Carpal Osteolysis: An Unusual Entity Treated by Renal Transplantation

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Dear Sir,

Carpal and tarsal osteolysis (CTO) is a severe disorder of unknown etiology characterized by gradual resorption and lysis of bones of the hands and feet, beginning in the carpal and tarsal bones [1]. The diagnosis is based on the X-ray abnormalities and both sporadic and familial cases have been reported [2,3]. The nephropathy appears more frequently in the sporadic form and it does not have specific features, appearing during childhood or adolescence, with progression to end-stage renal disease (ESRD) in a variable period of time [4–7]. We report here a case of a patient with sporadic CTO and renal failure that received a cadaveric renal transplant after treatment with continuous ambulatory peritoneal dialysis (CAPD). To our knowledge, there are only two previous cases successfully transplanted [8, 9].

Case Report

A 14-year-old girl was admitted to the hospital to receive a renal transplant. Her previous medical history was unremarkable. At the age of 7 years, she consulted somewhere else because of slow growth, pes cavus and limitation of flexion and extension of both wrists. The physical examination at that time revealed height 107 cm ($p < 3$), weight 17.800 kg ($p < 3$). A rather distinctive and asymmetric face with micrognathia, retrognathia and hypertrichosis was evident. Small hands with ulnar deviation of the fingers and nodules at the level of proximal interphalangeal joints of the right hand were observed. Flexion and extension of both wrists were mildly limited. Routine laboratory data, including tests for renal function and urinalysis, were normal.

X-rays of the wrists showed absence of the second line of the carpal bones with no other abnormalities. Karyotype was normal.

Eight months before admission, she developed vomiting, generalized edema, dyspnea and oliguria and was admitted at another hospital. She looked severely ill and pale. Blood pressure was 160/96 mm Hg ($p > 95$). Heart rate was 140 bpm. Laboratory tests showed hemoglobin 80 g/l (8 g/dl), creatinine 1,096 $\mu\text{mol/l}$ (12.4 mg/dl), urea 11.7 mmol/l (313 mg/dl), sodium 135 mmol/l, calcium 1.57 mmol/l (6.3 mg/dl), total serum proteins 48 g/l. Serum phosphate was not reported. X-ray of the abdomen showed decreased-

Fig. 1. a X-ray of the right wrist showing osteolysis with almost complete resorption of the carpal bones, extending to the distal end of the cubitus and the lower radial epiphysis. No bone fusion is observed, b X-ray follow-up after transplantation, showing progression of bone abnormalities with lysis of the distal cubitus.

sized kidneys and a renal echography revealed the presence of atrophic kidneys. X-rays did not show extraskkeletal calcifications. Her diagnostic workup ruled out the existence of an obstructive uropathy as the cause of her renal disease. The diagnosis of ESRD was made and the patient was placed on a CAPD program. After 8 months on CAPD, she was referred to our hospital to consider a renal transplant. When she came to our center, her height was 141 cm (-3 SD), weight 31 kg, blood pressure 140/90 mm Hg. There were no new findings in the physical examination. Radiological studies showed worsening of the abnormalities described previously, with total absence of carpal bones (fig. 1 a). She received a cadaveric renal transplant. Ciclosporin and prednisone were administered as immunosuppressive agents. Her recovery was uneventful, except for mild hypertension that was controlled by small doses of beta-blockers and diuretics. At a follow-up visit 9 months later, her serum creatinine was 106 $\mu\text{mol/l}$ (1.2 mg/dl). New X-rays of the wrists showed clear evidence of progression of the bone abnormalities with lysis of the distal cubitus (fig. 1b).

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Discussion

In our patient, the diagnosis of CTO associated with nephropathy (Thieffry-Shurtleff syndrome) [10] was supported by the presence of ESRD of uncertain etiology and characteristic radiological abnormalities in spite of the absence of previous arthritic episodes. Severe hypertension, azotemia and slight abnormalities in the urinary sediment are usually present. Most of the cases evolve to ESRD in a short span of time.

Although pathological changes are not uniform, vascular abnormalities are among the most commonly reported findings. Bennett et al. [6] described the histological findings at necropsy in a patient who died before developing ESRD. There were interstitial changes and abnormalities in the vasculature most prominent in the small vessels. Some glomeruli were thrombosed and necrotic with crescent formation. Although the pathogenesis has not been satisfactorily explained, those histological changes suggest a primarily vascular disease. Furthermore, there are some case reports describing vascular changes in other territories, such as the myocardium, skin and synovial cartilage. An abnormal immune process has also been postulated to explain the pathogenesis.

In our case the deterioration of her renal function was probably quite rapid, as is suggested by her normal renal function when she was 7 years old and the absence of radiological signs of secondary hyperparathyroidism. Only 13 cases of CTO associated with nephropathy have been reported [6, 8, 9, 11]. We have found only 2 other cases in the literature reporting successful renal transplants in patients with CTO and ESRD [8, 9]. Although clinical improvement after transplantation has been described [9], it is not known whether or not there is regression of bone lesions after the implantation of a renal graft. Interestingly, the X-ray findings of our patient clearly worsened, as is shown in figure 1.

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