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TEST YOURSELF: ANSWER

A child with painless left wrist swelling

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Discussion

Radiographs (Fig. 1) showed remodeling with an irregular expanded contour of left distal ulnar epiphysis and left lunate in particular at its ulnar aspect. Distal ulnar metaphysis was spared. The trabecular and cortical continuity of the excrescences with the parent bones were shown better on the computed tomography images (Figs. 2–4). Magnetic resonance imaging confirmed the bony enlargement (Fig. 5) and showed complete cartilage covering of the involved bones (Fig. 6). There was no involvement of distal radial epiphysis or remainder of the carpal bones. On the basis of the imaging findings, the lesions likely represented osteochondroma of the distal ulnar epiphysis and the lunate (an epiphyseal equivalent). A diagnosis of dysplasia epiphysealis hemimelica was made. The patient was followed-up clinically and remained largely asymptomatic at 2-year follow-up. The epiphyseal enlargement remained stable.

Dysplasia epiphysealis hemimelica is a rare non-hereditary developmental bone dysplasia with an estimated prevalence of 1 in 1,000,000 [1]. It is characterized by osteochondromatous growth arising from the epiphysis in a hemimelic distribution (involving more commonly the medial or lateral aspect of the ossification center). It was first described by Mouchet and Berlot as 'tarsomegaly' in 1926. Subsequently, Trevor described the condition in 1950 and in 1956 Fairbank coined the most widely used term "dysplasia epiphysealis hemimelica". The condition typically affects children, usually in the lower limbs in particular the knees and ankles. Males are affected three times as common as females [1]. Involvement of the upper limb is uncommon and less than 40 cases have been reported in the literature [2]. Patients may present to clinical attention with pain, swelling, deformity, limb length discrepancy, or functional impairment.

The diagnosis is mainly radiological and hinges upon the predominant epiphyseal involvement in a hemimelic distribution as reflected by its name. Azuoz et al. [3] classifies the condition in three forms: the localized form, affecting only one epiphysis, the classic form, affecting more than one bone in a single extremity, and a generalized form, affecting the whole limb. As illustrated in our case, radiography is useful to look for other sites of involvement. Cortical continuity with the epiphysis is better shown on computed tomography while magnetic resonance imaging is of use in confirming the osteocartilaginous nature of the lesion, as well as delineating its relationship and effect on adjacent cartilage, ligaments, and neurovascular structures. Complications include local pressure symptoms and internal derangement. Malignant transformation has never been reported in the literature [4].

Treatment options included expectant management (as in our case) or surgical excision [5]. Prognosis depends on the size and location of the lesion, extent of articular incongruity, and involvement of nearby structures.

In summary, knowledge of the typical epiphyseal location and hemimelic distribution should allow diagnosis of this rare condition even in an atypical location radiologically. Computed tomography and magnetic resonance imaging are of help in confirming the osteocartilaginous nature and depicting any local complications.

The case presentation can be found at https://doi.org/10.1007/s00256-017-2858-4

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