



Cryosurgery in the excision of a giant local recurrent sacral chordoma: a case report and literature review

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Received: 13 April 2017 / Revised: 27 November 2017 / Accepted: 16 December 2017
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Abstract



Purpose Chordoma is a low-grade malignant tumor with recurrence and metastasis tendency that originates from embryonic notochordal remnants. The sacrococcygeal region is the most commonly involved site. The aim of this paper is to report the results of the use of cryosurgery in the excision of a giant recurrent sacral chordoma and review of pertinent literature.

Methods A 64 years old female patient with a locally recurrent sacral chordoma came to our attention after ten interventions performed at another institute. A surgical treatment was performed using argon cryosurgery.

Results In this case, it was not possible to perform a marginal or wide excision, but it was intralesional with the removal of three major blocks. The fragments sent for the histological analysis measured total $35 \times 30 \times 8$ cm with a weight of 4.260 g.

Conclusions Given the gelatinous structure of the tumor and the possibility of contamination of operatory field, cryosurgery may be indicated when previous surgeries and the dimensions of tumor mass do not allow a resection with negative margins improving radiotherapy efficacy in the local control of tumor.

Keywords Local recurrent chordoma · Sacral chordoma · Cryosurgery · Cryotherapy

Introduction

Chordoma is a low-grade malignant tumor with recurrence and metastasis tendency that originates from embryonic notochordal remnants [1]. The incidence of chordoma is 0.08 per 100,000 individuals [2]. The sacrococcygeal region is most commonly involved site, followed by skull base and

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mobile spine [3]. It represents only 1–4% of all primary bone tumors [4], but the 40% of all primary sacral tumors [2].

Sacral chordoma has a male/female ratio of 2–3:1 and the majority of affected patients are in the fifth to seventh decades [5]. It has a local recurrence percentage of 43–85% [6–14] and most occur within 3 years from surgery [5].

Local recurrence of sacral chordoma after surgery is mostly related to the level of tumor and to the margin of resection.

In particular, S3 represents a watershed: oncologically appropriate resections above that level often include sacral nerves whose loss of function (i.e., sphincteric and sexual functions) is poorly tolerated by the patients. Resections below that level are less disabling.

Concerning margins of resection, intralesional and marginal resections are associated to a higher risk of local recurrence than wide resection [5, 15, 16].

The treatment of local recurrent chordoma (LRC) involves both surgery and radiotherapy. In particular, for LRC when surgery is planned, wide en bloc excision should be performed. Instead, when wide en bloc resection is not feasible for tumor localization, partial resection must be performed. Post-operatively, high-dose radiotherapy should be administered to reduce local recurrence [17].

The reported case is a 64 years old female patient affected by giant local recurrent sacral chordoma that was treated by means of intralesional en bloc resection using argon cryosurgery at our institute.

Case presentation

The patient was referred to our clinic because of a huge locally recurrent chordoma presenting as voluminous plurilobate formations in the context of buttock muscles bilaterally and in the right adductor region (Fig. 1). Neurological examination revealed urinary and fecal incontinence, but preserved motor function to lower limbs.

The patient had other minor chordoma localizations in the right paravertebral lumbar region and superficially in the subcutaneous layer of the gluteal region.

In the previous 7 years, the patient underwent several treatments at another institution, which were ineffective and the tumor kept relapsing. Since the early beginning, she had radiofrequency ablation twice (6 years before) and then, after a year, the first excision attempt with intralesional margins. Histopathological examination on the surgical specimen was diagnostic for chordoma, so 66 Gy in 33 fractions of conventional external beam radiation therapy was given as adjuvant treatment. Since then, the tumor relapsed locally seven times, and further debulkings were performed in each occasion (Fig. 2). The patient also had a course of chemotherapy with imatinib which did not provide any benefit.



Fig. 1 Axial and coronal RMI showing the preoperative extension of the tumor in the buttocks bilaterally

The case was discussed during the multidisciplinary oncology meeting at our institution and a further excision with the use of intraoperative argon cryosurgery as local adjuvant was offered to the patient. The patient accepted and written consent was obtained.

Tumor stretched along the posterior region of the thigh bilaterally and transversely at the level of the sacrococcygeal region. The probes of cryosurgery were used to facilitate the identification of adequate margins of resection (Fig. 3). Given the extent of the tumor, it was not possible to perform a marginal or wide excision, but there was an en bloc intralesional with the removal of three major blocks. It was, also, carried out at the terminal portion of the coccyx. The sciatic nerve was identified, isolated and preserved bilaterally (Fig. 4). The surgical specimens sent for histopathological examination (which confirmed diagnosis of chordoma) measured total $35 \times 30 \times 8$ cm and collectively weighted 4.260 g (Fig. 5). After surgery, the patient reported a worsening of fecal continence and a deficit of the anterior tibialis of the right leg. Bowel, bladder, motor, sensitivity and sexual function are, in fact, the most frequently compromised

Fig. 2 Sagittal and axial CT scan showing the outcomes of the previous intervention on the sacrum

