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# Coccydynia, a rare presentation of invasive sacrococcygeal chordoma in a young adult patient

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## ABSTRACT

Chordoma is a slow-growing malignant bone tumor that develops out of the embryonic notochordal residues and is rare. This is most frequently localized in sacrococcygeal joints, where the localized pain may go unnoticed or be mild in nature, and this may put off a diagnosis. Definitive diagnosis requires the use of histopathology with the assistance of immunohistochemistry. We describe a case of a 45-year-old male with progressive pain and swelling of the gluteal area since few months. Excision biopsy showed lobules of big polygonal physaliphorous cells in a myxoid stroma and had hemorrhage and necrosis. Mitotic activity was low. A chordoma provisional diagnosis was established based on the histomorphology. To prevent recurrence and achieve better outcomes, multidisciplinary management, which consists of surgical excision and long-term follow-up, is necessary.

## 1. INTRODUCTION

Chordoma is a slow growing, slow but locally aggressive malignant bone tumor that occurs as a result of remains of the notochord in the embryo. It represents fewer than 5 percent of the total number of primary bone tumors and in the axial skeleton it most often is involved. The most commonly affected anatomical region is the sacrococcygeal region and then the clivus and vertebral bodies [1]. Chordoma is linked to high levels of morbidity despite its sluggish growth pattern through local invasion, high rate of recurrence as well as resistance to traditional chemotherapy, and therefore early diagnosis and proper management is important.

Chordomas normally happen in adults around the fourth to seventh decades of life, and have a predominance among the male. Nevertheless, the cases have been reported in the broad age range. These tumors are clinically usually diagnosed as slowly growing masses that are related to blunt pain, neurological or bowel and bladder dysfunction depending on their location and size. Sacrococcygeal chordomas are often asymptomatic for long periods, leading to late diagnosis and subsequent manifestation at an advanced stage, when tissue invasion in the area has already occurred [2].

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**Keywords**  
sacrococcygeal chordoma,  
gluteal mass

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A significant diagnostic challenge of chordoma is the fact that historically, it has a histological similarity with other myxoid tumors such as myxoid chondrosarcoma, metastatic mucinous carcinoma, or myxopapillary ependymoma [3]. Radiological observations can only be suggestive and not conclusive, and therefore, histopathology examination is the foundation of diagnosis. Characteristic physaliphorous cells within a myxoid stroma are a characteristic feature, but immunohistochemistry is essential for diagnostic confirmation. The expression of Brachyury, which is a transcription factor required in the formation of the notochordal, is a nuclear expression sensitive and specific to chordoma and helps distinguish it amongst its histologic counterparts.

Taking into consideration the infrequency of chordoma and the possibility of challenges in the diagnosis, reporting of chordoma will help in creating awareness and knowledge on the clinicopathological presentation of chordoma. This case is presented because it is rare with a classical histopathological

appearance and the value of early diagnosis on better patient outcomes.

## 2. CASE PRESENTATION

A 35-year-old man came with a chief complaint of pain on right gluteal area with a slowly growing swelling. The patient indicated that the mass had been gradually progressive over a long period, and an increased pain intensity triggered him to seek medical attention. No trauma history, fever, or weight loss, or constitutional symptoms. At the time of presentation, the patient did not complain of bowel or bladder problems, lower limb weakness, or sensory problems.

MRI lumbo-sacral spine was done, which showed a large, well-defined, homogeneously enhancing mass lesion in the sacral canal centered in the lower lumbar-sacral region. It shows causing compression and displacement of the cauda equina nerve roots. It shows intense post-contrast homogenous enhancement with canal expansion and sacral remodeling.

