MULTIPLE CYSTIC TUBERCULOSIS OF BONE (JÜNGLING'S DISEASE)

Report of a Case

WILLIAM GIRDWOOD, JOHANNESBURG, SOUTH AFRICA

In 1920 Jüngling reported nine cases of "osteitis tuberculosa multiplex cystoides," and observed that it occurred in adults, that it was practically confined to lesions of the hands and feet, and that there were no sequestra or sinuses. He considered that the cases were tuberculoid in origin and were caused by an avirulent tubercle bacillus. In some respects

FIG. 1
The right forefoot.

FIG. 2
The left hand.

they showed features closely related to Boeck's sarcoidosis. Other reports in the literature have appeared under the titles multiple cystic tuberculosis of bone (Thornton 1938), Besnier-Boeck-Schaumann disease and Perthes-Jüngling disease. Birdsong and L'Engle (1948) found reports of only fourteen cases in the literature. Since that date, several further cases have been described by Frost (1942), Fox (1944), Jacobsen (1936), Kreidberg and Downing (1948) and Law (1939). In some cases children have been affected. Miliary lesions affecting the flat bones have been reported by Meng and Wu (1942) and Kienböck (1931) has described multiple cystic tuberculosis affecting only the long bones. Turkish *et al.* (1949) pointed out that Jüngling, reporting this disease in 1920 and 1928, noted the following characteristics: 1) Gradual onset during puberty or thereafter. 2) Pain occurred early and was not severe. 3) The small bones of the hands and feet were affected. 4) Cystic degeneration was seen on radiographic examination. 5) No involvement of periosteum or joints was present. 6) Often there was involvement of skin in the affected area (sarcoid). 7) No tubercle bacilli were

discoverable. 8) Tuberculous skin tests were usually negative. 9) Histology showed epithelial and lymphocytic cells and occasional giant cells, but no caseation or tuberculous bacilli. 10) The disease took a slow course, tending to spontaneous improvement and even recovery. The reported cases of the disease in children had differed in that tuberculin skin tests were usually positive, guinea-pig inoculations or cultures were not infrequently positive, caseation was occasionally noted, associated skin lesions were only rarely seen, the long hollow bones were sometimes involved, and, although local lesions had a tendency to heal, the patient frequently died of generalised tuberculosis.

CASE REPORT

An adult Bantu patient complained of a swelling of the right metatarso-phalangeal joint of three months' duration. There was only slight pain. On examination the swelling of the foot was bony-hard. There was no attachment to skin and no tenderness, scars, sinuses or

local heat. In the left hand, in the region of the first metacarpal bone, there was a diffuse swelling without joint involvement. This was also bony-hard, not tender or hot, and without evidence of sinuses. In the neck there were multiple small shotty glands in the posterior triangle, with larger glands in the left submental and right submandibular region. There was a broad scar in the upper part of the left side of the neck, extending over the parotid region; the patient stated that this had discharged spontaneously in childhood.

Radiographs of the left forefoot (Fig. 1) showed destruction of the metatarso-phalangeal joint by a cystic trabeculated lesion. There was expansion of the bone and good definition of the cystic spaces. Erosion with bursting of the shell of the cortex was possibly present only in one area. The lesion was diffuse, extending through the whole of the proximal phalanx and proximally to the middle

FIG. 3

Section from metatarsal lesion showing tuberculous granulomatous tissue with giant cell.

of the metatarsal. Radiographs of the left hand showed similar but less extensive cystic changes in the first metacarpal (Fig. 2).

Differential diagnosis—Radiographically the lesion did not suggest a tumour. Syphilis, common in the Bantu, was unlikely, as sclerosis of an irregular nature with gross periosteal new bone is the usual feature. Pyogenic osteitis required consideration, but the trabeculated cystic appearance without gross sclerosis or bone abscesses was against this. Gout was considered, but there was no evidence of chalky calcified areas, and the clinical lesion was painless and did not resemble gout. The numerous cystic areas favoured a diagnosis such as medura-mycosis, but this was excluded by the absence of sinuses and induration. The bones were too well calcified for osteitis fibrosa cystica.

The remaining possibilities were tuberculous dactylitis and Boeck's sarcoidosis. Brailsford (1944) described a trabeculated cystic form of tuberculous dactylitis. The metacarpal lesion,

with changes confined to the bone without involvement of the joints, together with the shotty glands in the neck and an old neck scar over the left parotid, and an increased sedimentation rate, supported a tuberculous origin. With regard to Boeck's sarcoidosis, the lungs were perfectly normal, there were no skin nodules and no evidence of uveoparotitis; but the serum proteins showed a total of 8.4 grammes per cent (albumen 4, globulin 4.4). This state of the proteins is a common finding among Bantu population and cannot be regarded amongst them as specific for sarcoidosis.

Treatment—The lesion of the foot was excised completely and a bone graft from the right ilium was inserted. Fixation was obtained by the use of a probe extending from the tip of the right first toe through the distal phalanx and the graft into the small remaining part of the first metatarsal, supplemented by a plaster splint. After three months the probe and the plaster were removed, and the patient was encouraged to walk.

Description of specimen—The cut surface of the specimen had a light brownish fleshy appearance and contained numerous bony spicules and fibrous bands running through its substance. *Microscopically*, sections showed the specimen to consist, for the most part, of tuberculous granulation tissue, in which there were numerous giant cells of the Langhans type, with well marked follicle formation (Fig. 3). Foci of caseation, scattered irregularly throughout the granuloma, could also be seen. In addition, a few bony trabeculae and fibrous connective tissue were present. Very scanty acid- and alcohol-fast bacilli were observed. The histological features were those of a tuberculous dactylitis.

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