



Case report

Gorham-Stout-Disease (GSD) of the proximal humerus: A rare cause of shoulder pain

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Abstract

Gorham-Stout Disease (GSD) also known as vanishing bone disease is an extremely rare disorder. This benign self-limiting condition may affect any group of age and may involve any part of the skeletal system. Forty cases of GSD of the shoulder have been reported in the literature with only twenty-three cases involving humerus. We hereby report the case of a 49-year-old gentleman with Gorham-Stout Disease of the humerus.

Keywords:

Gorham-Stout, Shoulder, management.

Introduction

Gorham-Stout-Disease (GSD) is a rare bone disorder characterized by progressive bone osteolysis usually associated to angiomatous proliferation of lymphatic vessels. Bone loss can be localized or spread to soft tissue and adjacent bones.

This benign condition may involve any part of the skeletal system. A central location's predilection has been reported. Its topography may include ribs, spine, pelvis, skull, clavicle, and jaw. The etiopathogenesis is still unclear. GSD may be related to uncontrolled endothelial proliferation of vascular and lymphatic capillaries within the bone [1]. The management of GSD should be case based. Many factors could interfere with the decision making. The extended functional loss may indicate surgery in appropriate candidates.

Observation

A 49-year-old gentleman without significant co-morbidities presented with the complaint of mild, gradual onset, non-radiating left shoulder pain with progressive restriction of shoulder movements for 8 months following a fall from bicycle which did not improve after symptomatic treatment. Local examination revealed major instability of left shoulder joint with loss of active movements and intact distal neurovascular status. Plain X-rays of left shoulder revealed severe osteolysis of the proximal third of the humerus and scapula (Figure 1). The MR scan showed low intensity signals on T1 and hyperintensity on T2 signifying bone loss and vascular proliferation (figure 2). Ultrasound guided biopsy of the lesion revealed angiomatous proliferation. The diagnosis of GSD was made. Shoulder replacement was indicated due to severe osteolysis, shoulder instability and movements restriction. However, the patient preferred a conservative management.



Figure 1: Plain roentgenogram (Antero-posterior view) of left shoulder showing severe osteolysis of proximal humerus with involvement of the scapula.

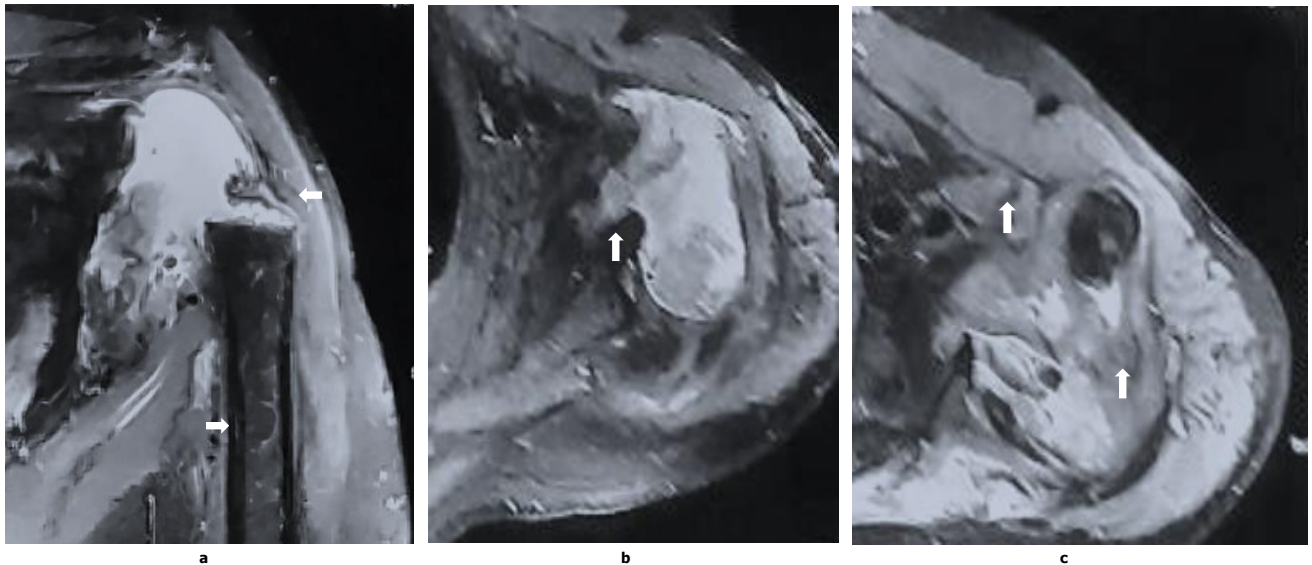


Figure 2: Left shoulder magnetic resonance imaging features
a: T2 weighted- Coronal of left shoulder showing spongy-like humerus aspect with almost complete head necrosis and diffuse hyperintensity in favor of vascular proliferation
b,c:T2 weighted- Axial showing extended necrotic process to shoulder's soft tissues

Discussion

Gorham-Stout Disease (GSD) of the shoulder is an extremely rare disease. Approximately 300 cases of GSD are reported in literature. A total of 40 cases of Shoulder GSD and only 23 cases of Humerus GSD have been reported in the literature [2,3]. Garbers et al reported a rare case of Gorham-Stout disease of both shoulders [4]. Its etiology is still unclear. The basic pathogenesis includes inappropriate activation of osteoclasts via stimulation of interleukin-6 and the receptor activator of nuclear factor-kappa B ligand (RANKL) that leads to spontaneous progressive osteolysis. The destructed osseous matrix is replaced later by fibrous tissue [5]. The diagnosis of Gorham's disease is based on clinical and radiologic features of bone loss with histological evidence of angiomatous proliferation [6]. CT scan and MR could contribute to the diagnosis. The lesion demonstrates T1 hypo-intensity and T2 hyperintensity on MRI. The vascular component of the lesion can be seen in fat suppressed T1 weighted post-contrast sequences. However, scintigraphy and positron emission tomography (PET) imaging with Fluorine-18-sodium (18F-NaF PET-CT) have higher diagnostic sensitivity and could be efficient in GSD diagnosis and treatment follow up [7]. Osteomyelitis, Paget's disease, Langerhans cell histiocytosis and hereditary multicentric osteolysis are the differentials for GSD. It is imperative to rule out malignancies before GSD diagnosis. The bone biopsy is always indicated. Shoulder GSD is a benign disease that significantly alters the patient's quality of life. The management of this entity is not standardized. The treatment is case based and nonconsensual. Aside from the pain relievers, several medications were proposed for GSD. This includes anti-resorptive drugs like bisphosphonates, angiogenesis inhibitors, low molecular weight heparin and steroids for the patients with mild to moderate symptoms. Some other molecules such as calcium, calcitonin, and vitamin D could be included in the treatment. Recently some inhibitors of the mammalian target of rapamycin (mTOR) were recommended in the GSD treatment. The results of medical therapies are variable [8].

Radiotherapy and surgery are considered for patients with severe symptoms. Recurrence rate is very low with resection and endo-prosthetic reconstruction compared to reconstruction with bone graft. Biological reconstruction combined with medications is not only effective in controlling the disease but also to halting its progression and recurrence. The prognosis depends on the lesion's severity and the location of the affected area. Mild form may remain stable for many years, while severe cases involving craniofacial and thoracic components can be associated with increased mortality [9].

Conflict of Interest: None

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