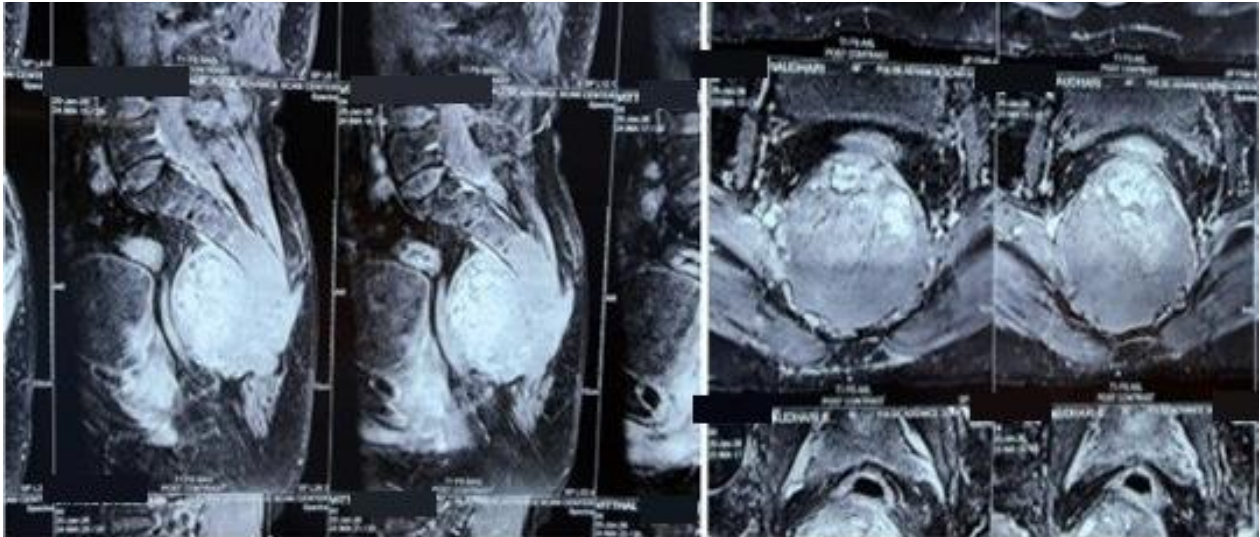


**Figure 1.** MRI of the lumbosacral spine with contrast.

Physical assessment showed that there was swelling in the sacrococcygeal and gluteal area. The mass was malicious in its development and was gradually growing in size. The palpation revealed a soft consistency of the swelling with the skin above it showing no sign of ulcer, discoloration, or sinus. There were no findings of local deficiency observed in the neurological examination, and no indication of compression of the surrounding pelvic structures

was seen. There was near-complete erosion of the sacrum and part of the coccyx.

C-arm image taken intraoperatively showed near-complete erosion of the sacrum and coccyx (Figure 2).

Intraoperatively, it was reported to be a soft, suckable tumor with areas of necrosis and hemorrhage and extensive erosion of surrounding soft tissue and bony elements (Figure 3).

Following the clinical results, a tentative diagnosis of a sacrococcygeal space-occupying lesion was taken into consideration, and the differentials were teratoma and soft tissue sarcoma. At the time of reporting, the radiological information was not obtained. Considering the clinical suspicion and progressive course of the lesion, an excision biopsy was to be carried out to make a definitive diagnosis. The removed sample was then sent to a histopathology lab.

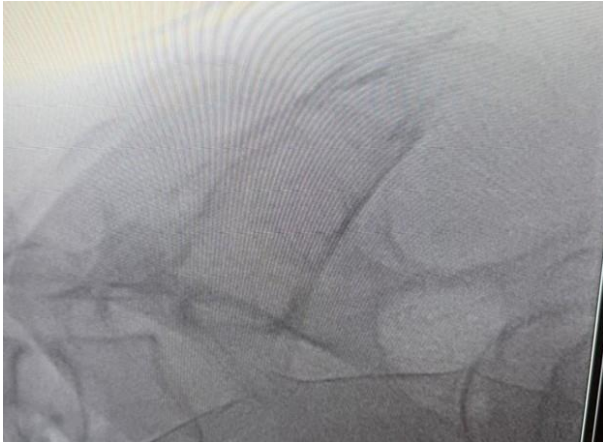


**Figure 2.** Intraoperative C-arm image showing erosion of the sacrum and coccyx.

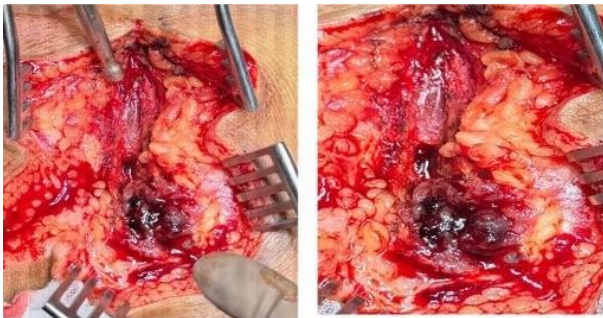


**Figure 3.** Intraoperative findings showing a suckable tumour with areas of necrosis.

### 3. HISTOPATHOLOGICAL FINDINGS

On gross examination of the specimen, several irregular pieces of soft tissue, grey-white in color to brown, were seen with a total size of about  $5 \times 5 \times 1$  cm. The tissues were observed to be weak and immature, and the whole specimen was sent to the microscope to make sure that proper sampling was done.

Microscopic observation has revealed a tumor of diffuse sheets and lobules of large polygonal cells embedded in a high myxoid stroma. The tumor cells were richly vacuolated, bubbly colored to eosinophilic, which gave the typical physaliphorous appearance. The nuclei were circular to oval, centrally located, vesicular, and had conspicuous nucleoli. Mitotic activity was not conspicuous, indicating a low proliferative index. This intervening stroma was of myxoid character and served to add to the gelatinous aspect of the tumor. Also, the areas of hemorrhage and necrosis in the tumor were detected, which revealed focal tissue destruction. The diagnosis based on the micromorphological appearance above was suggestive of chordoma.

### 4. IMMUNOHISTOCHEMISTRY

The immunohistochemical study is central in the diagnosis of chordoma, especially when the histologic characteristics mimic other myxoid tumors. Brachyury is one of the numerous markers that have become highly sensitive and specific nuclear marker to chordoma, indicating its derivation from remnants of the notochordal. Its expression is generally not found in histologic mimics like chondrosarcoma and metastatic mucinous carcinomas, and as such, it is a good diagnostic tool.

