Infantile peri-osteitis

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A 14-week-old girl was brought in with a history of painful swelling of both legs since the age of one month. The onset was insidious and was not associated with trauma or fall. There was no history of fever or associated constitutional symptoms. The birth history was normal and the infant was apparently asymptomatic until the age of one month. Examination revealed a healthy and alert infant. Both legs were bowed anteriorly and a uniform bony thickening of both tibiae was palpable throughout their lengths (figure 1). Both legs were extremely tender and the infant would withdraw both lower limbs and cry incessantly if any attempt was made to touch them. There was no increase in local temperature nor redness of the overlying skin. Knees and ankle joints were normal and demonstrated a full range of motion. Regional lymph nodes were not enlarged and other bones and joints were normal on examination. X-Rays of both legs revealed peri-osteitis of both tibiae with extensive subperiosteal new bone formation involving the entire diaphysis (figure 2).

Questions

- 1 What is the differential diagnosis of peri-osteitis in an infant?
- 2 What further investigations are required?
- 3 What is the likely diagnosis and treatment?

Figure 1 Clinical photograph showing bony swelling with anterior bowing of both legs

Figure 2 (A) Anteroposterior radiogram of both legs showing cortical thickening and subperiosteal new bone formation, (B) lateral radiogram of both legs

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Accepted 24 October 1997

Answers

QUESTION 1

The differential diagnosis of infantile periosteitis is given in box 1.

Infantile peri-osteitis: differential diagnosis

- osteomyelitis
- congenital syphilis
- scurvy
- hypervitaminosis A
- tumours: Ewing's sarcoma and metastatic neuroblastoma
- trauma
- idiopathic: infantile cortical hyperostosis (Caffey's disease)

Box 1

QUESTION 2

Serologic, biochemical and other laboratory tests are required to reach a diagnosis, and include a haemogram with complete leucocyte count and erythrocyte sedimentation rate, skeletal X-rays, bone scintigraphy to rule out other sites of involvement, serology (eg, VDRL and *Treponema pallidum* specific fluorescent antibody tests, which are sensitive for syphilis), and biopsy, if required to rule out malignancy.

In this child, the haemogram was normal except for a mild increase in erythrocyte sedimentation rate. Serum alkaline phosphatase was moderately elevated. Serologic tests for syphilis were non-reactive for the child as well as for the parents. Skeletal survey revealed similar lesions of cortical thickening and subperiosteal new bone formation in the mandible, left clavicle, right humerus, both ulnae and left radius, in addition to the lesions in both tibiae.

QUESTION 3

The clinical presentation, radiographic features and normal laboratory investigations are consistent with a diagnosis of infantile cortical hyperostosis or Caffey's disease. The exact aetiology and pathogenesis of the disease are unknown and treatment is largely empirical. The disease is reported to be self-limiting and spontaneous resolution is the rule.

Analgesics may be required in the acute phase to alleviate severe pain and tenderness. Corticosteroids are effective in alleviating the acute systemic manifestations but have no effect on the natural history of the disease nor do they hasten the healing process. Their use is controversial and they are best reserved for those infants who have extensive, severe or recurrent involvement. Antibiotic therapy or surgery is not indicated for this condition.

Discussion

Infantile cortical hyperostosis is a rare and poorly understood disease of early infancy that is characterised by soft tissue swelling and cor-

Infantile cortical hyperostosis: diagnostic features

- narrow age of onset (between one and six months)
- triad of irritability, swelling and bone lesions with cortical thickening and subperiosteal new bone formation
- mandibular involvement in most cases

Box 2

tical thickening of underlying bones. The first complete description of this clinical entity was given by Caffey and Silverman¹ in 1945.

The cause is unknown. Reports of familial occurrence suggest a possible hereditary factor, most probably an autosomal dominant trait. Other theories include an inherent defect of the arterioles of the periosteum causing local hypoxia, an allergic basis, and viral or bacterial infection causing acute inflammation.

Biopsy specimens reveal typical pathologic changes. In the early stages, there is a marked inflammatory process involving the periosteum and surrounding soft tissues. Gradually the inflammation subsides leaving behind a thickened periosteum and immature subperiosteal lamellar bone. A vascular fibrous tissue occupies the bone marrow spaces.

Disease onset is usually around the eighth week of postnatal life up to the age of six months.¹⁻³ The common clinical manifestations are hyperirritability and the presence of soft tissue mass over a long bone or commonly over the mandible. The swelling appears suddenly, is deep and firm and is usually tender initially. The tenderness gradually subsides and the soft tissue swelling merges into a uniform bony thickening of the entire shaft.

The mandible is the most common site of involvement (75–80%).^{2,3} In the limbs, the ulna is most commonly affected followed by the tibiae, clavicles, scapulae and ribs.² Other long bones are less commonly involved. More than one bone is commonly affected and if involvement is bilateral, it is often asymmetrical.

In early stages, the external surface of the involved bone is coarse, the underlying cortex is still visible and abundant subperiosteal new bone formation is seen. As the disease progresses, the new bone increases in density and becomes homogenous with the thickened cortex.

Several months, sometimes years are required for complete resolution. Spontaneous remission/exacerbations have been described at a later age. The overall prognosis is good, though residual deformities may occur, especially when the disease is severe with intermittent recurrences. Reported sequelae⁴ include fusion of adjacent bones such as the ribs, tibia and fibula, and radius and ulna; anterior bowing of the tibia or femur, limb length inequality. Other rare complications reported in literature are pleural effusion, diaphragmatic paralysis and exophthalmos.

Final diagnosis

Infantile cortical hyperostosis (Caffey's disease).

Keywords: peri-osteitis; infantile cortical hyperostosis; Caffey's disease

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Aortic valve mass and sudden blindness

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A 63-year-old man presented with a sudden and painless loss of vision in his left eye. He had no history of heart disease or arrhythmias, diabetes mellitus, hyperlipidaemia, hypertension or smoking. On admission his blood pressure was 160/90 mmHg, and his physical examination was unremarkable. Electrocardiography demonstrated normal sinus rhythm, with no signs of ischaemia or left ventricular hypertrophy. Ophthalmologic examination revealed no light perception in his left eye, and a pale and oedematous retina. The findings suggested a high probability of an embolic central retinal artery occlusion. Therefore 1.5 million units of streptokinase were given, with only a slight improvement of his vision. A transthoracic echocardiography (TTE) and carotid ultrasound performed the next day were interpreted as normal. The patient was discharged on warfarin therapy. While at home, he continued to have episodes of lightheadedness, dizziness and visual hallucinations once or twice a week. Transoesophageal echocardiography (TEE) was performed a few weeks after discharge (figure).

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Accepted 16 December 1997

Figure Transoesophageal echocardiography

Questions

- 1 What abnormality is shown in the aorta on the echocardiogram ?
- 2 What is the differential diagnosis ?
- 3 What is the most probable diagnosis, considering the echocardiographic findings and the patient's history and physical findings ?
- 4 What is the treatment of choice ?