

Sacral Chordoma: A Case Report with Radiographic and Histologic Correlation and a Review of the Literature

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ABSTRACT

A case of sacral chordoma clinically simulating pilonidal 'cyst' in a 47-year-old male is presented. The clinical presentation with radiographic and histologic features of this entity with post-treatment clinical follow up is presented with a review of the literature.

INTRODUCTION

Chordoma is a rare tumor of bone originally described by Virchow in 1857¹ and further characterized by Ribbert in 1894.² In the United States, <300 cases of chordoma are diagnosed each year with a reported incidence of 0.5 per million.³ Chordoma is a tumor of notochord cell derivation. Although the histogenesis of chordoma has been recently challenged,^{4,6} the traditional understanding of chordoma is that it arises from notochord remnants along the neuroaxis. The most common sites of involvement include the sacrum/coccyx (50%), the sphenoccipital bones of the skull (35%), and the vertebral bodies (15%). Chordoma of the sacrum occurs more frequently in males than in females (2:1) but the skull base and vertebral chordomas develop equally in both sexes.^{3,7} Although chordomas have been reported in every age group, the vast majority of patients are in the 6th and 7th decade of life.⁹ We report a case of a sacrococcygeal chordoma in a 47-year-old male presenting clinically as a firm, palpable midline mass over the sacrum mimicking a pilonidal cyst.

CASE HISTORY

The patient is an otherwise healthy 47-year-old white

man who presented to his family physician after having slipped and fallen in his hot tub, which resulted in persistent pain in the low back area. A routine lumbo-sacral radiograph showed a fractured coccyx and a subtle, ill-defined lucency involving the sacrum and coccyx. The fracture was treated conservatively and no follow-up study was obtained. Soon thereafter, as a result of continued and increasing pain, the patient consulted a general surgeon for a second opinion. The surgeon found on exam an area of induration and tenderness along the inner gluteal fold extending into the gluteal cleft. There was no drainage or punctum. A pilonidal cyst with associated inflammation was the primary clinical consideration, and an unsuccessful incision and drainage procedure was performed. Unaware of the findings noted on the initial radiograph but concerned as a result of the unusual clinical presentation and magnitude of pain, the surgeon then performed a wedge biopsy of the area. When 'chordoma' was returned as the diagnosis, MRI studies of the low back and pelvis were performed (Figures 1 and 2).

Radiographic Correlation

Magnetic resonance images were obtained in axial, sagittal, and modified coronal planes using various T1 and T2 weighted sequences. The study included both unenhanced sequences (Figures 1 and 2) and gadopentetate contrast enhanced sequences (not shown). A large, well-defined, lobulated mass was identified at the sacrococcygeal level (Figure 1). The mass measured approximately 13.1 cm x 10.9 cm x 9.7 cm and exhibited mild T2 hyper-intensity with the appearance of internal septations and lobulation. The mass exhibited heterogeneous intermediate signal on T2 weighted images (Figure 2). On subsequent contrast-enhanced radiographs, only mild, peripheral increased signal intensity was noted. Destruction of the lower sacrum and upper coccyx with insinuation into the right gluteal musculature was present along with anterior displacement of the rectum without invasion thereof.

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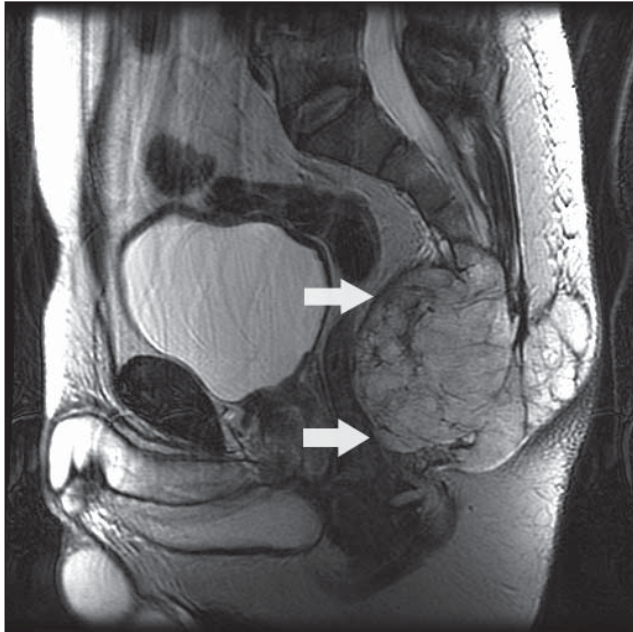


Figure 1. Sagittal T2 weighted unenhanced fast spin echo MRI – arrows indicate anterior border of mass.

