ORIGINAL STUDY

Subtotal Sacrectomy Followed by Maximally Tolerated Three-Dimensional Conformal Radiation for High-Level Chordomas with Neural Integrity: Technique and Outcome

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Abstract

Background data: Chordomas are the most common sacral tumors, usually treated by wide resections with free margins. Radical resections are likely followed by significant neurological deficits that may not be accepted for some patients who present with preserved neural functions.

Purpose: To present a local institutional experience in the technique and outcome of subtotal sacrectomy followed by maximally tolerated radiation therapy in a cohort of patients having high sacral (S1–S2) chordomas who presented with intact neurological functions and were not previously treated.

Study design: This is a retrospective case series.

Patients and methods: This is a retrospective case series conducted over 7 years, including 14 consecutive patients with high-level sacral chordomas who refused radical resection and underwent subtotal sacrectomy aiming to preserve as many neural structures as possible, followed by three-dimensional conformal radiotherapy given at a maximum dose tolerated by the surrounding viscera. The surgical resection, perioperative management, including radiotherapy techniques, and their functional and oncologic outcomes were reviewed.

Results: The mean age was 44.6 ± 4.7 years, and 10 (71.4%) cases were males. Patients complained of sacral pain for 2–9 months before diagnosis, and all were continent with no gross sensorimotor deficits. Resections were performed through posterior-only approaches using T-shaped or inverted U-shaped incisions. Surgeries went uneventful, with two incisions indicated for further plastic management. Negative margins were achieved in one case. Bladder/bowel functions were lost postoperatively in 28.6% of cases. Ten cases had disease progression at a mean duration of 62.4 ± 27.9 months. The median survival was 99 months. The 5-year absolute survival rate was 78.6% and the 5-year disease progression was 42.9%. The overall survival in this study was found to be 35.7%, with a median follow-up of 6.3 years.

Conclusion: Despite the promising outcomes at 5 years, the used strategy was associated with sphincteric dysfunctions in a substantial proportion of patients and failed to achieve an adequate overall survival. We recommend radical resections for young patients to improve survival. Subtotal sacrectomy followed by three-dimensional conformal radiation can be a valid alternative for older patients with shorter life expectancy (2021ESJ250).

Keywords: Three-dimensional conformal, Chordoma, Neural-preserving, Outcome, Sacrectomy

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Introduction

Chordoma is a locally malignant, slowly growing tumor of the midline axial skeleton arising from remnants of the notochord and representing 1–4% of primary bone tumors [1]. However, it is the most frequently encountered primary malignant neoplasm of the sacral region [2], which usually presents late owing to its slow course and subtle symptoms [3]. Such tumors used to be considered not susceptible to radiotherapy or chemotherapy [4], with en bloc resection including free margins regarded as the standard care of management to prevent recurrence and tumor progression [5]. However, in high sacral chordomas reaching S2 or higher, total sacrectomy with free margins can result in 100% bowel/bladder dysfunction and lost plantar flexion, necessitating ankle orthosis in up to 40% of cases [6]. In circumstances when the likely postoperative deficits are not accepted by the patient, subtotal sacrectomy may be an alternative surgical strategy with the drawback of a higher possibility of recurrence [7].

The evidence of radiotherapy has been established for sacral chordoma in the cases of marginal (R1) and intralesional (R2) surgical resection as a complementary treatment [8]. Confirmatory studies and analyses revealed that there is no survival benefit of adding external beam radiation therapy to negative surgical margin, while improved survival rates were confirmed by adding high doses of RT more than 65 Gy and proton beam therapy [9,10] and using carbon ion therapy [11] and intensity-modulated radiation therapy [12].

Many reports in the literature discussing chordomas have represented heterogeneous cohorts of patients, including primary and secondary lesions, affecting different spinal locations, treated via diverse modalities, various surgical techniques, and unequal radiation doses, making outcome measures uninterpretable [4,13–23].

This study aimed to present local experience in subtotal sacrectomy provided to patients with sacral chordoma who presented with intact neurological functions and did not accept radical resections for the sake of neurological integrity. We also present functional and oncologic outcomes over the follow-up period.

