Massive Osteolysis of the Upper Extremity A Case Report and Review of Literature

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ABSTRACT: A case report of massive osteolysis or Gorham's syndrome, was presented. This was the case of a 17 year old Filipino male who sustained a complete transverse fracture at midshaft of the left humerus. The patient was placed in cast and healing progressed normally. Four months later, he sustained an apparent refracture after forceful exertion. The fracture failed to heal and the humerus underwent massive osteolysis necessitating radical amputation.

INTRODUCTION

Massive osteolysis, a rare phenomenon involving the spontaneous dissolution of bone, was first extensively described by Gorham[1] in 1955. The first case, however, was documented by Jackson in 1838. Subsequent reports have used varying terms, such as disappearing bone disease, phantom bone, cryptogenic osteolysis and progressive bone atrophy[2].

A recent comprehensive review has documented only 98 cases of massive osteolysis from world literature[3]. Among Filipinos, only one case had so far been reported. This involved a 13-year old girl with progressive lysis of the pelvis associated histologically with massive angiomatous destruction. The present report is only the second case of massive osteolysis documented among Filipinos, and it demonstrates the diagnostic and therapeutic enigma that characterized this disorder.

CASE REPORT

A 17-year old male of full Filipino parenthood was first seen at the National Orthopaedic Hospital three and a half years ago for an injury to the left arm sustained in a basketball game. X-rays revealed a complete transverse fracture through the midshaft of the humerus without any evidence of pre-existing pathology. Simple plaster immobilization after closed reduction was done and the patient was followed up on an outpatient basis. After two months, repeat x-rays showed satisfactory alignment of fragments with advanced callus formation. External immobilization was discontinued and full range of motion of the involved extremity initiated. Two months later, the patient complained of swelling and pain of the same arm after forceful exertion. X-rays taken were interpreted as refracture of the left humerus with otherwise unremarkable bone findings. A sugar-tong plaster splint was reapplied and the patient was followed up regularly. Subsequent radiographic examinations, however, showed absence of fracture healing and progressive thinning of the humeral cortices. The ipsilateral clavicle and proximal radius later also demonstrated deossification. The patient was admitted for thorough work-up on April, 1988. CBC, serum proteins, calcium and phosphorus, BUN-creatinine and urinalysis were all within normal limits. Alkaline phosphatase was slightly elevated. An angiogram done on the left upper extremity was considered unremarkable.

Open biopsy of the humerus revealed thinned out cortical bone consisting of trabeculae interspersed with connective tissue and distended blood vessels; cystic areas filled with blood and surrounded by fibrous tissue were also noted. Such findings were interpreted as histologically compatible with an aneurysmal bone cyst. Clinically, however, massive osteolysis was suspected, and the patient was subjected to radiation therapy in an effort to diminish vascularity prior to a contemplated local resection and bone grafting procedure.

Five months after admission, x-rays revealed progressive disappearance of the entire humeral shaft, with subsequent dissolution of the glenoid fossa and proximal radius. At this point, local surgery was deemed futile. Refusing a proposed ablation of the upper extremity, the patient was instead sent home in an arm brace. For the next one and a half years, the

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patient remained pain-free and attended school with limited active motion of the left arm. In June 1990, however, he developed spontaneous pain of the left shoulder, for which analgesics gave transient relief. After three weeks, the pain grew in intensity and was accompanied by fever. The patient was then admitted to the Children's Medical Center. He was fairly nourished and ambulatory but markedly withdraw. Vital signs were within normal limits, except for moderate-grade fever. Examination of the head, neck and chest revealed no abnormalities, although there was recept for mild tenderness over the area of the left clavicle. The left arm was atrophic with preternatural mobility along its length; it was, however, nontender and showed no overlying skin abnormalities. The ipsilateral shoulder and elbow had a finding of slight limitation of motion. Forearm and hand function were essentially unremarkable. (Fig 1,2,3,4,5) CBC revealed a leukocytosis of 22,000, while other blood examinations were normal. Tuberculin testing and chest radiograph were both negative. A septic work-up, including Widal and malarial testing, done for continuous high-grade fever during the first hospital week, were also negative. Fever,however, lysed with intravenous cephalosporins. The patient was discharged with minimal symptoms 10 days after admission. Two weeks later, he was readmitted for ablation surgery. Psychiatric counselling had been carried out in the interim. A thoracoscapular (forequarter) amputation was performed on July 27 1990. Surgical margins were considered only marginal over the most proximal extent of the lesion, as massive involvement of the entire shoulder girdle was noted. Gross examination of the amputated limb showed absence of the scapula and clavicle except for their medial borders. Remaining bone tissue, consisted of very thin wafers of cortex surrounded by fibrous tissue cystic fluid and modest amounts of blood. The humerus was totally absent with only some remnants of the elbow joint remaining. The ulna was likewise nearly completely, and the radius was markedly thinned out. The remaining radial diaphyses contained serosanguinous fluid and had scanty marrow tissue. Histologic examination revealed decalcified bone mostly made up of hyaline and scattered around by dilated small to medium sized, thin-walled blood vessels. Findings were multiple hemangiomatous process which consistented with the clinical impression of massive osteolysis (Fig.6).

