



Case Report

Sacro-coccygeal chordoma: A diagnostic challenge in surgical pathology

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ABSTRACT

Chordomas are rare, slow-growing, locally invasive malignant tumors with a very low incidence rate. The present case report of sacrococcygeal chordoma in a 46-years female who presented to our institute, was diagnosed on histopathological grounds, and subsequently confirmed with immunohistochemistry. This report also reinforces the role of histopathological examination and Brachyury expression in the diagnosis of chordoma.

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INTRODUCTION

Chordomas, arising from notochord remnants, are uncommon tumors with a reported incidence of 8.4 per 10 million population. The usual age of onset is in the 5th and 6th decades. Males are more commonly affected than females, with a reported male-to-female ratio of 1.6:1. Chordomas are commonly seen arising from the axial skeleton, with 29.2%, 32.8%, and 32% of the tumors arising from the sacrum, mobile spine, and skull base, respectively.¹ When arising from the lower spine, the 4th and 5th sacral vertebra tend to be the common sites of involvement.² Though considered

malignant, these are slow-growing but locally aggressive tumors with a low to late metastasizing tendency. Patients usually present with a large mass associated with pain and pressure symptoms.³

The primary histological differential diagnosis for chordoma is chondrosarcoma, and it is essential to differentiate the two as this has treatment implications. A differentiation between the two can be done with immunohistochemistry (IHC) panels including keratin and epithelial membrane antigen (EMA), which is positive in the case of chordomas. These markers, however, are not always reliable, as keratin may not always be positive. An important discovery in this regard was made when Brachyury, a transcription factor associated with notochord development, was found to be present in chordoma.⁴

Here we present a case of Brachyury positive sacrococcygeal conventional chordoma in a female with a relatively younger age at diagnosis. Written informed consent was obtained from the patient for this publication, and ethical approval was taken from Institutional Review Committee (IRC) of Nobel Medical College for this case report.

CASE REPORT

A 46-year-old female presented with a history of intermittent lower back pain for 12 years and swelling in the sacrococcygeal region for 6 months associated with bilateral lower limb pain. On physical examination, a vague, deep-seated, diffuse swelling measuring 6 cm x 5 cm was palpated in the sacrococcygeal region. It was firm to hard in consistency, painful, and tender. Magnetic Resonance Imaging (MRI) demonstrated a large, heterogeneously enhancing sacrococcygeal mass with significant extraosseous components causing mass effect and invading both gluteus maximus muscles. The imaging findings suggested a possible neoplastic etiology.

Image-guided core biopsy was performed. Histopathological examination of the core biopsy obtained showed well-demarcated tumor cells arranged in cords and small nests embedded within abundant extracellular myxoid matrix (fig. 1). The individual cells were epithelioid with clear to light eosinophilic, round nuclei, dispersed chromatin, and inconspicuous nucleoli. Some cells showed abundant bubbly cytoplasm imparting physaliphorous cells like morphology (fig. 2). A few atypical mitotic figures were also identified. The impression was made as conventional chordoma. To confirm the diagnosis, immunohistochemical analysis for Brachyury was done, which showed strong nuclear positivity in the tumor cells (fig. 3).

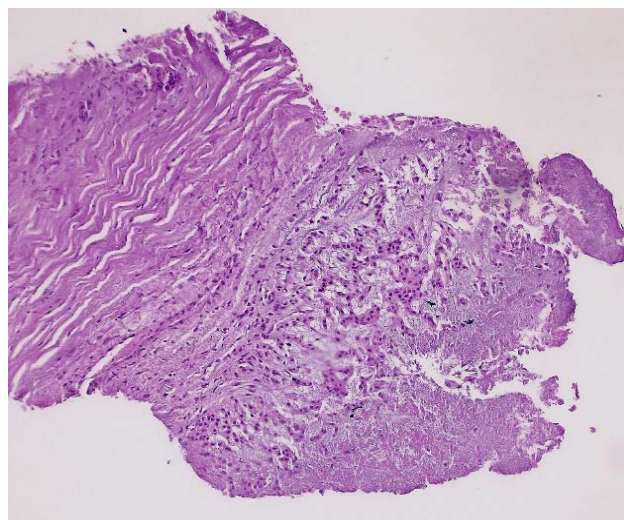


Figure 1: Chordoma. Cords of tumor cells embedded in a myxoid stroma. [H&E; 100x]

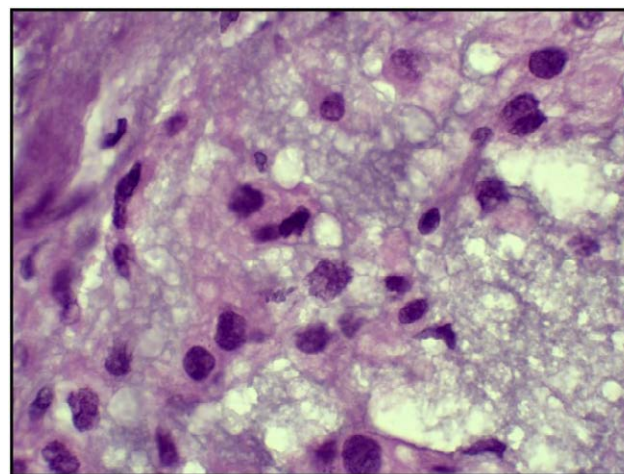


Figure 2: Chordoma. Large polygonal tumor cells with abundant bubbly cytoplasm (Physaliphorous cells). [H&E; 400x]