Patients and methods

This is a retrospective case series including consecutive high-level sacral chordoma cases that underwent subtotal sacrectomy followed by three-dimensional (3D) conformal radiotherapy over a period of ~7 years.

Patient selection and preoperative assessment

The medical records of patients admitted between November 2008 and December 2015 into the Departments of Neurosurgery and Surgical Oncology with the diagnosis of sacroccygeal lesions were revised on June 2021 (date of data collection). All histopathologically confirmed chordomas presented at high levels reaching S1–S2 segments were included as potential candidates. This histologic diagnosis was established postoperatively in most cases because only some surgeons performed a preoperative fine-needle trucut biopsy. Cases that received initial management (surgery or irradiation) elsewhere were excluded. Moreover, cases approved to perform radical (R0) resections were excluded from the study. Exclusion criteria implemented for final eligibility are presented in Table 1.

All selected cases had signed ‘informed refusal’ forms declining radical sacrectomy because of the inevitable postoperative sensorimotor/sphincteric deficits. Patients also signed informed consent accepting the likely recurrence and possible need for redo surgeries. Records of neurological examination, laboratory investigations, and radiologic assessments, including metastatic workup, were reviewed. All cases were treated with rectal enemas and surgical areas were shaved, followed by a shower using povidone-iodine shampoo in the night before surgery. Intravenous antibiotics were administered within 60 min from the surgical incision. An indwelling urinary catheter was inserted routinely for continuous bladder drainage after induction of anesthesia.

Table 1. Eligibility criteria for the study group.

<table>
<thead>
<tr>
<th>Cases</th>
<th>Numbers</th>
</tr>
</thead>
<tbody>
<tr>
<td>Total cases with sacral lesions presented over 9 years</td>
<td>84</td>
</tr>
<tr>
<td>Confirmed sacral chordomas not previously treated elsewhere</td>
<td>41</td>
</tr>
<tr>
<td>Excluded cases</td>
<td></td>
</tr>
<tr>
<td>Cases of low chordomas (below S2)</td>
<td>15</td>
</tr>
<tr>
<td>Cases presented with sensorimotor deficits</td>
<td>5</td>
</tr>
<tr>
<td>Cases considered inoperable (with distant metastasis)</td>
<td>2</td>
</tr>
<tr>
<td>Cases refused any form of surgical intervention</td>
<td>2</td>
</tr>
<tr>
<td>Cases accepted radical sacrectomy despite predicted neurological and sphincteric affection</td>
<td>3</td>
</tr>
<tr>
<td>Eligible cases with neural-preserving sacrectomy for high sacral chordomas</td>
<td>14</td>
</tr>
</tbody>
</table>
**Preoperative planning**

With the help of the neuroradiologist, the pelvic computed tomography (CT) scan with 3D reconstruction views and the lumbosacral MRI were reviewed. Although the tumor was not visible in 3D CT views, these images were important to check for the sacroiliac joint integrity and the details of the bony surface of the sacrum. Bone osteotomy lines in coronal and sagittal planes were designed and correlated to the bony surface details and surrounding landmarks to guide intraoperative bone cutting. According to right/left and anterior/posterior deferential extensions of the tumor, the obliquity of coronal and sagittal cuts was determined, respectively. All osteotomy lines were intended not to extend proximal to S1/S2 interface to avoid injuring proximal sacral roots contributing to ankle movements, sensations, and sphincteric functions. Tumor portions extending proximal to osteotomy lines were evaluated intraoperatively for possible debulking or marginal resections.

**Surgical technique**

All cases received hypotensive general anesthesia for better hemostasis and underwent an operation in a prone position with flexed hips and knees. Intraoperative fluoroscopy was not used in any of the cases. The skin was prepared and draped from the level of the last rib to the middle of the intergluteal cleft. Two types of surgical incisions were performed by operating surgeons as per revised records (Fig. 1). A wide inverted U-shaped skin incision was performed in six cases with its two limbs extending into the middle of each buttock down to the coccyx level, while its dome is located just below the intercrestal line. A full-thickness skin flap based distally was then raised with the help of monopolar electrocautery to expose the region of the sacrum and sacroiliac joints. In eight cases (with preoperative Trucut biopsy), T-shaped skin incisions were performed with the vertical limb extending from above the intergluteal cleft in the midline up to the L5/S1 level and the transverse limb extending in a curvilinear manner between the upper margins of both sacroiliac joints. The vertical limb of the incision was planned to include the tract of trucut biopsy allowing for tract excision during the operation to avoid the risk of tract seeding by the tumor.

