



Clear Cell Chondrosarcoma of the Sacrum

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Abstract

The authors report on a rare case of clear cell chondrosarcoma arising from the sacrum in a 26-year-old man. The patient was treated with *en block* excision of the tumor and reconstruction with instrumentation and bone grafting. There was no local recurrence or metastasis of the tumor at a follow-up examination 30 months postoperatively. To our knowledge, this is the first reported case of clear cell chondrosarcoma occurring in the sacrum.



Introduction

Clear cell chondrosarcoma is a rare malignant bone tumor of a cartilaginous origin. It is characterized by large tumor cells with clear cytoplasm arranged in sheets [1,6-8,10,11]. It is more common in men than in women with a ratio of 2:1 [1]. The age range is wide; however, most patients are in the third or fourth decades of life [1]. Clear cell chondrosarcoma usually involves the epiphyseal ends of the long bones, particularly the proximal femur and humerus [1-3,6-11]. Lesions arising from flat bones are rare [1,5,8].

Case Report

A 26-year-old man presented with a one-month history of increasing pain in his left buttock and calf. The patient could not stand or walk without assistance due to severe pain, and complained of urinary incontinence. He had no previous medical history of systemic illness.

Physical examination revealed soft tissue swelling and tenderness over the left side in the posterior aspect of his sacrum, but no mass was palpated. Neurological examination showed a decreased sensation to touch in the left S1 to S5 dermatomes. The patient had three fifths of his normal strength in the left hamstrings muscles, and the extensors and flexors of his left ankle and great toe. The patellar tendon reflex was normal, but the Achilles tendon reflex was absent in the left leg. Rectal examination revealed extremely weak sphincter tone.

Radiographs of the lumbar spine were normal, but radiographs of the pelvis revealed a large osteolytic lesion in the left sacrum (Fig. 1).



Fig. 1 An anteroposterior radiograph of the pelvis shows a large osteolytic lesion in the left sacrum (arrows).

Computed tomography of the sacrum demonstrated an intrasacral expansive lesion with cortical destruction, extending into the retroperitoneal spaces (Fig. 2). Angiography showed hypervascularity of the lesion which was supplied by vessels from the lateral sacral and the superior gluteal arteries.



Fig. 2 A CT scan shows an intrasacral expansile lesion with cortical destruction, extending into the retroperitoneal spaces.



A core-needle biopsy yielded a tumor tissue consisting of round to polygonal cells with clear cytoplasm arranged in sheets, multinucleated giant cells, and aneurysmal bone cyst components. The preoperative presumptive diagnoses included giant cell tumor of bone, aneurysmal bone cyst, chondrosarcoma, and clear cell chondrosarcoma. The patient subsequently underwent an *en block* excision of the left half of the sacrum using combined anterior and posterior approaches. The pelvis was stabilized with a sacral bar and Zielke's rods, and autogenous bone grafting from both fibulae was performed between the remaining sacrum and the left iliac crest.

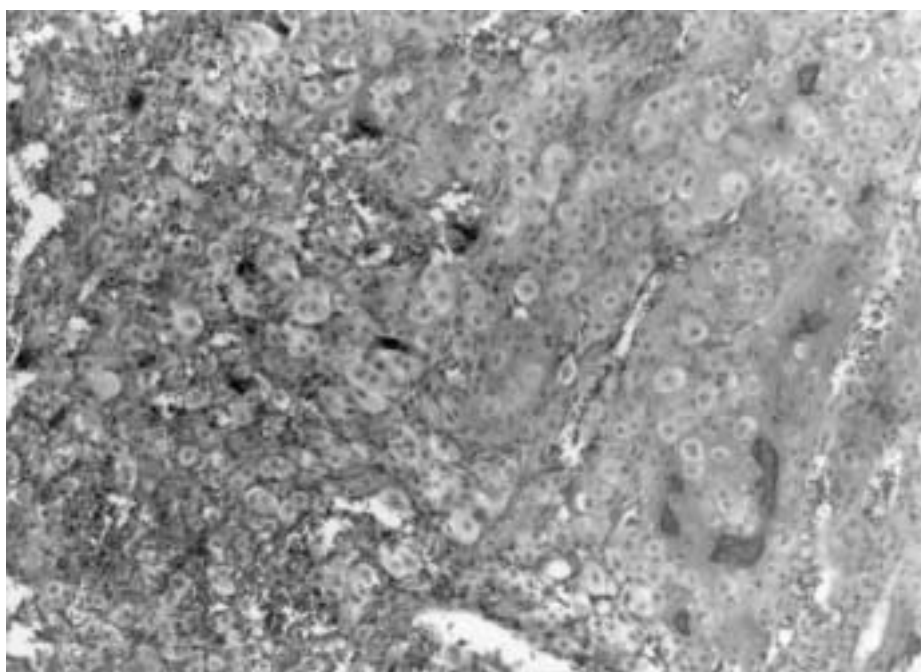


Fig. 3 *Histological specimens excised at surgery show numerous, round tumor cells with clear or slightly eosinophilic cytoplasm, arranged in sheets (hematoxylin and eosin, original magnification x 200).*

Grossly, the tumor was friable and brown to yellow in color, measuring 8x8x6 cm in size. It contained multiple small hemorrhagic cysts. Histological, the tumor showed sheets of tumor cells with clear or slightly eosinophilic cytoplasm in a scant stroma (Fig. 3). Cytoplasmic borders were distinct. The cells had round nuclei placed



centrally in the cytoplasm. Mitotic figures were infrequent. Chondroid islands with calcification were occasionally observed (Fig. 4). Secondary aneurysmal bone cyst components were observed in some areas. Periodic acid Schiff's stain demonstrated glycogen particles in the cytoplasm of the tumor cells. The histological diagnosis was clear cell chondrosarcoma.