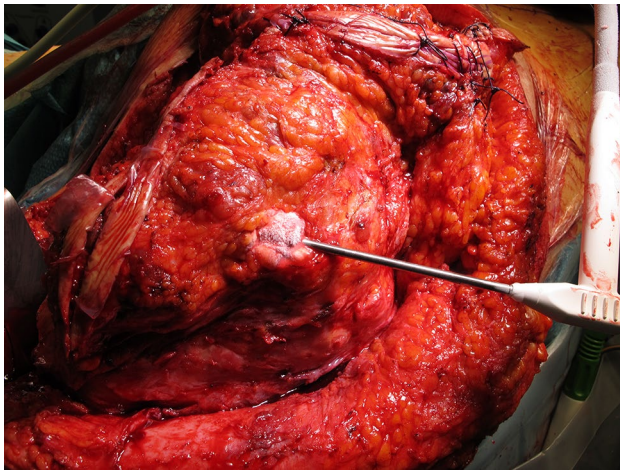


Fig. 3 Intraoperative picture of the freezing cycle of tumor mass



Fig. 5 Surgical specimen composed of three major blocks

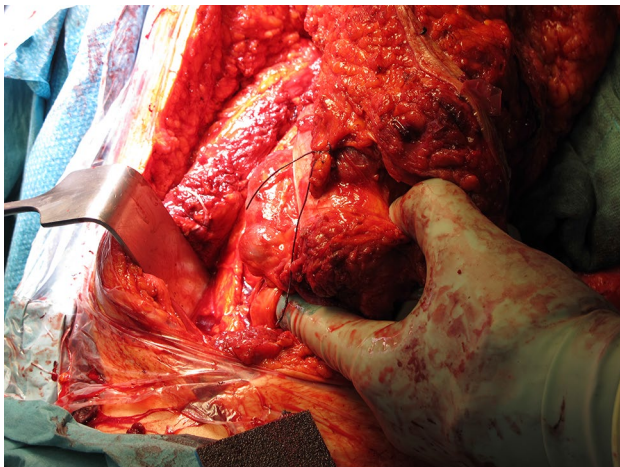


Fig. 4 Intraoperative picture showing dissection of the sciatic nerve

functions after sacral resection [18]. The early postoperative course was complicated by ischemic necrosis of the edge of

the wound that was debrided and an advanced dressings vacuum system was applied. Wound ischemia was not related to the cryoablation procedure, but to an impaired blood supply to the skin since excision involved the subcutaneous tissue involved by the tumor. The wound was revised 2 months later and ultimately healed properly.

During this latter procedure, cryosurgical excision was performed in another small localization of chordoma (right paravertebral lumbar region) not affected by the previous surgery. After healing of the wounds, patient received an additional course of proton therapy.

A 6-month follow-up MRI showed no recurrence of disease in the region where cryosurgery was performed.

Discussion and conclusion

Cryosurgery was firstly used by Cahan for the treatment of benign and malignant tumors [19, 20]. Subsequently, Marcove developed its use in bone tumors [21] suggesting

the alternation of three freeze cycles interrupted with a period of thawing [22].

De Vries et al. first introduced cryosurgery as new treatment modality for chordoma in the sacrococcygeal region. They treated four patients affected by sacrococcygeal chordoma from 1974 to 1980 without resection. Two patients had extensive tumors (greater than 10 cm) and were treated only palliatively. Two other patients who had smaller tumors (< 10 cm) had curative cryosurgical treatment. Both patients were disease-free 10 and 7 years after cryosurgical treatment [23].

In a study conducted on forty-three patients who underwent surgery for resection of sacral chordoma from 1990 to 2005, five patients were treated with liquid nitrogen cryosurgery using three cycle of freezing followed by a period of thawing. In all cases, surgeon submitted tissue for frozen section analysis from areas felt to have a close margin. When a positive margin was encountered on frozen section, the surgeon decided to extend the resection margins. Only in one case, there were positive margins at the final pathology examination, so the surgeon decided to remove more tissue near the positive margin a week later [24].

In another case series, five cases of recurrent chordoma and one recurrent mixopapillary ependymoma were treated with cryoablation. Four tumors were treated for local control, with no evidence of disease at 3, 6, 12 and 15 months follow-up [25].

According to the Chordoma Global Consensus Group, a prior history of piecemeal excision, prior high-dose RT and tumor capsule violation are exclusion criteria for resection with curative intent. A debulking surgery plus radiotherapy, instead, is indicated to achieve the local control of disease [26].

In our case, previous intralesional surgeries caused the spreading of tumor cells producing the appearance of several relapses of disease. We decided to perform an intralesional excision using cryosurgery in a patient in whom surgery intents are local control of disease and support to adjuvant therapies.

Chordoma presents an expansile, lobulated structure, with the cut surface revealing a blue/slate-gray gelatinous matrix alternate to other areas that exhibit a more solid chondroid texture. Due to its gelatinous structure, cryosurgery, through solidification of the mass, seemed to be a valuable tool in decreasing the likelihood of malignant cells spillage in the operatory field during piecemeal removal of the tumor.

In conclusion, cryosurgery may be used to increase the strenght of the margins when curative intent by performing a resection with negative margins is no longer an option (i.e., in cases of previous intralesional surgeries, huge tumor masses, high-dose radiation therapy).

Ultimately, this might result in improved radiotherapy efficacy when combined multidisciplinary treatments are planned.

Acknowledgements The authors thank Carlo Piovani and Cristiana Griffoni for their helpful collaboration in imaging storage and data management.

Funding No funds were received in support of this study.

Compliance with ethical standards

Conflict of interest The authors declare that they have no competing interests.

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