Intracortical Chondrosarcoma: a Case Report

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Abstract- Chondrosarcoma is the second most common primary mesenchymal malignant tumor of the bone. The most common form is central chondrosarcoma and the rarest is intracortical chondrosarcoma. Here, we describe the clinical, pathological, and imaging features of a case of intracortical chondrosarcoma as well as the outcome of surgical treatment. This is the third case reported in the literature.

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Introduction

Malignant bone tumors are rare neoplasms with an incidence of around one to two new cases per 100,000 individuals per year. After osteosarcoma, chondrosarcoma is the second most common primary malignant bone tumor, comprising 11% to 22% of bone sarcomas (1,2). Chondrosarcoma is most commonly seen in pelvis, proximal femur, ribs, skull, humerus, tibia, and scapula and rarely in other parts of the body.

Anatomically chondrosarcomas are classified into central, periosteal and soft tissue chondrosarcomas. Central type of this tumor develops within the bone cavity, and peripheral type arises from cartilage cap of a pre-malignant osteochondroma during malignant transformation within the tumor. The central type is usually an elongated, well-delineated osteolytic lesion with fusiform expansion of the shaft and thickening of the cortex. Endosteal scalloping or focal cortical destruction and cortical thickening are characteristic of central chondrosarcoma (8,9). Periosteal and soft-tissue chondrosarcomas are rare types contributing to 1%-2% of all cases (4).

Histologically chondrosarcomas have a characteristic bimorphic pattern on microscopy, with islands of variable differentiated hyaline cartilage interspersed with sheets or nests of undifferentiated small round cells (5). Based on additional histological features, they can be subgrouped into the conventional, clear cell, myxoid (chondroid), dedifferentiated and mesenchymal, which differ in their clinical behavior and prognosis (12). The conventional variety is the most common and the mesenchymal variant is the least.

A review of the literature shows that intracortical chondrosarcoma is very rare. The first case was reported in 2003 by Babinet *et al.*, which was located in the proximal femoral diaphysis and histologically belonged to the conventional category (3). Thöm *et al.*, described the second case of intracortical chondrosarcoma in 2009, which was in the distal femur and was dedifferentiated, histologically (6). Here we report the third case of intracortical chondrosarcoma, which arose in the distal part of the femur, and had a conventional histology.

Case Report

A 44-year-old woman was referred to our center in January 2009 because of a suspicious lytic lesion in the distal femur seen on radiograph. She presented with a three-year history of dull left knee pain. It had become worse during the previous 4 months. Physical examination was normal except for tenderness on medial condyle of the left distal femur. Laboratory tests were within normal ranges. On anteroposterior radiograph there was a small regular lesion, about 1 cm in diameter, in the medial cortex of the left femoral condyle (Figure 1). Furthermore, Tc99 bone scan revealed increased uptake in that area (Figure 2). We also studied the magnetic resonance imaging features of the lesion and found that it was intracortical and exhibited intermediate signal intensity similar to adjacent muscle on T1 weighted images and high signal intensity on T2 weighted images. There were few void signals present signifying intratumoral calcification. We found some

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medullary invasion with a sclerotic rim surrounding the tumor but there was no soft-tissue mass on the MRI images (Figures 3, 4 and 5).



Figure 1. Anteroposterior radiograph of the distal of the femur showing a regular small lesion in the medial cortex (white arrow)



Figure 2. Tc99 bone scan revealing an isolated increased uptake area in the distal part of the left femur

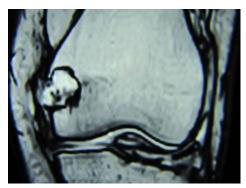


Figure 3. Coronal T2 weighted MRI showing a hyper-intense signal abnormality in the cortex of medial epicondyle with some void signals and a sclerotic rim separating the lesion from the medulla

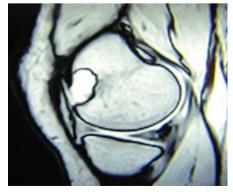


Figure 4. Sagittal T2 weighted MRI showing a hyper-intense signal abnormality in the cortex with a sclerotic rim in the medulla

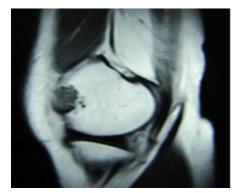
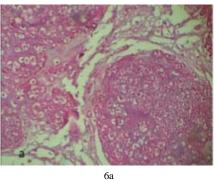
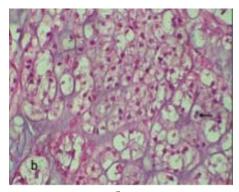


