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CASE REPORT

A case of infantile scurvy treated only with vitamin C : A forgotten disease

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Scurvy is a rare disease occurring because of a nutritional deficiency of vitamin C. In the paediatric age group the disease is usually characterized by musculoskeletal manifestations. Treatment is straightforward and consists of vitamin C administration. However, if the patient is left untreated, scurvy may be life-threatening. We report here the case of a 16-month-old infant with scurvy. After proper treatment, the complaints disappeared in a very short time period and the boy grew up as a completely normal child during the 12 years follow-up. Nowadays only few physicians have experience with this disease, and ascorbic acid deficiency can thus easily be overlooked. With this paper we aimed to remind of the efficacy of vitamin C administration for patients with scurvy.

Keywords : scurvy ; vitamin C ; musculoskeletal manifestations ; infant.

INTRODUCTION

Scurvy is a rather rare disease resulting from a nutritional deficiency of vitamin C. In the literature, there are some reports about scurvy in adults (*4,5*) but there is scarce mention of infantile scurvy (*6*). This is mainly because breast-feeding has become universal (*1*). In the paediatric age group, scurvy occurs in children with severely restricted diets. Musculoskeletal manifestations are prominent (*2*). In this age group, scurvy may become life-threatening (*6*). With this paper we aimed to remind of the efficacy of vitamin C administration for patients with this rare disease.

CASE REPORT

A 16-month-old boy was admitted to our institution because of swelling and tenderness of his limbs. He was refusing to walk. Thorough physical examination revealed that he had bilateral pseudoparalysis of the lower limbs. Arthralgia was obvious in his knees but there were no joint effusions. His gums were swollen and bleeding. There was no history of trauma. The mother stated that she could not breast-feed her baby because she did not have enough breast milk. The baby was fed only

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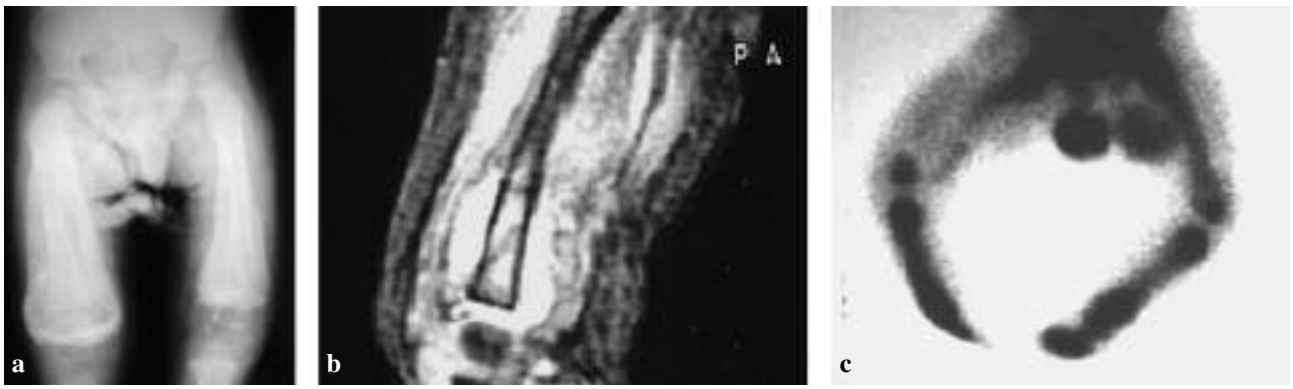

Fig. 1. — Radiograph (a), MRI (Magnetic Resonance Imaging) (b) and scintigraphy (c) of the patient before treatment.

with fruit juice, water, commercial puddings and baby food, made at home. He took no vitamin supplements. Radiographs of the femurs revealed severe elevation of the periosteum at the distal ends, cortical thinning with periosteal hypertrophy, metaphyseal spurs, ground glass osteoporosis, subepiphyseal infarction (Corner Sign) and a dense margin around the epiphyses (Wimberger's ring) (fig 1a). Laboratory tests, including white blood cell count, erythrocyte sedimentation rate and C-Reactive Protein, showed no evidence of infection nor arthritis. Magnetic Resonance Imaging (MRI) showed a diffuse symmetrical abnormal signal around the distal femoral regions (fig 1b). Bone scintigraphy with Tc-99m showed markedly increased club-shaped technetium uptake in both femurs (fig 1c). These findings were thought to support a diagnosis of infantile scurvy.

Supplementation with 1.5 g/day of oral vitamin C was started immediately. The parents were educated about dietary modifications. Within 10 days the periodontal problems resolved and swelling of the gums was markedly improved. The patient did no longer need analgesics and could move again without any pain. He was discharged from hospital after two weeks of treatment. One month later, radiographs of the femurs revealed calcification of the subperiosteal haematoma. The osteoporosis was also improved. The child started walking without pain. He was doing well and free of clinical symptoms at a follow-up of 12 months. The radiological findings resolved almost totally.

It has been 12 years since the child was first admitted in our institution and during this period the child grew up fine and he is still doing well. Radiographs of the femurs are normal (fig 2).