After elevating the skin and subcutaneous tissues, the lumbosacral facia was elevated bilaterally from its spinous attachments and divided transversely on each side to expose the sacrum and lamina of L5. Subsequent steps were similar in all cases, starting with bilateral identification and division of the medial attachments of the gluteus maximus and the sacrotuberous ligaments with a 2-cm safety margin. The piriformis muscle was then identified and divided together with sacrospinous and sacrococcygeal ligaments to enter the retroperitoneal space bilaterally. At that point, blunt dissection using a dry gauze was started to separate the soft tissues anterior to the sacrum without breaching the tumor pseudocapsule until the anterior tissues were separated completely distally to proximally. An abdominal towel gauze was then passed anteriorly to the sacrum to protect retroperitoneal structures.

Laminectomy L5 and the removal of posterior sacral elements were then performed to expose the thecal sac down to the level of S3 roots. The second sacral roots were preserved in all cases. However, S3 roots were assessed carefully for functionality and extent of being involved by the tumor. Unilateral S3 resection was decided in 10 patients (eight on the
right and two on the left side), whereas bilateral resection was performed in four cases. The unequal extension of the tumor bilaterally allowed for the preservation of single S3 roots in most patients. All sacrificed roots were extensively infiltrated and seemed nonfunctioning for the operating surgeon. The thecal sac was then ligated and divided below the level of neural preservation to expose the posterior sacral surface.

Any tumor extension to the ala of the sacrum or parts of sacroiliac joints or ilium was removed using a rongeur, leaving healthy bone without violating more than one-third of the sacroiliac joints. Sacral osteotomy was carried out through normal bone proximal to the upper extension of the tumor with the abdominal towel keeping the retroperitoneal structures safe. During osteotomy, proximal and distal to-and-fro movements of the hand carrying the osteotome helped the surgeon estimate the completion of bone cutting until the sacrum was fully mobilized. The anococcygeal ligament was then divided to free the distal end, which was elevated posteriorly and upward using Allis tissue forceps to expose the underlying space. Careful separation of the sacral piece was done by separating any residual attachments under direct visual control to finalize tumor extraction (Fig. 2).

None of our cases were indicated for spinal or spinopelvic stabilization. The posterior pelvic repair was performed using polypropylene mesh to support the rectum and avoid a future sacral hernia, followed by tight closure of the overlying facia if not invaded by the tumor (Fig. 3).

Protocol of postoperative management

Patients passed their postoperative stay in a regular ward with drains in place for the first 48–72 h. Urinary catheters were removed the next postoperative day to evaluate spontaneous voiding. Catheters were reinserted again in cases unable to pass urine. Only the prone and lateral positions were allowed by operating surgeons for the first few days to a week. Daily dressings were done once for dry wounds and at least twice when significant soaking was noticed. During the postoperative period, prophylaxis against deep venous thrombosis was conducted, including good hydration and mechanical and pharmacological measures. A pneumatic intermittent compression device was used when available (nine cases) from day 1 after surgery. Alternatively, above-knee compression, elastic stockings (five cases) were applied with regular manual massages to calves thrice daily. Enoxaparin sodium 40 mg subcutaneously once per day was started 12 h after surgery in all patients until achieving full mobilization. On the 10th to 12th day postoperatively, sutures were removed in dry healed wounds. Ambulation was allowed when tolerated with the aid of a four-legged adjustable walking frame. Cases with wound issues were managed in combination with plastic surgery.

