

Successful en Bloc Resection for Femoral Head Clear Cell Chondrosarcoma Without Biopsy: A Case Report

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Abstract

Background: Clear cell chondrosarcoma (CCCS) is a rare, low-grade, malignant chondrogenic bone tumour. This tumour commonly occurs at the epiphysis of long bones, particularly in the proximal femur.

Case presentation: This report describes a 58-year-old man with right hip pain since 5 months. Plain radiography, magnetic resonance imaging (MRI), and computed tomography (CT) revealed the characteristic appearance of chondroid mineralisation in the right femoral head, suggesting typical CCCS. Although a biopsy is the gold standard for definite diagnosis before treatment, wide resection with removal of the biopsy tract is thought to negatively affect surgical margin and postoperative hip function. En bloc resection without a biopsy and a hip hemiarthroplasty were performed instead. The pathological diagnosis was CCCS, and an adequate surgical margin was obtained. No local recurrence or distant metastases were found, and postoperative function was excellent at the final follow-up.

Conclusion: The femoral head is a typical location of CCCS. Wide resection with adequate margins is the main treatment strategy for CCCS. When radiological features are typical, performing an en bloc resection without performing a biopsy is an acceptable treatment that may improve patient outcomes.

Background

Clear cell chondrosarcoma (CCCS) was first described as a clear cell variant of chondrosarcoma by Unni et al. (1). CCCS is extremely rare, accounting for only 2.5–2.7% of all chondrosarcomas (2, 3). CCCS affects men more frequently across widely distributed age groups (4). This lesion characteristically occurs at the epiphyses of long bones, especially in the proximal femur and humerus (5). Radiological and pathological diagnosis is challenging due to its rarity; this can commonly lead to inadequate surgical treatment. Patients who underwent initial curettage had a high rate of local recurrence; however, patients who underwent wide resection had a low recurrence rate (4). The initial surgical margin of surgery for CCCS was correlated with the oncological outcome (5).

Biopsy has been the gold standard for diagnosis and management of malignant bone tumours; unfortunately, tumour cell seeding in the biopsy tract is a risk factor of local recurrence. Therefore, removal of the biopsy tract is widely accepted (6). This often requires sacrifice of more healthy tissues and may result in poor postoperative limb function.

In this case report, we describe a typical case of CCCS occurring in the right femoral head that was highly suspected using only radiological methods. The patient was successfully treated with an en bloc resection without requiring a biopsy of the lesion.

Case Presentation

A 58-year-old man reported with right hip pain since 5 months. The patient visited a nearby clinic, and an abnormality in the right femoral head was found after taking a plain radiograph. The patient was referred

to Osaka city university hospital During the initial visit, plain radiography revealed fuzzy, irregular calcifications on the femoral head with an indistinct distal margin (Fig. 1). Magnetic resonance imaging (MRI) was taken with iso signal intensity on the T1-weighted image (T1WI: Fig. 2a) and with heterogeneously high signal intensity with a focal enhancement of spotty areas (T2WI: Fig. 2b). The lesion was peripherally enhanced with contrast material on the T1 fat-suppressed image. This decreased spotty signal intensity in central areas, which then revealed matrix mineralisation (Fig. 2c). Computed tomography (CT) also revealed a honeycomb-like calcification on the femoral head (Fig. 2d). No cortical destruction was observed. The tumour size was 40 mm × 30 mm × 42 mm. At this point, CCCS was highly suspected as being the cause. En bloc resection and a hip hemiarthroplasty (Fig. 3) were performed through invasive mini-incision anterolateral approach of detaching the anterior third of the gluteus medius to exposing hip joint (7). Two days after the surgery, the patient could walk with a walker aid. There were no adverse effects of surgical site infection and hip dislocation in his clinical course. The gross findings of the resected femoral head tumour showed a mixture of white and yellow lesions with well-defined borders. Chondroid tissue and ossification were also visible. The surface cartilage of the femoral head was smooth and intact, without evidence of a pathological fracture (Fig. 4a). Microscopic examination of the resected specimen showed that the tumour was composed of sheet-like proliferation of atypical cells with abundant clear cytoplasm, swollen nuclei, and scattered bone formation (Fig. 4b). These pathological diagnoses were consistent with the typical presentation of CCCS; a 2-cm wide margin was obtained according to the evaluation system of the Japanese Orthopedic Association (8).

At the final follow-up 32 months after surgery, no local recurrence or metastases were observed. Postoperative functional outcome was measured using the International Symposium on Limb Salvage score (9). The patient scored 30 points, the highest possible outcome.

Informed consent was obtained from the patient for publication of data and photographs.

