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Original Article

Sacrococcygeal chordoma-illustrative cases and our experience

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Abstract: Chordoma is an uncommon malignant bone tumour of low metastatic potential, the commonest site of which being sacrum. We intend to report two cases of giant sacrococcygeal chordoma managed surgically. The first patient presented with natal cleft swelling since past 3 years which on examination had a size of 12*10*14 and was mildly tender, non reducible, non pulsatile and non fluctuant. The swelling had a variegated surface and extended from sacral region till 2 cm above anal verge. The second patient presented with low backache with radiation to the left lower limbs along with numbness in posterior aspect of left thigh. Physical examination in the second patient was near similar to that in first case except the decreased perianal sensation with otherwise normal neurology in the second patient. The imaging and histopathology was consistent with sacroccocygeal chordoma in both cases. Both patients underwent wide margin resection with preservation of both S2 and right S3 roots. Effective management of sacrococcygeal chordoma requires early diagnosis, accurate preoperative staging, definitive and adequate surgical resection with proved tumour-free cut margins while in those declining surgery, radiotherapy can be considered as an alternative.

Keywords: Sacrococcygeal chordoma, wide rsection

Introduction

Chordoma is a relatively uncommon tumour that accounts for 1% to 4% of all malignant bone tumours and has an annual incidence of 0.1 per 100,000 people [1, 2]. Sacrum is the most commonly affected site. Other common sites are the base of the skull and vertebral bodies. Giant sacrococcygeal chordoma has rarely been reported in Indian literature with none in a patient with PPRP. The biological behaviour of chordoma is characterized by a slow aggressive local growth with a low tendency in metastasizing to distant sites such as lungs, bones, soft tissue, lymph nodes, liver, and skin. Although it is considered to be of low metastatic potential, up to 40-60% of a patient are, however, reported to develop distant metastases throughout their disease [3, 4]. Usual clinical feature include low back pain not responding to non-steroidal antinflammatory medications, typically worsening on sitting, which may be accompanied with urinary and fecal complaints. Histopathology remains the diagnostic standard with usual management involving surgical excision, although conservative management has been described for patients declining surgery [5, 6]. We present our experience with 2 cases of surgically treated giant sacrococcygeal chordomas.

Methodology

The study was approved by an ethical committee bearing approval n umber - AIIMS ORTHO/2022/023. Prior informed consent was obtained from all the patients.

Case 1

A 46-year-old male presented came to OPD with chief complaints of swelling over the sacrococcygeal region for the past 8 months and pain and discomfort in sitting. The pain gradually increased and swelling significantly increased in the last 3 months as difficulty in passing urine and constipation for 3 months. Physical examination showed a giant mildly tender, non-reducible, non-pulsatile, non-fluctuant

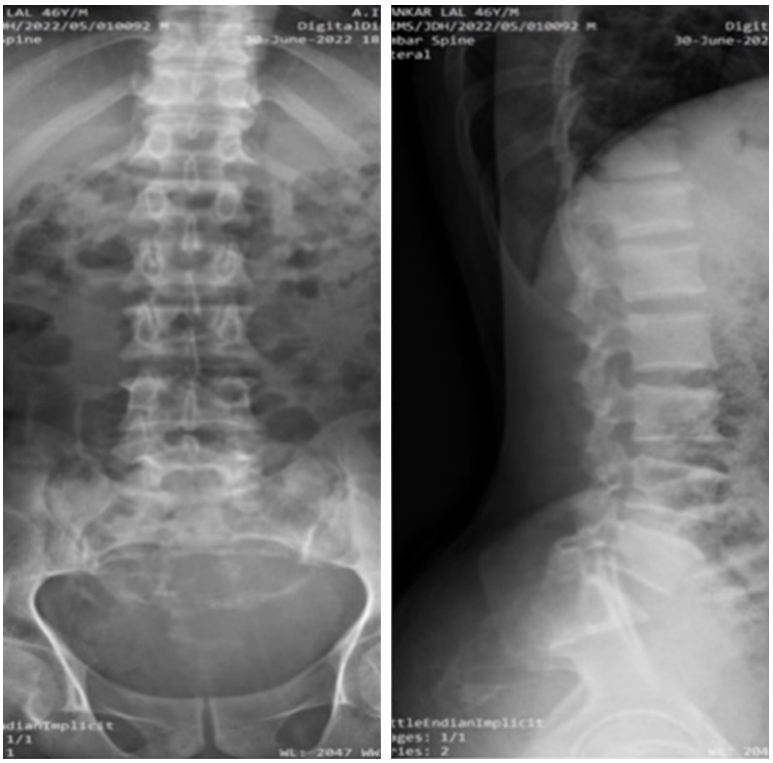

Figure 1. Anteroposterior and lateral X-ray of Lumbosacral spine showing lytic lesion in the sacrum.