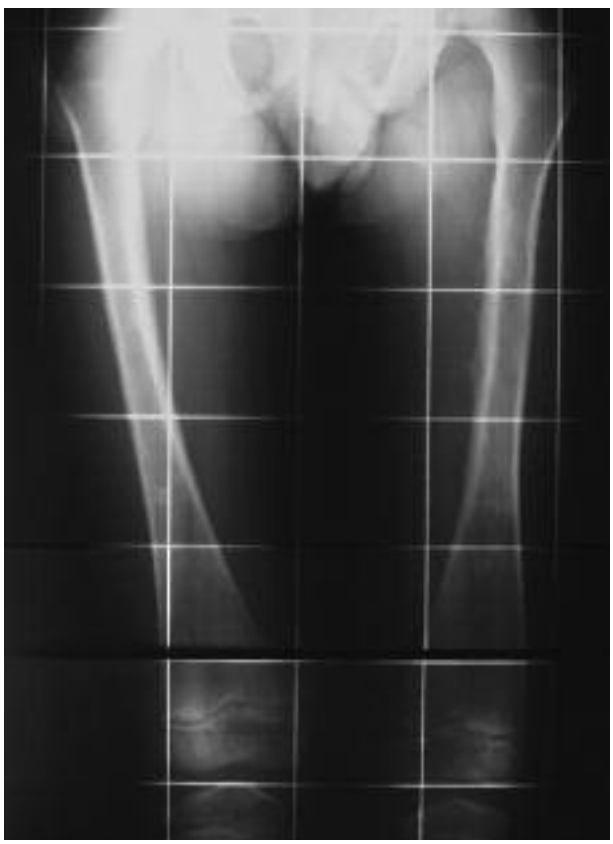

Fig. 2. — Radiograph of the femurs 12 years after the initial treatment.

DISCUSSION

Scurvy is an ancient disease. It has already been described in the Ebers papyrus written 1500 BC (*1*). Unlike most mammals, human beings are unable to synthesize vitamin C. Human breast-milk has a high concentration of ascorbic acid (vitamin C) provided that the mother does not have a vitamin C deficiency (*7*). That is why scurvy is rarely seen in the paediatric age group. Infants with strictly restricted diets are at risk for the disease. Front *et al* in 1978 and Weinstein *et al* in 2001 reported cases of scurvy in the paediatric age group, treated with vitamin C (3,*7*). Fatal infantile scurvy was reported by Mimasaka *et al* (*6*).

Vitamin C has a key role in the synthesis of collagen. Some studies attribute the capillary fragility responsible for the clinical symptoms of the disease to deficiency of pericapillary collagen (*4,2*).

Only few physicians have experience with the disease nowadays and thus ascorbic acid deficiency can easily be overlooked. The most distinguishing features of the disease are : a history of nutritional problems, irritability, swollen and bleeding gums, pseudoparalysis, pain in the limbs and arthralgia. Musculoskeletal manifestations are prominent in infants. Routine laboratory tests are not very helpful but are necessary for the differential diagnosis. An ascorbic acid level below 2.5 mg/l is critical for the disease but even a lesser level is not always an indication for scurvy ,because the deficiency must be maintained for more than 5-6 months before clinical symptoms occur (*1,2*).

Radiological features are very important for the diagnosis. Elevation of the periosteum due to subperiostal bleeding, cortical thinning, metaphyseal spurs, ground glass osteoporosis, subepiphyseal infarction (Corner sign) and a dense margin around the epiphyses (Wimberger's ring) can be seen (*1*). MRI is not necessary for the diagnosis but can be helpful for differentiation from other diseases. Radioisotope bone scan can also be useful due to the specific uptake pattern (*3*).

The treatment of scurvy is very simple. Supplementation with 1-1.5 g/day of oral vitamin C for 2 weeks is a remarkably effective treatment. Early diagnosis is vital as the disease may sometimes be life-threatening. We strongly recommend considering scurvy in the differential diagnosis when there is a history of nutritional problems and above mentioned clinical and radiological findings exist.

REFERENCES

1. **Edeiken J, Hodes PJ.** *Roentgen Diagnosis of Diseases of Bone.* Wilkins-Williams, Baltimore, 2nd ed, 1973, pp 418-426.
2. **Fain O.** Musculoskeletal manifestations of scurvy. *Bone Joint Spine* 2005 ; 72 : 124-128.
3. **Front D, Hordoff R, Levy J, Benderly A.** Bone scintigraphy in scurvy. *J Nucl Med* 1978 ; 19 : 916-917
4. **Halligan TJ, Russell NG, Dunn WJ, Caldroney SJ, Skelton TB.** Identification and treatment of scurvy : a case report. *Oral Surg Oral Med Oral Pat Oral Radiol Endod* 2005 ; 100 : 688-692.
5. **Kieffer P, Thannberger P, Wilhelm JM, Kieffer C, Schneider F.** Multiple organ dysfunction dramatically improving with the infusion of vitamin C : more support for the persistence of scurvy in our welfare society. *Intensive Care Med* 2001 ; 27 : 448.
6. **Mimasaka S, Funayama M, Adachi N, Nata M, Morita M.** A fatal case of infantile scurvy. *Int J Legal Med* 2000 ; 114 : 122-124.
7. **Weinstein M, Babyn P, Zlotkin S.** An orange a day keeps the doctor away : scurvy in the year 2000. *Pediatrics* 2001 ; 108 : e55.