IJO Indian Journal of Orthopaedics

CASE REPORT

Year : 2006 | **Volume** : 40 | **Issue** : 1 | **Page** : 55-56

Gorham's Disease - *A case report*

Deepali Jain, Vijay Kumar, RK Vasishta, Mandeep Singh Virk, Mandeep S Dhillon

Department of Histopathology and Orthopaedics, Postgraduate Institute of Medical Education and Research, Chandigarh, India

Click **here** for **correspondence address** and email

How to cite this article:
Jain D, Kumar V, Vasishta RK, Virk MS, Dhillon MS. Gorham's Disease - *A case report*. Indian J Orthop 2006;40:55-6

How to cite this URL:
Jain D, Kumar V, Vasishta RK, Virk MS, Dhillon MS. Gorham's Disease - *A case report*. Indian J Orthop [serial online] 2006 [cited 2008 Nov 27];40:55-6. Available from: http://www.ijoonline.com/text.asp?2006/40/1/55/34436

Introduction

Gorham's disease (disappearing bone disease, massive osteolysis, idiopathic osteolysis, essential osteolysis, progressive atrophy of bone, spontaneous absorption of bone, phantom bone, hemangiomatosis/lymphangiomatosis of bone, progressive osteolysis) is a rare disorder characterized by proliferation of vascular channels that results in destruction and resorption of osseous matrix [1]. Since the initial description of the disease in 1954, fifty years have elapsed but still the precise etiology of Gorham's disease remains poorly understood. Herewith we report a case of Gorham's disease with its histopathology features.

Case report

A young girl came to our institution in 2002 at age 18 complaining of right forearm pain for one and half years. Her pain was not responding to conventional analgesics. Radiographs showed complete loss of bone of proximal one third of the ulna. In addition there were ill defined radiolucent foci of osteolysis present with in the distal humerus and proximal two third of radius involving intramedullary and subcortical regions, resembling findings seen in patchy osteoporosis. The intervening elbow joint afford no protection to extension of the disease [Figure - 1]a,b. A biopsy from the same area revealed few dead bony fragments along with skeletal muscle and hemorrhage. Magnetic resonance imaging (MRI) showed cystic replacement of distal humerus and proximal radius and ulna with no significant soft tissue component. T1-weighted-spin echo MRIs showed uniformly low signal intensity in the involved bones, whereas increased signal intensity was observed in T2-weighted-spin echo images. There were vanishing borders suggestive of hemangiomatous lesion. Subsequently surgery was planned and above elbow amputation had been done including right distal humerus area and forearm. On gross examination,only a small fragment of humerus remained intact. Proximal part of the right radius and ulna was virtually absent. Microscopy showed numerous ectatic variable sized thin walled vascular channels surrounded by meager hematopoetic elements and adipose tissue [Figure - 2]a, b.At one focus there was increased osteoblastic and osteoclastic activity seen. Based on all these findings a diagnosis of Gorham's disease was given.

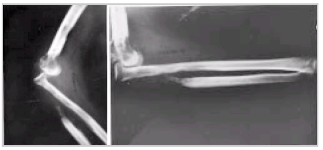

Fig 1a. An oblique view of elb...

On follow up patient was doing well for two years after treatment.

