MULTIPLE PSEUDO-CYSTIC TUBERCULOSIS OF BONE

Report of a Case

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The term multiple psuedo-cystic tuberculosis of bone was suggested in 1952 by Komins to avoid existing confusion in the nomenclature of certain sarcoid lesions of bone and some forms of osseous tuberculosis. It applies to characteristic multiple cyst-like bone lesions seen in tuberculosis of children; it differentiates these lesions from those due to sarcoidosis which Jungling (1920) had grouped under the term osteitis multiplex tuberculosa cystica; it also differentiates them from the multiple osseous tuberculous lesions seen in adults and to which the term disseminated bone tuberculosis has recently been applied by Alexander and Mansuy (1950). Komins suggested that the term osteitis multiplex tuberculosa cystica (or cystoides) be discarded but that a modification—osteitis multiplex cystoides sarcoidosa—be retained for the osseous lesions of sarcoidosis.

Multiple pseudo-cystic tuberculosis of bone is uncommon. Komins collected only thirty-one cases in a review of the English literature up to 1952; he emphasised, however, that some cases might have been lost in the confusion of terminology since Jüngling, who considered sarcoidosis as a form of tuberculosis, first used the term osteitis multiplex tuberculosa cystica in 1920 to describe a number of cases, many of which were sarcoid in nature.

Nineteen of Komin's thirty-one patients were two years of age or under, six were one year or under, and all were under fifteen. Eighteen out of twenty-one cases were in non-whites. The patient to be described in this report was of Indian extraction and was ten months old when he died. This case is of interest for three reasons: 1) the extensive involvement of the bones, particularly the skull; 2) the presence of a bone lesion at birth; and 3) the possible, though perhaps not likely, co-existence of syphilis.

CASE REPORT

The boy was born in Northern Manitoba of Indian parents. The father had been under treatment for syphilis for two years. The mother was not investigated until the patient was several months old; her blood Wassermann reaction was strongly positive seven months after the patient's birth. Her lungs showed no clinical or radiographic abnormality; a Mantoux test was positive. Three siblings were alive and well; one sibling died of undetermined causes at the age of four months. There was alleged to be no contact with tuberculosis, but such information is quite unreliable among many Indian populations.

The patient was normal at birth except for a swelling of the fifth right metacarpal. He was breast-fed almost exclusively and received no added vitamins. Progress was slow; he soon became weak, pale and irritable. At two and a half months he developed a chronic purulent nasal discharge, anorexia and occasional vomiting. At three and a half months bilateral cervical swellings appeared and he developed a cough; about three weeks later swellings appeared almost simultaneously on the forehead, at the proximal phalanx of the left index and ring fingers, below the right elbow anteriorly, and on the lateral aspect of the right upper arm. At five months the nasal discharge became bloody, and it was noted that the congenital swelling of the right fifth metacarpal, which previously had remained unchanged, also began to enlarge.

FIG. 1

Lung fields at age seven months, showing paratracheal lymphadenopathy and diffuse parenchymatous lesions.

FIG. 2

Skull at age eight months.

Clinical examination—On admission to St Boniface Hospital at the age of six and a half months he was a grossly malnourished, pale, irritable baby weighing only 12 pounds 13 ounces. He was too weak to sit up or even to support his head. There was intermittent sanguino-purulent nasal discharge but no local ulceration was evident. The liver and spleen were both palpable 4 and 2 centimetres below the costal margin respectively, smooth and somewhat firm. There was marked bilateral cervical lymphadenopathy. Several bony swellings were noted, particularly in the upper extremities and skull (Figs. 1 to 4). There was a prominent swelling almost in the centre of the forehead, 1.75 centimetres in diameter by 1 centimetre in height; it was soft and fluctuant, and was attached to the skull but not to the skin. Firm fusiform swellings were present in the fifth metacarpal of the right hand, and in the proximal phalanx of the index and fourth finger of the left hand. Other bony swellings were noted at the upper

FIG. 3

FIG. 4

Figure 3—Upper limbs at age eight months showing multiple pseudo-cystic lesions. Swelling of the fifth metacarpal was present at birth. Figure 4—Lower limbs at age seven months, showing early pseudo-cystic lesions in the tibia and fibula.

end of the ulna on each side and at the lower end of the right ulna. A mobile, slightly fluctuant mass, sized about 1.5 by 2.5 centimetres, was felt at the medial aspect of the right upper arm. *Investigations and additional findings*—The Wassermann reaction was negative on two occasions but a single Kahn test was weakly positive (1:8 dilution). It was felt that congenital syphilis could not be excluded, particularly in view of the family history and some of the clinical findings. A three weeks' course of penicillin (total dose 2.3 million units) was therefore given. The child showed improvement in appetite, was much less irritable and the nasal discharge disappeared, but there was no appreciable gain in weight.

In the meantime a diagnosis of tuberculosis was made on the basis of a strongly positive Mantoux reaction (1:1,000), radiographs showing extensive bilateral pulmonary changes suggestive of tuberculosis, the presence of acid-fast bacilli morphologically identical with mycobacterium tuberculosis on smear and culture in pus obtained from the swelling on the forehead, and a biopsy of a cervical gland showing necrosis, Langhans' cells and a histology

compatible with a diagnosis of tuberculosis. Dihydro-streptomycin was started (total dose 14 grammes) and he was transferred to a sanatorium. Additional laboratory findings (at age seven months) were as follows: haemoglobin 53 per cent, red blood corpuscles 3.6 m., white blood corpuscles 28,950 (polymorphonuclears 59 per cent, lymphocytes 39 per cent, monocytes 2 per cent); platelets 750,000. Cerebro-spinal fluid normal except for 11 lymphocytes per cubic millimetre. Liver function tests: cephalin cholesterol flocculation + + + +; thymol turbidity + + + +; Takata-Ara + + + +. No tubercle bacilli were grown from three stools.

