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CASE REPORT

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Abstract We report the case of a clear cell chondrosarcoma (CCCS) occurring in the femoral head of a 14-year-old skeletally immature boy. Radiographic examination revealed a well-defined, osteolytic lesion in the epiphysis of the femoral head. Given the patient's age and the radiographic appearance of the lesion, chondroblastoma was high on the differential diagnosis. A frozen section was performed at the time of open biopsy was felt to be consistent with either chondroblastoma or CCCS. CCCS in a skeletally immature patient was felt to be unlikely, so curettage and bone grafting was performed. Final pathology review, however, confirmed the diagnosis of CCCS. The patient was taken back to surgery 4 weeks later for a wide resection and hemiarthroplasty.

Keywords Clear cell chondrosarcoma · Chondroblastoma · Skeletally immature · Femoral head · Radiograph · CT · MRI

Introduction

Clear cell chondrosarcoma (CCCS) is an uncommon variant of chondrosarcoma first described by Unni et al. in 1976 [1]. It is composed of chondrosarcomatous tissue characterized by large, round-to-oval cells with abundant clear cytoplasm and scattered giant cells. It most commonly occurs in the epiphysis of the proximal femur or proximal humerus of patients in the third to fifth decade of life. We report on a case of CCCS mimicking a chondroblastoma in a skeletally immature 14-year-old boy.

Case report

A 14-year-old boy presented with complaints of mild to moderate pain in his left groin for 6 months. He denied recent trauma and the remainder of his past medical history was normal. Physical examination was unremarkable except for slight irritability and decreased range of motion of the left hip. Radiographs revealed a well-defined, osteolytic lesion in the epiphysis of the femoral head (lost). Computed tomography (CT) revealed a lytic lesion involving one third of the femoral head with a thin, well-defined sclerotic rim (Fig. 1). There was no internal matrix mineralization. A radionucleotide bone scan showed intense uptake throughout the femoral head (not shown). Magnetic resonance imaging (MRI) showed edema

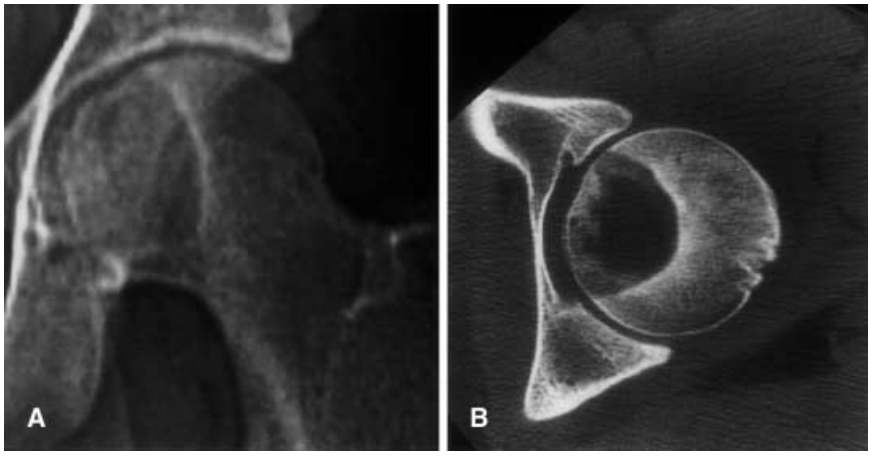

Fig. 1 **A** Scout image from CT scan. There is a large, well-defined lucency with a sclerotic rim in the medial femoral head. **B** Cross-sectional image from CT. The subchondral cortex is thin and somewhat irregular

