Epiphyseal lesion of the humerus in a child

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Answer

Solitary epiphyseal enchondroma of the humeral greater tuberosity

Discussion

The original description of the enchondroma by Jaffe and Lichtenstein hypothesized that these lesions result from dislodgement of normal chondrocytes from the physis, thereby leading to their common placement within the metaphysis or metadiaphysis of long bones as the physis grows away from the stranded cells; this idea was elaborated by Brien and coauthors [1, 2]. The relationship of enchondroma

formation to physeal growth therefore makes the epiphyseal form of this lesion an unlikely entity, contributing to its rarity. Potter and coauthors asserted that whereas aberrancy of the primary ossification center produces conventional, metadiaphyseal enchondromas, errors in the secondary ossification center result in epiphyseal enchondromas [3].

Solitary, radiolucent lesions of the epiphyses are uncommon in the pediatric population. The most common is chondroblastoma, after which osteomyelitis, giant cell tumor, enchondroma, and chondroblastic osteosarcoma may be considered [4]. Intraosseous ganglia and aneurysmal bone cysts may occur, however, they are more common in older patients.

Our patient's radiograph of the proximal humerus demonstrates a well-circumscribed, lucent lesion containing some faint stippled calcifications; a small avulsion fragment is likely the result of the inciting trauma, which initiated the patient's work-up. MRI reveals a well-marginated lesion that has low signal on T1-weighted sequences and high signal on T2-weighted sequences. Chondroblastoma commonly exhibits substantial peripheral marrow edema and a low to intermediate signal centrally on T2-weighted sequences in 80–90 % of cases [5]; this patient's imaging suggests a more indolent lesion. Based on his clinical presentation and radiographic imaging, chondroblastoma was the presumed diagnosis although higher marrow signal would normally be expected about the lesion's periphery on T2 sequences.

Our patient was taken to surgery, where open biopsy was performed (Fig. 1). Frozen section suggested a benign, cartilaginous histology, therefore the entire lesion was curetted and the deficit was filled with cancellous allograft bone. Six months following surgery, the patient had returned to full activity and was without symptoms (Fig. 2).

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Fig. 1 Intraoperative radiograph with fluoroscope demonstrating successful access of the lesion with curette

The gross specimen consisted of fragments of well-circumscribed, lobulated mature blue-gray cartilage and frozen section indicated a benign diagnosis. On low power, some of the lobules of cartilage were bordered by mature bone and contained focal calcifications (Fig. 3). The chondrocytes were variably spaced, but the tumor had overall low cellularity. No mitoses were seen, and the chondrocytes were small and nondescript (Fig. 4). No focal areas of myxoid degeneration were seen. These findings are consistent with a solitary, benign enchondroma of the epiphysis [6].

Whereas the metadiaphyseal enchondroma is one of the most common bone lesions encountered by orthopedic oncologists, Potter and coauthors' investigation demonstrates the rarity of these lesions in the epiphyses. Their study period spanned 55 years at two tertiary medical centers and revealed only 33 epiphyseal enchondromas [3]. They described ten lesions of the proximal humerus, two were located within the greater tuberosity. Johnson and coauthors reported a series of 12 solitary, epiphyseal enchondromas that were located in the proximal humerus, distal femur, and proximal tibia; two of these lesions progressed to chondrosarcoma [7].

Single events of solitary, epiphyseal enchondromas have been reported including one of the proximal humerus and one of the proximal femur [8, 9]. Enchondromatosis involving the epiphyses is much more common [10]. A mutant parathyroid hormone receptor that causes constitutive activation of Indian hedgehog has been identified in humans

and is shown to result in the formation of multiple enchondroma-like lesions in mice [11].

Histological findings of enchondromas within the long bones differ slightly from enchondromas of the small bones in the hands and feet, in that the latter tend to be more hypercellular [6]. However, neither should show extensive osseous destruction or soft-tissue extension, which should be considered features of malignancy [12]. Older lesions may show extensive calcification with accompanying cystic degeneration, but no extensive myxoid change or necrosis should be seen in this benign tumor [6, 13].

Whereas management of conventional enchondromas is generally confined to observation, epiphyseal lesions are often symptomatic. In Potter's series, 23 patients reported pain, three presented with pathologic fracture, two complained of a mass, and five were asymptomatic. Of the 33 patients in their series, 26 underwent curettage with bone-grafting resulting in one recurrence. Six patients had resection of the involved epiphysis after skeletal maturity and one had en bloc resection of the entire distal fibula. The authors acknowledged that the magnitude of treatment was, in general, unnecessary, however, management decisions were based upon radiographic indications of a more aggressive condition, chondroblastoma, and the diagnosis of enchondroma was unexpected. Similar to the patients described in Potter's series, the decision to take our patient to surgery was made initially on the premise that his lesion was most likely a chondroblastoma.

In summary, epiphyseal enchondromas are uncommon, and these lesions are often mistaken for chondroblastoma. Although a high index of suspicion would be required to identify these lesions preoperatively, in patients presenting with chondroblastoma-like findings on radiographs without antecedent, pre-trauma symptoms, and lacking characteristic surrounding marrow edema on MRI, this diagnosis should be considered and non-operative management with serial radiographic follow-up may be a prudent initial course.

Fig. 2 Postoperative radiograph at 6 months following surgery

Fig. 3 Low-power (×4, H&E) histological image showing lobules of mature cartilage (*small arrows*) with focal adjacent enchondral ossification (*large arrows*)

Fig. 4 Low-power (×20, H&E) histological image showing small, benign appearing chondrocytes (*small arrows*) and low overall cellularity

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