

Massive Osteolysis—Gorham's Syndrome

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Massive osteolysis of bones is rarely encountered in children. Gorham's syndrome is characterized by a spontaneous onset of rapid destruction and resorption of bones resulting in severe deformities with subluxation and instability of joints(1). There are only around 60 cases of this unusual disorder reported in literature(2) and none from India.

Case Report

A 10-year-old male child, born of a non consanguineous marriage, presented with dull aching pain and depression over the right infraclavicular region of six months duration. There was no history of trauma or family history of similar illness. On clinical examination there were no dysmorphic features. The right anterior chest wall was flattened with drooping of the right shoulder. The clavicle on the same side was not

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clearly visible and the sternum and the rib cage on both sides anteriorly were very soft to palpate. The rest of the skeletal system was normal. Systemic examination was otherwise unremarkable.

The hematological and routine biochemical parameters, including serum calcium, phosphorus and alkaline phosphatase were normal. Mantoux test was 10 mm positive. Radiographs of the chest showed marked osteoporosis and a fracture of the right clavicle with marked thinning and absence of the anterior halves of the upper six ribs on the right side. Osteoporosis and thinning of the ribs were also evident on the left side (*Figs. 1 & 2*). Bone scan revealed an increased uptake of tracer activity at the lateral end of clavicle, and third and fourth ribs on the right side. Bone biopsy from the right clavicle showed that the marrow spaces were widened and replaced by thin walled capillaries with no evidence of regeneration, and was suggestive of angiomatosis.

While in hospital, the patient developed a right sided hemorrhagic pleural effusion. The tubercular antigen in the pleural fluid was positive. The child was given a protective anterior corset for the chest and treated with antitubercular drugs.

Discussion

Idiopathic massive osteolysis was first described by Jackson in 1838. Gorham *et al.* in 1955, found its association with a vascular abnormality(3). This non-familial disorder is characterized by spontaneous absorption of bones in a unicentric fashion, the process stabilizing spontaneously over years or progressing relentlessly until practically all osseous tissue has disappeared.

Different forms of idiopathic osteolysis are differentiated on the basis of clinical,

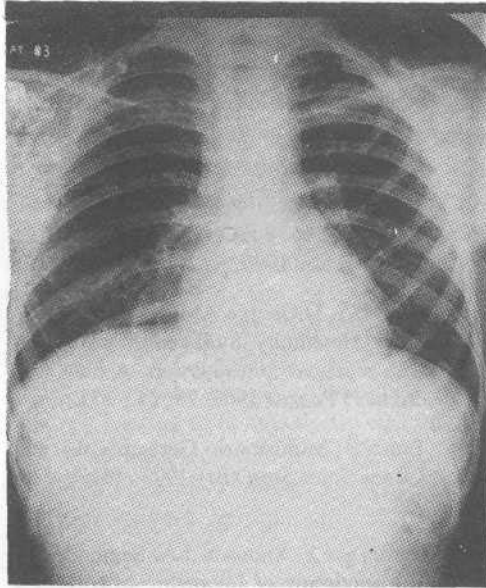


Fig. 1. Disappearing anterior ends of the ribs.

radiological and genetic criteria. Types I and II are the dominant and recessive varieties, respectively, and are usually multicentric. Types III and IV are unicentric non-hereditary forms, the former associated with nephropathy and the latter with hemangiomas. In the Winchester syndrome (Type V) osteolysis is associated with contractures, short stature, nephropathy and corneal clouding(4,5) and is inherited in an autosomal recessive manner.

Our patient presented with increasing deformity of the right chest wall over a period of a few months. Serial X-rays showed gradual thinning and subsequent disappearance of the anterior ends of the upper 6 ribs. The clavicles and sternum also appeared markedly thinned out. There was no history of trauma, or evidence of infection or inflammation thus ruling out secondary causes of osteolysis. The biochemical and metabolic profile of the patient were normal



Fig. 2. Lateral X-rays showing thinning of the ribs with disappearance of the sternum.

thus establishing the idiopathic nature of the disease. The exact pathogenesis of this disorder is still obscure but Gorham *et al.* concluded that angiomas seen histologically, as in our patient, led to active hyperemia and osteoclastic activity(2). The abnormal vessels may also infiltrate the skin, subcutaneous tissues, underlying deep tissues and mediastinum resulting in secondary pulmonary complications such as hemorrhagic pleural effusion as in our patient.

Diagnosis of this condition is based on the clinical picture and radiological findings of loss of bone mass in the initial stages and concentric shrinking of the bone ends in the later stages(5,6). However, histological evidence of abnormal vasculature with absence

of regeneration conclusively establishes the diagnosis.

Various forms of treatment such as ionizing radiation, amputation or local resection, embolization, use of diphosphonates, calcitonin and mithramycin have been tried, but none found to be definitely successful(7).

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Profile of Childhood Disability

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At least 1 in every 10 children is born with or acquires a physical, mental or sensory impairment(1). The lay public is generally uninformed as to the cause, prevention and treatment of childhood disability. They are often filled with misinformation, superstition and fear about the condition(2). With the present technology, at least 50% of all disabilities may be prevented or post-

poned(3). The present study was carried out to identify the types and various causes of childhood disability.

Material and Methods

Four hundred and seventy four children of both sexes in the age group 0-14 years, attending the Department of Physical

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