MJBU, Vol 13, No. 1&2, 1995

MASSIVE OSTEOLYSIS OF GORHAM

Mea'ad K. Hassan M.B.CH.B, C.A.B.P, Hana Al-Sadoon M.B.CH.B, DMRD

Mea'ad K. Hassan M.B.CH.B, C.A.B.P, Lecturer in Dept. of Paediatrics

Hana Al-Sadoon M.B.CH.B, DMRD, Department of radiology, Basrah Maternity and child hospital.

INTRODUCTION

Vanishing bone disease or massive osteolysis of Gorham is relatively a rare disease, recognized usually in childhood, but more than one third of patients are over the age of 35[1]. A family history is not apparent. The exact nature of the disease is unknown, the possibility that this condition may represent a neoplastic proliferation of haemangiomatous (or lymphangiomatous) tissue has been suggested in the past. Apparently any bone can be affected and the degree of osseous destruction generally increases relentlessly over a period of years and although the degree of osseous deformity may become severe, serious complications are not frequent[2].

CASE REPORT

A nine-year old boy was referred to Basrah maternity and child Hospital in December 1994, his complaint was inability to walk.

His present illness started when he was 6 years old, at that time he had pain in the right hip joint following trauma one month before. The pain and limitation of movement was progressing one which involved other joints of the body. After one year following the trauma he was completely bedridden. There was no significant systemic manifestations apart from weight loss

and anorexia. Past history was not contributory. Parents were not relatives, he has 7 siblings, all were healthy.

Physical examination revealed a cahexic child. His weight and sitting height were < 3rd centile. There was a chest deformity with prominent lower sternum, pronounced middle thoracic kyphosis, widened, slightly tender wrists and fixed flexion deformity of both knee joints.

Complete blood picture, ESR, uric acid and renal function tests were normal. Serum alkaline phosphetase was slightly elevated. Roentenograms showed almost total destruction of the whole pelvis and the hip joint with faint outline of the residual femoral head, pathological fracture of the femoral neck, evidence of generalized osteopenia and extensive absorption of many bones (resorption of the clavicle, scapula, right upper ribs, humerus head) and pathological fractures of many ribs, the vertebrae can barely be seen.

DISCUSSION

This peculiar clinical entity generally present with gradual disappearance of bone resulting in fracture and deformity[3]. The most dramatic aspect of massive osteolysis is its radiographic appearance, laboratory analysis may show unremarkable results, with slight elevation of serum alkaline phosphatase. Arteriography and lymphangiography have shown no connection of these lesions to either vascular or lymphatic circulations[2]. Scintigraphy may demonstrate an area of decreased uptake corresponding to the site of diminished or absent bone tissue. In the early stages of the disorder, the pathologic feature resemble those of skeletal hemangioma, in later stages, vascular fibrous tissue replaces the angiomatous tissue[2]. There are difficulties not only in the diagnosis but also in its therapy. No treatment has been really successful[4].

In some cases major operative procedures to stop further progress were performed, bone transplantation was tried but the result was unsuccessful, in few patients radiotherapy was used to stop the disease, but was difficult to assess the results of radiotherapy since the disease can stop progressing spontaneously[4].

REFERENCES

1. David Sutton. Textbook of radiology and imaging. 4th edition.

2. Donald Resnick, Gen Niwayama. Diagnosis of bone and joint disorders. Vol. 3, W.B. Saunders Co. 1981. 3023-3038.

3. G. vasudeva lyer, Ammu Nayar. Massive osteolysis of the skull, case report. J. Neurosurgery. 1975; 43: 92-94.

4. G. Hedyden, O.D., L.G.Kindblon, and J. Moller Nielsen. Disappearing bone disease, a clinical and histological study. The J. of Bone and Joint surgery. 1977; 59-A, 57-61.

Fig 1. Shows generalized osteopenia, total destruction of the whole pelvis and both hip joints with faint outline of residual femoral head and pathological fracture of femoral neck.

Fig 2. Shows generlized osteopenia with extensive resorption of right upper ribs and pathological fracture of many ribs.

