Pediatr Radiol (1998) 28: 234–236

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Skeletal tuberculosis: dactylitis and involvement of the skull

Received: 3 July 1997
Accepted: 5 November 1997

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Abstract A re-emergence of tuberculosis (TB) is occurring world wide in both developed and developing countries. The clinical picture caused by infection with *M. tuberculosis* may simulate many other disease entities and may result in unnecessary investigations with a delay in diagnosis and treatment. Skeletal TB tends to be isolated to one anatomical site. We report a 6-year-old boy with disseminated skeletal TB with dactylitis resembling sickle cell anaemia and lytic lesions similar to those which are often seen in neuroblastoma, Langerhans' cell histiocytosis and leukaemia. The clinician should be aware that TB can mimic almost any disease and recognise the radiographic appearances of skeletal tuberculous lesions.

Introduction

Tuberculosis (TB) is an important cause of childhood morbidity and mortality in many developing countries. The advent of AIDS has caused an exacerbation of TB in developing countries and a re-emergence of the disease in industrialised countries [1]. Symptoms of tuberculosis can be varied and may simulate many other disease entities. Skeletal TB, which was the most frequent site of extra-pulmonary TB prior to the availability of effective chemotherapy, now occurs only rarely and is restricted to one anatomical site [2]. We present a child with disseminated skeletal TB with signs which simulated other conditions.

Case report

A 6-month-old boy presented with a 1-week history of coryza, coughing and fever together with a swelling over the left forehead. Physical examination revealed severe growth retardation with mass, length and head circumference below the third percentile for age. A 3×3 cm cystic swelling was present on the left forehead. The fourth finger of the left hand and the dorsum of the left foot were swollen. Cervical lymphadenopathy and mild respiratory distress with bilateral crepitations and rhonchi were noted. A mildly enlarged liver and tip of the spleen were palpated. Further physical examination was normal. The white cell count was 10.6×10^9/l, Hb 5.2 g/dl, MCV 65 fl and platelets 220×10^9/l. Urea, electrolytes and liver enzymes were normal except for a LDH of 458 u/l (normal < 350 u/l). A Mantoux skin test was negative (0 mm).

Signs suggestive of right lower lobe pneumonia were seen on the chest X-ray. Skeletal X-rays demonstrated multiple lytic lesions with slightly sclerotic borders in the cranial vault, humeri, femora, tibiae, radii, ulnae, metatarsals, metacarpals and phalanges (Figs. 1–3). Tc-99 bone scintigraphy did not show increased uptake in these lesions.

The histology of tissue obtained on biopsy of both the mass in the left frontal area of the skull and the cervical gland was typical of TB. *M. tuberculosis* was cultured from gastric washings. Tuberculosis meningitis was ruled out by a normal spinal fluid tap.

Discussion

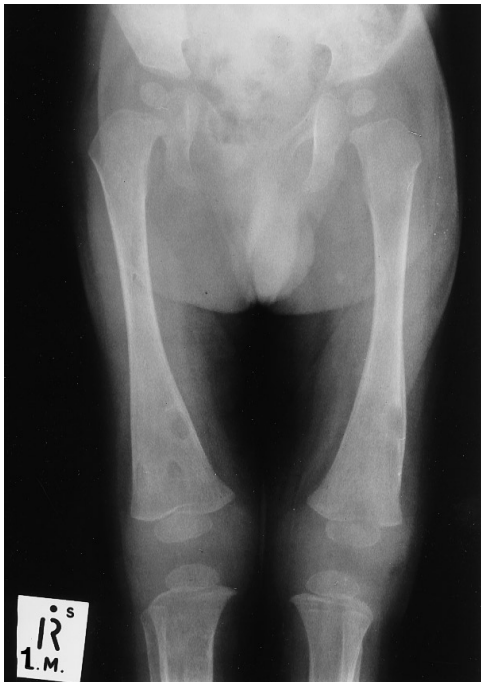
The outcome of lympho-haematogenous spread of tuberculous bacilli, which is part of the primary complex, is determined by the quantity of bacilli released and by host resistance [3], and may result in active metastatic tuberculous lesions in extra-pulmonary sites. Skeletal lesions typically begin as an endarteritis in the metaphysis of the long bones. Granulation tissue erodes and destroys bone by pressure necrosis and direct bacterial action with progressive demineralisation and limited osteoblastic activity. Cortical destruction or extension

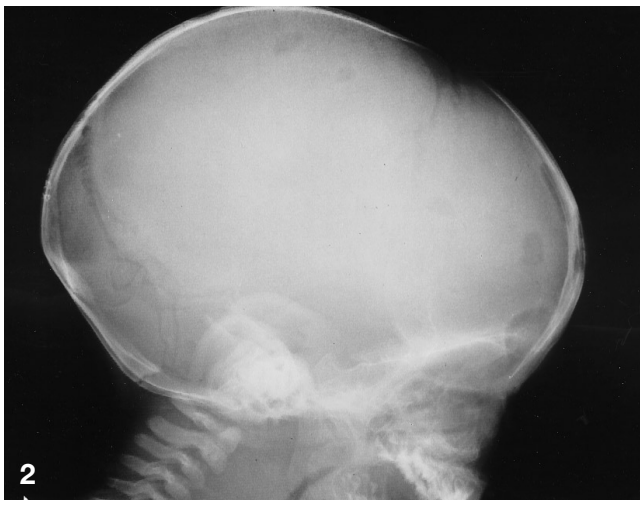
through the epiphysis into the joint space may ensue. Initial repair is by fibrous tissue, but bony defects may persist for years.

