DYSPLASIA EPIPHYSEALIS HEMIMELICA (TREVOR'S DISEASE) OF THE DISTAL RADIUS

N. LEVI, S. E. ØSTGAARD, B. LUND

The authors report a rare case of dysplasia epiphysealis hemimelica (DEH) or Trevor's disease of the radius in an Eskimo from Greenland. DEH is indistinguishable microscopically from osteochondroma, however its epiphyseal site of origin differentiates it from the osteochondroma, which characteristically originates in the metaphysis.

Key words : dysplasia epiphysealis hemimelica ; Trevor's disease ; upper extremity ; radius.
Mots clés : dysplasie épiphysaire hémimélique ; maladie de Trevor ; membre supérieur ; radius.

INTRODUCTION

Dysplasia epiphysealis hemimelica (DEH) or Trevor's disease is a rare osteocartilaginous overgrowth involving one or several epiphyses or ossification centers on one side of the body. The incidence is less than 1/1 million. It was first reported by Mouchet and Belot in 1926 as a tarsal bone disorder which they called tarsomegaly (7). Trevor, in 1950, reported eight cases of the disease with lower limb involvement and recognized a definite entity which he called tarso-epiphyseal aclasis (10). In 1956, Fairbank added 14 cases to the literature and proposed the more generalized term of dysplasia epiphysealis hemimelica as the tarsal bones are not the only site of involvement (4). Kettelkamp *et al.*, in 1966, noted that one of their cases involved the entire, rather than half of the epiphysis as hemimelic would suggest and the term unilateral epiphyseal dysplasia has been suggested (2, 3, 5). Approximately 100 cases have been reported (1, 2, 3, 6). Involment of an upper extremity is extremely rare with fewer than 15 examples published in the English and French literatures (1). We report a patient with the disease involving the distal radius.

CASE REPORT

A 66-year-old male eskimo was referred with a 51 years history of a swelling in the distal part of his left forearm. Apart from glaucoma he had no previous medical history. There was no family history of bone dysplasia. The patient noticed the swelling when he was 15 years old. It had however been painless until 6 months before his referral and the swelling had been increasing during this last period. On examination a 3×4 cm hard and nontender focal mass on the volar aspect of the distal end of the radius was found. Radial and ulnar pulses were normal. Neurological examination of the hand was also normal. Plain xray of the left wrist revealed a 2×2 cm mass arising from the ulnar side of the radial epiphysis (Fig. 1). Tc-99m bone scintigraphy did not reveal any other bone pathology. A sagittal STIR-sequence with transverse T1 weighted image with gadolinium contrast revealed an intermediate signal intensity without enhancement (Fig. 2). Open biopsy showed an osteochondroma. The tumor was surgically removed and the patient made an uneventful recovery.

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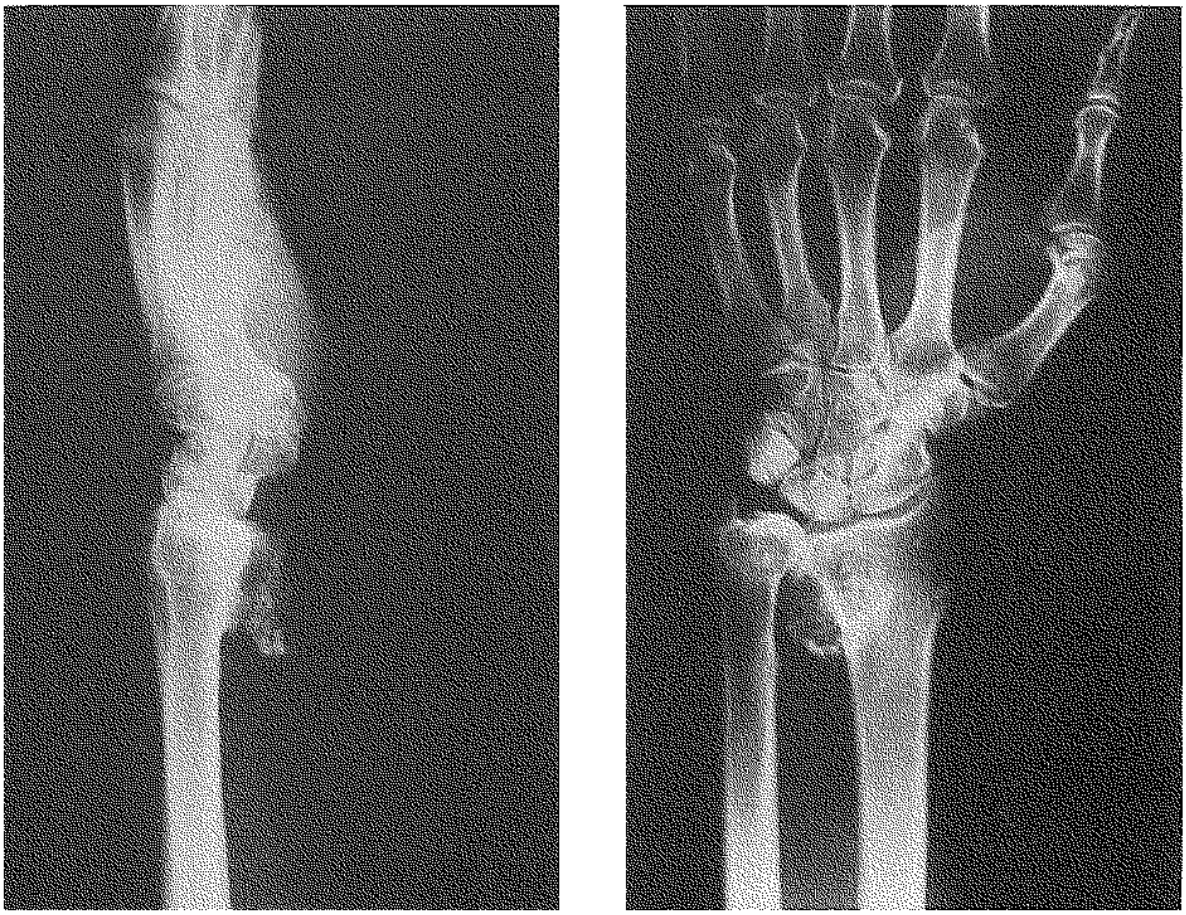

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Fig. 1.

