www.jmscr.igmpublication.org *Impact Factor 1.1147*
ISSN (e)-2347-176x

IGM Publication Journal Of Medical Science And Clinical Research
An Official Publication Of IGM Publication

A Rare Case of Gorham's Disease of the Radius with Implant in Situ: A Case Report

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ABSTRACT:

Gorham disease is a rare condition characterized by intraosseous neoplastic proliferation of hemangiomatous tissue with progressive, massive osteolysis. We present a pathologically proved case of Gorham disease that involved the left parietal bone in a 35-year-old woman. Imaging studies included conventional radiography of the left wrist and distal forearm.

Gorham disease—also known as massive osteolysis of Gorham, disappearing bone disease, vanishing bone disease, phantom bone disease, and Gorham-Stout syndrome. Gorham's disease is a rare disorder characterized by proliferation of vascular channels that results in destruction and resorption of osseous matrix. The precise etiology of Gorham's disease still remains poorly understood. There is no malignant, neuropathic, or infectious process involved in the etiology of this disorder. In this report, we present the unique involvement of the distal radius with an implant in situ.

Key words: *Gorham's Disease, Massive Osteolysis, Implant In Situ, Radius*

INTRODUCTION

Gorham-Stout syndrome was first reported by Jackson in 1838 in a 12-year-old boy with complete osteolysis of the humerus. In 1955, Gorham and Stout further characterized the main pathologic features of this rare disease as nonmalignant intraosseous proliferation of hemangiomatous or lymphangiomatous tissue that caused massive osteolysis. Gorham's disease, fortunately, is an extremely rare disorder of the musculoskeletal system. Gorham's disease is progressive in most patients; however, in some cases, the disease process is self-limiting.

We report herein a case of massive osteolysis of the radial shaft with a dynamic compression plate in situ.

CASE REPORT

A 35-year-old woman had an asymptomatic deformity over the left wrist region that progressed over a period of 1 year. The patient had sustained a Galleazi's fracture of the left radius following trauma due to Road-traffic accident 1 year ago. A conventional radiograph of the forearm was obtained, and patient underwent an Open Reduction, Internal Fixation with a Dynamic Compression Plate. She followed the normal post operative protocol and normal activities were started 2 months after the surgery. She started complaining of pain in the wrist and distal radius region which was dull aching and vague. She noticed a deformity over the left wrist joint which progressed gradually.

Physical examination revealed no palpable mass or tenderness over the left wrist, but a large area of softening and depression over the left distal radius about 7 cm long from wrist joint was found. The laboratory results were unremarkable. Conventional radiography of the forearm and wrist showed a large, sharply demarcated bony defect without marginal sclerosis of the left distal radius [Fig. 1(a)&1(b)]. A chronic but still active destructive process of the distal 1/3rd radius was considered on the basis of the clinical history and imaging findings. To search for other possible similar bony lesion in the body, a Technetium-99 m methylene diphosphate (Tc-99 m MDP) whole-body bone scan was undertaken, which revealed no other lesion. The preoperative differential diagnosis included juvenile Paget disease at destructive stage, eosinophilic granuloma, & Gorham's disease but none of them correlated well with the clinical and radiologic features except Gorham's disease. Biopsy confirmed the diagnosis as it showed fragments of dead bone and numerous small vascular channels and skeletal muscle.

We planned a surgical resection of the affected radius and replace it with a fibula strut graft and subject the patient to radiation therapy. The patient did not wish to undergo any surgical treatment at this point in time due to family commitments. The patient received symptomatic treatment in form of analgesics, bisphosphonates, interferons and radiation. On follow up after 6 months there was no spread of the disease and seemed to be self limiting. Now its 13 months

since time of presentation and the disease has not progressed. She is doing her daily activities with the use of a hard brace as a cautionary means.

