

Vanishing bone disease involving the pelvis

ABSTRACT

Vanishing bone disease is a rare condition characterized by progressive osteolysis of the bony structures by vascular tissue and their replacement by fibrous, vascular connective tissue. A 38-year-old lady who had a previous history of angioma of the left iliac bone presented with vague symptoms of a limping gait and pain during walking since 2 years duration. The radiologic findings were suggestive of degenerative changes. A possibility of metastatic disease was also considered. However the biopsy and Positron emission tomography (PET) scan ruled out any active disease. Like in most other cases this was possibly a self-limited disease where bone resorption had spontaneously arrested.

Key words: Gorham's disease, Gorham Stout syndrome, Massive Osteolysis, Vanishing Bone disease

INTRODUCTION

Vanishing bone disease is a rare condition of unknown etiology affecting the skeletal system that was first described by Jackson in 1838.^[1] Gorham and Stout first presented it as a single disease entity with varying localization as a large series in 1954.^[2] Since then, this disease has acquired several synonyms like Gorham's disease, Gorham-stout syndrome, massive osteolysis, phantom bone disease, disappearing bone disease, essential osteolysis, etc.

The disease appears generally in the second and third decades of life with an equal sex distribution. The skeletal structures affected include the innominate (hip) bones, thorax, spine, maxilla and mandible although no area of the skeleton is immune.^[3] The clinical picture is characterized by progressive osteolysis of the bony structures by vascular tissue and their replacement by fibrous, vascular connective tissue.^[4] Radiologically, there are progressive osteolytic lesions suggestive of bone resorption. There are no reported cases of metastasis to date, although polyostotic and monostotic lesions have been described.^[4,5,6] In some cases, the process has spontaneously arrested^[1,7,8,9] and in others progression of disease^[5,6] has resulted in death of the patient. We report a young lady with vanishing bone disease affecting the pelvic bone that had spontaneously arrested.

CASE HISTORY

An 18-year old young unmarried girl presented in 1983 with gradually worsening pain in the left

hip and difficulty in walking since 6 months. A radiolucent lesion was noted on a plain radiograph in the left iliac bone. A diagnosis of angioma of the left iliac bone was made by incisional biopsy. Due to the benign nature of the disease and unmarried status, the girl refused surgery and preferred to be kept under observation. Twenty years later in 2003, she presented with difficulty in walking, a limping gait and pain in the right thigh radiating to the legs while walking; but no pain at rest. Trendelenburg's test was positive and the right limb was longer than the left. There was also evidence of scoliosis to the right. Routine blood investigations were essentially within normal range. Plain radiogram and computerized tomographic scan of the pelvis [Figure 1] showed almost complete osteolysis (absence) of the left bony pelvis. The major portion of ischium and pubis on left side was also not visualized. The sacrum also showed a few lytic lesions with some fragmentation. There was erosion of the right iliac bone, fragmentation of the pubic bone, fracture of the acetabular margin, and widening of joint space on the right side. A whole body bone scan with ^{99m}Tc-MDP (Methylene diphosphate) demonstrated increased focal concentration of radiotracers in right acetabular area, head of right femur, right half of symphysis pubis along with medial portion of ramus of right pubic bone and right half of body of sacrum. There was gross photo deficient area over the left iliac bone [Figure 2]. The radiological findings were suggestive of degenerative changes. The possibility of metastatic disease was also considered because of the absence of left hemipelvis. A core biopsy from the right iliac bone revealed bony trabeculae and fatty marrow with no

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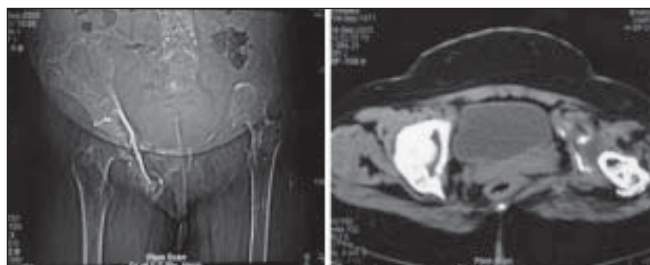


Figure 1: Xray and CT scan of Pelvis showing almost complete osteolysis of left bony pelvis