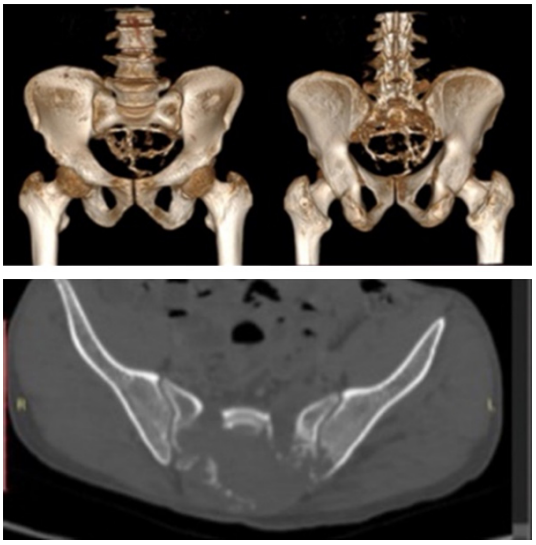

Figure 2. CT scan of Lumbosacral spine showing near-complete destruction of sacrum and involvement of left iliac bone.

swelling measuring 11*13*10 cm. In MR, the lesion appeared central lytic destructive mass (~8×14×13 cm) is noted infiltrating the entire sacrum & coccyx, and extending epidural space, bilateral neural foramina, left sacroiliac joint & left iliac PSIS (**Figures 1**, **2**). Anteriorly it is extending into presacral space as a cystic component and infiltrates the left pyriformis. It is abutting & displacing the right pyriformis, urinary bladder and proximal rectum (& recto-sigmoid Junction) anteriorly. Posteriorly it extends into paravertebral sacral muscles with mild post-coccygeal extension. The sacrococcygeal tumour was considered to be a chordoma based on the imaging results and preoperative histopathopatholgy. Tumour cells displayed, strong diffuse immunopositivity for EMA and CK and focal weak positivity for Brachyury.

The patient underwent surgery in form of wide excision and lumbopelvic fixation with 6 pedicle screws and pelvic reconstruction with 14 hole recon plate and left fibular strut graft (**Figure 3A-C**). At 18 days following surgery, the patient presented with a superficial infection over the suture site due to soakage resulting from dead space underneath for which local debridement was done along with empirical antibiotics. successfully. At 9 months follow-up, there was no evidence of recurrence of tumor or infection.

Case 2

A 19-year-old female came to OPD with the chief complaint of complaints of low backache with radiation to the left lower limb over the last 4 months and numbness in the posterior aspect of the left thigh for the last 2 months. The pain aggravated on sitting and on rising from the sitting position. Physical examination showed a giant mildly tender, non-reducible, non-pulsatile, non-fluctuant swelling measuring 10*8*12 cm with variegated surface and heterogenous consistency extending from the sacral region till 3 cm above the anal opening. The patient had decreased perianal sensation with otherwise normal neurology. In MR, a 40×60×40 mm lesion was seen involving the S3 vertebra, part of S2 & S4 vertebrae, extend-

