

Dysplasia Epiphysealis Hemimelica of The Trochlear Epiphysis

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Dysplasia epiphysealis hemimelica is an uncommon disorder primarily involving the epiphyses of the lower extremities. We report imaging findings of a case of dysplasia epiphysealis hemimelica involving the trochlear epiphysis of the humerus. Conventional radiography showed irregular ossification in the area of trochlear epiphysis, the finding thought to be diagnostic of this disorder. However, other imaging modalities, such as double contrast arthrography, computed tomography and MR imaging, provided valuable information about the nature and extent of the epiphyseal pathology.

Key words : dysplasia epiphysealis hemimelica, elbow, MR imaging

Introduction

We report imaging findings of a case of dysplasia epiphysealis hemimelica involving the trochlear epiphysis of the humerus. Dysplasia epiphysealis hemimelica (Trevor's Disease) is a rare skeletal disorder in childhood characterized by asymmetrical osteocartilagenous overgrowth of one or more epiphyses^{1,2)}. Lower extremity involvement is far more common than upper extremity involvement. Histological findings are identical to those of osteochondroma arising from the metaphysis. In a review of the English literature, 23 reported cases showed upper extremity involvement; the most frequent site of involvement was the carpal bones and less commonly glenoid, proximal humerus and distal ulna³⁾. Involvement of the trochlea has been reported in a case with generalized form⁴⁾, but to our knowledge, a solitary lesion in this area has not been previously reported.

Case report

A 8-year-old boy presented with a complaint of left elbow pain of one month duration. As the pain persisted despite the conservative treatment in a hospital, he was referred to our hospital for further evaluation and management. The patient had a past history of fall from a tree

at the age of 5 years when he sustained mild trauma in the back without any injury to the elbow. The family history was unremarkable. Physical examination revealed a cubitus valgus on the left. There was no associated swelling, muscle atrophy, restriction of motion and alteration in length of the limb.

Conventional radiographs of the left elbow showed irregular ossification corresponding to the site of trochlear epiphysis (Figure 1). Cubitus valgus deformity was also noted. No abnormalities were detected in other joints. Double contrast arthrography of the left elbow clearly demonstrated enlarged trochlear epiphysis containing irregular ossification (Figure 2). Computed tomography also revealed irregular ossification in the same region. Bone scintigraphy showed slightly increased uptake over the lower end of the left humerus. T1-weighted (TR 400 msec, TE 20 msec) MR images showed enlargement of the trochlear epiphyseal cartilage with irregular ossification demonstrating high signal intensity of fatty bone marrow (Figure 3 A). A gradient echo (TR 300 sec, TE 20 msec, flip angle 30 degrees) sequence revealed similar findings with clear distinction between the cartilage and ossifications (Figure 3 B). The diagnosis of dysplasia epiphysealis hemimelica was suggested on the basis of these imaging findings.

At surgery, trochlear epiphysis of the distal humerus was found to be elevated. The surface of the lesion was covered by the smooth cartilage with patchy areas of discoloration, where the cartilage appeared to be thin. The lesion was partially removed to restore the normal epiphyseal contour. Histopathology of the specimen showed lamellar bone with bone marrow tissue surrounded by cartilage.

Postoperative radiography of the elbow showed residual ossifications in the trochlea, although they were smaller in size compared to the preoperative examination. Cubitus valgus deformity was found to be improved. The patient was asymptomatic for three years after the surgery, but the radiography showed regrowth of ossifications of the trochlea epiphysis resulting in recurrent cubitus valgus deformity.

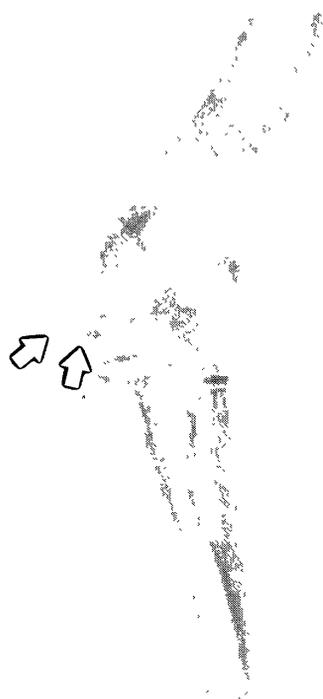


Fig 1 A



Fig 1 B

Figure 1 Anteroposterior (A) and lateral (B) radiographs of left elbow demonstrate irregular ossification at the site of trochlear epiphysis (arrows).



Fig 2

Figure 2 Double contrast arthography showing an enlarged trochlear epiphysis containing irregular ossifications (arrows).

