Carpal-tarsal osteolysis

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Abstract. Two cases of carpal-tarsal osteolysis are described. It is believed that clinical and radiological findings of the patients are compatible with the dominant type of the disease. Although case 2 does not have the indications of nephropathy, it is rather too early to decide whether he has it or not, because of his age.

Carpal-tarsal osteolysis was first described by Froehlich and Corret in 1937 [1]. Since that time there have been about 25 reports in the literature and different terms have been used to denote this rather rare condition.

The disease begins as an arthritic episode in early childhood and may simulate juvenile rheumatoid arthritis. The syndrome is characterized by progressive shrinkage of bones of hands and feet. It leaves severe deformities and disability in adult life. Associated renal findings and hypertension have been described repeatedly in the literature. In nearly all of them the initial renal symptoms have generally appeared in adolescence and less frequently in childhood.

We report two new cases. One of them had nephritic findings which began in early childhood. The other was without nephropathy.

Case 1

A 7-year-old boy was admitted to the hospital because of deformities of his hands and feet.

It was learned that his hands and feet were swollen and there was increased sensitivity to touch when he was 6 months old. At the age of two, he developed walking difficulties with recurrent swellings of the wrists. His left wrist and ankles were especially affected, becoming progressively worse, and flexion contractures of the hands were observed.

His mother, father and his eldest brother were all physically and mentally normal. There was no family history of any disease and the parents were unrelated.

Fig. 1. Case 1: There is apparent resorption of the proximal portion of the second to fifth metacarpals. The carpal bones are totally resorbed.
Fig. 2. Case 2: The carpal bones are totally absent while radius and ulna show gradual tapering and bowing distally. The most marked change in the wrists is the destruction of the distal ends of the radius and ulna and the proximal ends of the metacarpals. The same changes although milder can also be observed in elbows. Because of apparent mutilative changes in wrists and elbows (the so-called arthritis mutilans) the joints mentioned are dislocated.

Fig. 3. Case 2: Most of the tarsal bones have vanished. Some of them and the proximal portions of the metatarsals are destroyed.