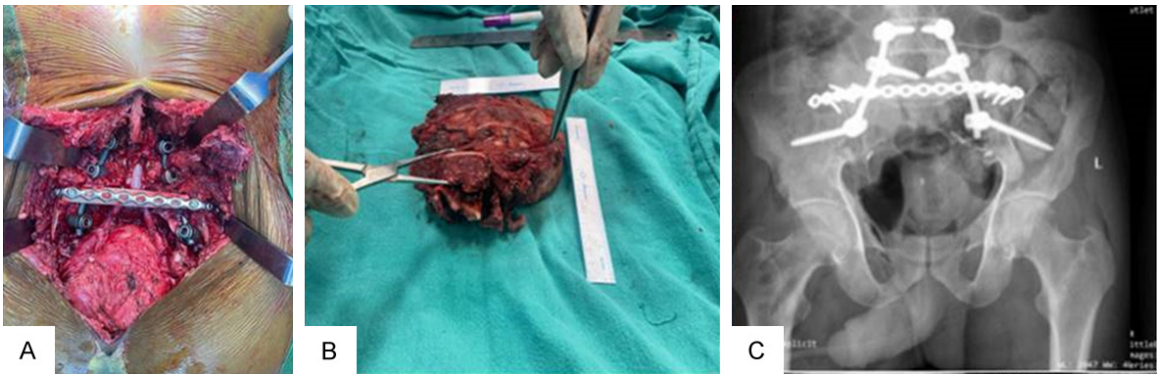

Figure 3. A. Intraoperative image showing lumbosacral fixation with pelvic ring reconstruction using non-vascularized fibula. B. Excised chordoma mass. C. Poste operative radiograph showing lumbopelvic fixation.

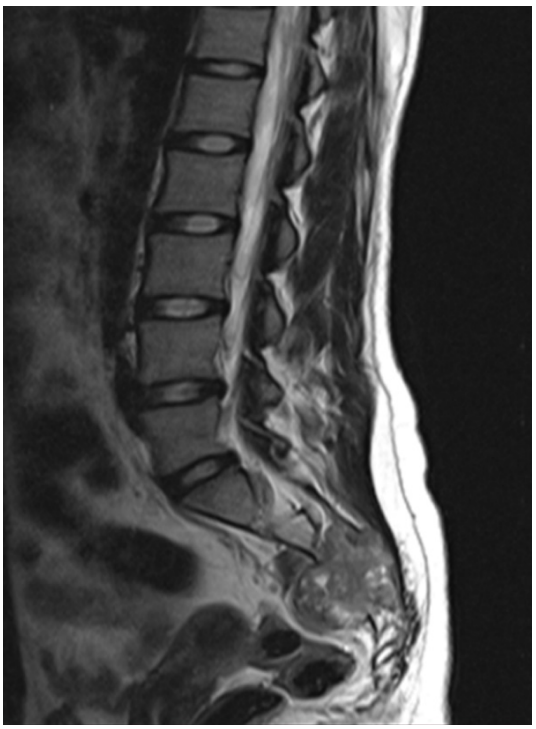

Figure 4. Preoperative T2 weighted MRI showing mass involving the lower part of the sacrum with anterior pelvic cavity extension.

ing into the pelvis and abutting mesorectal fascia, left SI joint, obliterating central sacral canal, bilateral sacral neural foramina and nerve roots (**Figure 4**). On preoperative histopathology, the tumour cells exhibited diffuse immunostaining for vimentin and EMA along with the majority of tumour cells showing pancytokeratin immunopositivity. Brachyury shows foci of nuclear-positive tumour cells. S100 showed variable and scant expression. The tumour cells were immunonegative for desmin, myogenin, CD34, CD99 (non-specific, weak), TLE-1, SALL4 & WT-1. The sacrococcygeal tumour was considered to be a chordoma based on the imaging results and preoperative histopathopatholgy.

The patient underwent surgery in form of wide margin resection-middle sacrectomy with R0 margins-with preservation of both S2 and both S3 roots. Hypotensive anaesthesia and local vasoconstrictors were used to control intraoperative bleeding and 1 unit of PRBC was transfused intraoperatively. Post-resection, bilateral Sacroiliac joints were found to be stable intraoperatively. A postoperative biopsy confirmed the diagnosis of a sacral chordoma which weighed up to 405 g and was measured as 7.6×11.2×8.9 cm (758 cm^3) (**Figures 5**, **6**).

At 15 days following surgery, the patient was discharged uneventfully. The pain was relieved postoperatively and the patient recovered well, maintaining bowel and bladder control. 1 month after surgery she developed sharp shooting bilateral radiating pain for which she was observed and managed conservatively with analgesics. In the 9 months of follow-up, no evidence of recurrence or neurocompromise was found.

Discussion

Chordoma accounts for less than 5% of all bone tumours and it is observed especially among 30 to 60 years with a predilection for the male sex [2].