In the current case, immunohistochemical confirmation with Brachyury was recommended to support the impressions of the histopathology [4]. Nevertheless, the immunohistochemical work-up results were either pending or not available when the results were reported. However, the typical appearance with the microscope and clinical presentation made a diagnosis of sacrococcygeal chordoma highly favored.

### 5. DISCUSSION

#### Overview of chordoma

Chordoma is a bone malignant neoplasm that is rare and which develops out of the remnants of the embryonic notochord. These residues remain along the axial skeleton and justify the preference of chordoma for the midline structures. The sacrococcygeal area is the most frequently affected area, accounting for almost half of all cases, and then next comes the clival and mobile spinal areas [5]. The chordomas have slow but consistent growth, and local invasion is very likely, but distant metastasis is

unlikely to occur quickly. Though they are largely indolent, they present tremendous treatment challenges because of anatomical limitations, as well as their local recurring tendencies.

### Clinical correlation

In clinical practice, chordomas have an insidious onset, and the symptoms appear after months or years. The presenting complaint is most often pain, which is most often accompanied by a slowly growing mass. In the sacrococcygeal lesions, the patients can complain of pain in the glute, sitting difficulties, or local pressure effects. In severe cases, tumors press on the neighboring neural structures, causing neurological impairments or bowel and bladder difficulties. The patient in the case had gluteal pains and a slowly progressive mass, which is typical of the clinical behavior of sacrococcygeal chordoma [6]. The tumor is slow growing, and as a result, it is usually diagnosed later because the symptoms are mild and nonspecific, and the lesion may reach a significant size before being detected.

### Histopathology

Diagnosis of chordoma still relies on the histopathological examination. The characteristic microscopic appearance is the presence of physaliphorous cells - huge polygonal cells with excessive vacuolated bubbly cytoplasm - in sheets or lobules in a myxoid stroma. The nuclei are generally vesicular, centrally located, and round to oval-shaped. The mitotic activity tends to be low, indicating a slow rate of growth of the tumor. In the current case, the histomorphology characteristics observed, such as background of myxoid, inconspicuous mitosis, hemorrhage, and necrosis area, were a strong indication of chordoma [7].

Nevertheless, chordoma is morphologically similar to a number of other myxoid neoplasms, and it should be carefully differentiated. Myxoid chondrosarcoma does not contain physaliphorous cells and is usually characterized by chondroid differentiation. Signet ring cells will also be found in metastatic mucinous carcinoma, but the carcinoma will tend to exhibit more cytological atypia and clinical features of an initial tumor in another location. Although myxopapillary ependymoma is also found in the sacrococcygeal region, papillary architecture and glial differentiation are found, which, in this instance, were not evident.

Histopathological correlation is, therefore, necessary to reduce the diagnostic opportunities.

### Immunohistochemistry role

Immunohistochemical tests are important in the diagnosis of chordoma, especially in those cases that have overlapping histological characteristics. A transcription factor in the notochordal development, namely, Brachyury, is regarded as the most sensitive and specific immunohistochemical marker of chordoma [8]. The expression of its nucleus also differentiates chordoma and histologic mimics (chondrosarcoma, sarcoma, and metastatic mucinous carcinoma), which are usually negative and negative for Brachyury. In the current case, an immunohistochemical study of Brachyury was recommended as a measure to corroborate the histopathological outcome, which showed the sensitivity of ancillary examinations to obtaining diagnostic reliability.

### Management and prognosis

Wide surgical excision with negative margin is the main treatment mode of chordoma, which has the highest likelihood of providing local control of the disease. Nevertheless, it may not be possible to fully resect the tumor because it may be located too close to vital neurovascular vessels. High doses can be done first or as an adjuvant to radiotherapy, especially proton beam radiotherapy, which is believed to enhance local control, especially in individuals who have residual disease or positive margins [9]. Local relapses are also connected to chordomas, though they are accompanied by aggressive management relative to high recurrence rates, thus the need to carry out prolonged surveillance. Follow-up is also crucial in the long-term because the recurrence can take place several years following the initial treatment. Early detection and a multidisciplinary approach to treatment are some of the important issues in enhancing patient outcomes.

## 6. CONCLUSION

Chordoma is a rare notochordal malignant tumor that grows slowly but behaves aggressively locally and has a high recurrence rate. Clinical awareness of the disease is the key to its early detection because a late diagnosis is associated with disease progression and morbidity. Diagnosis is still based on the histopathological examination, where characteristic

physaliphorous cells and myxoid background give significant diagnostic hints. Confirmation of the diagnosis should be done through an immunohistochemical test on Brachyury to make an absolute diagnosis and to differentiate chordoma and its histological imitators. The best approach to management is a multidisciplinary approach that includes pathologists, surgeons, and oncologists. The positive results of chordoma patients can be enhanced by a high index of suspicion and precise pathological diagnosis.

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