MULTIPLE PSEUDO-CYSTIC TUBERCULOSIS OF BONE

Report of a Case

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Multiple pseudo-cyst formation is an uncommon manifestation of skeletal tuberculosis. Confusion has been caused by the failure to differentiate between this form of osseous tuberculosis, the solitary foci of diaphysial tuberculous osteitis, tuberculous dactylitis, and the bone lesions of sarcoidosis.

This confusion has largely been due to Jüngling (1920, 1928) who, believing them to be of tuberculous etiology, gave the name osteitis tuberculosa multiplex cystica (afterwards changed to cystoides to indicate their pseudo-cystic nature) to the lesions he found in the small bones of the hands and feet in certain cases of sarcoidosis. Since then cases of tuberculous cystic bone disease with lesions quite unlike those associated with sarcoidosis have been given the eponymous title or described as examples of osteitis tuberculosa multiplex cystica or cystoides.

This anomaly has not passed unnoticed, for Ellis (1940), Sweet and Abramson (1941), Golding (1950), Alexander and Mansuy (1950), and Komins (1952), among others, have drawn attention to it. That confusion still exists, however, is shown in a recent paper by Girdwood (1953) in which an atypical case of tuberculous dactylitis was described as an example of Jüngling's disease.

Komins (1952) has attempted to clear up the muddle in the early literature. He accepted thirty-one cases of multiple cystic tuberculosis of bone in childhood from the English literature, reported three more of his own, and suggested that the bone lesions of sarcoidosis should be called osteitis multiplex sarcoïdosa.

Additional cases have since been reported by De Pape (1954) and by Murray (1954). Murray described the characteristic features, of which the most important were: the predilection for childhood; the tendency for the lesions to affect the long bones symmetrically and simultaneously and to progress and regress in harmony; the rarity of joint involvement and the favourable prognosis if it did occur; the lack of early bone reaction with later cortical expansion and fusiform enlargement; and the frequent occurrence of sinuses without sequestrum formation.

The case to be reported agrees with these criteria and demonstrates the effect of streptomycin on this type of bone tuberculosis.

CASE REPORT

A Chinese boy aged three years was admitted to hospital with a four months' history of discharging sinuses in the right orbit, the left foot and both forearms. All the sinuses had been preceded by painless swellings which had either broken down spontaneously or had failed to heal after incision. He was the only child of intelligent healthy parents of good social standing. He had not suffered from any serious illness before, and there was no history of contact with tuberculosis.

Clinical examination—He was a healthy looking child, normal in size for his age. The temperature and pulse were normal. There were painless, fusiform bony swellings of the distal ends of both forearms, with two sinuses on the left wrist and one on the right. There was a swelling of the distal part of the left foot with two sinuses. There was a sinus, involving the upper eyelid, at the outer canthus of the right eye. On examination of the chest there was diminution of air entry and dullness on percussion at the right apex.

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Radiological examination—A radiograph of the chest (Fig. 1) showed collapse and consolidation of the right upper lobe, with enlargement of the hilar lymph nodes; the appearances were typical of a primary tuberculous complex. There was a destructive lesion of oval shape in the posterior part of the right sixth rib. The radiographs of the forearms demonstrated multiple cyst-like lesions. The areas of destruction were oval, with regular, clearly defined margins. There was cortical expansion and in some areas fusiform enlargement of the bone with a little subperiosteal new bone formation (Figs. 2 and 3).

Radiological examination of the remainder of the skeleton revealed similar lesions in many of the bones (Figs. 4 to 12).

A diagrammatic representation of the radiological findings (Fig. 13) indicates the tendency for the lesions to be symmetrically and centrifugally situated with reference to the axial skeleton and for the distal parts of the limbs to be affected.

Investigations—The Mantoux test was strongly positive with 0.1 millilitre of 1/1,000 old tuberculin. The Kahn test was negative. The serum calcium was 9.5 milligrams per cent, and serum protein 7.3 grammes per cent. The albumen-globulin ratio was normal. The erythrocyte sedimentation rate was 20 millimetres in the first hour (Westergren).

Sputum was not available, but repeated examinations of gastric washings and faeces were negative for acid-fast bacilli. Curettage of the sinuses and culture of the scrapings, on two occasions, gave a poor growth of staphylococcus aureus. No fungus was grown, and direct smear and culture were negative for acid-fast bacilli.

Biopsy from right ulna—The naked eye appearance was that of a soft granuloma which shelled out of the bone with ease, leaving a smooth cavity. There was no pus or caseous material. Culture of the specimen yielded no acid-fast bacilli or other organisms.

Histologically the specimen consisted of inflammatory granulation tissue replacing bone and bone marrow. Numerous multinucleated giant cells were present and the inflammatory tissue was studded with a number of characteristic "tubercles" each consisting of giant cells,
Fig. 2
Right radius and ulna.