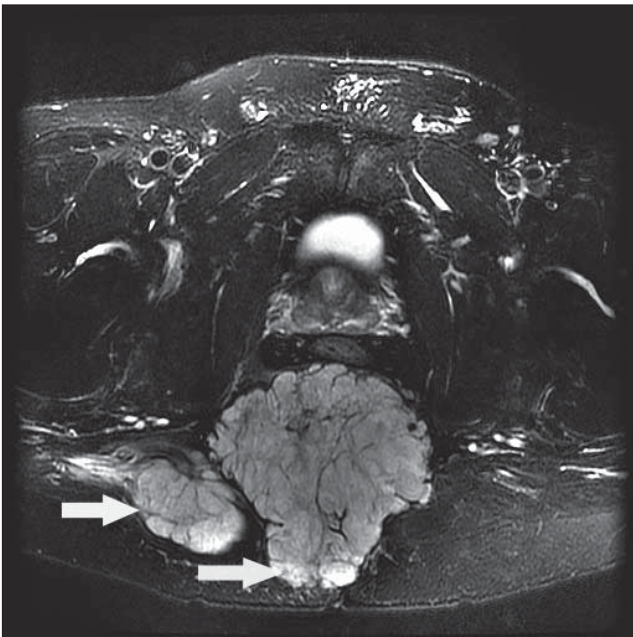


Figure 2. Unenhanced axial T2 fast spin echo with fat saturation – arrows indicate right lateral and posterior borders of lobular mass.

Histologic Correlation

The wedge biopsy of the subcutaneous component to the sacral mass showed irregular nests and cords of tumor cells supported by a basophilic, mucoid matrix. The cells displayed round to oval nuclei, ample eosinophilic cytoplasm, and normal nuclear/cytoplasmic ra-

tios. Many of the cells showed prominent cytoplasmic vacuoles (Figure 3). No necrosis or mitotic figures were identified. A special histochemical stain for Alcian Blue at pH 2.5 (for acid mucin) highlighted the background matrix (Figure 4). Immunohistochemical studies for a pan-keratin cocktail and S-100 were also performed on the formalin-fixed tissue and showed the tumor cells to be immunoreactive for antibodies to these epitopes.

DISCUSSION

Chordoma is a tumor that comprises <1%-3% of primary bone tumors^{3,7} and is generally considered a slow-growing, locally invasive neoplasm with a variable capacity to metastasize.^{3,7,9,12} The reported incidence of associated metastasis ranges from 7% to 48%.^{3,7,9} Chordoma occurs predominantly within the bony pelvis (sacrum and coccyx) and the sphenococcyx areas of the skull (primarily the clivus). Historically, chordomas were thought to arise from embryonic notochord ‘remnants’ (termed “physaliferous” cells or “bubble bearing” cells¹) present along the length of the neuroaxis at developmentally active sites. These sites are the ends of the neuroaxis and the vertebral bodies. In exceptionally rare instances chordomas can also occur in extra-axial locations.^{3,7,9} Recent studies⁴⁻⁶ suggest that chordomas actually arise from pre-existing benign notochord cell ‘tumors’ with inherent neoplastic potential and not from remnants of the notochord. Histologically, chordoma is a tumor composed of epithelioid cells arranged in cords and nests. These cells display characteristic irregular intra-cytoplasmic vacuoles and are usually supported by a mucoid-like matrix that appears blue-gray on formalin-fixed tissue stained with hematoxylin and eosin and brilliant blue when stained with Alcian blue at pH 2.5. Immunohistochemistry of chordoma shows reactivity to antibodies for keratins (epithelial markers) and S-100 (neural tissue marker). The primary differential diagnosis is that of metastatic carcinoma with a striking similarity to metastatic renal cell carcinoma. Successful treatment involves the complete radical resection of the tumor mass. Although chordoma is resistant to the effects of conventional radiation therapy,^{3,7} some institutions combine the use of post-operative high LET radiation (linear energy transfer) with surgery to attempt to increase disease-free intervals involving cases in which the tumor was not completely resected.⁸ One unique approach in incompletely resected or unresectable tumors is the use of percutaneous radiofrequency ablation of the tumor.¹¹ Irrespective of therapy, long-term studies of sacral chordomas show a 5-year survival rate of 51% and a 10-year survival rate of 35%.^{3,9,10} Of those