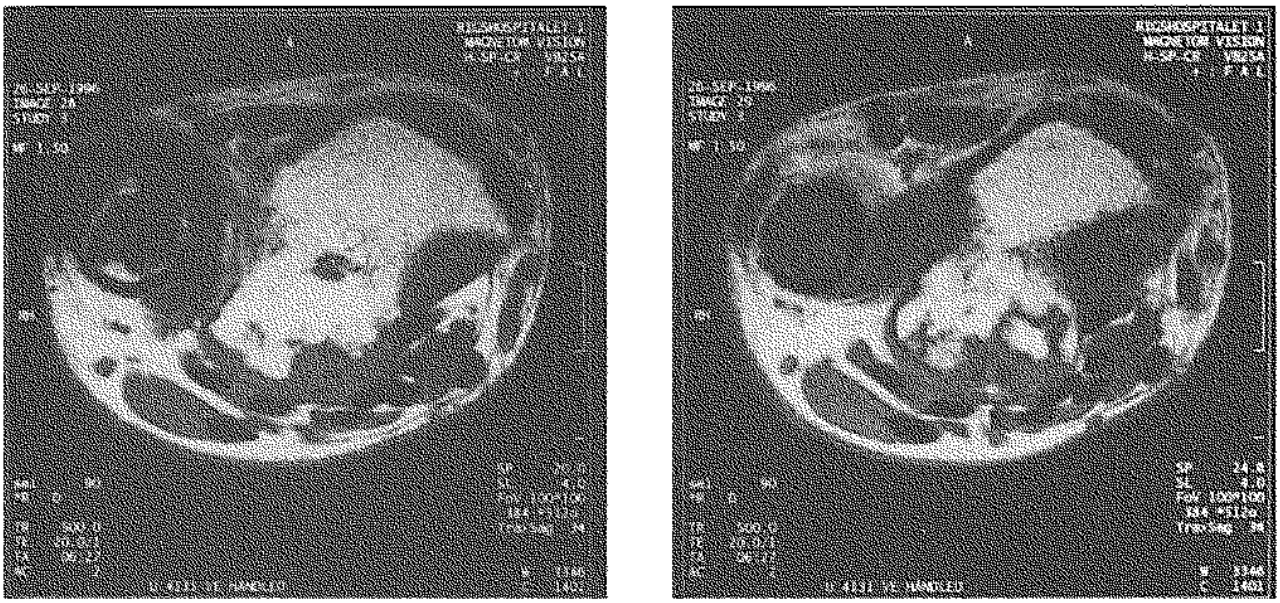

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Fig. 2.

DISCUSSION

DEH usually becomes evident in children and young adults and is more common in men than in women with a ratio of approximately 3 to 1 (5, 9). Typical clinical manifestations include swelling and less commonly pain and deformity (5, 9). The talus, distal femoral and distal tibial epiphyses are the commonest sites of disease (3, 5). Multiple bones (in a single extremity) are affected in 60-70% of cases and the disorder can be classified into one of three forms (8, 9). The localized form describes monoarticular involvement. The classical form presents with involvement of more than one area in a single limb. In the general or severe form, there is involvement of a whole lower extremity from the pelvis to the foot or ankle. Kettelkamp *et al.* reviewed 57 cases of DEH in 1966 and found that only one side of the body was affected (5). However more recent studies have documented bilateral DEH (3, 6). The histopathologic features of the lesion are identical with those of an osteochondroma (6, 9). Malignant transformation has never been reported (2, 6, 9). Although histologically identical, DEH and osteochondromas are separate clinical entities and are distinguished by their different location : osteochondromas arise from the metaphysis or diaphysis whereas DEH arises from the epiphysis (2, 6). Although the prognosis of this disease is generally regarded as good, with benefit from local excision of the lesion, persistent deformities and secondary osteoarthritis are evident in some cases (5, 9).

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SAMENVATTING

N. LEVI, S. E. ØSTGAARD, B. LUND. Dysplasia epiphysialis hemimelica (ziekte van Trevor) van de distale radius.

De auteurs beschrijven een geval van dysplasia epiphysialis hemimelica of Trevor's aandoening in de radius van een eskimo in Groenland. Deze aandoening is niet te onderscheiden van een osteochondroma. Nochtans is een osteochondroom zelden epiphysair gelocaliseerd.

RÉSUMÉ

N. LEVI, S. E. ØSTGAARD, B. LUND. Dysplasie épiphysaire hémimélique (maladie de Trevor) du radius distal.

Les auteurs présentent un cas de dysplasie épiphysaire hémimélique (DEH) ou maladie de Trevor au niveau du radius distal chez un esquimau du Groenland. Cette dysplasie rare est impossible à distinguer microscopiquement d'un ostéochondrome ; cependant, son site d'origine épiphysaire le distingue de l'ostéochondrome, qui naît typiquement au niveau métaphysaire.