# Dedifferentiated Sacral Chordoma: a case report

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**Abstract**

Sacral chordoma is the most common primary bone tumour bone in the sacrum. Though the incidence of chordoma is rare. Chordoma is divided into subcategories such as conventional chordoma, chondroid and dedifferentiated. Within these three categories the rare. Among the rarity, women and young adult were the least population to be diagnosed. We presented a case of a young female with dedifferentiated sacral chordoma.

Keywords: sacrectomy, chordoma, dedifferentiated, chondroid

## Introduction

Chordoma is a rare malignant, slow growing and locally aggressive notochordal tumour, which is often diagnosed in the late fifties and male predominant.(1)The incidence is less than 0,1/100000 per year.(2) Despite its rarity, chordoma is one of the main tumours found in the sacrum. 29-50% of the chordoma was found in the sacrum. Physical findings are varied according to where in the spine does the tumour grows which extended from abdominal discomfort, defecation and voiding difficulties to paraplegia and paraesthesia. It was diagnosed with radiology imaging either CT scan or MRI and histopathology. Chordoma is divided into several subtypes such as conventional (the most common), chondroid, poorly differentiated and dedifferentiated or sarcomatous chordoma (chordoma associated with high grade sarcoma, which account for only 5% of cases and have the worst prognosis).

Currently, the best treatment for chordoma is surgical wide excision tumour removal which associated with high mortality and morbidity due to the involvement of the neurovascular anatomy. Chordoma is not sensitive to neither radiotherapy nor chemotherapy though there were several studies that shown there is a little benefit in treating chordoma with radiotherapy.(2) Treatment with chemotherapy which is insensitive in chordoma is being developed as targeted therapy,(2)

In this case, a young adult female patient was diagnosed with conventional chordoma. Thus, S3-S5 sacrectomy and wide excision was performed. The histopathology study following the surgical procedures showed dedifferentiated chordoma. This study aims to present and discuss about sacral chordoma.

## Case presentation

A 21-year-old female with no prior medical history presented with buttock pain. Pain is characterised as a sharp excruciating pain that radiates to the legs and anal region. There was no motoric deficit, but patient was having difficulties in defecation and voiding. The patient had no visible abnormalities in the sacrum or abdominal region. Abdominal distention was palpated without any palpable mass either in the anterior or posterior part of the pelvic. Lasseque test was positive in both side of the leg, bow string test tested positive in

both leg, motoric deficit was absent in this patient whilst pain was presented predominantly in the buttock. Patient underwent MRI studies which showed a mass in the level of S3 of the sacrum.

Core biopsy was performed, and pathology study showed a low grade chordoma Thus, surgical wide excision sacrectomy was scheduled. Patient refused treatment as there were risk of multiple complications and

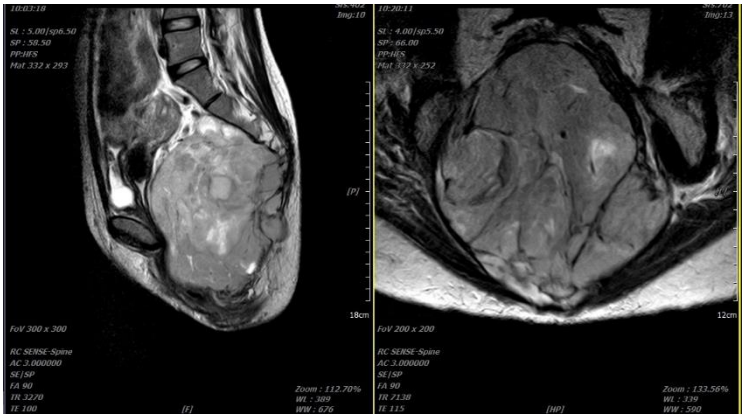

*Figure 1 Solid lobulated lession on the level of S1-S2 to the coccygeus with hemmorage component inside, followed by destruction of ventral side coccygeus, which ventrally displace uterus and bladder, dorsally filled the pelvic space (12,7 x 13,5 x 14,6)*

decide to undergo chemotherapy instead. After underwent six sessions of chemotherapy and open biopsies in other hospital for a year, the patient decided to undergo the surgical wide excision as there was no significant improvement in her condition. Follow up MRI prior to surgery for evaluation and preparation.

