Case report

Late local recurrence, at 19 and 17 years, of sacral chordoma treated by en bloc resection

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\textbf{ABSTRACT} \\
Sacral chordoma (SC) is a malignant bone tumor with high risk of local recurrence (LR) even after en bloc resection, generally in the first 10 years after resection. We report two cases of late LR, at 17 and 19 years. Two male patients, aged 45 and 53 years, presented with large SC needing a combined approach for en bloc resection. Surgical margins were safe for the first patient and borderline for the second. The patients had yearly follow-up. The first patient developed LR on the posterior wall of the right acetabulum and the second developed LR in the right sacro-iliac notch, at 17 and 19 years, respectively. These two cases of very late LR of SC advocate for yearly screening of patients even more than 20 years after resection.
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\textit{Level of evidence:} IV (case report).

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1. Introduction

Sacral chordoma (SC) is a malignant bone tumor with a high risk of local recurrence (LR)\textsuperscript{[1–3]}. En bloc resection is the only possibility to attempt to eradicate the tumor and to reduce LR risk\textsuperscript{[4–7]}. Despite progress in adjuvant treatments, no study has shown non-operative therapy to be able to eradicate the tumor\textsuperscript{[8,9]}. SC is a slow growing tumor, and LR can occur and manifest itself numerous years after surgery. There are few long-term studies on the treatment of SC that have had more than 20 patients and no study reported series with more than 2 years’ follow-up years or LR detected more than 15 years after resection\textsuperscript{[10]}. Only Zukerberg and Young\textsuperscript{[11]} described a case of later chordoma metastasis to the ovary more than 30 years after primary resection.

We report two cases of late LR of SC at 17 and 19 years after combined en bloc resection\textsuperscript{[4]}. At the time of writing, at a mean follow-up of 20.5 months after LR resection, neither patient had relapsed or died.

2. Case reports

2.1. Case 1

A 45 year-old male presented SC revealed by low back pain and sphincter dysfunction without sensorimotor deficit, and confirmed in November 1992 by CT-guided biopsy. Extension assessment on injected pelvic CT scan revealed a 120 × 110 mm SC invading both sacroiliac joints as far as the L5–S1 disc (Fig. 1). There was no invasion of gluteus maximus muscles or the skin and no pulmonary metastasis on chest CT. The SC did not extend beyond the sciatic notches.

In December 1992, a sacrococcygectomy without neoadjuvant or adjuvant treatment was performed, including both sacroiliac joints and the lumbar-sacral junction, on combined approaches. The dural sac was ligated at L5–S1. To stabilize the pelvis, lumbo-sacral osteosynthesis was associated, without bone graft in view of the high risk of surgical-site infection and the possibility of subsequent adjuvant radiation therapy (RT) that could impair a bone graft. Histological analysis reported a lobulated non-necrotic SC with wide surgical margins without mitosis.

Two weeks after the procedure, the patient presented intestinal occlusion needing surgery, due to an intestinal loop blocked in the sciatic notch. Postoperative course also included disassembly of the osteosynthesis, due to the absence of primary bone graft, requiring two tibial-bone autografts and change of instrumentation. Bone fusion was finally obtained. The patient suffered from severe bladder and bowel dysfunction needing daily intermittent self-catheterization and preventing return to work. Subsequent

Abbreviations: RT, radiation therapy; CT, computed tomography; LR, local recurrence; MRI, magnetic resonance imaging; SC, sacral chordoma; STIR, short-tau inversion recovery.

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Fig. 1. Case 1. A. Sagittal T2-weighted pelvic magnetic resonance imaging showing 120 × 110-mm sacral chordoma (SC). B. Radiograph of the tumor after resection showing invasion up to the lumbosacral junction.

Fig. 2. Case 1. (A) Axial T1 and (B) coronal T2 short-tau inversion recovery (STIR)-weighted images illustrating local recurrence (LR) 19 years after resection. LR invades the right hip and posterior acetabular wall.

Fig. 3. Case 1. A. Postoperative anteroposterior pelvic radiograph after resection of the local recurrence (LR). Right total hip replacement was associated to ring reinforcement for the acetabular reconstruction. Lumbo-iliac instrumentation remained from the first procedure because of total resection of both sacroiliac joints. B. Surgical specimen. LR on posterior wall of the acetabulum.
standard follow-up was performed every 4 months during the first year, every 6 months during the next two years, then annually. Each follow-up consultation included neurological and scar assessment and radiology on contrast-enhanced pelvic MRI.

