Radiological and laboratory features of infantile cortical hyperostosis

A case report

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Summary. *We describe a boy aged nine months with infantile cortical hyperostosis in association with cystic fibrosis. Symmetrical periosteal thickening was present in the clavicles, ribs, femora, humeri, ulnae and radii. Periosteal hyperostosis of the humerus developed in association with an increase in the levels of serum alkaline phosphatase a month before the appearance of hyperirritability and soft tissue swelling about the hip joints. The condition gradually resolved during the following eight months.*

Résumé. *Nous rapportons ici le cas d'un nourrisson âgé de neuf mois présentant une hyperostose corticale infantile associée à une fibrose kystique. Un épaississement périosté a été observé symétriquement dans les clavicules, les côtes, les fémurs, les humérus, les cubitus et les radius. Les hyperostoses périostées de l'humérus sont survenues en association avec une augmentation du taux des phosphatases alcalines un mois avant l'apparition d'hyperirritabilité et de gonflement des tissus mous aux alentours des articulations coxo-fémorales. Un retour graduel à la normale a été observé dans les huit mois.*

Introduction

In 1930, Roske reported an infant who suffered from hyperirritability with thickening of bones and painful swelling of the overlying soft tissues [8]. In 1945, Caffey and Silverman described 4 examples of the

same disorder and termed the condition “infantile cortical hyperostosis” [2]. Other reports have followed [3, 7, 9], but little is known about the radiological findings before the onset of symptoms. We report an affected infant in whom the radiological changes of periosteal thickening and the levels of serum alkaline phosphatase were serially checked during the course of the disorder.

Case report

In February 1989, a nine-month old boy, who had been admitted to the paediatric ward with intractable diarrhoea and repeated pulmonary infection due to cystic fibrosis, was referred to our orthopaedic clinic on account of irritability when changing diapers. His temperature was 38.5°C and physical examination revealed soft tissue swelling and tenderness in his right thigh. Hyperirritability was noticed when the right leg was handled. Radiographically, symmetrical periosteal thickening was demonstrated in the clavicles, ribs, humeri, ulnae and radii (Fig. 1), femora and tibiae (Fig. 2). No other members of the family had a similar condition. There was no evidence to suggest the battered child syndrome or scurvy. A serological test for syphilis was negative as were cultures of the blood and hip joint fluid. The serum alkaline phosphatase level was elevated. Anaemia with an iron deficiency pattern and thrombocytosis were present. Serum calcium and phosphorous levels and liver and renal function were normal. A diagnosis of infantile cortical hyperostosis was made; the clinical and radiological features gradually subsided over a 5-month period following the onset of symptoms.

Serial radiographs including bilateral views of the humeri had been obtained during the management of the child's cystic fibrosis. Retrospectively, these radiographs revealed sequential changes of periosteal thickening in the humeri before and after the appearance of the clinical symptoms. The humeral cortical index, as defined by Barnett and Nordin [1], was calculated as follows: the thicknesses of the medial and lateral humeral cortices were measured and the sum of these thicknesses divided by the total width of the right humerus at the midpoint of the shaft (Fig. 3). These indices varied from 0.25

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Fig. 1. Radiograph showing symmetrical periosteal thickening of the humeri (arrows), ulnae (arrow) and radii (arrow)

Fig. 2. Radiograph showing symmetrical periosteal thickening of the femora (black arrows) and tibiae (white arrow)

to 0.75 over an eight-month period. In January, one month before the onset of the symptoms, cortical thickening began to increase sharply and peaked in February (Fig. 4). Thereafter, it declined gradually until April and then more rapidly, reaching the initial levels in July. The serum alkaline phosphatase level also started to increase one month before the onset of the symptoms, peaking in March and decreasing to the baseline levels after April (Fig. 5).

Discussion

Infantile cortical hyperostosis is a benign disorder of unknown origin that usually has its onset before the age of 6 months. The condition presents with general irritability, soft tissue swelling and tenderness of the affected parts. Radiographs reveal a periosteal reaction in the bones which underlie the swelling.

Those most commonly involved are the mandible, clavicle, ribs and the long bones of the limbs [2, 7, 8]. The absence of mandibular involvement, as in the patient whom we investigated, is uncommon, although not unprecedented. Wilson reported changes in the left radius and ulna without mandibular involvement [10] while Finsterbush and Hussein reported three

Fig. 3. Diagrammatic representation of the measurement and calculation of the cortical index

affected persons, only one of whom had a periosteal reaction in the mandible [5].

There have been no reports on infantile cortical hyperostosis accompanied by cystic fibrosis. This latter condition is extremely rare in Japan although it is one of the most common lethal inherited disorders in Europe and North American. Cystic fibrosis is characterised by the production of abnormally viscous

Fig. 4a, b. Radiological changes of the humerus before and during the symptoms. **a** Left: early, middle: mid, right: late January, **b** Left: early, middle: mid, right: late February. Cortical thickening (arrows) appeared in mid January **a**: middle and progressed in February (**b**)

Fig. 5. Cortical index and serum alkaline phosphatase level

mucous secretions in the lungs and digestive tract [6]. In our patient, the presence of fever, an elevated erythrocyte sedimentation rate and repetitive pneumonia due to cystic fibrosis led us to infer that an infectious agent might be responsible for infantile cortical hyperostosis, although no bacterial or viral agents could be identified.

There have been few reports of presymptomatic radiographic changes in infantile cortical hyperostosis. In their review, Finsterbush and Rang [4] reported that the hyperirritability was sometimes present early in the course of the disorder and that it could precede the soft tissue swelling by a few days or weeks. They also stated that the swelling usually began before the roentgenographic changes appeared in the underlying bone [4]. Retrospective radiological analysis of our patient revealed that periosteal hyperostosis had already commenced, together with elevation of the serum alkaline phosphatase level, one month before the onset of symptoms.

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