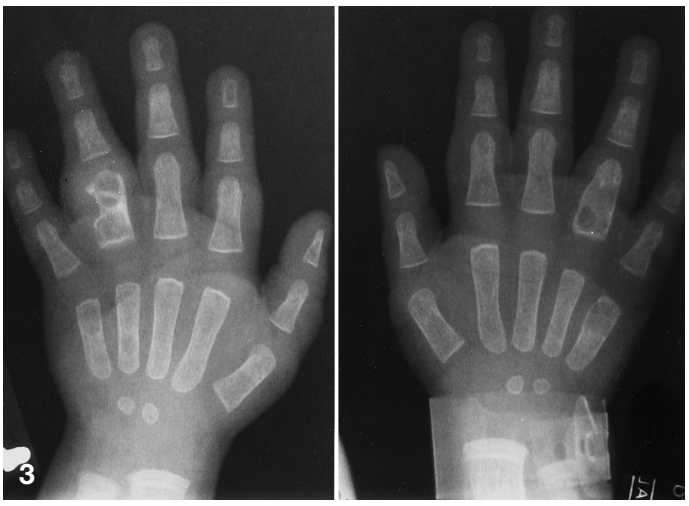
Dactylitis may present within 4–6 weeks after infection, as the principal feature or as a minor manifestation of widespread TB. The phalanges are most commonly involved with an indurated, fusiform, and usually painless swelling on the dorsal surfaces of the hands or feet. Prior to anti-tuberculous chemotherapy, ballooning of the cortex (spina ventosa) and widespread destruction of all the layers of bone was common [4]. The classical radiographic appearance of spina ventosa can easily be distinguished from the dactylitis associated with Hb-S disease but may cause confusion clinically. Sickle cell dactylitis is characteristically bilateral and dissolution is followed by new bone that appears irregularly sclerotic. In TB, radiographic findings do not reveal osteoporosis or new bone formation, and the phalanges are expanded and the trabecular markings are destroyed which results in a ballooned-out appearance. These two conditions frequently have a high prevalence in the same area. It is important that physicians who treat patients with sickle cell disease be aware that the dactylitis and anaemia may be due to TB. The further differential diagnosis of tuberculous dactylitis includes congenital syphilis, sarcoidosis and haemangioma.

Lesions of the skull due to TB may be discovered by chance, or present as a firm, localised, painless swelling, which may initially be mistaken for a bone tumour. The typical radiographic appearance is that of single or multiple well-demarcated osteolytic lesions with little or no peripheral reaction. Supportive evidence for the diagnosis of tuberculosis is needed because the X-ray appearance of the lytic tuberculous lesions must be differentiated from those of histiocytosis, leukaemia, metastatic tumours and infections such as syphilis, osteomyelitis, blastomycosis and coccidioidomycosis.

The multiple skull and skeletal lytic lesions in this patient immediately suggested either Langerhans' cell histiocytosis or neuroblastoma. The radiological features of these conditions may be indistinguishable from those of skeletal TB. Histological confirmation is necessary before starting treatment because malnourished children may have a negative Mantoux test in the presence of TB, as was the case in this child. Malnourished children are usually immunocompromised and, therefore, the im-







Figs. 1, 2 Lytic lesions in the femora, proximal tibiae and skull vault

Fig. 3 Lytic lesions in the metacarpals and phalanges with a spina ventosa appearance of the proximal phalanges of the ring fingers

munological status was not further evaluated. A chest X-ray, examination of gastric aspirates, and lymph node or skeletal lesion biopsy may be necessary to confirm TB.

In this patient, chest radiographs revealed opacification in the mid and upper lobes of the right lung and the left upper lobe. *M. tuberculosis* was cultured from gastric aspirates, and lymph node and bone biopsies confirmed tuberculosis histologically. Cultures for other bacteria and fungi and investigations for syphilis, histiocytosis or malignancies, including bone scan and marrow and skin biopsies examined by light and electron microscopy were negative. The child was treated with anti-tuberculous drugs for 6 months and was well 12 months later at follow-up. The radiographic appearances had improved and only small lytic areas remained. The combination of severe growth retardation and extensive TB would normally be a strong indication to establish the HIV status of the patient. In a developing country, however, children are often referred over long distances without parental accompaniment so that the mandatory consent to perform HIV testing cannot be obtained. In view of this patient's clinical improvement and continued good health 1 year later, it was deemed unnecessary to perform the HIV testing which was not done at diagnosis.

The old dictum that TB can mimic almost any other disease entity remains true and clinicians need to be aware of this.

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