Post-operative course was uneventful. As of last follow-up 8 months after amputation, the patient has attained satisfactory one-handed function, attended school and engaged in otherwise normal activity. Radiographs of the thoracic cage have shown on evidence of lytic change.

DISCUSSION

Massive osteolysis or Gorham's disease has been defined as a syndrome characterized by a non-familial, histologically benign vascular proliferation originating in bone and producing complete lysis of all or a portion of the bone[4]. It has also been termed a predominantly monocentric osteolysis affecting as one or a group of contiguous bones[5]. Although still incompletely understood, Gorham's original description associating extensive skeletal dissolution with angiomatosis is presently well established[1,4,6,7,8,9]. The exact etiology of massive osteolysis is unknown. Gorham[9] has originally postulated that angiomas

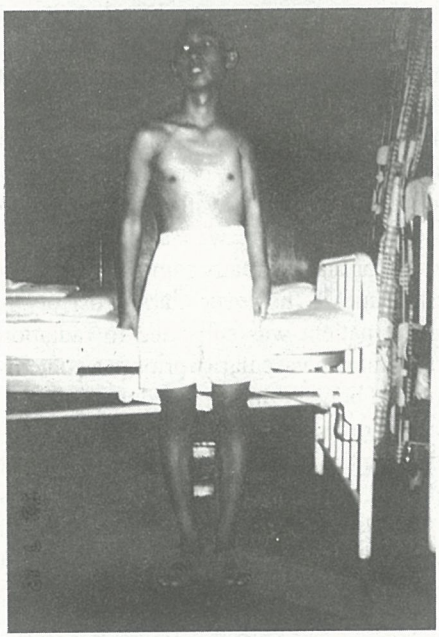

Fig.1. The patient 3 weeks prior to amputation. Note atrophic left upper extremity with absence of shoulder girdle.

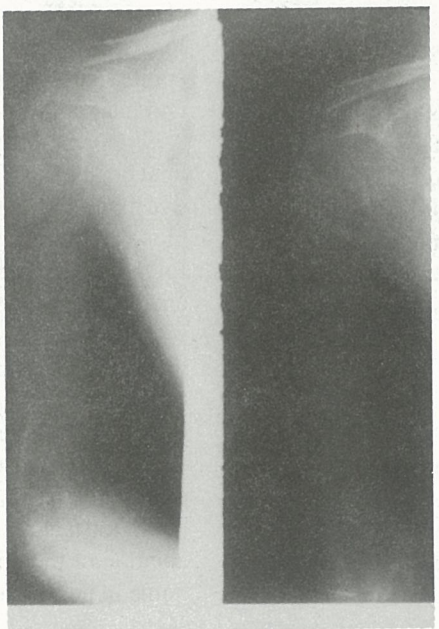

Fig.2. Radiographs one year after left arm fracture, showing massive dissolution of humerus, humeral head still visible.

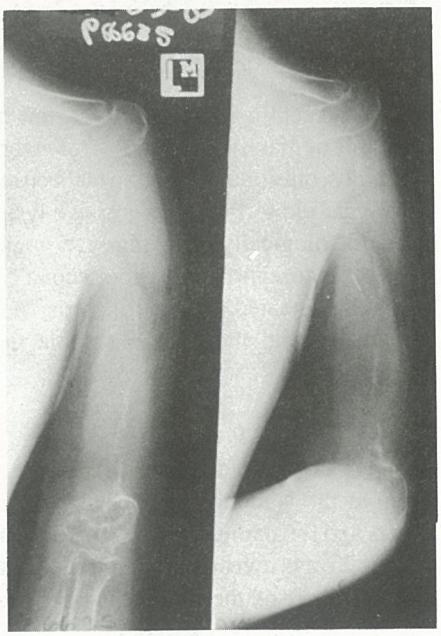

Fig.3. Follow-up radiographs 18 months from onset showing total humeral lysis with involvement of shoulder girdle and proximal ulna.