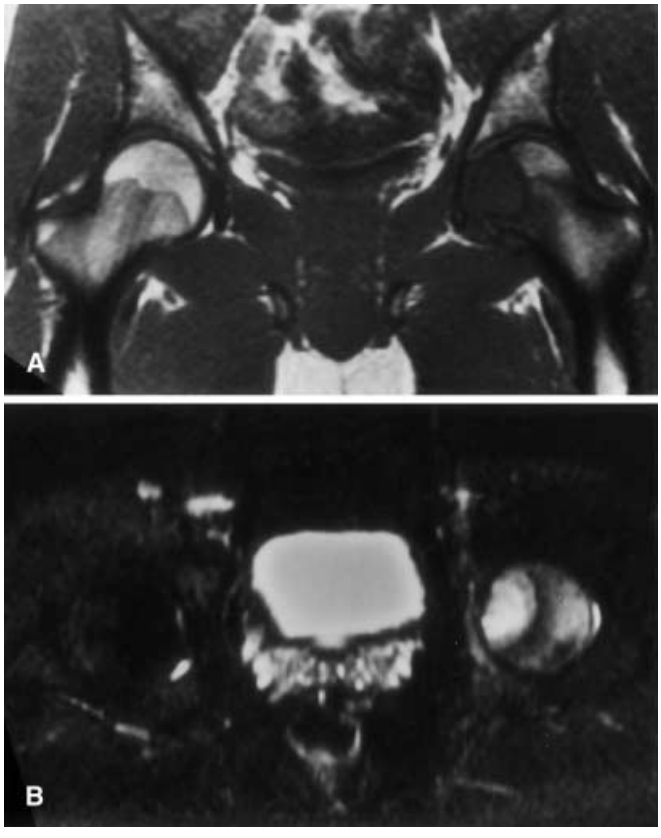

Fig. 2 **A** Coronal T1-weighted MR image (500/11). The lesion is well defined. It involves both the epiphysis and metaphysis of the proximal femur. The surrounding marrow is lower in signal intensity than in the normal right proximal femur. This represents marrow edema. **B** Axial inversion recovery MR image (5000/68/150). The lesion has high signal intensity. The surrounding marrow also is increased in signal intensity, while the normal right femoral head decreases in signal intensity as a result of fat suppression

around the lesion (Fig. 2). The patient's age and the radiographic appearances suggested a preliminary diagnosis of chondroblastoma.

The patient underwent open biopsy through an anterolateral approach. Review of the frozen section revealed a myxo-cartilaginous lesion containing giant cells and delicate calcifications. This resulted in a differential diagnosis that included chondroblastoma and CCCS. Given the patient's age and the presence of edema on MRI, it was felt that CCCS was unlikely. Thus, the curettage and bone grafting was completed as initially planned. In order to prevent possible contamination of the iliac crest bone graft harvest site, the iliac crest was draped separately from the operative site at the hip. Also, separate instruments and new gloves were used. These precautions are taken with all bone tumor cases, whether presumed benign or malignant.

Final review of the pathology revealed a lesion characterized by a complex matrix that included chondroid material with delicate calcification, in addition to the formation of small islands of woven bone (Fig. 3). As in the frozen section, characteristic cells with clear cytoplasm and scattered multinucleated giant cells were present. A final diagnosis of CCCS was rendered. The patient was taken back to surgery 4 weeks later for a wide resection of the left femoral head and neck followed by a hemiarthroplasty. Residual tumor was found in the specimen. Surgical margins were negative for tumor, and the patient had an uneventful postoperative recovery. At 24 months after surgery the patient has no evidence of disease and a hip hemiarthroplasty that is functioning well.

Discussion

CCCS is a rare variant of chondrosarcoma first described by Unni et al. in 1976 [1]. It accounts for less than 2% of all chondrosarcomas and classically occurs in the epiphysis [2, 3]. The proximal femur is the most common site, followed by the proximal humerus, distal femur and proximal tibia. The tumor has also been reported to occur in the pelvis, skull, scapula, ulna, phalanges, talus, rib and vertebrae [3, 4]. CCCS most commonly occurs in adults in their third to fifth decade [1, 2, 3, 5, 6, 7]. The youngest patient in a series of 47 cases from the Mayo Clinic was 14 years of age [3]. In a review of the English language literature, the youngest reported case of a CCCS occurred in the pelvis of a skeletally immature 12-year-old boy in Japan [8]. No other reports of CCCS occurring in skeletally immature individuals were found.