Discussion And Conclusions

In our case, radiological imaging with plain radiography, MRI, and CT revealed a typical presentation of CCCS in the right femoral head. Therefore, an incisional biopsy was not thought to be needed, and an en bloc resection and hip hemiarthroplasty were performed without a preceding biopsy. Pathological diagnosis confirmed the presence of CCCS, and a clear surgical margin was achieved. The postoperative limb function of the patient was excellent at the final follow-up.

When a suspicious malignant bone tumour is located in the femoral head, incisional biopsy is planned from the lateral side of the proximal femur through the femoral neck to prevent tumour contamination into the hip joint space (10). To remove biopsy tract that potentially contain the tumour cell, hip replacement with a tumour prosthesis is necessary after en bloc resection, resulting in poor postoperative function rather than hip hemiarthroplasty. Therefore, after considering the surgical margin and the postoperative function of the patient, en bloc resection without performing a biopsy may be an acceptable treatment for radiologically typical CCCS.

Collins et al. (11) exhibited some characteristic radiological features of CCCS on plain radiographs, CT, and MRI, which closely matched those in our case. Specifically, the CT scan showing matrix mineralisation with characteristic chondroid appearance was in accordance with our case. The World Health Organisation (12) has stated that CCCS is a primary bone tumour involving the ends of the bone mostly in the proximal femur and humerus. Radiographically, giant cell tumour of the bone, chondroblastoma, chondrosarcoma, chondroblastic osteosarcoma, and osteonecrosis of the femoral head were all differential diagnoses (3). Every experienced orthopaedic oncologist in specialised centre highly might be able to suspect the presence of CCCS in the femoral head, just as in our case.

Histological diagnosis is important to initiate the treatment for bone tumours. A core needle biopsy sometimes results in the wrong diagnosis compared to an incisional biopsy, usually because of insufficient material quality (6). Therefore, Nakayama et al. (5) recommend an incisional biopsy for suspected CCCS instead of a core needle biopsy. However, when an incisional biopsy is selected for preoperative diagnosis, surgeons need to remove the tumour and the potentially contaminated area covered with healthy tissue.

CCCS is a low-grade, malignant chondrogenic tumour that is resistant to chemotherapy and radiotherapy, with surgical removal using adequate margins being the main treatment strategy (4). The surgical margin is highly correlated with local recurrence and lung metastases (2–5). Itälä et al. (2) demonstrated that the 5-year and 10-year survival rates were 100% and 89%, respectively. Nakayama et al. reported that the 5-year and 10-year overall survival rates were both 89%. Local recurrence and lung and bone metastases were discovered over 5 years after the initial diagnosis (2, 3, 5). Orthopaedic oncologists must be aware that careful long-term follow-up is necessary in order to catch any recurrences or metastases that form.

In conclusion, if there are typical radiological findings, en bloc resection without a prior biopsy may be an acceptable treatment for CCCS located on the femoral head.

Abbreviations

CCCS

clear cell chondrosarcoma

CT

computed tomography

MRI

magnetic resonance imaging

Declarations

Ethics approval and consent to participate: Not applicable

Consent for publication

Written informed consent was obtained from the patient's next of kin for publication of this case report and any accompanying images.

Availability of data and materials

All data obtained is available within the manuscript.

Competing interests

The authors declare that they have no competing interests.

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Authors' contributions

MH and HN were responsible for study design and writing. NO, YO, and AT were responsible for data collection. All authors read and approved the final manuscript.

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Figures



Figure 1

Plain radiography revealed an osteolytic lesion with chondroid matrix mineralisation in the right femoral head.