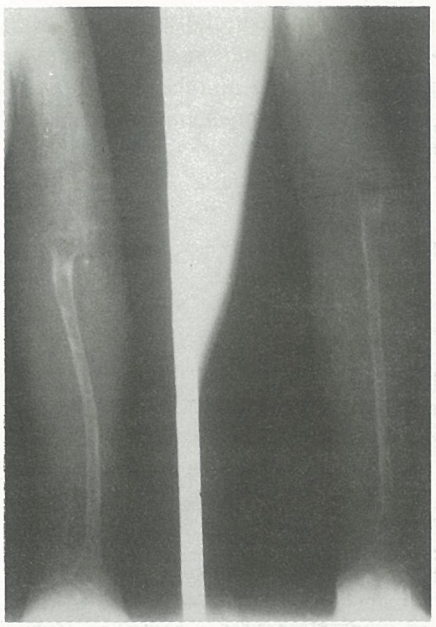

Fig.4. X-rays of upper extremity just prior to amputation. The entire humerus and most of ulna have been lysed. The radius is markedly thinned out.

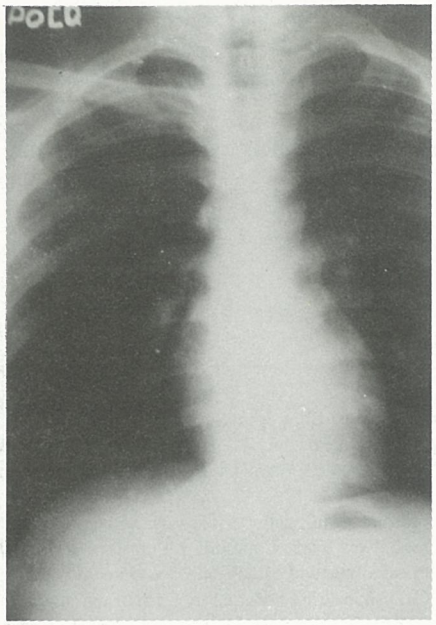

Fig.5. Chest radiograph before amputation showing disappearnce of left shoulder girdle with no further involvement of chest cage.

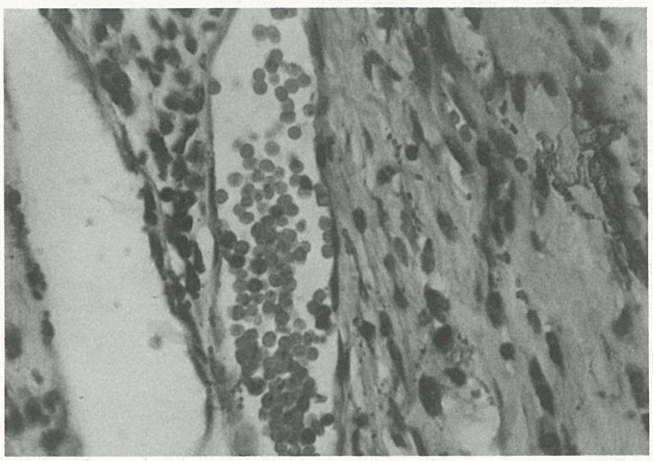

Fig.6. Microscopic view of specimen showing thin-walled vascular channels filled with blood and surrounded by fibrous connective tissue.

may increase local oxygen tension by acting as a shunt. Alternatively, Thompson[10] has proposed the role of pressure from unrestricted growth of granulative tissue in causing bone resorption. Other studies have attributed bone destruction to abnormal activity of perivascular monocytes, endothelial cells and, more notably, osteoclasts[4,11]. However, the search for a definitive pathologic mechanism for this unusual syndrome continues.

Gorham's[1,8] classical histopathologic description of massive osteolysis depicted bony destruction as anastomosing vascular spaces lined by endothelium and surrounded by fibrous stroma. This pattern of angiomatous proliferation, usually involving blood vessels but also lymphatics, has been consistently demonstrated in subsequent reports[9,11-14].

The clinical, radiologic and histologic picture documented in the present case fits closely to the previous descriptions. As exemplified by our patient, the most common age of occurence is the teenage years[9]. However, the disease is known to affect a wide range of ages from one month to 75 years, with a mean below 30. No particular sex or racial predilection has been noted. A clear history of trauma is seen in over half of cases, and pathologic fractures are a common occurence[9,11-14].

Although the disease affects almost any bone, a propensity for involvement of the shoulder and pelvic girdles has been observed. The arm and shoulder region is the primary site of involvement in 26 percent of cases. Adjacent bone extension, as in the present case, is seen in 76 percent, although the process is monocentric[4, 9, 12,14].