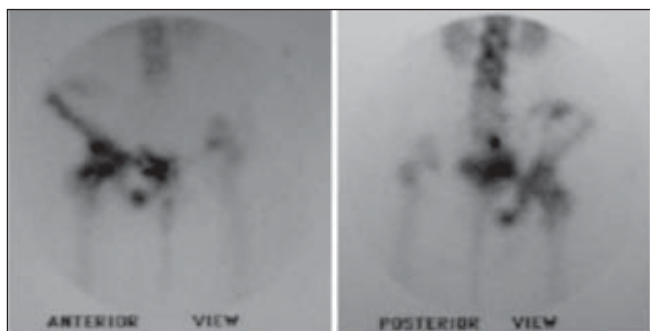


Figure 2: Bone scan showing gross photo deficient area over the left iliac bone

haematopoietic elements. To differentiate degenerative changes from active disease, PET scan using 18-Fluorodeoxyglucose (FDG) was done which showed no evidence of increased tracer uptake in the osteolytic sites. There was normal physiological uptake and non-visualization of the left hemipelvis. Thus it was concluded that the radiological findings were due to degenerative changes. She was reassured about the self limiting nature of the disease and provided symptomatic treatment along with a walking aid as support and kept under regular follow up.

DISCUSSION

Gorham's syndrome is an extremely rare condition of the bone, with fewer than 200 cases reported in literature.^[10,11] Although the disease is quite well described, it does not occur often enough to be recognized early, taking many years for the condition to be recognized. In most instances, the condition is not recognized until a fracture occurs following trivial trauma and bone healing is delayed. Clinically the course of the disease appears to be divided into two phases, an active phase of bone resorption and lysis and a second, generally dormant phase. The succession of phases is unpredictable, taking a few months to many years.^[12]

A literature review reveals that there is no consensus to date regarding the optimum treatment for this condition. Several factors influence treatment decisions, which range

from symptomatic treatment alone to aggressive surgical resection with reconstruction. Radiation therapy also can be quite effective with 30-45 Gy to the involved site as seen in one series which reported 14 of the 22 cases with successful outcomes.^[5] However, this is difficult to prove in the face of a disease that is known to spontaneously arrest.^[5,10] Moreover, the complications of radiotherapy in children and young adults should also be considered, including the potential for malignant transformation, growth restrictions and damage to tooth formation and eruption. Agents that inhibit bone resorption (bisphosphonates, calcitonin) have also been used with variable success.

Our case report was a clinical diagnostic dilemma, where there was an initial history of angioma of the left iliac bone which had destroyed the left side of pelvis and like in most other cases this was possibly a self limited disease where bone resorption had spontaneously arrested. Though the radiological findings were not conclusive, they did suggest the possibility of an active disease. The biopsy and PET scan findings ruled out any active disease. Currently there is no data in literature regarding FDG uptake in active Gorham's disease and this could be one of the first cases where PET has helped in deciding the management of such a case.

REFERENCES

1. Jackson JBS. A boneless arm. *Boston Med Surg* 1838;18:368-9.
2. Gorham LW, Wright AW, Shultz HH, Maxcon FC Jr. Disappearing Bones. A rare form of massive osteolysis: Report of two cases: One with autopsy findings. *Am J Med.*1954;17:674-82.
3. Pedicelli G, Mattia P, Zorzolli AA, Sorrone A, De martino F, Sciotto V. Gorham Syndrome. *JAMA* 1984;252:1449-151.
4. Cherrick HM, King OH Jr, Dorsey JN Jr. Massive osteolysis (disappearing bone, phantom bone, acute absorption of bone) of the mandible and maxilla. *J Oral Med* 1972;27:67-74.
5. Dunbar SF, Rosenberg A, Mankin H, Rosenthal D, Suit HD. Gorham's Massive Osteolysis: The role of radiation therapy and a review of the literature. *Int J Radiat Oncol Biol Physics* 1993;26:491-7.
6. Holroyd I, Dillon M, Roberts GJ. Gorham's disease: A case (including dental presentation) of vanishing bone disease. *Oral Surg Oral Med Oral Pathol* 2000;89:125-9.
7. Campbell J, Almond HG, Johnson R. Massive osteolysis of the humerus with spontaneous recovery. *J Bone Joint Surg (Br)* 1975;57:238-40.
8. Gorham WL, Stout AP. Massive osteolysis (acute spontaneous absorption of bone, phantom bone, disappearing bone) *J Bone Joint Surg (Am)* 1955;37:985-1004.
9. Heyden G, Kindblom LG, Nielsen JM. Disappearing Bone Disease: A Clinical And Histological Study. *J Bone Joint Surg (Am)* 1977;59:57-61.
10. Swelstad MR, Frumiento C, Garry-McCoy A, Agni R, Wiegel TL. Chylotamponade: An unusual presentation of Gorham's Syndrome. *Ann Thorac Surg* 2003;75:1650-2.
11. Fontanesi J. Radiation Therapy in the treatment of Gorham Disease. *J Pediatr Hematol Oncol* 2003;25:816-7.
12. El Mofty S. Atrophy of the mandible (massive osteolysis). *Oral surg, Oral Med, Oral Pathol* 1971;31:690-700.