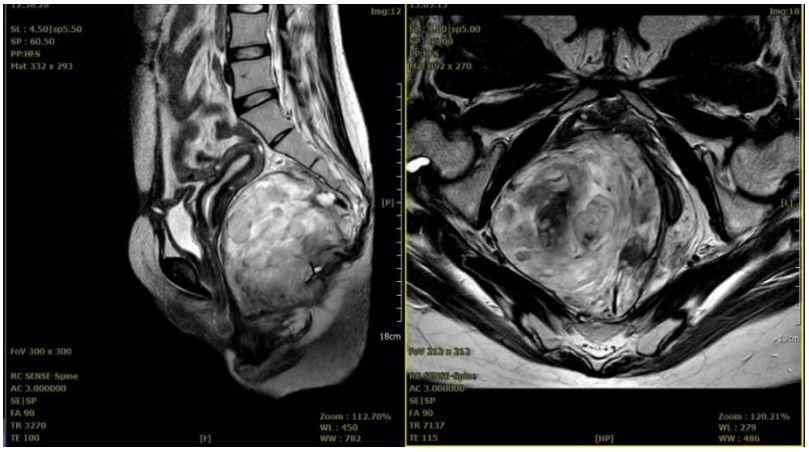

*Figure 2 Primary bone mass of the vertebral body on the level of S4-S5, and coccygeus, which widened to the pelvic space on the level of S2 (was at the level of S1-S2) smaller in size (110,7 x 114,8 x 106,3)*

Sacrectomy was performed on the level of S3. Pathology study of the sample showed a dedifferentiated chordoma. After surgery, the patient has no significant neurological deficit.

