

Dysplasia Epiphysealis Hemimelica Of Lateral Aspect Of The Distal Ulnar Epiphysis

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Abstract

Dysplasia epiphysialis hemimelica (DEH) is a rare developmental disorder 1 affecting one or more epiphyses of the long bone and/or short bones of the carpus or tarsus 2. It usually affects the lower limb but upper limb involvements have been reported. Involvement of the ulna is very rare 3. Microscopically this lesion comprises of osteocartilaginous exostosis. The purpose of this case report is to report a atypical case of a patient who had the lateral aspect of the distal ulnar epiphysis involved which has not been reported before, review the literature and discuss the management.

CASE REPORT

A 15-year-old female presented to the orthopaedic clinic with a long history of swelling and pain in the left wrist. Clinically there was no limitation of the range of motion of the wrist or shortening of the limb and the right wrist was unaffected without any symptoms.

Radiographs of the wrist showed an enlarged lateral half of the distal ulnar epiphysis. The height of the distal ulnar epiphysis was more than the styloid process of the ulna. The styloid process appeared normal and unaffected. There was superior contouring of the enlarged part of the epiphysis. This enlarged portion of the epiphysis abutted against the medial part of the distal end of the radius. There was also minimal subluxation of the distal radioulnar joint.

Figure 1

Figure 1: Lesion affecting the lateral part of the distal ulna



Figure 2

Figure 2: Lateral view of the wrist



The distal end of the radius and the carpal bones were normal. We made a diagnosis of Dysplasia epiphysealis hemimelica.

DISCUSSION

Dysplasia epiphysealis hemimelica is a rare developmental disorder of the childhood with a reported incidence of about 1:1000,000 ¹. It was first described by Mouchet and Balot in 1926 ³. It generally manifests between ages of 2 to 14 with a male to female ratio of 3:1 ⁴.

Majority of the case report about this condition are regarding the lower extremity. The common presentation is that of a painless bony swelling with or without joint deformity. Rarely upper limb is involved. Although there is no definite genetic involvement, Hessinger et al ⁵ described 7 cases in 12 generations of one family. The lesion usually affects one half of the epiphysis medial or the lateral side hence the term

hemimelica in its description.

Histologically, the lesion is indistinguishable from an osteochondroma ⁶.

The aetiology of the disorder is unknown. Malignant transformation has been reported.

Our case was atypical because of several reasons. Few cases of DEH involving the distal ulna are reported ^{2,3,7}. With none involving the lateral half of the distal ulnar epiphysis. The presentation at 15 yrs was quiet late with pain where pain is not a constant feature.

Surgery is often required if there is pain or functional impairment due to deformity or compression of the adjacent articular structures ¹. Our patient did not wish to undergo surgery as the pain was not intolerable. At a recent follow-up visit, more than a year after her initial presentation she is completely pain free.

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