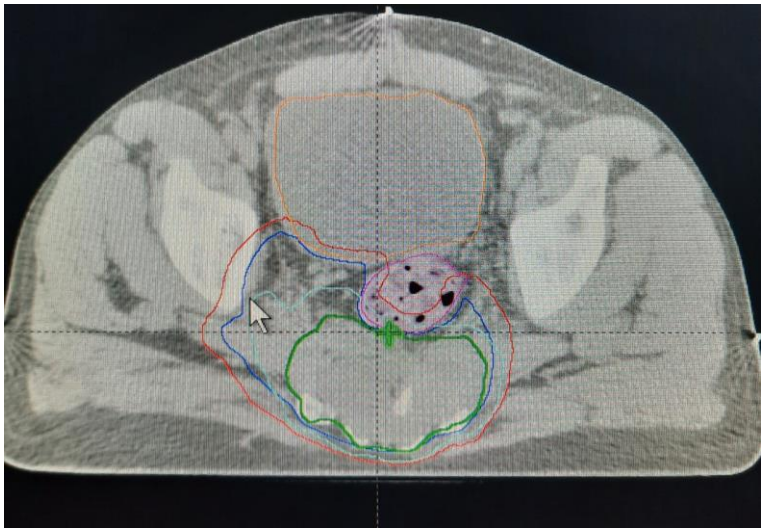

Figure 3: Axial CT-scan image from treatment planning system, showing delineation of CTV1 (blue), CTV2 (bold red), PTV1 (red) and PTV2 (cyan)

The patient was treated with dynamic conformal arc therapy using two arcs. Treatment was done in simultaneous integrated boost (SIB); a total dose of 56 Gray (Gy) was prescribed to the PTV1, and a total of dose of 70 Gy was prescribed to PTV2, all in 33 fractions, five days a week. PTVs were covered by the 95% isodose volume, and maximum hot spots were <110% (Fig 4).

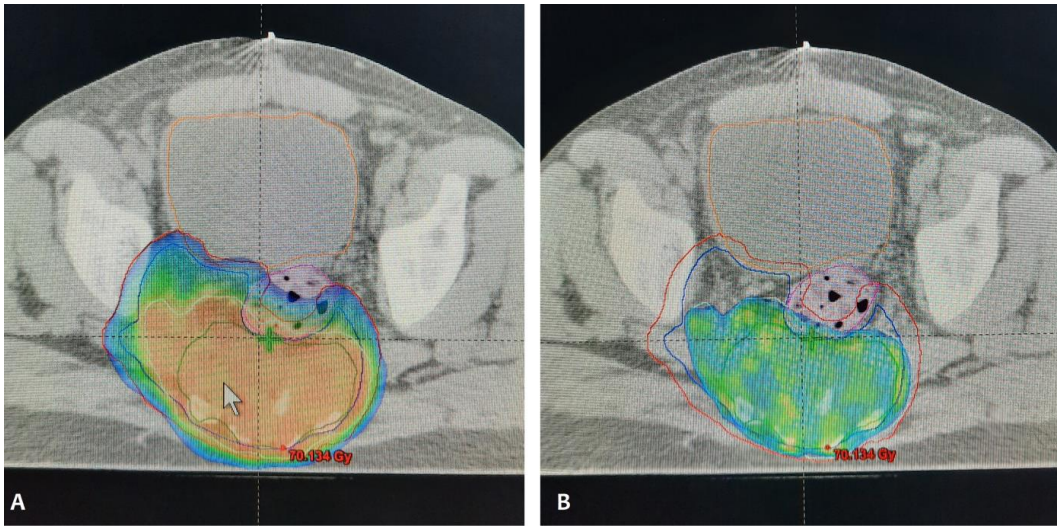

Figure 4: Axial CT-scan images from treatment planning system, showing dose distribution in the PTV1 (A) and PTV2 (B)

Evolution was marked by a significant improvement in symptoms. With a follow-up of 5 years, patient presents actually no symptoms with a radiological stability of the residual disease after surgery.

DISCUSSION

Chordoma is a rare tumor that develops from embryonic notochordal cell remnants. It has a slow growth rate that hinders its early diagnosis [1]. In 50% of all cases, this tumor involves the sacral region, and it affects men twice as often as women [5]. It is typically seen in the 5th and 6th decade with a mean age of 55 years [6]. Pain and radiculopathy due to compression of the sciatic nerve or iliolumbar trunk was reported in up to one third of these patients, and up to one third of patients presented with symptoms urinary tract [7].

The initial treatment of sacral chordomas consists of wide local excision when possible [8]. Complete excisional surgery, although desirable, is not feasible in many cases because of anatomic constraints to surgical access and the proximity of adjacent critical normal structures [9]. For this reason, postoperative radiation therapy is frequently used, despite the fact that chordoma is a very radioresistant tumor [10].

Newer RT techniques, including stereotactic radiosurgery and charged particle irradiation, have been used to target the bone lesion while reducing the radiation exposure to the surrounding nerve roots and the cauda equine [11].

CONCLUSION

Sacral chordoma is a rare tumor. The aim of treatment is to improve clinical symptoms and prevent disease progression. Wide en Bloc surgery is generally impossible at this level, hence the importance of performing excisional surgery which aims to decompress the nerve structures and improve symptoms. The main role of radiation therapy is to prevent possible progression of the disease.

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