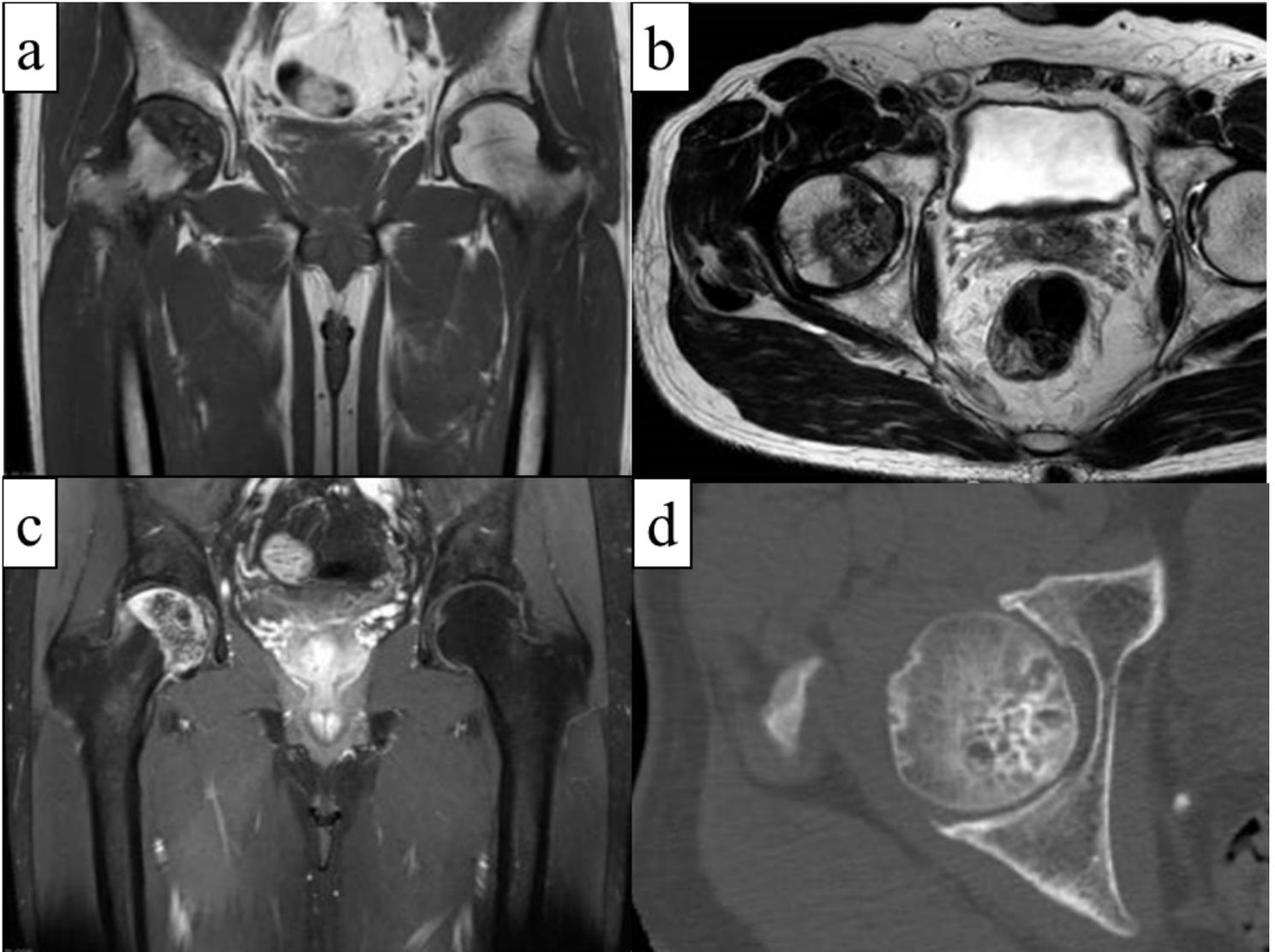


Figure 2

a. Magnetic resonance imaging (MRI) using a coronal T1-weighted image (T1W1) of the tumour showed iso signal intensity in the right femoral head. b. On an axial T2-weighted image (T2WI), heterogeneously high signal intensity with a spotted area was visible. c. The tumour was peripherally enhanced with contrast material on a T1 fat-suppressed image with a focally decreased spotted area. d. The matrix mineralisation with characteristic chondroid appearance was evident in the right femoral head.



Figure 3

Postoperative plain radiographs showing the hip hemiarthroplasty performed after en bloc resection.