DISCUSSION

Gorham disease is a nonhereditary, nonmalignant and a disease with no sex predilection. Its prognosis is unpredictable and its etiology unknown. Also the correct treatment protocol is unavailable. Most cases occur in children and young adults (usually less than 40 years of age) and no definite inheritance pattern has been reported.[1] Diagnosis is often delayed in most cases as laboratory studies are usually within normal limits. A high index of clinical suspicion together with characteristic radiographic and histopathological findings are helpful for making an early accurate diagnosis. The natural history of Gorham's disease is unpredictable and, in some cases, spontaneous regression has been reported.[5] 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. More than 150 cases have been documented in the literature[6]. Among them, the pelvic and shoulder regions were most frequently involved, although any bone may be affected. The radius is among the least common locations of involvement that too with an implant in situ.[1],[2],[7],[8],[9]

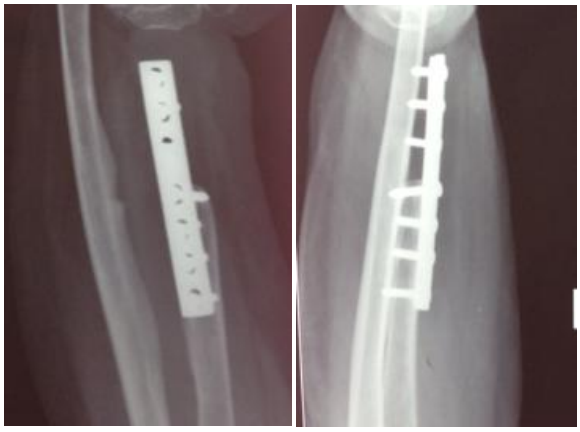
The pathological process involves replacement of the normal bone by an aggressively expanding angiomatous or lymphatic tissue, sometimes both.[10] Pathologically, there are two stages of the disease process: the first with vascular proliferation followed by a second stage in which residual fibrous tissue replaces resorbed bone. [11] Although the normal levels of serum calcium and alkaline phosphatase seen in the disease suggest lack of osteoclastic activity, recent studies showing elevated levels of IL-6 suggest that osteoclasts do play an important role in the disease process. [12]

The treatment of Gorham's disease is controversial. Medical treatment with estrogens, magnesium, calcium, vitamin D, vitamin B12, fluoride, calcitonin, cisplatin, actinomycin D, thalidomide, somatotrophin, interferons, amino acids, placental extracts, and transfusions of placental blood have proved unsuccessful. [13],[14] Long-term bisphosphonates, although a therapeutic option, have not been found to have any significant effect on the disease process. [15] Surgical treatment generally involves resection of the affected bone, with or without replacement prostheses or bone grafts. [16],[17] Amputation has also been done in a number of patients. [4] 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.[3] Diaphyseal involvement poses a challenge to the treating surgeons. The treatment of diaphyseal disease has been either amputation or radiotherapy followed by bone grafting. Despite the fact that bone grafts

have been found to resorb, some studies have shown promise with fibular grafting. [5],[16]

Due to the rarity of the condition a high index of clinical suspicion is needed to arrive at an early, accurate diagnosis. Radiation therapy early in the disease course prevents disease extension, halts the process and may even result in recalcification.



CONCLUSION

Gorham's disease is a rare musculoskeletal disorder. Its diagnosis is usually delayed and often missed as not many physicians have opportunity to treat this rare disease entity in their clinical practice. Gorham's disease is a rare, peculiar musculoskeletal disorder in which the affected bone virtually disintegrates and is replaced by vascular fibrous connective tissue. The etiology of Gorham's disease is still speculative. Its clinical presentation is variable, largely depending upon the site of skeletal involvement. The natural history and prognosis of this disease are unpredictable and no effective therapy is known. [18]

We must take a thorough history and perform a complete physical examination for all patients with such presentations. Other diagnoses, such as infection and cancer, must be ruled out by appropriate blood tests and radiographic studies. A definitive diagnosis must be established by performing a biopsy of the offending lesion. In recent years, most patients have been treated with surgery and/or radiation therapy. Certain patients like ours do well without surgery also but need to be regularly followed up.

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