

Sacral Chordoma: A Case Report

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Abstract

Chordoma is a rare malignant tumor of musculoskeletal system and it only accounts for 1-4% of Primary bone Tumor. It originated from the remnant of notochord and the most frequently found in Sacrum. The treatment of choice is wide surgical excision with or without radiotherapy.

A 48 year-old male with chief complaint of lump at sacral region since 6 months with a size equal to a chicken egg. Physical examination showed a solid tumor with a diameter of 10 cm x 10 cm, there were no clinical neurological compromises found. A CT scan confirmed a tumor with a size of 10 cm x 14 cm x 14 cm that extended to pelvic cavity, later Fine Needle Aspirations suggested the mass as a chordoma. A wide excision with adequate margin was performed by a spine surgeon and with a digestive surgeon confirmed there was no abdominal viscera involvement.

Surgical wide excision is still the mainstay treatment of the chordoma. Although it is the treatment of choice, it comes with its own disadvantages due to the lower nerve root being resected. The neurological symptoms found after-surgery in our patient was loss of sensation and erectile dysfunction post-surgery.

Keywords: Sacrum, Malignancy, Tumor, Orthopedic Oncology

Introduction

Chordoma is a rare malignant tumor of musculoskeletal system and it only accounts for 1-4% of Primary bone Tumor and accounts for 20% of the tumor in spine. It originated from the remnant of notochord.⁽¹⁾⁽²⁾ It has male predominance (2:1), with the most common age group between 50 and 60 year-old. Chordoma most common found in clivus and sacrococcyx region. Bakker et. Al reported in English study, 45% of the cases involves sacrum or coccyx and 26% involves the Skull base. It is an indolent malignant tumor but shows local aggressiveness to surrounding vessels and nerves. It often spread to adjacent vertebra but spares the intervertebral disk space unlike vertebral osteosarcoma and chondrosarcoma.⁽²⁾⁽³⁾⁽⁴⁾ Chordoma historically does not respond well to conventional radiotherapy and cytotoxic chemotherapeutic agents, therefore NCCN Guideline 2021 recommends surgical excision with or without adjuvant radiotherapy in resectable cases and radiotherapy in unresectable cases, as a local control.⁽²⁾⁽⁵⁾

Sacral chordoma can be classified using Enneking system and marginal or wide excision can be applied to them. However the complex anatomy surrounding the sacro-coccyx region, makes it challenging to plan the surgery, such as the nerves related to lower extremity, sexual, bowel and bladder function, Iliac vessels, and retroperitoneal viscera. Usually the surgery planned in two stage, with anterior approach to create the desired margin, and to evaluate the retroperitoneal structure and perform partial sacrectomy from the anterior. Then the second stage was to complete the sacrectomy and tumor

removal from the posterior approach. Posterior only approach maybe feasible for cases involving the S3 or below.⁽⁶⁾The first successful combined anteroposterior resection for a presacral tumor was performed in 1945 by Bowers at Walter Reed Army Medical Center. An initial posterior approach for a presumed chordoma was unsuccessful because of extensive vascular collateralization⁽⁷⁾

Recent findings about the molecular process involved in chordoma have uncovered the new novel agents for treatment in advanced and relapse cases need gene mutation screening and immunohistochemistry, so that agents such as Imatinib, Dasatinib, and Erlotinib can be more specific in targeting the growth factors and pathway that contributes to the tumor growth in specific patient.⁽²⁾

Case Report

A 48 year-old male with chief complaint of slow growing and lump in sacral region for over 6 months. Accompanied by dull pain, discomfort while sitting and difficulty in voiding and incontinence preceding the chief complaint. Physical examination showed a solid tumor, immobile, with a bone like consistency with size of approximately 10 cm x 10 cm, there were no