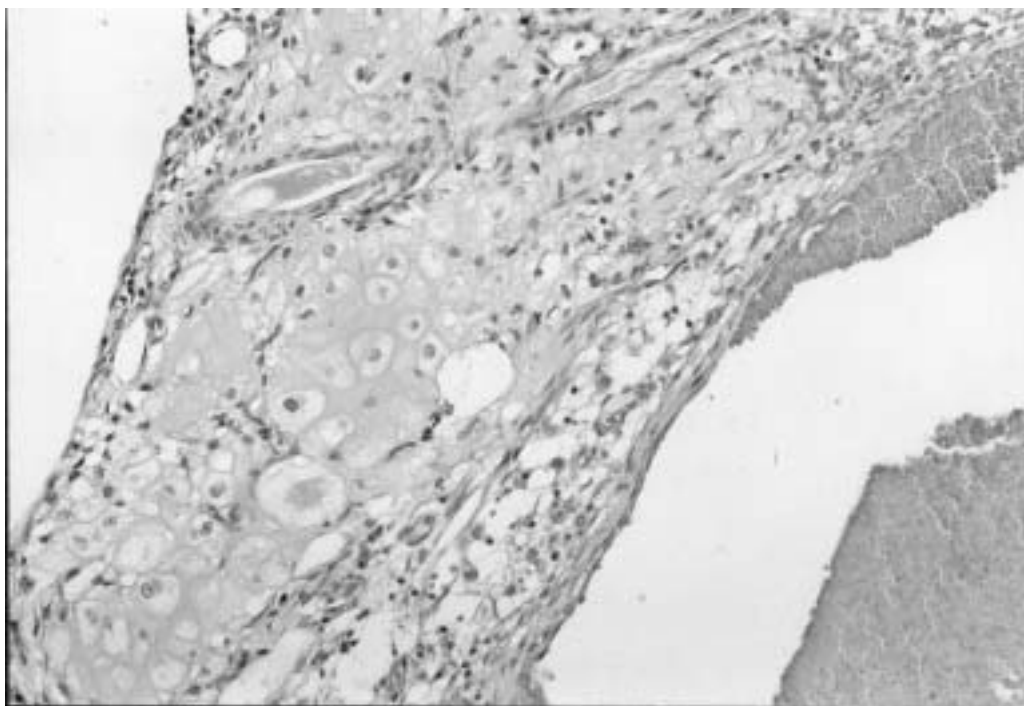


Fig. 4 Areas showing chondroid islands and aneurysmal bone cyst-like vascular spaces (hematoxylin and eosin, original magnification x 400).

The postoperative course was uneventful. At a follow-up examination 30 months after surgery, there was no evidence of local recurrence or distant metastasis of the tumor. The patient could walk without crutches, although neurological deficits of the left S1 and S2 distribution remained.

Discussion

Clear cell chondrosarcoma is rare and accounts for only 2% of all chondrosarcomas [8,9]. The tumor has a marked propensity to affect the epiphyseal ends of the long



bones [1-3,6-11]. Approximately 60% of the tumors are located in the proximal femur, followed by, in descending order, the proximal humerus, the distal femur, and the proximal tibia [1,8,10]. Flat bones such as the spine, ribs, pelvis and skull are infrequently involved [1,5,8,10]. The sacrum is a rare site for clear cell chondrosarcoma with no cases having been described in the English-language literature, to our knowledge.

Whether clear cell chondrosarcoma represents a subtype of chondrosarcoma or is the malignant counterpart of chondroblastoma is unknown. Mirra [8] believes that clear cell chondrosarcomas develop from longstanding, undetected chondroblastomas, because both tumors involve the epiphyseal ends of the long bones and share similar radiological features. In addition, clear cell chondrosarcomas occasionally contain chondroblastoma-like cells [6].

Radiographic appearance of clear cell chondrosarcoma is nonspecific. More than 60% of clear cell chondrosarcomas show purely osteolytic lesions, and the remainder contain areas of calcification within the tumor [1]. Bone expansion is commonly seen. Cortical destruction is observed in approximately 20% of the tumors [1]. Thus, biopsy is necessary to establish the diagnosis in most cases.

Histological features of clear cell chondrosarcoma are characterized by large clear cells arranged in sheets, with areas of conventional chondrosarcoma [1,6-8, 10,11]. In addition, the tumor might contain areas resembling aneurysmal bone cyst, giant cell tumor of bone, osteosarcoma, osteoblastoma, and chondroblastoma [1,2,6,8,11]. These secondary structures are potentially a frequent source of misdiagnosis. Our case typically showed areas of clear cells and areas of secondary aneurysmal bone cyst.

Clear cell chondrosarcoma is a low-grade malignancy, and Bjornsson et al. reported



that the overall mortality was 15% in a series of 47 patients with the condition [1,4]. However, since a high recurrence rate (more than 80%) is reported in patients who underwent simple excision and curettage, *en block* excision of the tumor is recommended as the minimum operative procedure [1,9]. Hartwright et al. [4] reported a patient with clear cell chondrosarcoma, who developed a local tumor recurrence after 19 years after the original occurrence. Close monitoring over an extensive period of time is recommended for all cases of clear cell chondrosarcoma.

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