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Cystic Tuberculosis of Bones

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Multiple osteolytic lesions involving the axial and extra axial skeleton are a rare form of tuberculosis (TB) in children(1). It is seen in patients with rapidly acquired immunity and in those with good resistance(2). There are scanty reports of such lesions in pediatric patients and therefore we wish to present this unusual form of tuberculous osteomyelitis which showed a favorable response to treatment.

Case Report

A 5-year-old boy presented with fever, slowly progressive and painless soft swellings over the forehead, left cheek, right mid-axillary region, both legs and right thumb of 6 months duration. Systemic examination revealed a soft hepatomegaly of 2 cm. Investigations included a positive Mantoux test (20 × 20 mm) and an ESR of 55 mm at 1 hour. Hemogram, liver function tests and abdominal sonography were normal. Skeletal survey revealed fifteen lesions. Well defined rounded osteolytic lesions with sclerotic margin were seen in the right frontal and parietal bones, body of left mandible, right 8th and 9th ribs and without sclerosis on both tibiae and fibulae (*Fig. 1*). Lateral margins of the D_2 and D_3 vertebral bodies and their left pedicles were eroded. Osteolytic lesions were also seen in the left ischial bones, distal half of the right humeral diaphysis and the left radial shaft. Characteristic fusiform expansion described as spina ventosa was noted in the left first metacarpal.

A ^{99m}Tc MDP bone scan showed uniformly increased tracer concentrations in

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Received for publication: June 16, 1992; Accepted: December 4, 1992

Fig. 1. Xrays of the legs before treatment. Arrows indicate the multiple cystic lesions.

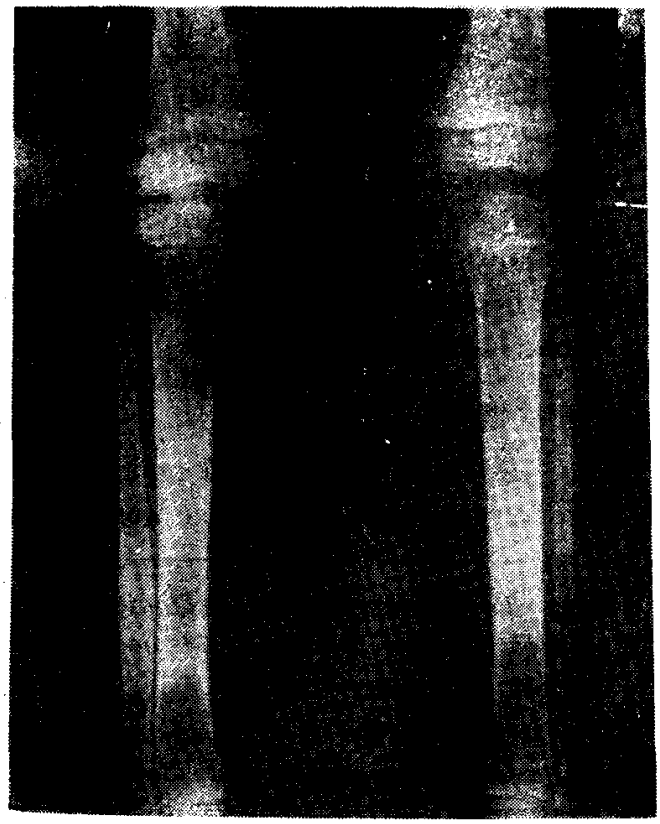

Fig. 2. Xrays of the legs after treatment. There is significant improvement of the lesions.

both legs and left foot, rest of the skeleton being normal. Bone marrow aspiration was inconclusive but a bone biopsy demonstrated Langhan's type of giant cells. Acid fast bacilli were grown from the aspirate obtained from the frontal swelling.

The child was treated with streptomycin, INH, rifampicin and pyrazinamide for 2 months, and INH, rifampicin and ethambutol were continued for further 16 months. The swellings subsided within 4 months with radiological improvement (*Fig. 2*); and there were 3.5 kg weight gain and 6 cm height gain after 8 months of therapy.

Discussion

Cystic form of TB was first described as osteitis tuberculosa multiplex cystoides by Jungling in 1920(2). The lesions are symmetric, variable in size with little reactive proliferation and may be accompanied by sclerosis(3). Such multiple cystic lesions can be confused with histiocytosis(4).

The lesions consist of widening and diffuse rarefaction of the diaphysis. The cortex is eroded from within and gets thickened externally. This periosteal reaction results in a fusiform appearance of tubular bones. Children of 5-10 years of age present with dactylitis. Lesions of small tubular bones are called as 'spina ventosa', *i.e.*, spine like projections, puffed as full of air(2).

Cystic lesion involving only the skull, frontal and parietal bones are more common, and were described by Reid in 1842(5). They can be circumscribed or spreading, and account for 0.2-1.3% of all TB osteitis. Half the cases occur by 10 years of age and 75-90% manifest by 20 years. Recent Indian reports include 2 cases by Bhandari *et al.*(6) and one by Tata *et al.*(7). The youngest case with skull le-

sions was described in a 4-month-old(8). Mohanty has studied a large series, the youngest being a boy of 3 years(9).

Multiple osteolytic lesions mimic a variety of childhood malignant conditions like neuroblastoma, histiocytosis, lymphoreticular malignancies, syphilitic, pyogenic or mycotic osteomyelitis. Though the tubercular lesions are disseminated, the response to therapy is excellent and prognosis is good as was seen in our case.

Acknowledgements

The authors wish to thank the Dean, Seth G.S. Medical College, K.E.M. Hospital for allowing us to publish this case report and the Department of Pathology, Tata Hospital for their kind cooperation.

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Cohen Syndrome

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In 1973, Cohen *et al.*(1) described three cases of a syndrome, the major features of which were obesity, hypotonia and mental retardation with facial, oral, ocular and limb anomalies. Eighty cases have since been reported(2) and inheritance is autosomal recessive(3).

This report profiles two cases that have most of the major abnormalities and is the first report from India after the syndrome has been recognised.

Case Reports

Case 1: A ten-year-old male child of non-consanguinous parentage was referred

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Received for publication: October 20, 1992; Accepted : December 8, 1992