

Case Study

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Sacral Chordoma: A Pitfall in Surgical Management

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Abstract

Chordomas are rare primary, locally invasive tumour of the bone which derived from notochordal remnants. Currently, the mainstay of treatment of chordomas is surgical resection. Despite the clear advantages of adequate surgical margin, the locally advanced nature of chordomas makes wide resection of the tumour difficult as they are often in close proximity with the surrounding vital organs. The published literatures of sacral chordomas mainly focus on the approach of surgery, reconstruction post-resection, long-term survival and reports on successful surgical resection. We report a case which highlights the pitfall in the surgical management of a sacral chordoma. Our patient developed delayed bowel perforation which may be associated with the sacrum osteotomy.

Keywords: chordoma; sacrum; surgery; pitfall; outcome.

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Introduction

Chordomas are rare primary, locally invasive tumors of the bone.^{1,2} They are derived from notochordal remnants, affecting mainly the spine (32.8%), cranium (32.0%) or sacrum (29.2%).² The incidence of chordomas is 0.08 per 100,000 people and they constitute 1-4% of all bone cancers.²

They normally affect men of age more than 40 but younger women suffering from chordomas have been reported in the literature.^{2,3} Although metastasis is uncommon, chordoma is difficult to treat as the patient normally presents late and the tumor is often insensitive to chemotherapy and radiotherapy, making neo-adjuvant and adjuvant therapy not feasible in the management of chordoma.² Currently, the primary mode of treatment is surgical resection of the tumor. Due to the locally invasive nature of chordomas, a large tumor is difficult to resect due to its proximity to the surrounding neurovascular structures, bowels and urinary bladder.¹

The published literatures of sacral chordomas mainly focus on the approach of surgery, reconstruction post-resection, long-term survival and reports on successful surgical resection.¹⁻⁷ Here, we would like to highlight a pitfall in the surgical management of a sacral chordoma.

Case Report

Mdm L, a 65-year-old Chinese lady with no known medical illness, presented with 3 years history of low back pain with right-sided lumbar radiculopathy. She was treated as lumbar spondylosis with right-sided radiculopathy as out-patient for 2 years. However, the pain worsened, and it started to affect her activities of daily living and sleep despite on multiple oral analgesics. She subsequently developed numbness and weakness of her bilateral lower limbs, associated with urinary incontinence.

There was no bowel incontinence. Clinically, there was no palpable mass per abdomen and at the lower back. There was L5/S1 myotomal neurological deficit at bilateral lower limbs. Plain radiographs of the lumbosacral spine revealed a large, ill-defined soft tissue shadow overlying the sacral region.

Magnetic resonance images showed a lobulated mass measuring 10.4 (width) x 13.0 (anteroposterior) x 11.2 (craniocaudal) cm arising from the second sacral vertebra with post-contrast heterogenous enhancement, in keeping of a sacral chordoma (Figure 1).

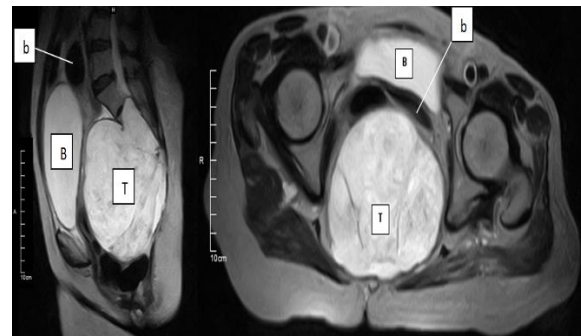


Figure 1 shows the magnetic resonance images of the patient's pelvis. In the T2 sagittal view (fast spin echo without contrast), the tumor (T) can be seen arising from the S2 vertebra, causing mass effect anteriorly onto the urinary bladder (B) and bowels (b). In the T2 axial view, it is clearly shown that the bowels are compressed between the urinary bladder (B) and the tumor (T).