Discussion

The etiology of Gorham's disease is still very speculative, the prognosis unpredictable, and any effective therapy still unknown. The pathological process is the replacement of normal bone by an aggressively expanding but non-neoplastic vascular tissue [2],[3] , similar to a hemangioma or lymphangioma. Wildly proliferating neovascular tissue causes massive bone loss. In the early stage of the lesion, the bone undergoes resorption, and is replaced by hypervascular fibrous connective tissue and angiomatous tissue. Histologically, involved bones show a non-malignant proliferation of thinwalled vessels; the proliferative vessels may be capillary, sinusoidal or cavernous. In late stages, there is progressive dissolution of the bone leading to massive osteolysis, with the osseous tissue being replaced by fibrous tissue. The stimulus that generates this change in the bone is unknown [3] . The clinical presentation of Gorham's disease is variable and depends on the site of involvement. The shoulder [4] , and the pelvis [5] are the most common sites. It often takes many months or years before the offending lesion is correctly diagnosed. Gorham's disease can involve men or women and any age group. No familial predisposition has been found [6] . A high index of clinical suspicion is needed to arrive at an early, accurate diagnosis. The medical treatment for Gorham's disease includes radiation therapy [7] , anti-osteoclastic medications, and interferon [8] . In most cases, Gorham syndrome is only diagnosed after a pathological fracture. However in the present case, patient presented with chronic pain at the site of osteolysis. According to Patel [9] the humerus, radius and ulna are rare sites for involvement though few case reports are there in the literature.

Histopathologically, hemangiomatous or lymphangiomatous tissue can be observed [2] . Therefore the differential diagnosis of angiomatosis is significant; the latter has, however, a multiple extraostotic origin, in contrast to Gorham syndrome. The differential diagnosis also includes other forms of osteolysis such as familiar osteolysis, macro osteolysis or essential osteolysis.

Usually, the prognosis depends on complications, such as neurological deficits or pleural effusion. The life expectation is not altered if the extremities are involved. Several therapeutic modalities have been used in the management of Gorham's disease [9] . Surgical treatment options include resection of the lesion and reconstruction [10] . In general, no single treatment modality has proven effective in arresting the disease. It is worth noting that the success rate after the use of a bone graft is low. Most surgeons,

based on their personal experience, have observed that the bone graft undergoes dissolution.

References

1. Gorham LW, Wright AW, Shultz HH, Maxon FC, Jr. Disappearing bones: a rare form of massive osteolysis: report of two cases, one with autopsy findings. *Am J Med.* 1954;17:674-682
2. Dickson GR, Hamilton A, Hayes D, Carr KE, Davis R, Mollan RA. An investigation of vanishing bone disease. *Bone.* 1990;11:205-210
3. Devlin RD, Bone HG 3rd, Roodman GD. Interleukin-6: a potential mediator of the massive osteolysis in patients with Gorham-Stout disease. *J Clin Endocrinol Metab.* 1996;81:1893-1897
4. Remia LF, Richolt J, Buckley KM, Donovan MJ, Gebhardt MC. Pain and weakness of the shoulder in a 16-year-old boy. *Clin Orthop.* 1998;347:268-271, 287-290.
5. Hejgaard N, Olsen PR. Massive Gorham osteolysis of the right hemipelvis complicated by chylothorax: report of a case in a 9-year-old boy successfully treated by pleurodesis. *J Pediatr Orthop.* 1987;7:96- 99.
6. Gorham LW, Stout AP. Massive osteolysis (acute spontaneous absorption of bone, phantom bone, disappearing bone): its relation to hemangiomatosis. *J Bone Joint Surg (Am).* 1955;37-A:985-1004.
7. Gutierrez RM, Spjut HJ. Skeletal angiomatosis: report of three cases and review of the literature. *Clin Orthop.* 1972;85:82-97
8. Hagberg H, Lamberg K, Astrom G. Alpha-2b interferon and oral clodronate for Gorham's disease. *Lancet.* 1997;350(9094):1822-1823
9. Patel DV. Gorham's disease or massive osteolysis. *Clin Med.* 2005; 3: 65-74
10. Turra ST,Gigante C, Scapinelli RA. 20 year follow up study of a case of surgically treated massive osteolysis. *Clin Orthop.* 1990,250:297-302.

Correspondence Address:
Deepali Jain
Department of Pathology, All India Institute of Medical Sciences, New Delhi, 110029
India
Login to access the email ID

Source of Support: None, **Conflict of Interest:** None

Figures

[Figure - 1], [Figure - 2]