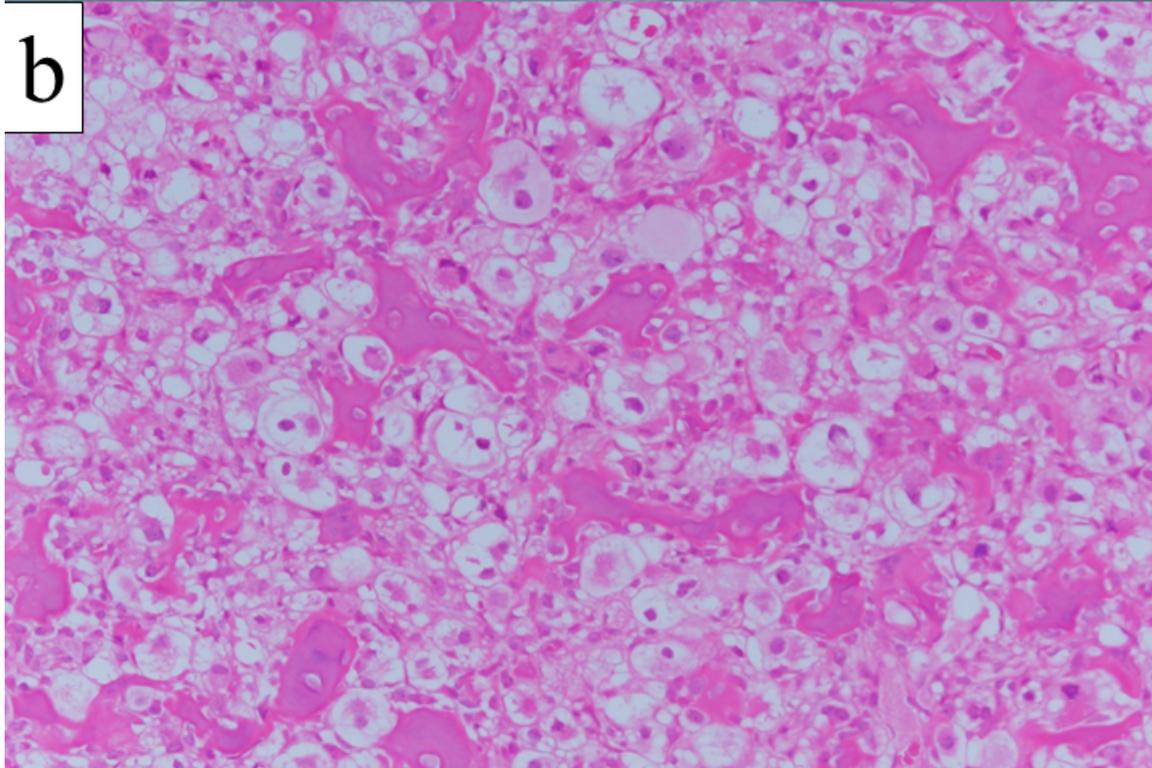
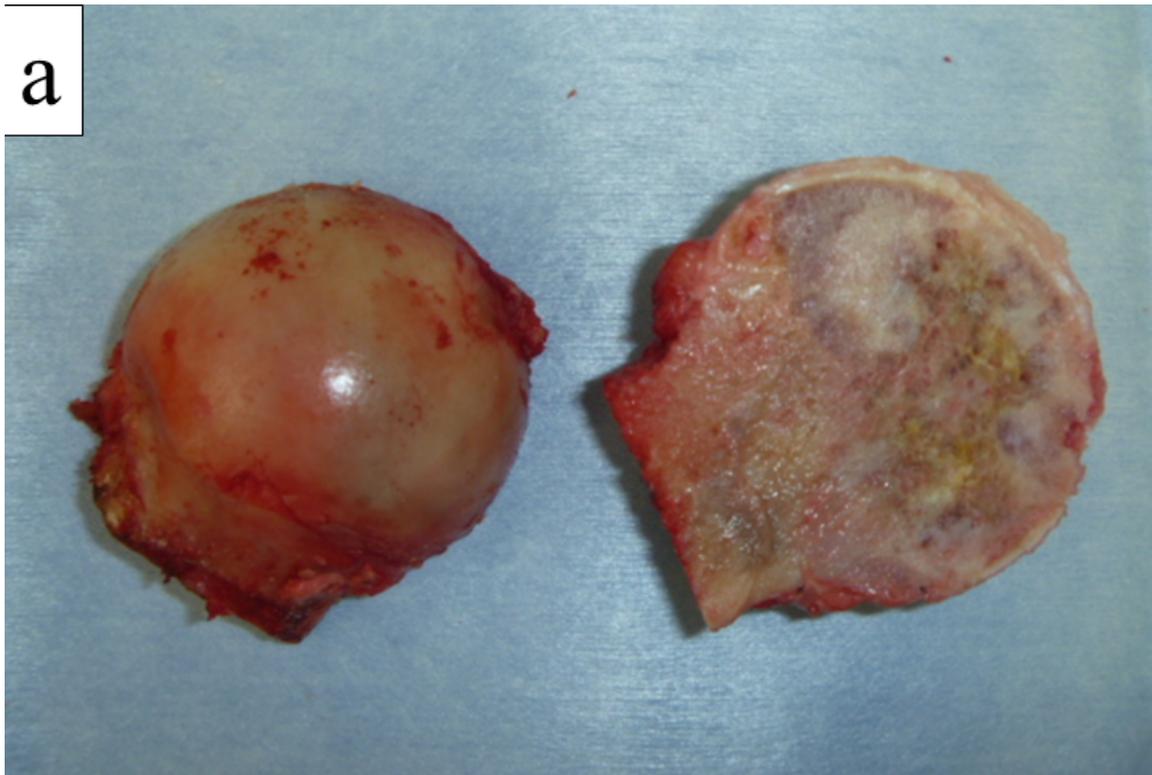


Figure 4

a. Grossly, the tumour was composed of chalky-white areas of mineralisation and a chondroid matrix with a well-defined border. The hip joint surface was smooth. b. Microscopic findings showed that the tumour cells had a well-defined border and clear cytoplasm. New bone formation was observed. The pathological diagnosis was clear cell chondrosarcoma (magnification: $\times 200$).