Sung *et al* based on 30 MRIs in sacral chordoma patients reported characteristic findings as

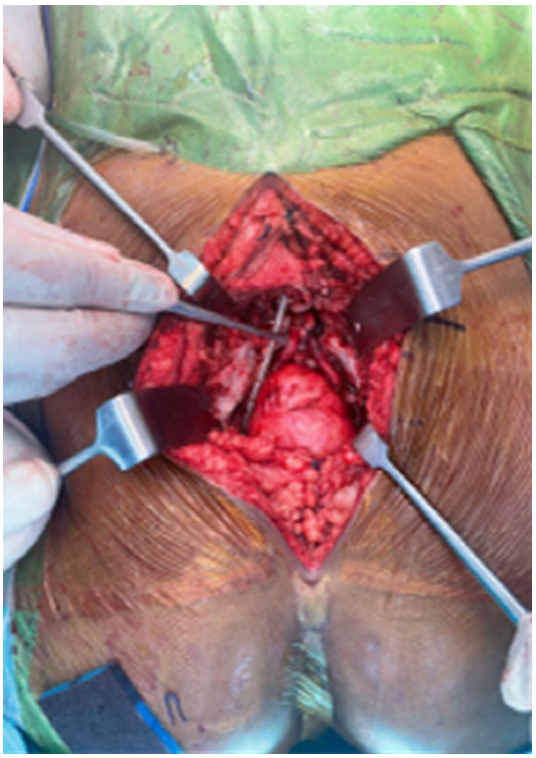

Figure 5. Intraoperative image after tumour resection.

a sacral lesion with heterogeneously high signal intensity with criss-crossing septa on long-repetition-time imaging, well-encapsulated pseudopodia-like or lobulated appearance, and gluteal muscle infiltration. MRI in all the cases was consistent with this finding and invariably had some degree of gluteal muscle infiltration. Some of the common differentials for the sacrococcygeal region located pathologies are chordoma, giant cell tumour, neurofibroma, teratoma, metastasis, myeloma, osteoblastoma, aneurysmal bone cyst, lipoma, osteosarcoma and chondrosarcoma, anal fistulas, sacral dermatoid and post-partum lesions [9, 10].

Histology shows multiple lobules of the abundant myxoid matrix with cords, strands, and solid nests of physaliferous cells, which show enlarged atypical nuclei and eosinophilic cytoplasm with variable-sized vacuoles. Dedifferentiation and development of high-grade sarcoma areas are possible. Xu et al showed that the specimen for giant chordoma were positive for cytokeratin, epithelial membrane antigen, Ki-67, S100, and vimentin [7]. The brachyury gene has been implicated in familial chordomas [8]. In both the cases that had IHC profiles carried out in the institute, specimens were positive for CK, EMA and variably for brachyury. Its histological assessment is often delayed due to non-typical signs and symptoms of disease with a frequent clinical diagnosis of pelvic or vertebral and irradiated pain due to discogenic or unspecific pathology, as has also been in our third case.

Surgery is the primary treatment for this rare bone tumour due to the sluggish rate of biological growth and the low prevalence of metastatic dissemination [11]. Both cases were operated. The extension of margins is a very important prognostic factor correlated with the incidence of local relapses and overall survival. Local recurrence has been significantly associated with an increased risk of metastases and tumour-related complications [3, 12]. In both case, the margins were free from tumour cells. Although, this potentially curative margin-free “en bloc” resection is often very hard to obtain due to the anatomical sites of origin of chordoma such as the Sacro-coccygeal area where often a complexity of structures such as bones, nerves, mesentery etc. are involved. In both cases, it was found to abutt the anorectal fascia for which intraoperative help of a general surgeon was sought to prevent perforation, which is a common complication while dissecting in this region for the tumour. Rectal involvement is rare in these cases due to tough anorectal fascia [7, 13].

Because sacral chordomas are often big, the cavity and soft-tissue defects that occur are widespread, which can contribute to issues such as sacrococcygeal skin necrosis, infection, and delayed or non-healing [1, 7, 13]. We did not have any of above described major complications, although, we preferred a delayed suture removal as a precautionary measure. We had neuritic pain in the second patient radiating to the bilateral lower limb for which she needed hospitalization but was managed conservatively with intravenous morphine.

