Asymptomatic grade-2 central chondrosarcoma of the distal femur with non-aggressive radiological features

This paper discusses the case of a 57-year-old man with an incidental finding of a radiologically non-aggressive chondroid lesion and concomitant osteonecrosis in the left distal femur. The final resected specimen showed a grade-2 chondrosarcoma. This case illustrates that long-term follow-up is necessary for non-aggressive chondroid lesions. If surgical management is considered, resection with an adequate margin is superior to intralesional curettage.

Case report

A 57-year-old man initially presented to an orthopaedic surgeon for mild left knee sprain in October 2010. He had no prior history of knee pain. Physical examination was unremarkable except for tenderness over the medial joint line. X-ray showed an incidental cartilaginous lesion at the intercondylar region of the distal femur, and also a lytic lesion in medial femoral condyle (Fig 1a and 1b). The distal femoral lesion appeared well-delineated, with speckled chondroid features and no cortical erosion or soft tissue shadow. The lytic lesion in the medial femoral condyle was subchondral at the medial femoro-tibial articulation, and compatible with subchondroid osteonecrosis of distal femur.

The blood count, renal and liver function test results, and calcium and phosphate levels were unremarkable. Magnetic resonance imaging showed a well-circumscribed lesion of 1.5 cm x 1.5 cm x 1 cm (craniocaudal x anteroposterior x transverse section) within the distal femur (Fig 1c-1f). It was heterogeneously hypointense to surrounding...
Bone on T1-weighted imaging, and heterogeneously hyperintense on T2-weighted imaging with patchy heterogeneous contrast enhancement. The lesion at the medial femoral condyle was subchondral with joint cartilage collapse. It was homogeneously hypointense to surrounding bone on T1-weighted imaging, and hyperintense on T2-weighted imaging with contrast enhancement.

Technetium-99 bone scan and spectrometry showed a hot lesion in medial femoral condyle but no uptake was noted in the intercondylar region. The provisional diagnosis was a chondroid lesion in intercondylar region of the distal femur and concomitant osteonecrosis in the medial femoral condyle. Image-guided biopsy of the intercondylar lesion revealed a chondroid lesion with myxoid matrix embedding cells with nuclear atypia, which was therefore suspicious of malignancy. The patient was then referred to us for further management.

We performed a computer-navigated complete resection of the chondroid lesion, with 1-cm margins in all directions. The knee joint was resurfaced with a total knee prosthesis. Nine months postoperatively, he was asymptomatic and walked unaided. Follow-up imaging (computed tomographic [CT] scan thorax and bone scan) showed no distant recurrence.

Microscopically, the resected specimen was composed of chondroid cells with permeation into the native lamellar bony trabeculae. There was increased cellularity, with myxoid changes, and mild nuclear pleomorphism with binucleation (2 nuclei within a cell). Mitotic figures were inconspicuous. The overall picture was compatible with a grade 2 over 3 chondrosarcoma. There was a clear margin of 8 mm (Fig 2).

Discussion

Histologically, chondroid tumours are a large family of neoplasms with varying degrees of chondroid differentiation. Chondroma, either enchondroma or osteochondroma, is the commonest benign chondroid tumour and chondrosarcoma is the commonest malignant counterpart. Chondrosarcomas\(^1\) form a heterogeneous group ranging from the least aggressive grade 1 (well-differentiated) to the most malignant grade 3 (poorly differentiated), all based on the histological features such as cellularity and nuclear atypia.

While chondrosarcomas are arbitrarily divided into three grades, in reality they show a continuum of pathology ranging from benign chondromas to the most malignant grade-3 lesions. To differentiate a grade-1 chondrosarcoma from one that is benign based on histology alone is known to be very difficult. Therefore, the clinical and radiological features often become pertinent for an accurate diagnosis.

Weiner\(^2\) stated that the likelihood of having a chondrosarcoma in any cartilaginous lesion increases in the presence of the following features: adulthood, growth after skeletal maturity, pain, axial skeleton (pelvis or scapula) involvement, size longer than 6 cm, calcification more in the central part of lesion, endosteal erosion of more than 2/3 of the cortex, periosteal reaction, and an associated soft tissue mass. In our patient, the clinical and radiological features did not point towards malignancy, yet the final histology was a grade-2 chondrosarcoma. Based on this, we would advise longer follow-up for any chondroid lesion, even when it looks innocent. When in doubt, histological assessment should be considered.

No imaging is reliable in differentiating enchondromas and subtle low-grade chondrosarcomas. Increased isotope uptake has been reported for both types of lesions.\(^3\) In the future, \(^18\)F-fluoro-2-deoxyglucose positron emission tomography (PET) integrated with CT may have the potential to differentiate enchondromas, low-grade and high-grade chondrosarcomas. Studies have shown that using a standardised uptake value (max) of 2.0 as the dividing line between benign and malignant lesions yields a sensitivity of 90%, specificity of 100%, and accuracy of 97%. To date, however, recourse to
PET scans as part of the routine investigation for this disease entity continues to be debated.

Chondrosarcomas are generally not radiotherapy nor chemotherapy sensitive, and surgery remains the mainstay of treatment. It is also known that local curettage ablation carries a higher risk of local recurrence; around 10% of recurrent lesions also show an increase in the malignancy grading. In Gitelis et al’s report, the local recurrence and metastatic rates of grade-1 chondrosarcomas were 33% and 9%, respectively. In grade-2 tumours, respective rates were 38% and 33%, and in grade-3 tumours, they were 19% and 44%. The recurrence is usually related to the adequacy of surgical clearance. Ozaki et al also found that intralesional surgery led to more local relapse, but none of the local recurrence cases in his series developed metastatic lesions.

In fact, metastasis of low-grade lesions can develop without any local recurrence. The Mayo Clinic reported that local recurrence led to more distant metastasis and was associated with a decrease in overall survival. In their series, 5/12 patients (grade 1 and stage 1A) with local recurrence had distant metastasis, all of whom died from the disease. The difference in survival may not be significant in the first 5 years, but becomes more obvious after 10 years of follow-up. In general, an en-bloc resection of the tumour with an adequate margin is considered to be a much safer procedure.

Some argue that grade-1 chondrosarcomas behave more like aggressive benign lesions rather than true malignant ones. Recent reports have also shown that intralesional curettage of these low-grade lesions lead to low local recurrence rates. Furthermore, these locally recurrent tumours were still amendable to re-resection. The argument for curettage is based on superior functional outcomes compared to en-bloc resection. Although we concur with this statement, it should be noted that most of these reports were retrospective reviews with outcome analyses based on the postoperative histology. Also, the follow-up periods were not long enough for all recurrences to be clinically manifest. In reality we usually do not have the final grading before definitive surgery. In Normand et al’s series, three out of eight grade-1 chondrosarcoma patients turned out to have higher grades than expected, and two of them died of the disease. This is similar to our patient, who had clinical and radiological features of a benign chondroid lesion (enchondroma or grade-1 chondrosarcoma), yet received a final histological diagnosis of a grade-2 chondrosarcoma. When considering surgical removal of chondroid lesions, we therefore advise resection with an adequate margin in order to avoid disaster following histological upgrading of the tumour at final sectioning.

To conclude, while intralesional curettage is still commonly performed for chondroid lesions with radiologically benign features, our case illustrates that finally, histological upgrading may be encountered. We recommend that chondroid lesions should be removed en-bloc with adequate margins. For patients who opt for a conservative approach with watchful waiting, long-term follow-up is mandatory.

References