Carpal and tarsal osteolysis

L. Lemaitre¹, J. Remy¹, M. Smith¹, J. P. Nuyts², J. Cousin³, M. O. Farine⁴ and P. Debeugny⁵

Departments of ¹Pediatric Radiology and ²Pediatrics, Hopital Calmette; ³Department of Pediatrics, Hopital Saint Antoine; and Departments of ⁴Pathology and ⁵Pediatric Surgery, Cité Hospitalière, Lille, France

Abstract. This report describes the symptoms, clinical course and radiological features in three cases of idiopathic carpotarsal osteolysis. Before signs of extensive osteolysis were noted, one of our patients showed flattening, loss of harmonious curvature and minimal osteoporosis of carpal and tarsal bones, all early radiological signs which have not been described previously. We discuss the findings, classification and our ideas as to the cause of the disorder.

Key words: Idiopathic osteolysis – Carpal and tarsal bones – Capillary proliferation – Nephropathy

Carpal tarsal osteolysis is a rare disorder of unknown aetiology. There is a gradual resorption of bone beginning in the carpus and tarsus. It is frequently associated with similar metacarpal and metatarsal lesions. Bone lysis may spread and involve the proximal limb segments. This variety of osteolysis may be familial and/or associated with life-threatening nephropathy.

There being no particular clinical, biochemical or histological features that are specific for the disease, the main criterion for classification is radiology [14]. The radiologist must therefore know the radiological signs of the condition in order to avoid diagnostic errors and prevent inappropriate treatment.

An analysis of the radiological findings in three recently examined cases is presented. Special emphasis is laid on the early evidence of incipient osteolysis.

Case reports

Case 1. This 11-year-old boy with no previous history of disease was admitted to the Saint Antoine hospital in February 1979 for an evaluation of pain in the hands and feet.

The clinical history began in January 1978: nodules appeared in the palms, at the level of distal interphalangeal joints and at the right medial malleolus. Several biopsy specimens were taken from various sites, resulting in the diagnosis of infantile digital fibroma.

Following cryotherapy, the nodules receded, but the wrists and feet became painful. The first radiological assessment in May 1978 (Fig.1a) was negative. However, persistent clinical complaints led to another radiological evaluation in August 1978 (Fig.1b), which showed carpal osteolysis but no lesions in the feet. A "rheumatological" appraisal was made in January 1979, which excluded polyarthritis. During hospitalization, the boy was in good general condition. He had no fever, but had considerable limitation of flexion and extension of both wrists. The feet hurt but no objective change was detected there. There were no associated malformations and the family history – only the parents and the elder brother were examined – revealed no hereditary factor. The rest of the clinical examination was normal, in particular the blood pressure (100/60 mmHg).

The laboratory results were as follows: ESR 3/4; 4,660,000 red cells/mm³; Hb 8 mmol/l; (5 800 white cells/mm³; the phosphorus/calcium balance was normal: Ca 2.60 mmol/l, P 1.10 mmol/l, alkaline phosphatase 257 IU; normal renal function: urea 6 mmol/l, creatinine 50 µmol/l; no proteinuria. Tests for inflammation were negative – CPR, Rose-Waller and LE-cells. Chromatography of urinary oligosaccharides and mucopolysaccharides was normal.

The various radiological examinations carried out demonstrated rapid deterioration up to May 1979, then relative stability to the present time. The carpal lesions appeared first and were more severe than the tarsal ones. There was a general lysis of all the carpal bones together with similar lesions at the bases and epiphyses of the metacarpal bones. At both ends of the proximal and middle phalanges, at the distal end of the radius and on the phalangeal tufts, the lesions were bilateral and roughly symmetrical (Fig.1 a and b). In the feet, lysis was less serious, involving the right tarsal navicular, the joint surfaces of the right and left second and third cuneiforms and the distal ends of several phalanges (Fig.2b and c). The rest of the skeleton did not present any radiologically detectable anomaly.

Case 2. This 3-year-old boy was admitted to the Calmette hospital in April 1978 because of painful swelling in both wrists.

At various times during the early period of his life, different complaints resulted in assessment of skeletal age, which actually facilitated the subsequent analysis of both onset and development...
of the osteolytic lesions. However, the diagnosis was not made at the onset of disease. In May 1976, a positive tuberculin skin test was documented and the skeletal age assessed. In September 1976, the skeletal age was reassessed because of failure to walk at normal age. In November 1976, he developed a definite, permanent slight limp and painful swelling of the dorsum of the right foot. Films showed irregular joint surfaces involving the calcaneus and cuboid bones (Fig. 4a) with no carpal lesions. Normal laboratory assays included ESR, blood cell counts, leucocyte pattern and fibrinogen values. A proteinuria of 0.30 g per 1 was found. After immobilization for 3 months in plaster, the child still suffered from an intermittent limp, but he had no remaining pain, oedema or local inflammatory signs (March 1977). On account of the extensive radiological lesions involving the right cuboid, calcaneus and talus (Fig. 4b) and in spite of laboratory assays still showing no signs suggestive of inflammation, the condition was diagnosed as "osteo-arthritis".

Three months later, in June 1977, the discovery of failure to grasp adequately with the left hand led to a radiological evaluation which was considered as normal, although at review minimal changes in shape of the left capitate and hamate were noticed (Fig. 3a). These findings were shown to have become more marked when another radiological study was made in October 1977 on account of the development of pain in the joints (Fig. 3b).

Hospitalization in April 1978 followed because of painful swelling in both wrists. Functionally there was a limitation of extension and flexion of the wrists and pain on passive mobilization of the hands. A moderate degree of discomfort during walking persisted. On examination there was apparent swelling of the dorsum of the wrist on both sides but no detectable sign involving the feet. Repeat laboratory assays were once more normal: ESR, full blood count, antistreptolysins, CPR, fibrinogen, Hargraves' cells, Rose Waller, antinuclear antibodies within normal range. Renal evaluation was quite normal apart from persistent proteinuria, and the blood pressure was normal too. However, the radiological lytic lesions of the carpus and tarsus led us to propose the diagnosis of carpal-tarsal osteolysis (Fig. 3c and 4c).

The course was characterized by increasing lytic lesions of the carpus and tarsus (Fig. 3d and 5), followed by changes in the left elbow (Fig. 6a) and later on in the right (Fig. 6b). The right foot was severely deformed (Fig. 5) and an arthrodesis was performed in March 1980, with resection of the right cuboid. The microscopical study of specimen showed a bone structure characterized by osteoporosis of an unusual type with an adipose marrow, and fibrotic patches, which contained vascular proliferation and a great many capillaries, as well as a few, somewhat larger vascular cavities lined by fibrous tissue. Contiguous with the fibrocartilaginous tissue was a vascular proliferation of the angiomatous type.

Case 3. This 10-year-old girl was admitted to the Dunkirk hospital in February 1978 for evaluation of a swelling of both wrists. Intermittent oedema of the hands was first noticed at the age of 4 or 5 years, and she had had a dysfunction of the upper limbs for the