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## Images

### **Sacral Chordoma**

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A 63 years old woman, survivor of melanoma excision 15 years back, presented with pain in the lower back for the last one year. The pain was present on rest and leading her to get up from sleep. No radiation of pain or weakness in lower extremities was noted. She reported some perianal numbness and mild difficulty in voiding. She previously had bilateral Total hip replacements about 5 years back. Her neurological exam was otherwise unremarkable. Her MRI showed a large sacral mass from S2 below involving the sacroiliac joints and the gluteus maximus. She underwent a biopsy which revealed the tumour to be a Chordoma. Later she underwent a wide margin excision by a Sacrectomy. This was performed by a

combined anterior and posterior resection. A laparotomy was performed to ligate the branches of the internal iliac artery and veins supplying the tumour, followed by dissection of the tumour from the anterior structures. Resection margin was defined by passing K-wires from the front to back. This was followed by turning the patient prone and excising the tumour through a posterior elliptical incision to incorporate the previous biopsy site. Margins of Gluteus maximus were included in the resection to achieve a clear resection margin. Laminectomy of L5 and sacrum was performed and the dura was ligated and amputated distal to S1 nerve roots. The sacral resection was performed distal to S1 vertebra. A spanning construct using two screws

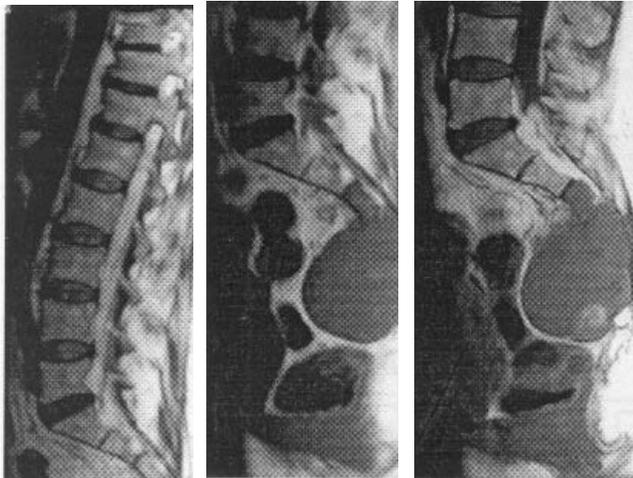


Figure 1. (A) Most MRI examinations usually do not cover all of Sacrum, therefore the same patient had the first MRI and Sacral tumour cannot be seen because of incomplete coverage, leading to another MRI (B and C) Complete Sacral MRI shows a mass in the distal part of sacrum in the following MRI examination.

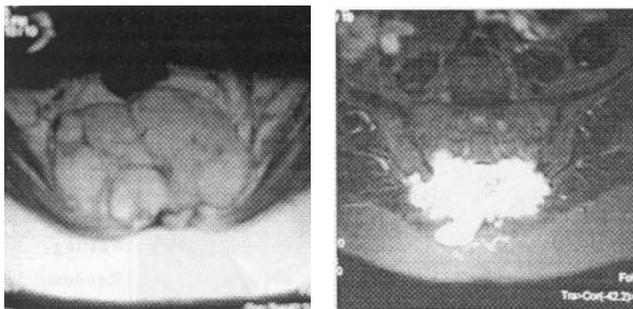


Figure 2. Involvement of the Gluteus Maximus muscles can be seen on MRI.

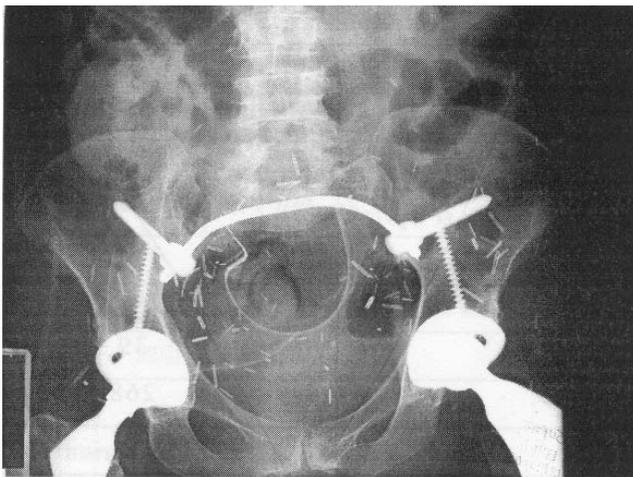


Figure 3. Post-resection X-rays shows absence of the distal sacrum, spanning construct for sacroiliac stabilization and previously done bilateral total hip implants. Multiple vascular clips can be seen in the operative field as well.

and rod were placed in the iliac bone to prevent any instability. The skin defect created by the tumour resection was covered by double rectus abdominis flaps which were harvested after placing the patient supine at the completion of the resection procedure and placing the flaps in the

pelvis. The patient was then again turned prone and the flaps were brought out posteriorly through the tumour resection wound defect and were used for coverage in the sacral region. The patient also underwent a colostomy in the initial part of surgery anticipating the post sacral nerve root resection bowel incontinence and the location of the surgical wound. Postoperatively the patient demonstrated some transitory additional neurological deficits (foot drop) apart from the anticipated bowel and bladder issues, but over the period of next few months was able to gradually restore herself to normal neurological status apart from the bladder and bowel incontinence (due to sacral roots resection). The histopathologic examination of the specimen confirmed it to be sacral chordoma and it showed clear resection margins. The patient is almost 2 years into her followup now and has not shown any signs of recurrence.

### Commentary

Chordoma is a locally aggressive but a slow growing and rare malignant tumour. It arises from ectopic rests of notochordal tissue and has a predilection for Sacrum (50%), skull base (35%) and mobile segments of spine (15%). It is uncommon in individuals below 40 years of age and the male to female distribution is 2:1. Because of its slow growth most sacral chordomas present with extension into the pelvis but usually spare the rectum. Metastasis is usually a late event. Most patients present with local pain or radiculopathy but are usually diagnosed late with a palpable pelvic mass or with bladder-bowel symptoms. Chordomas can be easily missed since they appear as a lytic lesion on X-rays and most MRI and CT scans do not scan the sacrum below S2. Biopsy should preferably be performed by the operating surgeon. A transvaginal or a transrectal biopsy is strongly condemned. Wide en bloc resection is crucial for cure as studies clearly show the combination of intralesional debulking and radiotherapy is associated with a high recurrence rate. The local recurrence of chordoma is associated with 21-fold increased incidence of tumour related death. Sacral resection is a very technically demanding procedure and require a multidisciplinary team approach. Usually an anterior followed by posterior procedure is required for vascular control and resection of the tumour. Trans-pelvic Rectus Abdominis myocutaneous flaps have positively contributed in decreasing the wound dehiscence after resection of large sacral chordomas.<sup>1</sup> Sacral resection below S3 are uncomplicated but resection through the sacroiliac joint are much more challenging. It requires the amputation of the dural sac at the level of resection which in high sacral resection will lead to bowel and bladder compromise and sexual inability. In resections of the dural sac below S3 no