Radiotherapy technique

All resected cases were referred to adjunctive radiotherapy once the healing of their wounds was complete. Planning of all patients was done using a CT simulator with CT cuts at 5-mm intervals to the whole pelvis after ordinary setup measures of the patients. Eye fusion was used with MRI pelvis slides to ensure the exact delineation of the gross target volume (GTV) of any residual soft tissue at the operative bed. Then, the clinical target volume, defined as the preoperative tumor size, was then delineated, as shown in the preoperative CT films. Finally, the planned target volume is generated by

Fig. 2. (A) The use of abdominal towel gauze for gentle gradual separation of the tumor. (B) Final appearance of the field following sacrectomy with the ligated thecal sac just distal to last preserved roots and posterior wall of the rectum. (C) The excised tumor after maximum possible safe resection.
adding a 10-mm homogenous margin to the clinical target volume.

The main limiting factors from increasing the dose beyond 60 Gy were the absence of the intensity-modulated radiation therapy technique in our institute and the dose constraints in surrounding organs at risk, namely, the bladder and rectum. The tolerance dose for the bladder and rectum is 60 Gy and for the ‘intestines’ is 45 Gy. All of the cases were positioned prone and treated with a three-field technique; planned target volume received a 60-Gy total dose, whereas the GTV may reach up to $105-109\%$ of the dose (i.e. $63-65$ Gy); this dose was our limit, as we could not reach beyond 60 Gy to the organs at risk.

Follow-up and outcome assessment

Follow-up schedules were given to the patients, including regular and emergency visits. Regular visits to the outpatient clinic were 2 weeks after discharge, every 3 months during the first year, every 6 months during the second year, and then annually afterward. Emergency visits were indicated in case of swelling, local pain, or neurological deficits. Data collected during follow-up visits included local, neurological, and radiographic examinations of the chest and pelvis. A detailed CT scan was performed for all cases postoperatively and in selected situations during follow-up (Fig. 4). MRI was performed postoperatively, then every 6 months during the first 2 years, and annually afterward, in addition to any time disease progression was suspected by the attending physician (Fig. 4). At the time of data collection, telephone calls for selected patients (i.e. dropouts) were performed with the preparation of a visit to the clinic when possible to complete follow-up data. The duration of follow-up was estimated as the time between diagnosis and the end of follow-up, which was either the time of data collection or the time of death. Death in this study was either due to disease progression (died of disease) or unrelated events (died from other causes). Functional outcome was evaluated at the end of the second year of follow-up and the end of follow-up according to Karnofsky performance status [24]. Oncologic outcome was determined by median overall survival (OS), 5-year absolute survival rate, progression-free survival (PFS) rate at 2 years, and 5-year disease progression. The median OS was calculated using the Kaplan–Meier method and was defined as the length of time from the date of diagnosis that half of the patients with chordoma were still alive (Fig. 5). PFS rate at 2 years was defined as the percentage of patients at the end of the second year postoperatively without signs of tumor progression. Five-year absolute survival rate was the percentage of patients alive 5 years from diagnosis, whereas 5-year disease progression was the percentage of patients with confirmed local progression at 5 years from diagnosis. Tumor progression was estimated by revising the documented MRI data immediately after surgery and during the follow-up period. Residual tumors in early postoperative MRIs were classified as either sizable (with reported longest diameter in MRI ≥10 mm) or
nonsizable (longest diameter <10 mm or not detected). Progression for sizable residuals was defined as an estimated increase of more than or equal to 20% (from MRI-reported dimensions) in the longest diameter of the lesion or in the sum of the longest diameters of two or more lesions during the follow-up. Progression of nonsizable lesions was defined as an MRI-confirmed appearance of one or more new lesions and/or unequivocal progression of a nonsizable preexisting lesion. These parameters were
guided by the RECIST criteria introduced in 2000 to assess changes in tumor burden in response to therapy [25]. Lesions that became invisible in follow-up MRI were considered complete remission, whereas those that remained detectable but did not fulfill criteria of progression were considered stable local residuals.

**Results**

**Demographics and clinical characteristics**

The current study included 14 cases of high sacral chordomas reaching S2 or higher that were continent with no gross sensorimotor deficits and underwent subtotal sacrectomy followed by maximally tolerated local radiotherapy. Their ages ranged from 38 to 51 years, with a mean of 44.6 ± 4.7 years. Ten (71.4%) cases were males, with a male to female ratio of 5 : 2. None of the selected patients had sensorimotor deficits preoperatively; however, six (43%) patients had some sphincteric disturbances, though they remained stool/urine continent. All cases presented with sacral pain lasting from 2 to 9 months (median of 4.4 months and interquartile range of 4) before diagnosis. Clinical presentations, patient characteristics, and tumor characteristics are presented in Table 2. In the six cases with some sphincteric disturbances, the lesion affected lower S1, whereas in other cases, lesions reached up to S1 or S2.