Despite treatment, deterioration was progressive. He developed a chronic purulent discharge from the right ear which showed acid-fast bacilli on smear and culture. More swellings appeared in the skull and elsewhere; draining sinuses developed at some of the swellings. His temperature, which had ranged between 99 and 100 degrees Fahrenheit, rose to 102–103 degrees in the final month and he died at the age of ten months.

Post-mortem findings—The body was emaciated and weighed only 9 pounds. Draining sinuses were present, as already noted. Caseating lesions were found in the skull, in both ulnae, in the right radius, in the second, third and fifth right metacarpals, and the first, second and third left metacarpals, in the proximal phalanges of the left index and ring fingers, and in the right tibia and fibula. The visceral organs involved were the lungs, liver, spleen and kidneys. There was general lymphatic involvement, affecting the cervical, axillary, inguinal and hilar glands on both sides, and the mesenteric and retroperitoneal glands. A few small tuberculomata were found in the cerebral hemispheres, the cerebellum and the pons; one was immediately sub-meningeal but the lepto-meninges appeared intact grossly.

The skull lesions affected the frontal and parietal bones. They consisted of caseous areas in the cancellous bone, from half a centimetre to two centimetres in diameter, with erosion and expansion of the outer table and in some areas also the inner table. In one area the abscess had broken through the inner table to become confined by the dura, whereas in others it had broken through the outer table to involve the subcutaneous tissues and skin; in these areas the skin was fixed and no longer freely mobile over the swelling. Similar lesions were seen in the short and long tubular bones: they consisted of oval or round caseous lesions in the medullary cavity expanding the cortex, involving the periosteum and even breaking through this and the skin to form a draining sinus.

Microscopic examination—The typical histology of tuberculosis was seen in nearly all sections of the organs mentioned, with numerous Langhans' giant cells, necrosis, and acid-fast bacilli. No Langhans' cells were found in the brain, nor in the fifth right metacarpal and proximal phalanx of the fourth left finger, but acid-fast bacilli were abundant. Many lesions were of the miliary type. No histological evidence of syphilis was found (these findings were corroborated by the Armed Forces Institute of Pathology, Washington, D.C.). There was an occasional lesion in the liver which at first glance suggested a gumma but proved tuberculous. The middle ears and mastoids indicated moderate tuberculous involvement but the nasal cartilages, choroid of the eye, vertebral bodies and all other organs not already mentioned showed no lesion.

DISCUSSION

The extensive involvement of the short and long tubular bones and skull, and the pseudo-cystic nature of these lesions, are associated with the low resistance, in this case, of the American Indian to tuberculosis. Other coloured races show similar tendencies (Widmann, Ostrum and Miller 1939, Komins 1952). Straus (1933) emphasised that the lesions in the skull are seldom seen under one year of age; this is because such lesions start in cancellous bone and there is little cancellous bone in the skull in the first year of life.

The history of a lesion present at birth offers interesting speculation—though nothing more than speculation because the patient was not seen until several months later, when many other lesions had already appeared. Radiographs of this congenital lesion of the fifth right metacarpal at seven months were suggestive of tuberculosis, and histological examination

at necropsy three months later showed focal diaphysial necrosis, partial erosion and expansion of the cortex, and thickening of the periosteum; no Langhans' giant cells, but abundant acid-fast bacilli, were present. It is impossible to say whether the initial lesion was tuberculous or whether tuberculosis had been superimposed on another condition such as syphilis.

Did this infant have syphilis? Unfortunately necropsy does not help us much here. No spirochetes were seen in the tissues and no lesions suggestive of syphilis were found at necropsy, but it is possible that the course of penicillin administered three months before death may have eliminated all evidence of this disease. On the other hand, despite the positive family history, the positive Kahn reaction and the temporary improvement after anti-syphilitic treatment, it is still doubtful whether the child was syphilitic when first seen. Not all babies born of syphilitic mothers have the disease, and according to Kampmeier (1943) approximately 20 per cent escape. The Kahn reaction was only weakly positive and the Wassermann reaction was negative on two occasions; the temporary improvement after penicillin treatment may have been due to its effect on secondary invaders. It is not quite so easy to explain the disappearance of the haemorrhagic snuffles. Haemorrhagic snuffles has been described in tuberculosis in the absence of syphilis, but then penicillin would not be expected to be beneficial (Hughesdon 1946). Nevertheless the lesions seen on admission and at necropsy were all typically tuberculous, with marked evidence of caseation and suppuration, with many draining sinuses. According to Kampmeier suppuration with sinus formation is never seen in uncomplicated syphilis. The congenital lesion may have been due to syphilis; but it was an isolated lesion whereas it has frequently been stressed that the bony lesions of congenital syphilis are symmetrical.

SUMMARY

1. A case of multiple pseudo-cystic tuberculosis in an American Indian infant is described.
2. The extensive lesions in the skull of this infant are considered to be of particular interest.
3. The possible significance of a bony lesion present at birth is discussed.

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