Fig. 3
On admission.

Fig. 4
After three months.

Fig. 5
Further radiographs at time of admission.

Fig. 6
Right humerus.

Fig. 7
Left humerus.

Fig. 8
Left radius and ulna.
FIG. 8
Lower limb bones at time of admission. Figure 8 - Part of left femur. Figure 9 - Left tibia and fibula. Figure 10 - Right tibia and fibula.

FIG. 11

FIG. 12
Right foot (Fig. 11) and left foot (Fig. 12) at time of admission.
epithelioid cells and fibroblasts. Occasionally areas of caseation were present in the inflammatory tissue (Figs. 14 and 15). Although acid-fast bacilli were not demonstrated in the section the histological evidence of tuberculosis was very strong (Dr H. A. Sissons).

**Treatment and subsequent progress**—Streptomycin (grammes \( \frac{1}{4} \) daily) and para-amino-salicylic acid (grammes 6 daily) were given for 100 days. The forearms and left lower leg were immobilised in light plaster splints. The child never looked ill and at no time had a cough. His appetite was good and he gained weight satisfactorily. The temperature and pulse remained normal. The erythrocyte sedimentation rate fell to 10 millimetres/hour two weeks after treatment had begun and thereafter remained below 5 millimetres/hour.

Three weeks after treatment by streptomycin was begun the sinuses began to heal; that in the orbit was healed at one month and the remainder were healed in two months.

Radiological examination of the skeleton three months after admission and shortly before the child’s discharge from hospital gave evidence that a considerable degree of healing had already taken place.

A further radiological examination seven months later showed that all the affected bones had regained their normal shape and that bone texture in most sites had resumed its normal appearance.

Radiographs of the right forearm taken at these times illustrate the process of healing (Figs. 3 and 4).

The child’s general condition remains good, but a recent radiograph of the lungs still shows partial atelectasis of the right upper lobe.

**DISCUSSION**

Despite the failure to isolate mycobacterium tuberculosis from the sputum, stomach, faeces or one of the bony lesions, the radiographic appearance of the lungs, the positive Mantoux reaction, the histological findings and the rapid disappearance of the bony lesions under streptomycin therapy are almost conclusive evidence of a tuberculous etiology in this case.

Most of the recorded cases of multiple pseudo-cystic tuberculosis of bone affecting children (Table I) have occurred in under-nourished coloured infants in tropical or semi-tropical countries, but in our experience the usual case of skeletal tuberculosis in these regions in no way differs from that found in more temperate and socially advanced parts of the world, and the child in this case showed no evidence of malnutrition.

Although analysis of the recorded cases shows that a number of children died of generalised tuberculosis the relatively benign character of multiple pseudo-cystic tuberculosis of bone, the tendency to spontaneous regression and the rarity of joint involvement with a favourable prognosis if it does occur, have been noticed by several observers. It is indeed a striking feature that such widely disseminated disease may cause so little constitutional disturbance and heal leaving so little evidence.

In pseudo-cystic tuberculosis of the bone the widespread distribution of the lesions, their simultaneous onset and the similarity of their radiographic appearances are evidence of haematogenous dissemination over a short time.

It is tempting to suppose that the organism in these cases might have a lowered virulence
Fig. 14
Photomicrograph showing granulomatous tissue with scattered multinucleated giant cells. (× 108.)

Fig. 15
Bone marrow, showing a tubercle consisting of two multinucleated giant cells surrounded by epithelioid cells and fibroblasts. (× 200.)
or be of a different type, but in cases in which there has been bacteriological proof the organism has not shown cultural or morphological differences and animal inoculation has not demonstrated any altered virulence.

Any explanation of this uncommon form of skeletal tuberculosis must inevitably be a pure hypothesis, but we believe that a possible solution may be found in a study of the time relationship of the initial bacillemia to the primary lung lesion. It is difficult to prove in man that bacillemia is a regular feature of a primary infection, but it is a constant finding in animal experiments (Wilson 1933), and in all reported necropsies in early cases of primary complex tubercles have been found widely distributed throughout the body.

According to Wallgren (1948) there is an interval of five or six weeks between the primary infection and the development of hypersensitivity to tuberculin. The usual lesion of bone and joint tuberculosis occurs after the establishment of an allergic state.

If on rare occasions the bacillemia should occur before the development of tuberculin sensitivity and bacilli were able to gain a foothold in the bones with sparing of other more vital organs, it is reasonable to suppose that the response might be quite different from that found after allergy had developed, and that the subsequent pattern and progress of the disease might be substantially altered.

SUMMARY

1. A case of multiple pseudo-cystic tuberculosis of bone presenting as a generalised disease of the skeleton in a child is reported.
2. The response to streptomycin and para-aminosalicylic acid is shown.
3. A possible explanation of the etiology of this rare disease is suggested.

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