**Operative results and complications**

The osteotomy level was done at nearly S1–S2 junction in all cases after exposure of the spinal canal widely and preserving as many as possible of the intact sacral roots. The osteotomy lines were designed as oblique as needed to accommodate the asymmetrical tumor extension bilaterally (Fig. 5). All surgical procedures were uneventful, with no reported visceral or vascular injuries. The mean blood loss was about 1441 ± 410 ml, as calculated by

![Fig. 5. A case of S1-coccyx chordoma. (A) Sagittal MRI showing proximal, distal, anterior, and posterior extents of the tumor. (B) Axial MRI shows the differential extent of the tumor to the right side. (C) The excised sacral specimen showing an asymmetrical shape of the specimen. (D) Postoperative 3D pelvic CT, showing the oblique osteotomy line, which was more proximal on the right to accommodate the tumor extension. CT, computed tomography.](image-url)
summing up the blood volume collected from the reservoir of the suction device and that estimated in soaked surgical gauzes and towels.

Two patients developed wound dehiscence during the postoperative hospital stay with no cerebrospinal fluid leak and were referred to plastic care for flap procedures. One of them had a T-shaped skin incision with ischemic gapped corners of the wound, whereas the other had a U-shaped incision and developed wound infection and delayed healing. The latter was diabetic for more than 10 years. The two patients underwent successful management by plastic surgeons with complete healing within 3–4 weeks. Other patients were discharged with clean, dry wounds 1–2 days following suture removal. Patients received radiation therapy 4–6 weeks after complete surgical wound healing.

There were no reported venous thromboembolic events during the postoperative hospital stay or in follow-up visits. The results of pathological examination of surgical margins, bladder/bowel control after surgery, and the 2-year functional and oncologic outcomes are presented in Table 3. All cases were of the chondroid chordoma subtype. Negative margins of excised specimens were histopathologically documented in only one (7%) case. In 71.4% of cases, bladder and bowel control could be preserved and remained so over 2 years of follow-up.

**Outcome results**

By the end of the second year of follow-up, most cases (about 71%) were able to carry on normal activity and work with some signs or symptoms of

<table>
<thead>
<tr>
<th>Cases</th>
<th>Margins</th>
<th>Bladder and bowel functions</th>
<th>KPS</th>
<th>Disease status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Positive</td>
<td>Normal</td>
<td>100</td>
<td>Stable local residual</td>
</tr>
<tr>
<td>2</td>
<td>Positive</td>
<td>Normal</td>
<td>80</td>
<td>Complete remission</td>
</tr>
<tr>
<td>3</td>
<td>Positive</td>
<td>Normal</td>
<td>90</td>
<td>Stable local residual</td>
</tr>
<tr>
<td>4</td>
<td>Positive</td>
<td>Normal</td>
<td>90</td>
<td>Stable local residual</td>
</tr>
<tr>
<td>5</td>
<td>Positive</td>
<td>Intermittent catheterization, digital rectal evacuation</td>
<td>70</td>
<td>Stable local residual</td>
</tr>
<tr>
<td>6</td>
<td>Positive</td>
<td>Normal</td>
<td>90</td>
<td>Stable local residual</td>
</tr>
<tr>
<td>7</td>
<td>Positive</td>
<td>Normal</td>
<td>100</td>
<td>Complete remission</td>
</tr>
<tr>
<td>8</td>
<td>Positive</td>
<td>Normal</td>
<td>100</td>
<td>Complete remission</td>
</tr>
<tr>
<td>9</td>
<td>Positive</td>
<td>Intermittent catheterization, digital rectal evacuation</td>
<td>70</td>
<td>Stable local residual</td>
</tr>
<tr>
<td>10</td>
<td>Positive</td>
<td>Normal</td>
<td>90</td>
<td>Stable local residual</td>
</tr>
<tr>
<td>11</td>
<td>Positive</td>
<td>Intermittent catheterization, digital rectal evacuation</td>
<td>60</td>
<td>Local progression</td>
</tr>
<tr>
<td>12</td>
<td>Negative</td>
<td>Normal</td>
<td>90</td>
<td>Complete remission</td>
</tr>
<tr>
<td>13</td>
<td>Positive</td>
<td>Normal</td>
<td>80</td>
<td>Stable local residual</td>
</tr>
<tr>
<td>14</td>
<td>Positive</td>
<td>Intermittent catheterization, repeated enemas</td>
<td>60</td>
<td>Local progression</td>
</tr>
</tbody>
</table>