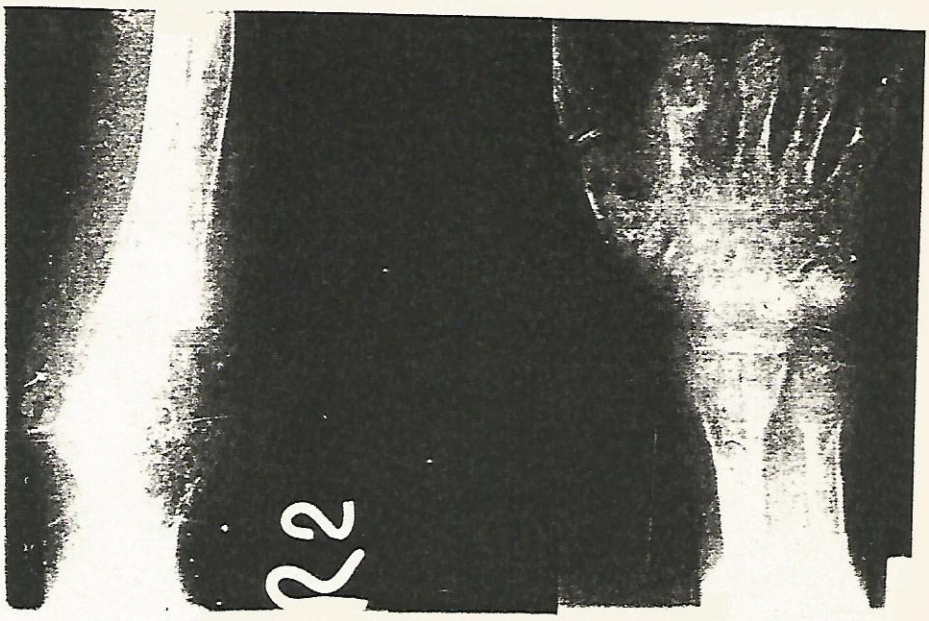

Fig 3. Shows severe decrease in bone density, wide lower metaphysis of radius and ulna with resorption of upper and lower ends of radius and ulna, lower humerus, carpal bones and the ends of metacarpal bones.

Fig 4. Shows severe decrease in bone density, the vertebrae can barely be seen, compressed biconcave vertebral disc space.

Fig 3. Shows severe decrease in bone density, wide lower metaphysis of radius and ulna with resorption of upper and lower ends of radius and ulna, lower humerus, carpal bones and the ends of metacarpal bones.

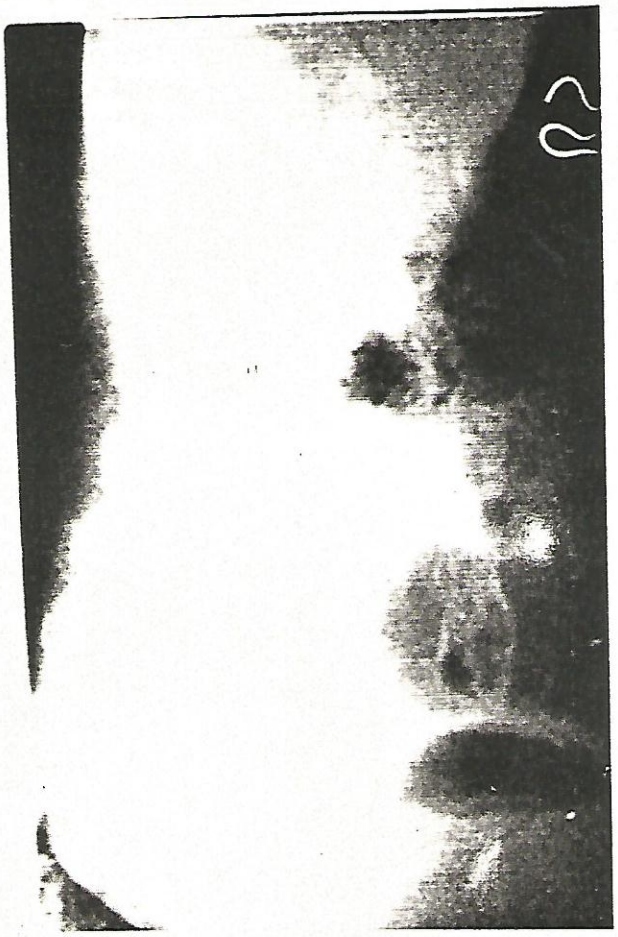

Fig 4. Shows severe decrease in bone density, the vertebrae can barely be seen, compressed biconcave vertebral disc space.

انحـــلال العظـم الحســيم لكـــورهـــام

هـذه المقالة تصف حـالة الطفل بـالغ من العـمر ٩ سنوات يعانى من انحلال العظم الجسيم وقد كان يعانى من هزال عضلى شديد مع عدم القابلية على التحرك وكان يشكو من هذه الاعراض خلال الثلاث سنوات التى سبقت دخوله المستشفى.