Figure 5. Sagittal T1 weighted MRI showing an iso-intense signal abnormality, similar to muscle in intensity, in the cortex

The patient underwent incisional biopsy for further diagnostic clues. On gross examination, the lesion was a grayish white, lobulated mass with some focal calcifications. Histopathologic examination of prepared sections showed sheets and nests of pleomorphic tumoral cells with round granular/vesicular nuclei & nucleoli, plump amphophilic, some mildly basophilic cytoplasm with distinct cytoplasmic borders, few mitotic figures and some tumoral cells with more than one nucleus per lacunae in chondroid basophilic matrix with focal calcification arranged in lobular pattern. This tumor invades bone and entraps degenerated residual bone tissue, confirming the diagnosis of conventional chondrosarcoma (Figure 6).



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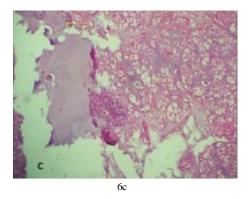


Figure 6. a: Photomicrograph showing pleomorphic chondrocytes with plump cytoplasm and one or more nuclei arranged in lobular pattern (X 63). b: High magnification showing mildly basophilic cytoplasm, vesicular nuclei & few mitoses (black arrow) (X250).
c: Photomicrograph of the margin of the tumor showing entrapment of residual bone (yellow arrow) (X 63)



Figure 7. Lateral radiograph of the knee 2 years after surgery: bone cement well in place with no recurrence

We conducted a marginal resection of the tumor. Biopsy tract was removed and curettage augmented by high speed burring at least 5 mm into normal bone. The cavity was filled with bone cement. The patient had a complication-free post-operative course and was discharged from the hospital in good condition. Two years after the operation, the patient is pain-free and there are no signs of recurrence on radiographs (Figure 7).

For this study, a written consent was obtained from the patient.

Discussion

Our case is the 3rd intracortical chondrosarcoma reported. We considered our case as an intracortical one because the tumor growth started within the cortex and then the mass was able to split it and continue to grow. This is possible given the location of the tumor in the metaphysis, which has a weaker cortex. Eventually, part of medial cortex was also pushed into the medulla. Furthermore, pathologic examination of the apparent sclerotic rim of the lesion shows the presence of cortical bone. As cortical bone is present on either side of the mass, the tumor is more likely to have arisen from the cortex. (Figure 6c)

Radiologically, we see a lytic lesion in the medial femoral condyle without any periosteal reaction. The differential diagnosis includes benign lesions such as osteoid osteoma, fibrous cortical defect, intra-cortical hemangioma, intra-cortical chondroma intracortical chondromyxoid fibroma, osteomyelitis as well as malignant conditions such as intracortical chondrosarcoma and intracortical osteosarcoma (11,10). Histopathologic examination of the lesion reveals a cartilaginous tumor and only intra-cortical chondroma and intra-cortical chondrosarcoma remain possible. Classically, intra-cortical chondroma radiographs show a round lucent lesion, marginal sclerosis and sometime intra-lesional calcifications. On pathology, intracortical chondroma shows benign appearing chondrocytes with no atypical features and rare binucleated chondrocytes (13). On the other hand, chondrosarcoma shows few mitotic figures & some tumoral cells with more than one nucleus per lacunae. More importantly bone invasion and entrapment of residual bone tissue by the neoplastic cells in pathology. Consequently, based on pathology, we were able to finalize the diagnosis as malignant intracortical chondrosarcoma and rule out chondroma.

Although a lytic intracortical lesion with regular borders, seen in this case, supports a benign bony tumor as the diagnosis, based on pathology we know that we are dealing with a malignant chondrosarcoma. It should be considered that in this case, the rim of sclerosis was the anteromedial cortex. This cortex has been displaced and invaded by tumoral cells.

In addition to the unique and interesting presentation

of this chondrosarcoma, our treatment plan led to an excellent outcome. We used curretage augmented with high-speed burring to remove the tumor and at least 5 mm of normal bone around it. The defected area was reconstructed with bone cement filling to avoid future fractures and help with early gain of range of motion. Use of bone grafts or prostheses were also potential reconstructive approaches but were not used. We did not see any signs of recurrences in our patient after 2 years. The patient also did not receive chemotherapy, radiotherapy, or adjuvant treatment but was under close observation. Such attention is very important in all cases with a previous history of malignant bone tumor.

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