KPS, Karnofsky performance scale.
the disease, whereas nearly 29% of cases were unable to work but could live independently at home and care for most of their personal needs. The median Karnofsky performance status was 90 (60–100) and 70 (0–100) at 2 and 5 years, respectively. The mean follow-up duration was 77.2 ± 23.1 months.

Ten (71.4%) cases had disease progression at a mean duration of 62.4 ± 27.9 months. The time of recurrence and final status at the end of follow-up are presented in Table 4. Two (14%) cases had a confirmed local disease progression before 2 years, and two cases developed metastasis with delayed progression (at 50 and 54 months) of initially stable residuals. The cases with early disease progression (<2 years) had postoperative residuals after maximal safe resections with the largest diameters of about 3.2 and 2.5 cm within the S1 vertebra, which could not be excised without sacrificing neural (S1 and S2 roots) functions. At the time of progression, these tumors were 5.3 and 4.1 cm, respectively. Both cases underwent salvage palliative management after informed consent, in the form of debulking surgery with an additional dose of 18 Gy over 10 fractions targeting the GTV ‘tumor recurrence’ with a 10-mm margin. One of these two cases died at 34 months, whereas the other survived but developed a very poor functional outcome at 5 years and died shortly after. The other eight cases with later disease progression refused further resections. Six of them were referred to palliative radiotherapy for pain control, putting into consideration the previous dose received (60 Gy) and the time gap. The other two cases decided to perform further surgical management elsewhere.

Overall, 86% of cases remained stable or underwent remission at 2 years postoperatively (2-year PFS rate, 86%). The median OS was 99 months (SE = 20.8, 95% confidence interval = 58.1–139.8) as measured by the Kaplan–Meier curve (Fig. 6). The 5-year absolute survival rate was estimated to be 78.6%. Six cases developed disease progression either locally or distally or both with/without death from the disease by 5 years (i.e. 42.9% 5-year disease progression).

Discussion

Although chordomas have been long considered a radioresistant tumor, radiotherapy was reported to be a useful modality when used adjunctively to surgery but not adequate as a freestanding therapy [26,27]. Although some authors have reported no survival benefits for postoperative radiation therapy [15], their patients received radiation doses between 25 and 60 Gy. This contrasts with our patients, who all received high doses of at least 60 Gy. Others [27], found that the addition of radiation therapy in subtotal resections can prolong the median time to recurrence from 8 months to 2.12 years. The authors have reported a PFS rate of 86% at 2 years in the current study. Similarly, some authors suggested that preoperative or postoperative irradiation permits marginal resection in high sacral chordomas allowing for neural function preservation [26].

Although the mean age and the median duration of symptoms were comparable to other recent reports [28], earlier studies have reported older ages and longer duration of symptoms [13,29]. This may be owing to the inclusion of smaller tumors at lower sacral levels rendering symptoms vaguer with late urinary symptoms. Males were almost double the females (5 : 2), which was close to other reports