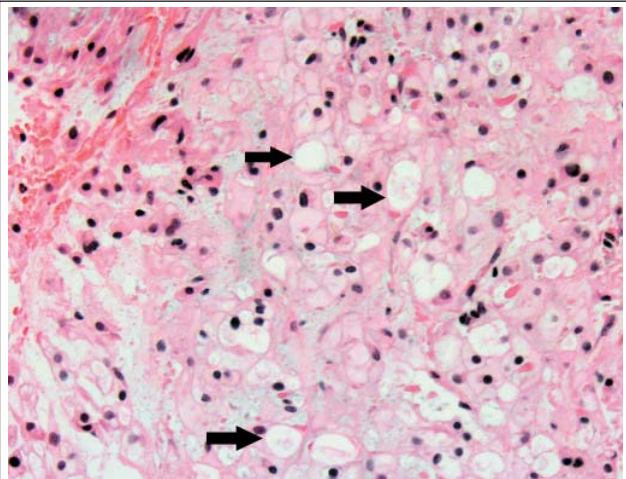


Figure 3. Biopsy - 20X. Arrows indicate 'bubble cells.'

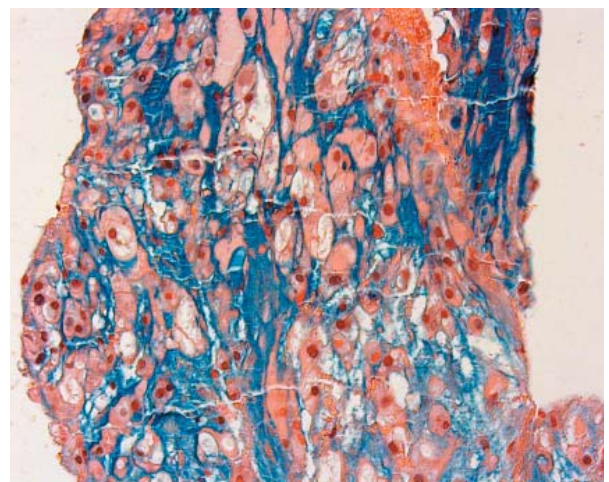


Figure 4. Alcian Blue. Myxoid cell matrix stains blue.

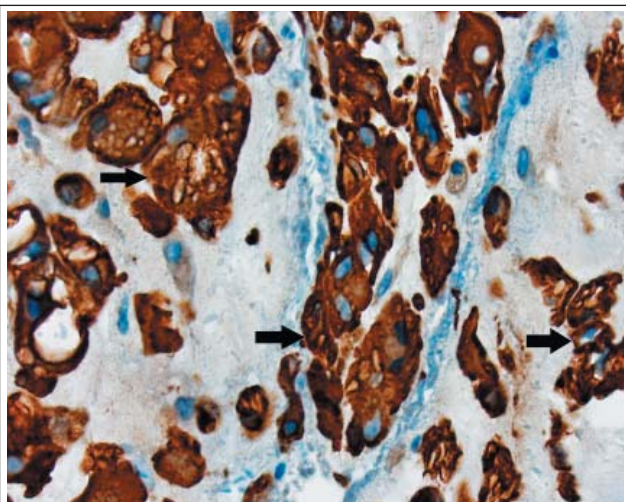


Figure 5. Immunohistochemistry for Pankeratin - 20X. Arrows indicate brown immuno-reactive cells.