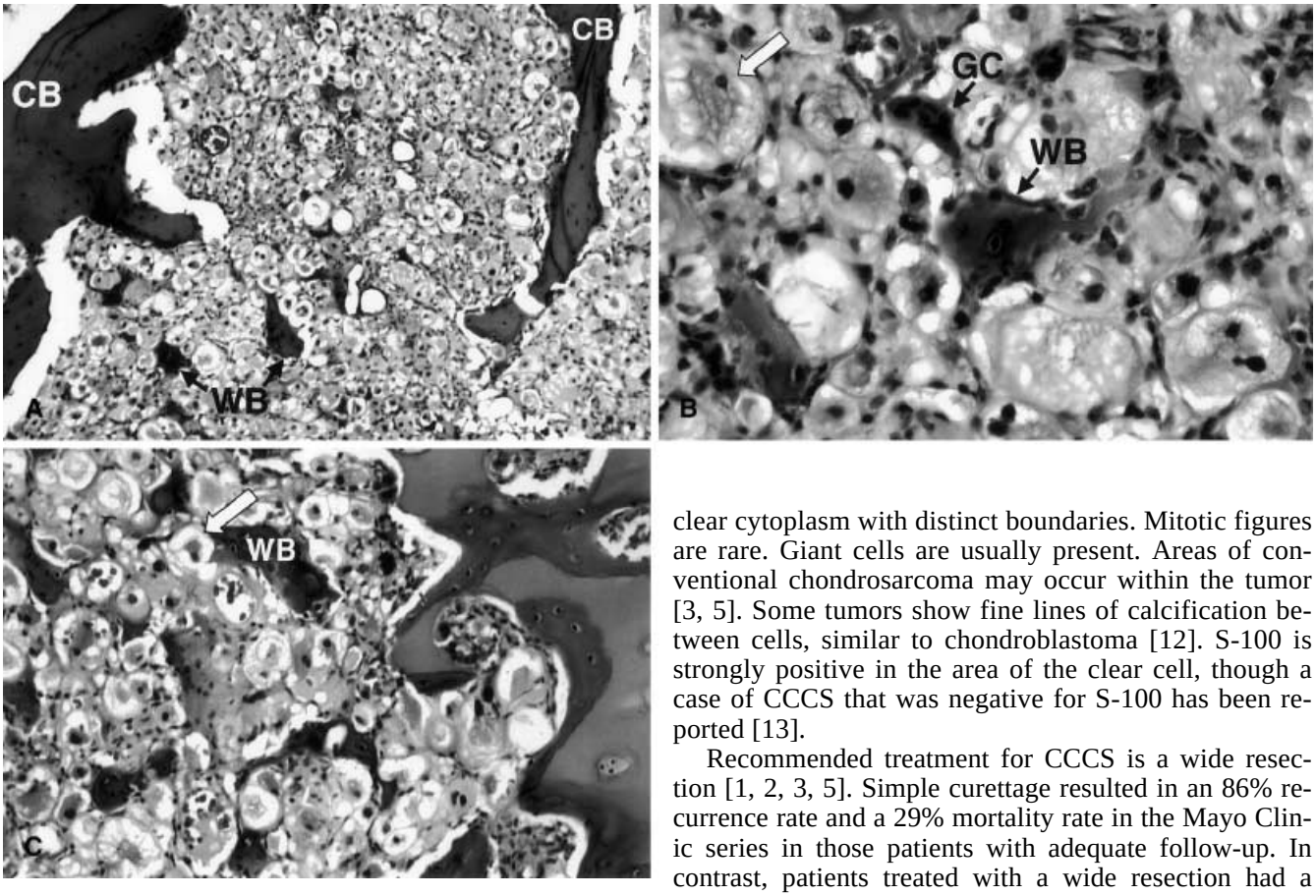

Fig. 3A–C Photomicrographs showing replacement of marrow space between cancellous bone (*CB*) by tumor (**A**: H&E, ×100), and classic features including chondroctyes with abundant cytoplasm (*white arrow*), islands of woven bone (*WB*), and osteoclast type giant cells (*GC*) (**B**: H&E, ×200; **C**: H&E, ×400)

The most common radiographic appearance for CCCS is a lucent lesion located at the epiphysis of a long bone [1, 3]. Areas of calcification within the tumor can occur. Twelve of 36 tumors in the Mayo series that had radiographs available for review had soft, fluffy areas of calcification rather than the discrete clumps typically seen in conventional chondrosarcomas. Expansion of the bone occurred in the majority of cases, though the degree of expansion was usually slight. A sharp interface between bone and tumor was usually present, though sclerotic and indistinct borders have also been described [1, 3]. MRI scans characteristically do not demonstrate edema around the lesion [9] as opposed to certain benign bone tumors, to include chondroblastoma [10, 11].

Grossly, the lesion does not have the characteristic translucent appearance of a low-grade chondrosarcoma but rather is either soft or more solid and gritty, depending on the amount of bone and calcification present [1, 3, 5]. The microscopic hallmark is the clear cell, which has a centrally placed, round nucleus surrounded by clear cytoplasm with distinct boundaries. Mitotic figures are rare. Giant cells are usually present. Areas of conventional chondrosarcoma may occur within the tumor [3, 5]. Some tumors show fine lines of calcification between cells, similar to chondroblastoma [12]. S-100 is strongly positive in the area of the clear cell, though a case of CCCS that was negative for S-100 has been reported [13].

Recommended treatment for CCCS is a wide resection [1, 2, 3, 5]. Simple curettage resulted in an 86% recurrence rate and a 29% mortality rate in the Mayo Clinic series in those patients with adequate follow-up. In contrast, patients treated with a wide resection had a 16% recurrence rate and an 8% mortality rate [3]. As with other low-grade chondrosarcomas, neither chemotherapy nor radiation therapy are indicated.

The interesting aspects of this case are the lesion's close resemblance to a chondroblastoma and the patient's young age. Because of its epiphyseal location, distinct borders and occasional calcifications, CCCS is known to mimic chondroblastoma [11, 14]. However, CCCS most commonly occurs in the third to fifth decade, whereas chondroblastoma occurs in the first and second decades [3, 12]. The previously published case in a 12-year-old boy occurred in the pelvis, not the classic epiphyseal location [8]. Thus, CCCS has not traditionally been included in the differential diagnosis of epiphyseal lesions in skeletally immature patients. In addition, the finding of significant marrow edema on MRI, as in our case, is felt to be a frequent finding with chondroblastoma [10, 11] but not CCCS [9]. This case highlights the rare occurrence of CCCS as an epiphyseal lesion in a skeletally immature individuals, indistinguishable on imaging from chondroblastoma. Also, marrow edema demonstrated on MRI does not preclude the diagnosis of CCCS. After biopsy confirmation of the diagnosis, treatment should consist of resection of the lesion with a margin of normal bone. As with other chondrosarcomas, long-term follow-up must be performed to screen for recurrence.

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