During the 19th year of disease-free survival in January 2011, the patient reported a recent right hip pain. Pelvic MRI and CT-guided biopsy confirmed LR in the posterior acetabular wall (Fig. 2), without mitosis on histology: extension assessment was negative. Using a Kocher-Langenbeck approach, en bloc resection of the hip and posterior acetabular wall was performed for total hip replacement with acetabular reconstruction by ring reinforcement and bone allograft (Fig. 3). Resection was transarticular, as the tumor was not invading the hip joint. The polyethylene acetabular component was cemented and the femoral stem with an alumina femoral head was impacted into the femoral shaft. Despite healthy margins, adjuvant RT was performed.

2.2. Case 2

A 53 year-old male presented with large non-metastatic SC revealed by pelvic pain and erectile dysfunction. In January 1997, histological diagnosis was established by surgical biopsy, which found typical non-necrotic SC without mitosis. MRI found a 130 × 120 × 90 mm SC with a superior limit invading the S2 vertebral body without invasion of the skin or sciatic notch (Fig. 4).

En bloc sacroccocygectomy was performed through both S2 sacral foramina and the dural sac was ligated at S1–S2, without neoadjuvant RT. Immediately postoperative course showed intestinal occlusion, managed by nasogastric aspiration. Surgical margins were borderline (<1 mm) [12] on the anterior and posterior side of the tumor and wide on the superior side.

Postoperative RT was performed, with a cumulative dose of 50 Gy. Because of the RT and partial sacroiliac joint resection, the patient developed severe aseptic left sacroiliitis. Intra-articular corticosteroid infiltration showed moderate efficacy, but the patient still walked with an antalgic limp. The patient showed the same bladder and bowel dysfunctions as patient 1, and was followed up clinically and radiologically in the same manner. At the time of writing, he was still unable to return to work.

During the 17th year of disease-free survival in March 2014, the patient described an unusual sciatic pain. Pelvic MRI confirmed the presence of a 27 × 16-mm lesion in the right sciatic notch behind the posterior column of the right acetabulum (Fig. 5). Ultrasound-guided biopsy confirmed LR. After neoadjuvant RT, the cancerous nodule was removed via a Kocher-Langenbeck approach with neurolysis of the right sciatic nerve. Surgical margins were intralesional as the nodule was in contact with the sciatic nerve, which was spared so as to conserve motor function. Histological analysis confirmed diagnosis of LR without mitosis or region of dedifferentiation but with a large area of necrosis.

3. Discussion

The occurrence of LR of SC is of poor prognosis, significantly reducing overall survival [3,13,14]. No case series of operated SC reported intervals greater than 15 years between surgical
resection and LR. Moojen et al. [15], in a retrospective study of 15 SCs, reported LR at a maximum 11 years, at a mean 7 years’ follow-up (range: 4–19 years). In the present study, LR at 17 and 19 years shows that retrospective studies have certainly underestimated LR, and that patients need regular follow-up to detect local relapse as early as possible even 20 years after primary surgery.

The two present cases had some similarities: invasion of the cephalic sacrum (above S2), and resection of a huge tumor. SC size and proximal invasion of the sacrum are controversial risk factors for LR [13,16]. For Fuchs et al. [6], resection level and tumor volume did not affect recurrence-free survival. According to Dhawale et al. [3], cephalic extension may impair the recurrence-free survival. The only recognized risk factor influencing LR is the quality of surgical margins [5,6,13,17], although other risk factors probably exist: tumor size and resection level seem to be potential factors. Insufficient sample size due to the infrequency of SC and short follow-up doubtless lead to underestimation of the rate of LR and may explain the inability to identify other risk factors.

Regarding case 2, although adjuvant RT did not prevent LR, it may delay onset. The interest of RT remains debated, as the majority of case series did not show any major impact on survival [4]. Nevertheless, a few authors, such as Moojen et al. [15], reported that patients who received immediate RT after resection, even with positive margins, showed significantly longer continuous disease-free survival than patients with wide margin resection without postoperative RT. Chen et al. [9] reported interesting results using high-dose proton/photon radiotherapy on inoperable SC, illustrating the ability of RT to achieve local control although not eradicating the tumor. The drawback of photon-beam RT is local impact on surrounding tissue. At present, RT is adapted to tumor control but the real ability of adjuvant RT to improve disease-free and overall survival remains unknown [6]. Systematic adjuvant RT could be useful even with wide surgical margins [18]; probably, despite wide margins, residual tumor cells may be left at the resection site.

Proton-beam RT could be an alternative to photon-beam RT [19,20]. Using heavy-particle-beam radiation, proton-beam RT, like carbon ion therapy, allows higher doses (> 60 Gy) to be delivered to radioreistant tumor tissue while limiting toxic impact on surrounding tissue [21].

The present study thus emphasizes that surveillance is mandatory even 20 years after primary surgery, even in case of safe margins. Thanks to rapid diagnosis, LR size is limited, allowing less invasive surgery than in the first procedure.

Disclosure of interest

The authors declare that they have no competing interest.

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