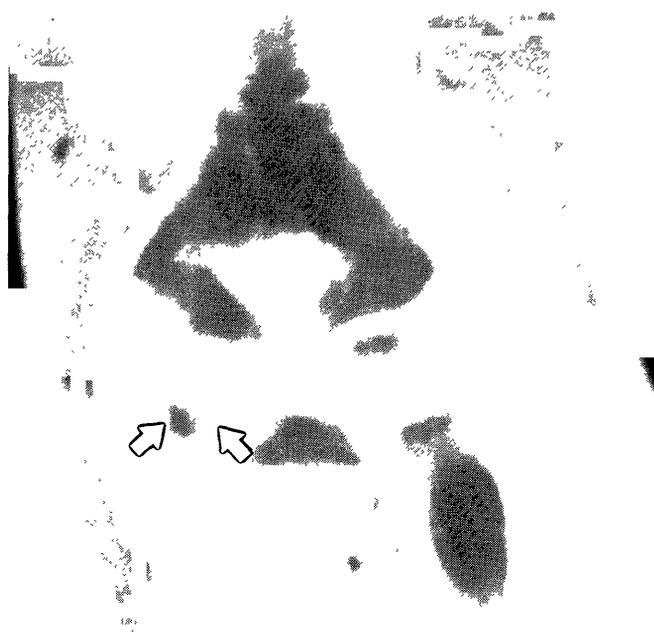


Fig 3 A



Fig 3 B

Figure 3 T1 weighted (TR 400 msec, TE 20 msec) MR image (A) demonstrates irregular ossification (arrows) in the enlarged trochlear epiphysis with high signal intensity of bone marrow. Outline of the cartilage and extent of the lesion (arrowheads) are more clearly depicted on gradient echo (TR 300 msec, TE 20 msec, flip angle 30 degrees) MR image (B).

Discussion

The initial clinical manifestations of dysplasia epiphysealis hemimelica include swelling and less commonly pain and deformity of the involved joints, usually localized to one side of the body. The diagnosis can be delayed in cases without pain, because majority of the patients are too young to notice the painless swelling. Secondary changes such as muscle atrophy, restriction of movements and growth disturbance in the involved extremity are frequently encountered in late stages. This patient had a short history of swelling and pain with minor degree of valgus deformity at the time of presentation. The absence of secondary changes in this patient can be attributed to the early presentation and diagnosis.

Dysplasia epiphysealis hemimelica is primarily diagnosed based on the findings on conventional radiography. The lesion appears as focal calcifications or ossifications in one side of the ossifying epiphysis, and is often associated with irregular enlargement of the affected epiphysis⁹. Later, the ossifications can become confluent with the adjacent bone and may appear as lobulated mass protruding from the epiphysis. The normal centers of ossification in the affected limb may appear prematurely and can be larger than those of the opposite side. The adjacent metaphysis may be widened and develop longitudinal striations. Conventional radiological findings are diagnos-

tic when enlarged epiphyseal cartilage is fully calcified or ossified. However, the initial manifestations which include a paraarticular soft tissue mass with small calcific foci do not exclude the other diagnostic possibilities, such as myositis ossificans, tumoral calcinosis, synovial osteochondromatosis, or other calcified and/or ossified tumors.

Imaging modalities including arthrography, computed tomography and MR imaging are useful in differentiating dysplasia epiphysealis hemimelica from the entities cited above. They also should be employed for the preoperative work-up to clarify the size and extent of the lesion. Arthrography, especially the double contrast technique, has been used to delineate an outline of the articular surface and to decide the intraarticular extent of the lesion⁶. Similar findings can also be obtained from computed tomography which has been found to be superior than arthrography as both intraarticular and extraarticular extent of the disease can be adequately demonstrated⁷. Bone scintigraphy confirms the localization of the disease at a single site. MR imaging findings of dysplasia epiphysealis hemimelica have been reported in several cases^{8,10}. MR imaging has the advantage of being a multiplanar noninvasive technique and having excellent soft tissue contrast. It provides great detail of the unossified cartilagenous mass as well as the status of the articular and growth plate cartilage. Potential cleavage plane between the mass lesion and parent epiphysis or

articular surface abnormality can also be imaged optimally^{8, 11)}.

In this patient all these details were obtained from MR imaging which appears to be superior to other modalities. MR imaging should be the first modality to be used when additional diagnostic informations are needed. In short, we report a rare case of dysplasia epiphysealis hemimelica involving the trochlea, discuss the relative role of various imaging modalities in diagnosis and management and emphasize the utility of MR imaging as the technique of choice in problematic cases.

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