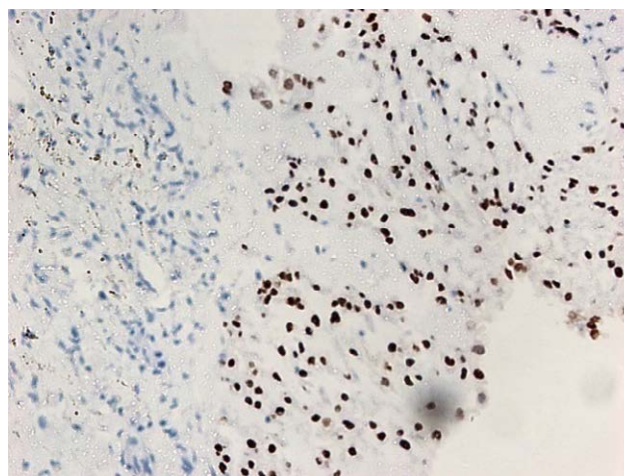


Figure 3: Immunohistochemistry shows strong nuclear positivity for Brachyury in tumor cells.

DISCUSSION

Chordomas, with an incidence rate of less than 0.1 per 100,000 per year, account for 1-4% of all malignant bone tumors.⁵ These tumors have a male preponderance with a median age of onset of 58.5 years, and patients generally present with a large mass (median size 8-9 centimeters) associated with pain and pressure symptoms.^{1,6-9} It originates from embryonic remnants of the notochord located along the neuraxis at developmentally active regions. Consequently, chordoma characteristically arises in the midline of the axial skeleton.¹⁰ Our case similarly involved the midline skeleton, highlighting its notochordal origin. Presenting features that have been reported in the literature include mass, sciatica, urinary tract infection, constipation, or cauda equina symptoms.¹¹

Chordomas often present with long-standing, nonspecific symptoms such as back pain, leading to delayed diagnosis. The prolonged history of intermittent lower back pain in our patient for over a decade reflects the slow-growing nature of chordomas. Symptoms usually arise due to mass effect on adjacent neural and soft tissue structures rather than early metastatic spread. The development of a palpable swelling and bilateral lower limb pain in our case likely corresponds to tumor progression with extensive extraosseous extension, which was also corroborated by MRI findings showing invasion into the gluteus maximus muscles. The most consistent MRI finding is a pronounced high signal on T2-weighted images, though this finding is non-specific.⁹

Histopathological examination remains the cornerstone of diagnosis. The classic microscopic appearance of conventional chordoma includes cords and nests of epithelioid cells embedded in an abundant myxoid matrix, with characteristic physaliphorous cells containing bubbly cytoplasm. These features were well demonstrated in the present case. However, distinction from histological mimickers—most notably chondrosarcoma—can be challenging, particularly in small biopsy samples, as both tumors share chondroid and myxoid features. This distinction is critical, as chordomas generally require more aggressive surgical management with wide excision, often combined with adjuvant radiotherapy, whereas treatment strategies and prognosis differ for chondrosarcoma.¹²

The belief that chordomas are notochordal in origin is further supported by the overexpression of brachyury in the tumor cells.¹³ A 2015 immunohistochemical study of over 5000 tumors, including 76 chordomas, found that brachyury expression was present in all the chordomas and none of the chondrosarcomas.¹⁴ Brachyury has since emerged as a potential pathologic marker for chordoma, with studies showing a sensitivity of 87-100% and an almost 100% specificity.⁴ Traditional epithelial markers such as cytokeratin and EMA are typically positive in chordomas but may show variable expression, particularly within chondroid areas. In a study including 12,399 non-chordoma tumors, 45 (0.36%) were found to have positive brachyury staining of variable intensity. Thirty-seven (0.3%), 7 (0.06%), and only 1 (0.01%) of the tumors were found to have weak,

moderate, and strong staining, respectively.¹⁵ It has thus been recommended that a definitive diagnosis of chordoma should include a triad of classic histopathological, MRI, and immunohistochemical findings.¹²

To our knowledge, there are fewer than 10 cases of chordoma reported from Nepal to date, of which this is the first case report demonstrating the utility of Brachyury IHC in its diagnosis. The utility of Brachyury is not just limited to diagnosis, but its role as a potential therapeutic target is also being explored, though the rarity of the tumor has limited significant progress.¹⁵ Although surgery and radiotherapy remain the mainstays of management, ongoing research into molecular pathways and immunotherapy, including Brachyury-targeted strategies, may improve outcomes for patients with chordoma in the future.

CONCLUSION

This case report emphasizes the importance of maintaining a high index of suspicion for chordoma in patients presenting with long-standing sacrococcygeal pain and mass lesions, even in younger females. It also highlights the indispensable role of histopathology supplemented by Brachyury immunohistochemistry in establishing an accurate diagnosis, which is crucial for guiding appropriate management and improving patient outcomes.

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