<table>
<thead>
<tr>
<th>Case</th>
<th>DOF (months)</th>
<th>Recurrence/local progression (TAT/month)</th>
<th>KPS at 5 years</th>
<th>Final status</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>99</td>
<td>Positive (79)</td>
<td>70</td>
<td>DOD</td>
</tr>
<tr>
<td>2</td>
<td>102</td>
<td>Positive (92)</td>
<td>80</td>
<td>DOD</td>
</tr>
<tr>
<td>3</td>
<td>56</td>
<td>Positive (54)</td>
<td>0</td>
<td>DOD</td>
</tr>
<tr>
<td>4</td>
<td>118</td>
<td>Positive (108)</td>
<td>90</td>
<td>DOD</td>
</tr>
<tr>
<td>5</td>
<td>51</td>
<td>Positive (50)</td>
<td>0</td>
<td>DOD</td>
</tr>
<tr>
<td>6</td>
<td>63</td>
<td>Positive (57)</td>
<td>70</td>
<td>DOD</td>
</tr>
<tr>
<td>7</td>
<td>97</td>
<td>Negative, stable residual</td>
<td>100</td>
<td>LWD</td>
</tr>
<tr>
<td>8</td>
<td>91</td>
<td>Negative, complete remission</td>
<td>100</td>
<td>DOC</td>
</tr>
<tr>
<td>9</td>
<td>86</td>
<td>Positive (81)</td>
<td>70</td>
<td>LWD</td>
</tr>
<tr>
<td>10</td>
<td>77</td>
<td>Positive (85)</td>
<td>80</td>
<td>LWD</td>
</tr>
<tr>
<td>11</td>
<td>61</td>
<td>Positive (23)</td>
<td>20</td>
<td>DOD</td>
</tr>
<tr>
<td>12</td>
<td>75</td>
<td>Negative, complete remission</td>
<td>100</td>
<td>NED</td>
</tr>
<tr>
<td>13</td>
<td>71</td>
<td>Negative, stable residual</td>
<td>70</td>
<td>LWD</td>
</tr>
<tr>
<td>14</td>
<td>34</td>
<td>Positive (22)</td>
<td>0</td>
<td>DOD</td>
</tr>
</tbody>
</table>

DOC, dead from other causes; DOD, dead of disease; DOF, duration of follow-up; KPS, Karnofsky performance scale; LWD, living with disease; NED, no evidence of disease; TAT, time after treatment.
According to Wei et al. [32], the tumors in our study were of types II and III. A combined anterior–posterior resection is recommended for these types when they are big tumors extending above the S1 level, whereas posterior-only resection is preserved for smaller tumors at lower levels [32]. All of our cases were operated via a posterior-only approach using inverted U-shaped or modified T-shaped incisions. The former seemed simpler, allowing wider exposure with comparable healing, whereas the latter was more suitable in cases with preoperative biopsy allowing for excision of the track of biopsy during surgery.

All surgeries went uneventful, with acceptable blood loss compared with other studies [28,30]. All cases retained gross sensorimotor functions as S2 roots were preserved in all. However, bladder/bowel functions could be retained in only 71% of cases. Four cases (with bilateral S3 root sectioning) lost their bladder/bowel control postoperatively. Previous studies have reported the importance of S3 roots in bladder/bowel functions. Todd et al. [33] have reported that bilateral preservation of S3 roots was associated with retained bowel/bladder functions in 100 and 69% of cases, whereas unilateral preservation resulted in retained functions in 67 and 60%, respectively. Another study reported that the S3 root was primarily involved in bladder contraction with predominance for the right side [34]. The predominant concentration of B fibers within the S3 root, representing the parasympathetic nerves that control bladder functions [35], may explain preserved sphincteric functions in cases with saved S3 roots.

With improvement in radiation technology, effective higher doses can now be safely delivered to residual or inoperable sacral chordomas with minimal toxic effects on nearby tissues [17,20,23,36,37]. In a recent study, therapeutic RT was associated with a significant prolongation of the OS in the case of positive surgical margins that were found to be close to that of patients with negative margins. The authors suggested that surgical excision with planned positive margins and postoperative therapeutic RT is an acceptable strategy to reduce surgical sequelae [9]. All of the patients managed in the current study received high-dose postoperative radiation therapy with the 3D conformal technique, respecting dose constraints to surrounding tissues with no reported radiation-related complications during the follow-up period.

