

Massive Osteolysis in Distal Shaft of Humerus: A Case Report on Vanishing Bone Disease

Sana Noman, Seema Faiz, Sohail Ahmed Khan and Amjad Sattar

ABSTRACT

Gorham-Stout disease is a rare disease characterised by endothelial proliferation of lymphatics and vessels. It is also known as vanishing bone disease. Increased vascularity and proliferation lead to progressive bony osteolysis. We report a case of a 17-year female diagnosed as having post traumatic Gorham-Stout disease. Being reported for the first time from our population, this will be an informative addition to the already available literature. On X-rays, initially there is osteopenia with patchy distribution followed by coalescence of osteopenic patches, erosion of cortex with soft tissue involvement, and finally bone resorption. MRI shows increased T2 signal with heterogeneous enhancement.

Key Words: Osteolysis. Tumors. Humerus.

INTRODUCTION

Gorham-Stout disease, also termed as vanishing disease of bone, is a rare disease with an unknown cause characterised by a proliferation of endothelial lined vessels and its replacement with fibrous tissue on histology.¹ It was initially described in a teenaged having a boneless arm.² Later on, this condition was comprehensively reviewed by Gorham and colleagues.¹ This condition is thought to be a developmental disorder rather than neoplastic and may be associated with lymphatic malformation of the skin.³ The dramatic increase in vascularity and associated vascular changes are responsible for massive bony osteolysis.^{4,5}

We report a case of this rare condition in a young female patient, which has not been reported earlier from our population.

CASE REPORT

A 17-year female presented for an MRI of left humerus at our Department. Four months back, she suffered trauma to humerus and a radiograph at that time showed fracture of distal shaft of humerus with laterally displaced distal fracture fragment (Figure 1a). Her follow-up radiograph after one month showed regional osteopenia and resorption of bone near the fracture site (Figure 1b) and progressive resorption leading to non-visualisation of distal shaft of humerus (Figure 1c). Later, a skeletal scintigraphy was performed that showed focal increased radiotracer uptake over head of humerus and mild increased radiotracer uptake in distal two-thirds of

shaft of humerus. Now, she complained of pain and was advised MRI of humerus that showed cortical and medullary erosion noted at mid and distal shaft of humerus resulting in its tapering with complete resorption of a segment of distal shaft (Figure 2a). Distal humerus showed intact articulation at elbow joint. Enhanced soft tissue thickening representing fibrous tissue was seen along the resorpted segment of bone (Figure 2b). Adjacent muscular atrophy was noted. No abnormal enhancement was noted, and increased vascularity noted in region of non-visualised segment of bone (Figure 2c).

These findings were labelled as vanishing bone disease and histopathology was advised that showed fragments of lamellar bone. Some places showed decreased bony trabeculae of small size. Intervening tissue showed increased proliferation of variable sized thin and thick blood vessels. The radiological findings were confirmed as Gorham's disease on histopathology.

DISCUSSION

Vanishing bone disease or Gorham-Stout disease is a rare vascular abnormality distinguished by its features of abnormal lymphatics.^{6,7} Sinusoidal proliferation of blood vessels and lymphatics is present leading to progressive bone destruction.³ Clinically, it can present at any age, however, young age group is predominantly affected with no characteristic gender distribution or inheritance pattern.⁸ Common presenting symptoms usually include pain and edematous changes in the region involved.⁹ However, this condition can remain completely symptom-free unless a pathological fracture develops.¹⁰

Idiopathic osteolysis has been described by five types.¹⁰ Gorham disease is a type of osteolysis that is included in non-familial type, rest of the types described are inherited and commonly present in childhood. Neuropathy has also been described with type III

*Department of Radiology, Dow Institute of Radiology,
Dow University of Health Sciences, Karachi.*

*Correspondence: Dr. Seema Faiz, Resident, Dow Institute of
Radiology, Dow University of Health Sciences, Karachi.*

E-mail: dr_seemashaikh@hotmail.com

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Figure 1: Serial images of a 17-year girl after trauma. (a) Radiograph of humerus shows fracture of distal shaft of humerus with displaced fracture fragments. (b) One month later, the radiograph shows resorption of distal shaft of humerus. No soft tissue swelling is evident. (c) Progressive resorption leading to non-visualisation of distal shaft of humerus.

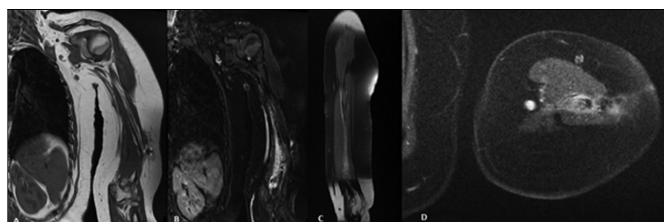


Figure 2: (a) Coronal T2 weighted image (b) Coronal STIR (c) Sagittal STIR MRI sequences shows pointed distal shaft of humerus with non-visualisation of small segment of distal shaft of humerus indicating its resorption. No abnormal marrow signal is identified in the rest of the visualised humerus. There is soft tissue thickening identified adjacent to site of resorpted bone likely representing fibroud tissue. The trochlea and capitellum are visualised and appear unremarkable. (d) Axial post show subtle enhancement in the soft tissue thickning.

osteolysis. Unlike type III, Gorham's disease is seldom associated to have neuropathy. Common reported sites involved by Gorham disease include skull, clavicle, scapula, pelvis, humerus, and ribs.¹⁰

Characteristically on plain radiographs, the disease can be separated into four phases, i.e. a) osteopenia with patchy distribution, b) osteopenic patches coalescence, c) erosion of cortex with soft tissue involvement by lesion, and d) resorption of bone.¹⁰ These radiographic characteristics are slow and progressive and may involve one or more bones. MRI typically demonstrates increased signal on T2-weighted images with features of heterogeneity. Post contrast enhancement may also be heterogeneous on MR images.¹⁰ Bone scan performed with Technetium 99m MDP, initially shows increased radiotracer uptake; and with ongoing resorption, this becomes negative.

Regarding treatment of this condition, mixed agreements are present and reported. Current treatment regimen usually indicates the use of medication against osteoclasts such as calcium and vitamin D supple-

mentation and bisphosphonate use, surgery to treat tumor and radiation therapy to prevent re-growth of tumor by inducing sclerosis.

Gorham disease is an uncommon disorder. Although international reports are frequent, to the best of our knowledge, this condition has not been reported from developing countries and no report exists from our country, making it an informative addition to already available literature.

Gorham's disease is a rare, locally aggressive osteolytic disease. The presence of bone resorption without any obvious cause, should undergo a thorough clinical examination coupled with follow-up radiological, laboratory, and histopathological investigations to provide an accurate early diagnosis of this incompletely understood condition, to estimate its occurrence, and to ascertain its prognosis.

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