There was no radiological evidence of distant metastasis. Core needle biopsy histopathologic examination revealed the diagnosis of sacral chordoma showing the evidence of physaliphorous cells with mildly pleomorphic nuclei with abundant eosinophilic cytoplasm and area of necrosis (Figure 3). Immunohistochemistry showed positivity for CK (AE1/AE3), EMA and S100.

A multi-disciplinary surgical intervention was performed, involving the orthopaedic surgeon, urologist, vascular surgeon, general surgeon, and anesthetist. A wide local excision was done via a combined anterior and posterior approach. Urologist, vascular surgeon and general surgeon were involved in the anterior approach. Both the sigmoid colon and rectum were carefully mobilized away from the sacral tumor and both ureters are slung with vessel loops (Figure 2).

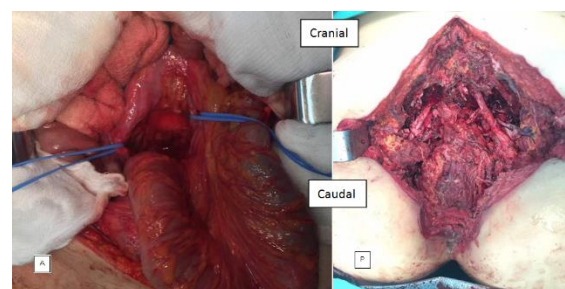


Figure 2 shows the combined anterior (A) and posterior (P) approach for wide local excision of the chordoma.

Anteriorly, the large bowels are mobilized away from the sacral tumor and both ureters are slung with vessel loops.

The left internal iliac vein was ligated through the anterior approach by the vascular surgeon as it was located adjacent to the tumor. The main tumour bulk was separated from surrounding tissues and S1 osteotomy was partially performed anteriorly using an ultrasonic bone scalpel. Osteotomies were completed at the upper border of S1 sacral foramen, through posterior approach. Part of the left posterior superior iliac spine was removed as the tumor is seen abutting the left sacroiliac joint. The left S1 nerve root was sacrificed.

Soft tissue reconstruction for the posterior wall defect was not performed. Finally, the bowels were carefully inspected for any iatrogenic injury and a defunctioning sigmoid colostomy was created for better wound care.

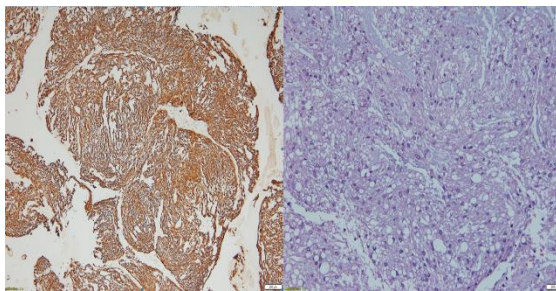
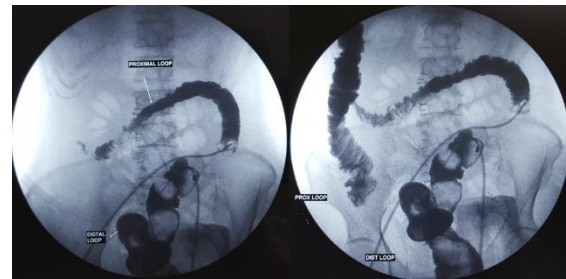


Figure 3 shows the neoplastic tissues consisting of tumor cells embedded within an abundant myxoid matrix, forming a lobular architecture separated by thin fibrous septa. The tumor cells display mildly pleomorphic nuclei with abundant eosinophilic, vacuolated, bubbly cytoplasm. Areas of necrosis are seen. No spindle cell is identified.