The earliest radiographic finding in Gorham's osteolysis is patchy osteoporosis. This is followed by concentric shrinkage of tubular bone with tapering at the end, and finally massive resorption and progressive contiguous bone involvement[15,16]. In the present case, early humeral osteoporosis was interpreted as deossification secondary to disuse. Osteolysis was detected only with cortical thinning and progressive radiolucency around the area of fracture.

The differential diagnosis of massive osteolysis include skeletal angiomas, angiosarcoma, essential osteolysis with nephropathy and hereditary osteolysis. Gorham's disease is distinguished from these entities by its massive and often rapidly progressive character, the lack of any genetic mode of transmission, and the absence of concomitant systemic illness or organ dysfunction[4,5].

The clinical course of massive osteolysis is variable. Some cases undergo spontaneous arrest after a number of years[4,9,14,17]. Our patient's rapidly progressive course resembles more closely cases observed to be "clinically malignant", with progressive dissolution both proximally and distally across two joints[9,10]. Death secondary to vital structure involvement occurs in 16 percent of reported cases, most commonly from spinal and/or chest wall lesions[4,12,18].

A consistently successful treatment of Gorham's disease remains elusive. Various modalities have been attempted, including hormonal, calcium and vitamin D treatment, ultraviolet radiation, chemotherapy, and bone grafting.Results, however, have been uniformly disappointing. Some authors have proposed more judicious conservative management such

as orthotic support[4,9,14,19]

Only radiation therapy has shown significant promise, with actual arrest of the disease and bone regeneration documented in a few cases[4,11,12]. Our patient, however, failed to show any positive response to irradiation. Amputation is accepted as definitive treatment, especially in cases with severe extremity involvement[14]. Rapid transarticular spreading of the disease in our patient left no other recourse and radical amputation can prevent life-threatening by extension of the disease into the chest wall.

REFERENCES

1. 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;37:985-1004.
2. Fornasher VL. Hemangiomatosis with massive osteolysis. J Bone Joint Surg(Br) 1970;52:444-51.
3. Silao JV, Montalban AM, Fontanilla JM. Gorham's massive osteolysis. Acta Med Phil 1979;15:89-94.
4. Choma ND, Biscotti CV, Bauer TW, Mehta AC, Licata AA. Gorham's syndrome: a case report and review of the literature.Am J Med 1987;83:1151-6.
5. Hardegger F,Simpson LA, Segmueller G. The syndrome of idiopathic osteolysis. J Bone Joint Surg(Br) 1985;67:89-93.
6. Enneking WF. Musculoskeletal tumor surgery. vol 2. New York: Churchill Livingstone 1983;1193-5.
7. Gorham LW. Circulatory changes associated with osteolytic and osteoblastic reactions in bone. Arch Intern Med 1960;105:47-64.
8. Gorham LW, Wright AW, Shultz HP Maxon FC. Disappearing bone; a rare form of massive osteolysis. Am J Med 1954;17:674-82.
9. Mendez AA, Keret D, Robertson W Ewen GD. Massive osteolysis of the femur (Gorham's disease); a case report and review of the literature. J Pediatr Orthop 1989;9:604-8.
10. Thompson JS, Schurman DJ. Massive osteolysis. Clin Orthop 1974; 103: 206-11.
11. Heyden G, Kingblom L, Nielsen M. Disappearing bone disease. J Bone Joint Surg(Am) 1977;59:57-61.
12. Cannon SR. Massive osteolysis; a review of seven cases.J Bone Joint Surg(Br) 1986;68:24-8.
13. Hambach R, Pujman J, Maly V. Massive osteolysis due to hemangiomatosis; report of a case of Gorham's disease with autopsy. Radiology 1958;71: 43-47.
14. Ross JL, Schinella R, Shenkman L. Massive osteolysis; an unusual cause of bone destruction. Am J Med 1978;65:367-2.
15. Johnson PM, McClure JG. Observations on massive osteolysis. Radiology 1985;71:28-42.
16. Torg JS, Steel HH. Sequential roentgenographic changes occurring in massive osteolysis. J Bone Joint Surg(Am) 1969; 51: 1649-55.
17. Sage MR, Allen PW. Massive osteolysis: Report of a case. J Bone Joint Surg(Br) 1974;56:130-5.
18. Dahlin DC, Unni KK. Benign vascular tumors. In: Bone Tumors, 4th ed. Springfield: Charles C. Thomas 1986;167-180.
19. Abell JM, Badgley C E. Disappearing bone disease. JAMA 1961;177:771-3.