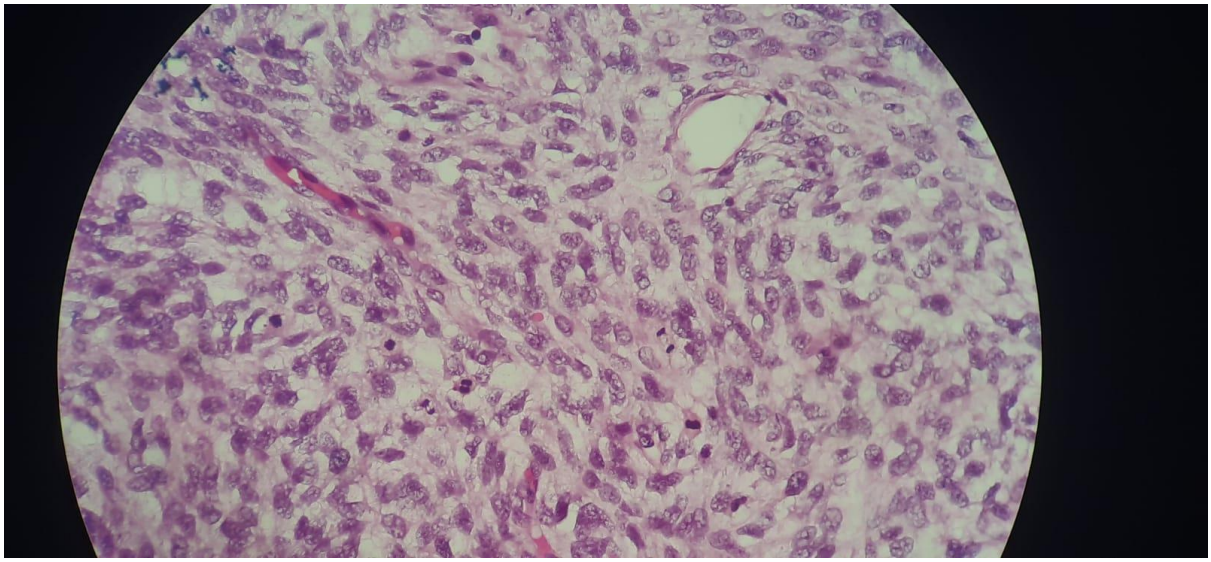

*Figure 3 Dedifferentiated chordoma*

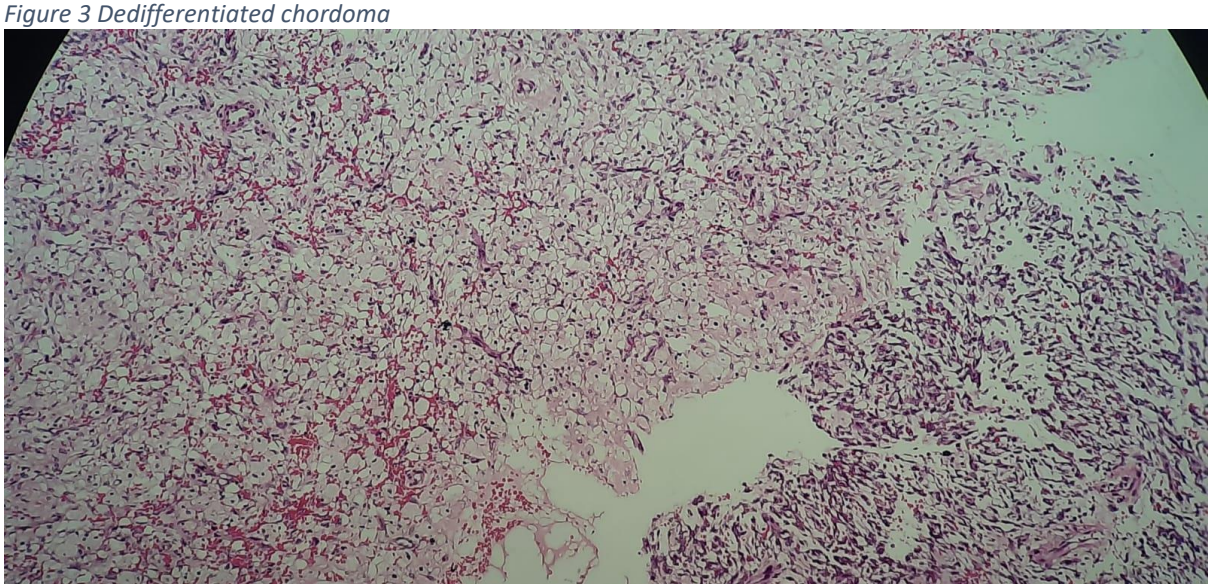

*Figure 4 Conventional Chordoma (Left) dedifferentiated chordoma (Right)*

**Discussion**

Chordoma is a rare malignant tumor of the bone that originated from the remnants of notochord. It is a slow growing, locally malignant tumor. Chordoma has an incidence rate of 0,84 cases per million persons per year, male predominant (0,1/100000) compared to female (0,06/100000). (3,4)While it is a rare tumor, chordoma is the most common primary malignant tumor found in the sacrum. It is commonly diagnosed in the late fifties. Female patients and those presenting in the younger age (<26 years) have higher incidence of skull base chordomas.(5)

Chordoma usually has a variety of clinical presentation in relation to which axial skeleton affected. Skull base chordoma usually presents with headache, cranial neuropathy, and endocrinopathy, while mobile spine and sacrum chordoma may present with localized pain, radiculopathy, myelopathy or even bladder or bowel dysfunction. (1,6)

Histologically, chordoma is categorized into three subtypes such as classical/conventional chordoma, chondroid, and dedifferentiated. The first two subtypes are low grade and locally aggressive while the latter shows aggressive behaviour.(1,7)

Chordoma is diagnosed through radiology imaging, X ray shows bone erosions with irregular calcium foci along lytic lesion. Computed tomography scan presents a low attenuation, hyperdense soft tissue mass with calcification, lytic lesions, and a locally destructive pattern. MRI is a better imaging modality which present a hypodense lesion in T1 flair. With contrast enhancement, lesion shows in T2 appears hyperdense in comparison.(8,9)

This patient is a twenty-year-old female. She first felt localized pain in the buttock that radiates to both legs. MRI showed a localize mass on the sacrum. Bowel and bladder dysfunction was later presented thus patient underwent laparotomy for colostomy followed by cystostomy. During the laparotomy, core biopsied was performed to determine the type of tumor. Pathology study showed conventional chordoma.

Patient initially was scheduled for surgical wide excision tumor removal. Patient refused treatment as she was informed of the risk followed by the surgery starting from lost of motoric function below the tumor level to death. The patient then decided to start chemotherapy.

Surgery Is the first line treatment in chordoma. Chemotherapy is not effective, hence targeted therapy is widely research. In sacral chordoma, resection either marginal or wide resections is recommended.(5,10) This treatment is often challenging as the location of tumor and surrounding anatomy, including neurovascular, which lies nerves contributing to lower extremity function, sexual function, bladder and bowel function, bony attachment as well as muscular attachment.(6) . This is a complex surgery which involves multidisciplinary team and has many potential complications such as wound dehiscence, neurogenic bladder and bowel, massive haemorrhage and sacropelvic instability. Hence, in many cases to resect the tumor up to the desired margins, nerve roots are sacrificed resulted in functional deficit varies to the involving nerve. (11)

Anterior and posterior approach of total sacrectomy is performed in sacral chordoma. Low sacral amputation often led to sacrifice the roots distal to S3, usually minimal deficit will occur except for perineal sensation and sexual function tough sphincter function is typically preserved.(9)

After six sessions of chemotherapy and another open biopsy performed in other hospital, around a year, the patient agreed to undergo the surgery. Re-evaluation MRI prior to surgery showed no significant change. Patient was still in pain, yet there was no motoric deficit. Wide excision with posterior approach on the level of S3 was then performed, and the tumor was then delivered to pathology. Pathology result showed a dedifferentiated chordoma. Patients was admitted for 3 weeks and discharged with no visible motoric deficit.

In contrast to conventional chordoma, dedifferentiated chordoma is a rare subtype of chordoma, with high grade sarcoma component. Moreover, it progresses rapidly and may create distant metastasis. This type of chordoma is well known for having the worst prognosis. Theories are made concerning the changes happened in conventional chordoma into dedifferentiated chordoma. the malignant transformation of conventional chordoma due to progression, or sarcomatous changes post irradiated chordoma.(12) Five years survival rate in sacral chordoma is 50% and ten-years survival rate is 30%. Long term follow up is needed to evaluate the neurogenic bladder and bowel.(13)

In conclusion, this is a rare case of a women in her twenties which was priorly diagnosed with conventional chordoma, went on a series of treatment, and finally had a sacrectomy. Dedifferentiated chordoma was showed in the pathology study of the removed tumor. The surgery resulted in no visible neurological deficit though patient follow up must be done to determine whether there is any recurrence of the tumor.

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