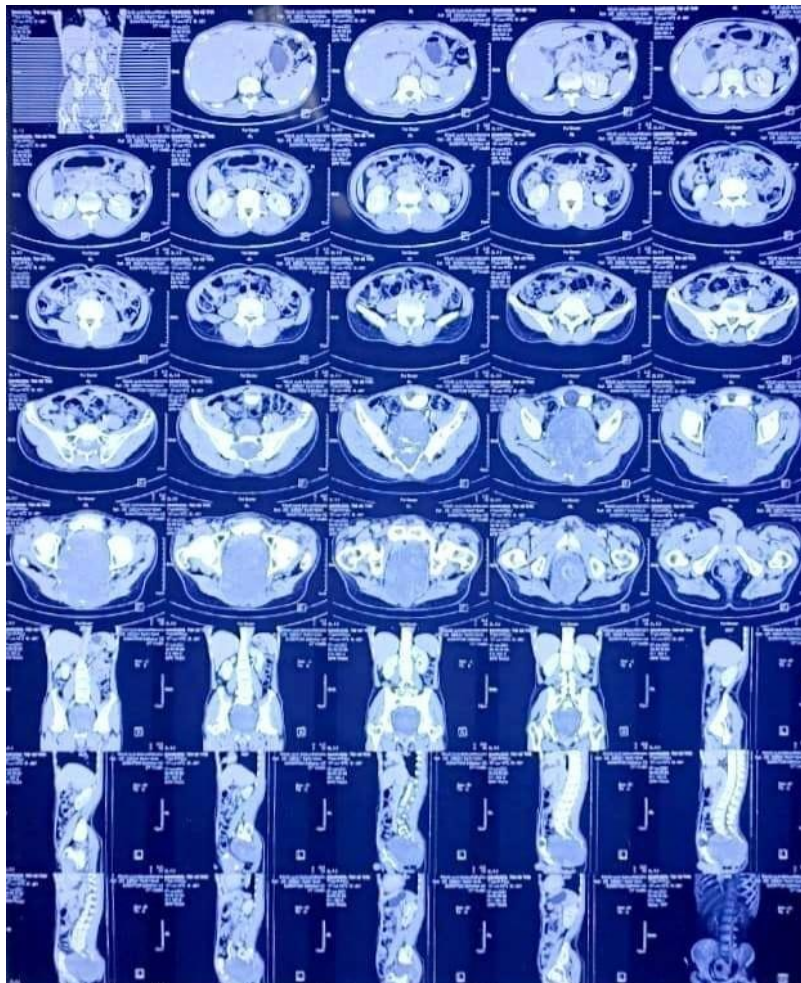


Figure 1 : CT Scan confirming the mass at sacral region neurological compromises found. We ordered a CT scan that confirmed a lobulated hypodense mass with a size of 10 cm x 14cm x 14cm that extended

to pelvic cavity and partially obliterated sacrum, and infiltrated the surrounding paraspinal muscles, and from CT scan there was no suggestive finding of metastasis to the liver, gall bladder, spleen, pancreas, and kidney. Fine Needle Aspiration finding suggested the tumor as a chordoma.

Due to the extension of the tumor and its close proximity to the retroperitoneal viscera, we planned a joint operation of wide excision of tumor by a spine surgeon from posterior approach and evaluation by digestive surgeon, to evaluate the involvement of visceral organ. From the spine surgeon to performed a wide excision with adequate margin, with expected outcome the loss of nerve root function at the level of the excision and from the digestive surgeon to evaluate the involvement of the abdominal viscera, and to

release the tumor from its surrounding from the anterior approach and if needed, to perform a resection and colostomy if the tumor invades the digestive tract. The tumor was resected with adequate margin with a size of 20 cm x 15 cm x 15 cm, and during the surgery the tumor did not invade the abdominal viscera and can be released

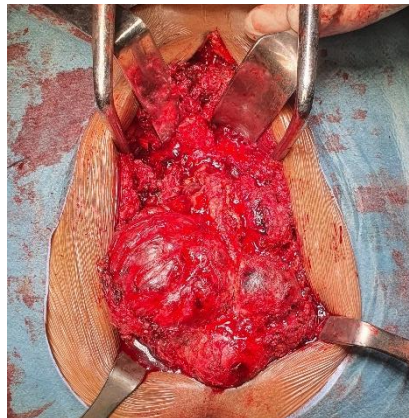


Figure 2 : Mass before resected, appeared to be lobulated..

both anteriorly and posteriorly, the sacrum was preserved above the S2 Level. Histopathological examination finding confirmed the tumor as a Chordoma.



Figure 3 : Mass after being resected, with dimension 20 cm x 15 cm x 15 cm.

A 6 month follow up, the patient is able to return to work, no complaint of pain at the site of the surgery and no weakness of the lower limb, bowel and bladder function was normal. There are problems of decreased in sensation near the groin region and mild erectile problem.

Discussion :

Sacral chordoma is a rare finding in daily practice, Half of case are found in sacrum, but it can occur anywhere in the spinal axis. It belong to the sarcoma family. It is a low-grade sarcoma but locally invasive and aggressive. 5 year survival of patients with sacral chordoma is about 50 %. This tumor is from notochord remnant from the mesoderm which in time will regress and become nucleus pulposus.

Chordoma commonly affect the age group of 40-60 year-old, and the case is twice more common in male than in female. The incidence is predicted to be about 1:1.000.000. Which is in line with our case, a 48 year-old male.

It is a slow growing tumor, and in our case the tumor is at the sacral region which will be hard to evaluate from physical examination and need more thorough examination due to the mass extended to the pelvic cavity and surrounding viscera. In our case the patient's complaint before he knew he have a growing mass was mainly due to the urinary symptoms.

The diagnosis are usually missed in plain X-ray, in our case there is some destruction of sacrum found in previous x-ray, therefore we order a CT-Scan to have a better view of the extent of the tumor. We proceed to histopathological fine needle biopsy to have a more targeted approach and it is in line with the CT-Scan and we established the diagnosis as a sacral chordoma.

Treatment of choice is still the surgical excision, with or without adjuvant radiotherapy, in resectable cases, while the radiotherapy as the treatment of choice if unresectable. In our case the tumor is resectable with acceptable margin and we did not perform the adjuvant radioteraphy.

The challenges during surgery was the close proximity of the tumor with retroperitoneal structures and also the vessels surrounding it. Therefore the posterior approach only have limited capability in evaluating the structured anterior to the tumor, therefore we suggest a planned, antero-posterior approach, by a join surgery by a digestive and spine specialist to perform the surgery.

The surgery usually need to sacrifice some nerve root from the sacral region, and it can affect the motor function, bladder and bowel function. If the tumor excision at higher than S1 level the motor dysfunction can occur.⁽⁸⁾In our case the sacral above the S2 level is preserved, therefore the motor function is preserved and there is only a slight dysfunction in bladder and bowel.

The 6-month follow up the patient is already back to his daily activity with only mild complain about his condition post-surgery. A scheduled follow up is needed to evaluate the patient is needed due to the possibility of recurrence.

Conclusion :

In our case sacral chordoma still have satisfactory outcome, treated with only surgical resection. The careful planning of surgical approach with a join surgery from digestive and spine surgeon need to be considered, and the level of which the excision performed can predict the outcome.

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