Via the combined anterior and posterior approach, we were able to achieve wide en bloc resection of the tumour. The estimated blood loss was 7 litres with a total of 10 pints pack cell and 3 cycles of DIVC regime (4 units of fresh frozen plasma, 6 units cryoprecipitate and 2 units of platelet concentrate) transfused. Post-operatively, the patient was recovering well until she developed fever on day 6 post-operation. It was associated with worsening leukocytosis and discharge containing fecal material was seen from the posterior wound. An intestinal gastrografin study, distal loopogram and ultrasound abdomen showed no obvious bowel perforation or intra-abdominal collection (Figure 4).



Distal loopogram in Figure 4 shows no obvious bowel perforation.

Her condition further deteriorated, and she was in septic shock with increasing amount of fecal discharge from the posterior wound. An exploratory laparotomy was performed on post-operative day 9, revealing a perforated ileum with concealed enteric contents, 60 cm from the duodeno-jejunal junction at the posterior aspect of the sacrum. A loop ileostomy was performed, but the patient succumbed to the complication on post-operative day 11.

Discussion

Consistent with the literature, the chordoma in this patient is large at the time of diagnosis.⁴ Wide resection is the surgical treatment of choice although seldom feasible due to its close proximity to the surrounding vital structures.¹ Nevertheless, achieving a safe surgical margin with en bloc resection is paramount in reducing the risk of recurrence.⁸ Yang et al demonstrates that surgical margin is the only modifiable predictive factor of local recurrence.⁸ In addition, patients with adequate surgical margins tend to survive longer than patients with insufficient surgical margin.

Study by Ji et al shows that 5-year survival rate of the patients with adequate surgical margin is 86% compared to 67% in those without adequate surgical margin.⁹ The 10-year survival rate difference is even more profound between patients with total resection (88%) and subtotal resection (31%).¹⁰ Despite the clear advantages of good surgical margin, achieving it in patients with large sacral chordoma is never easy and it risks iatrogenic damage of the vital neurovascular structures in close proximity.

In our patient, a combined anterior and posterior approach is used for two reasons. Anteriorly, neurovascular structures, the urinary bladder and bowels must be protected as they are in close proximity to the tumor. By mobilizing all the vital structures away anteriorly, we are able to achieve maximum tumour resection margin while reducing the risk of iatrogenic injury to the minimum. Secondly, the sacral bone needs to be osteotomised posteriorly as the tumor is obscuring bilateral sacral foramina over S1 anteriorly. As the posterior wound is close to the anus, we decided to perform a temporary defunctioning colostomy for better posterior wound care and to reduce the risk of surgical site infection. A reversal of colostomy is planned in four to six weeks, once the wound has healed considerably.

When the patient's condition deteriorates, an internal colostomy leakage is suspected but there is no radiological evidence to support the diagnosis. A perforated ileum is noted during an exploratory laparotomy when she is in septic shock. Despite the attempts to save the patient, she subsequently succumbed to the severe complication. In our opinion, the delayed bowel perforation is associated with the sacrum osteotomy.

The sharp edge produced by the ultrasonic bone scalpel serves as a risk which causes bowel perforation when the peristalsis resumes. Resection of the tumour through osteotomy should leave a smooth edge to avoid puncturing the internal organs. Coverage with soft tissue over the edges during flap closure over the posterior defect can be considered and planned together with the plastic surgery team.

Conventional photon-based radiation therapy is not useful in treatment of sacral chordoma. Nonetheless, latest studies of intensity modulated proton therapy (IMPT) show promising results, paving way for treatment of inoperable sacral chordoma.^{11,12} However, IMPT is not without its complications, including dermatitis, mucositis, pathological fractures and bowel perforation.¹³

Conclusion

Extra precaution must be practiced during resection of the chordoma and a thorough examination of all the vital organs must be made prior to closure of the wound intraoperatively. If there is any suspicion on bowel perforation, an exploratory laparotomy is often urgent for both diagnostic and therapeutic purposes.

Conflict of interest

This case report has been presented in the 48th Malaysian Orthopaedic Association Scientific Meeting and the abstract has been published in Malaysian Orthopaedic Journal.

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