Most of the chordoma studies in the literature are very heterogeneous, including chordomas of different locations, using various modalities of management and studying heterogeneous cohorts of primary and secondary chordomas. Surgery alone, proton/photon/carbon ions therapy, and surgery ± preoperative/postoperative photon therapy were all reported in the literature [4,5,11,12,14,16,18,19,21,22,38–44]. On the contrary, this study included a homogenous subgroup of patients. The authors found the 5-year survival rate to be comparable to some studies [11,12,16] but far below the others [4,19]. PFS at 5 years was 57.1% among our patients, which was comparable to the results from some studies [9,22] but inferior to the results of other studies [4,45]. In an analytic study exploring the prognostic factors affecting the survival of primary chordomas, they found age more than or equal to 60 years, distant metastasis, and nonsurgical therapy to be predictors of reduced
survival [46]. In another study comparing prognostic factors for primary and secondary sacral chordomas, not receiving radiation was found to be an independent predictor for worse local PFS [45]. The relatively young age of our patients and the early presentation might have a positive effect on the 5-year outcome. However, the OS in this study was found to be 35.7%, with a median follow-up of 6.3 years, where 28.6% (4/14) were living with the disease and 7.1% (1/14) had no evidence of the disease. This survival rate is considered significantly shorter compared with several studies reporting 52–84% survival at 10 years [5,22,39–41]. This was because 21.4% of patients died before 5 years, whereas 42.9% died between 5 and 10 years with a resulting loss of survival benefit after 5 years.

The authors believe that these survival results are inferior and not adequate for young patients who are otherwise healthy with a long expectancy of life. Although subtotal sacrectomy followed by 3D conformal radiation saved motor and sensory functions of operated patients, this strategy resulted in 28.6% loss of bladder/bowel functions owing to inadvertent root injuries, making it not a completely safe technique. For these reasons, it is believed that wide (R0) surgical resection with a secondary goal of neural preservation in young patients who presented to medical care with high sacral chordomas and neural integrity is a more appropriate management in favor of longer survival. Preoperative trucut biopsy for establishing the diagnosis should be implemented in all patients as it was proved to be a safe procedure with no additional risk of seeding of the tract when adequately excised through appropriately planned incisions. In addition, preoperative tissue diagnosis helps educate the patient about the nature of the disease and the likely outcome following marginal and intrasional resections, including multiple recurrences, repeated surgeries, and ultimately loss of function and thus can reduce resections of radical surgeries. On the contrary, older patients with comorbidities and shorter life expectancy may gain benefit from the relatively good 5-year survival of the used strategy having the advantage of neural preservation.

One of the limitations of the current study is the limited sample size, making it exposed to type II error. Second is the retrospective nature of the study. However, the selection of a subgroup of a relatively rare pathology to explore the outcome of their management is a hard task to perform prospectively. Third, no control group was available to compare the oncologic outcomes of radical free margin sacral resection with conservative high sacrectomy combined with postoperative high-dose radiation therapy. Finally, the lack of more advanced radiotherapy techniques, especially intensity-modulated radiation therapy or stereotactic RT, would enable delivering doses beyond 60 Gy to the GTV that may reach up to 75 Gy without increasing toxicity and give us more local control.

**Conclusion**

Despite the promising outcomes at 5 years, the used strategy was associated with sphincteric dysfunctions in a substantial proportion of patients and failed to achieve an adequate OS. We recommend radical resections for young patients to improve survival. Subtotal sacrectomy followed by 3D conformal radiation can be a valid alternative for older cases with shorter life expectancy.

**Ethical statements**

This study is done following the the World Medical Association Declaration of Helsinki, as a statement for ethical principles for medical research. All patients consented to the surgical intervention along with research consent to publish the medical data. The study was approved by our IRB.

**Conflict of interest**

There are no conflicts of interest.

**Abbreviations list**

- **CTV** Clinical target volume
- **DOC** Died from other causes
- **DOD** Died of disease
- **DOF** Duration of follow-up
- **EBRT** External beam radiation therapy
- **GTV** Gross target volume
- **IMRT** Intensity-modulated radiation therapy
- **KPS** Karnofsky performance status
- **LWD** Living with disease
- **OAR** Organs at risk
- **OS** Overall survival
- **PFS** Progression-free survival
- **PTV** Planned target volume
- **R0** Free-marginal (radical)
- **R1** Marginal
- **R2** Intraläsional

**References**