Pillai and Govinder noted in their review that commonest clinical presentation is low back pain not responding to non-steroidal anti-inflammatory drugs which worsens on sitting with upto one third having urinary complaints

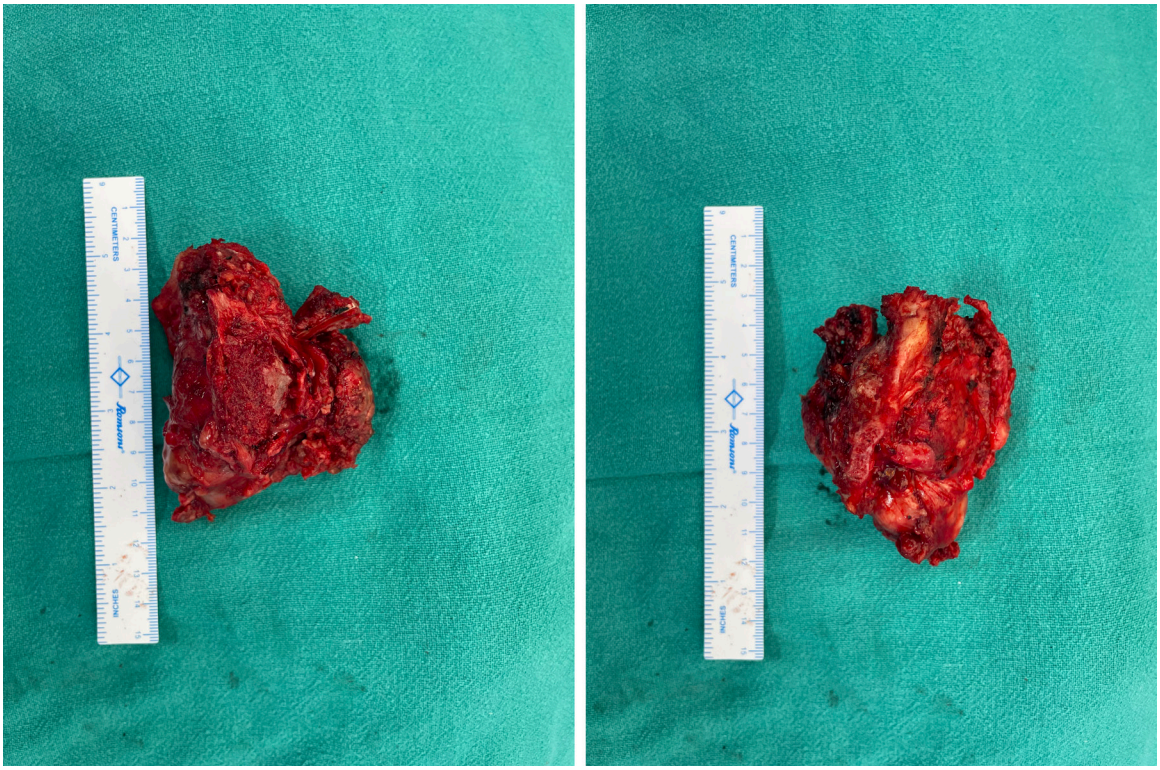

Figure 6. Excised tumour mass.

[10]. Although, radiology can guide towards treatment of patients presenting with such clinical complaints, histopathology remains the standard to make a diagnose of chordoma, differentiating it with other pathologies for which treatment may vary [14]. Ahmed et al noted in their systematic analysis that surgical resection with wide excision with or without ratiotherapy is most widely accepted treatment regimen for >60 months tumor progression free survival [5]. This is similar to observations of Walcott et al and Barber et al [9, 14].

Considering the above situation and the literature available we conclude that effective management of coccygeal chordoma requires early diagnosis, accurate preoperative staging, definitive and adequate surgical resection with proven tumour-free cut margins and close follow-up [1, 12]. In those declining surgery, conservative options can be considered such as radiotherapy with close follow-up. Meticuluous surgery and multimodal treatment options shall be considered in each case irrespective of age and shall be based upon the symptoms and involvement of tissues with an objective of having a tumor-free margins. Due to shrort to immediate term follow-up, it's difficult to comment on correlation of outcome with treatment regimen and age.

Our long-term experience is limited but literature has reported that despite the progress of current surgical techniques and some encouraging results with the use of targeted therapy, disease control and long-term patient prognosis are still poor and chordoma results generally in a long-lasting morbid disease [3, 15]. Nevertheless, the specific experience of the multidisciplinary team (surgeons, medical oncologists, radiotherapists, pathologists, radiologists) is a very important pre-requisite in succeeding to improve patients' quality of life and, hopefully, outcomes.

Conclusion

Giant sacrococcygeal chordomas are extremely rare entities and need special consideration in treatment. The risk of having complications such as heavy blood loss, hematoma formation, wound necrosis and bowel perforation can be prevented with proper planning.

Disclosure of conflict of interest

None.

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