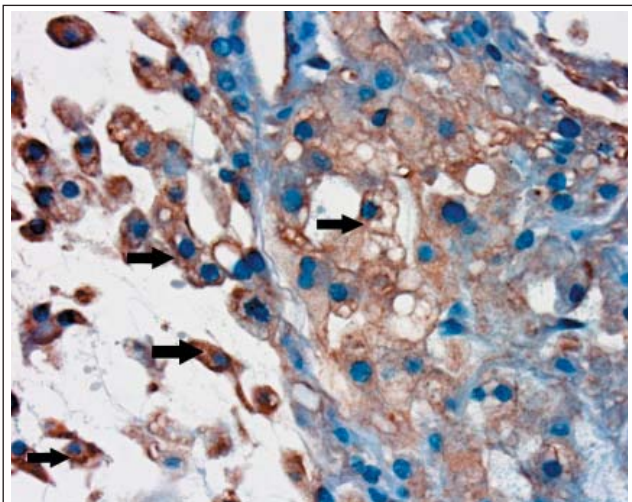


Figure 6. Immunohistochemistry for S-100. Arrows indicate brown immuno-reactive cells.

patients surviving for >5 years the recurrence rate for chordoma is 70%.⁸

CLINICAL FOLLOW-UP

The patient was referred to Johns Hopkins University Hospital, Baltimore, MD, for definitive treatment. The biopsy slides were reviewed by consulting Johns Hopkins' pathologists and the diagnosis of chordoma was confirmed. The Departments of Orthopedic Surgery and Neurosurgery made a consensus recommendation for complete resection of the tumor. Surgical intervention resulted in the en bloc removal of a 4.7 kg lobular, firm gray-tan tumor. The procedure required total S1 and S2 laminectomies with partial S3, S4, and S5 laminectomies, a high sacral amputation with osteotomy at the mid-S2 level, removal of coccyx, and partial bilat-

eral sacro-iliac joint removal. Reconstruction of the soft tissue in the area of the lower spine was accomplished using a gluteus maximus myofascial flap and superficial closure by a fasciocutaneous flap. The surgical pathology of the tumor was consistent throughout all areas of the mass and confirmed the biopsy interpretation. The surgical margins were considered focally involved by tumor. No post-operative radiation was given. The patient underwent extensive physical rehabilitation but was impaired by bowel and bladder dysfunction and erectile dysfunction. He remained free of radiographic evidence for tumor for 20 months. The most recent serial MRI studies obtained 20 and 26 months after radical resection of the tumor showed 2 separate areas of abnormal signal intensity involving the soft tissues adjacent to the right posterior margin of the original tumor mass,

the right ischium, and the anterior aspect of the right femoral head. These areas displayed slight increase in size between the 20th and 26th month consistent with recurrence of tumor.

SUMMARY

Chordoma is a rare neoplasm of notochord derivation that is hard to diagnosis and difficult to treat. Complete resection of the tumor is the only cure. Because of slow and insidious growth, sacral chordomas are usually large, bulky masses by the time of diagnosis. As a result, attempts at radical resection of the tumor are frequently inadequate. In addition, chordomas are resistant to the effects of traditional radiation and chemotherapeutic agents making standard adjuvant treatment a debatable and perhaps a controversial issue. Our case demonstrates a bulky 4.7 kg tumor arising in the sacrum with superficial extension of tumor into the pre-sacral subcutaneous soft tissues clinically simulating a pilonidal 'cyst.' Radical resection was attempted with the large tumor mass showing focal margin transaction indicating probable microscopic foci of residual tumor. Serial MRI studies obtained post surgically, however, were negative for evidence of tumor and no adjuvant therapy was given. The patient remained free of radiographic evidence for tumor for 20 months. The 2 most recent studies, at 20 and 26 months, now show changes consistent with tumor recurrence in several foci including pelvic soft tissues, ischium, and right femoral head. The clinical outcome of this case is typical for chordoma and reinforces the need for aggressive therapy in the treatment of chordoma. Aggressive therapy should include the attempt of complete resection of the tumor and, in those cases in which the surgical margins are involved by chordoma, additional consideration should be given to adjuvant high linear energy transfer (LET) therapy.

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