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Case Report

Gorham Disease of Distal Humerus Successfully Treated by Slight Shortening and Pin and Plate Fixation

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Abstract

Background: Gorham's disease (GD) is one of the scarce and idiopathic skeletal diseases which causes osteolysis following the proliferation of blood vessels. Progressive osteolysis GD of distal humerus with articular involvement and pathologic fracture has not been reported and our case is the first report of this disease involving distal humerus and its joints' surfaces.
Case Report: A 9-year-old boy, case of nonunion of medial condyle of humerus and pathologic fracture of distal humerus after minor trauma, was referred to our clinic and treatment started by casting but due to displacement and nonunion, we decide to operate him. Intraoperative finding was in favour of aneurysmal bone cyst (ABC) near fracture site; therefore, wide resection and fixation by medial tension band wiring (TBW), lateral plating, and fibular allograft application was done (post-operation pathologic result did not show microscopical features of this tumor) and 6 weeks later, he developed stress riser fracture above lateral plate; thus, plate removal was done and severe bone resorption was revealed. Therefore, another specimen was sent for pathology that showed hamartomatous and hemangiomatous lesion of bone. All findings were in favour of GD. He was operated another time and fixed by Persian fixation with small pins and plate, and early plate removal was done for prevention of stress riser fracture.
Conclusion: In cases of GD of distal humerus and pathologic fracture, Persian fixation is a good option for fixation and we suggest early device removal for prevention of stress riser fracture.

Keywords: Gorham Disease; Bone Resorption; Osteolysis; Humerus

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Background

Many disease may cause significant osteolysis that one of them is Gorham's disease (GD) (1). In 1838, for the first time, Jackson reported this disease (2). In 1954, Gorham et al. explained the pathological and clinical manifestation of the disease as an skeletal osteolytic process that was named "Gorham disease" (2). In GD, proliferation of blood vessels and large number of osteoclast causes bone destruction (3). Angiomatosis, rarely lymphatic proliferation, swelling of soft tissue, and loss of osteogenesis are the main features of disease (1). The pathogenesis and etiology of GD are unknown (4). Vanishing bone disease, phantom bone disease, and severe osteolysis disease are the other names of this disease (5). GD can affect any bone but generally affects shoulder and pelvis that have intramembranous ossification (6). Chemotherapy, radiotherapy, and surgery have been reported so far, but its treatment is so difficult and controversial.

According to our knowledge, more than 200 cases have been reported till now affecting the bones like skull, pelvis, ribs, foot, sternum, femur, spine, hand, radius, ulna, humerus, clavicle, and the scapula.

In this report, we describe a case of GD of distal humerus presented with pathologic fracture in a 9-year-old boy who developed stress riser fracture above plate after first surgery and then was successfully treated by slight shortening and Persian fixation (7); early device removal was done after union for prevention of stress riser fracture.

Case Report

A 9-year-old boy was referred to our clinic for non-healing supracondylar and medial condyle of humerus fracture. The patient had history of minor elbow trauma and elbow deformity without any workup and treatment 1 year prior to our visit. The patient came to our clinic with chief complaint of elbow pain after falling down. In physical exam, swelling, tenderness, varus deformity, bruise, and decreased range of motion (ROM) of elbow were revealed. Anteroposterior (AP) and lateral elbow radiography showed a new nondisplaced transverse supracondylar humerus fracture and nonunion of medial condyle of humerus with sclerotic border (Figure 1a). Initially, the patient was treated conservatively using a back slab and arm sling (Figure 1b). 6 weeks later, new X-ray of his elbow showed persistence of the fracture with displacement and noticeable bone resorption (Figure 1c).

Figure 1. Primary and 6 weeks later X-ray of elbow

Consecutively, the patient underwent surgery with posterior longitudinal incision and olecranon osteotomy. Intraoperative finding revealed cavity spaces in distal humerus filled with blood, bony resorption, and cortical thinning which was in favour of aneurysmal bone cyst (ABC) (Figure 2a). Therefore, resection of this tumor-like bony lesion, fixation by medial tension band wiring (TBW) and lateral reconstruction plate, and application of structural fibular allograft in bony defect were performed (Figure 2b and 2c).

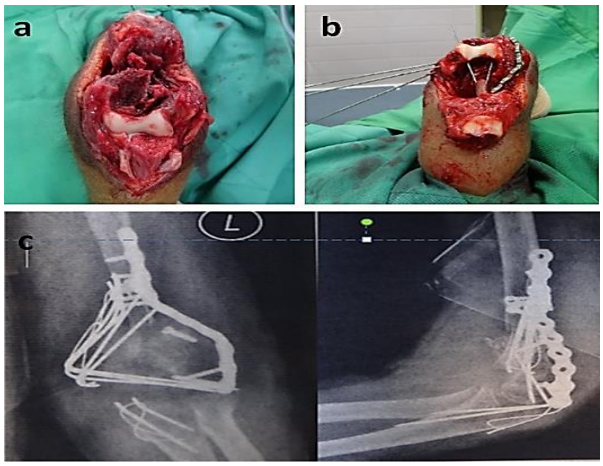

Figure 2. Intraoperative finding and fixation

Pathologic result did not show microscopical features of ABC. On 6 weeks post-operative follow-up, visit X-ray showed stress riser fracture above the plate (Figure 3a); therefore, he was reoperated through previous incision. Intraoperative findings revealed resorption of allograft and presence of cavity which was filled with blood and seroma (Figure 3b); thus, plate and all wires were removed and abnormal bone and soft tissue were sent for pathology which revealed hamartomatous and hemangiomatous lesion of bone. Metabolic laboratory tests were normal. Infectious workups such as culture, erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) all were negative. Magnetic resonance imaging (MRI) typically demonstrates increased signal on T2-weighted images with features of heterogeneity (Figure 3c).

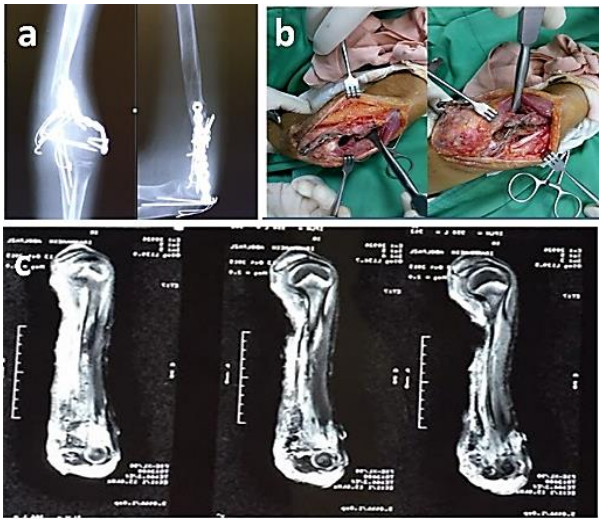

Figure 3. Stress riser fracture (a), intraoperative finding (b), magnetic resonance imaging (MRI) evaluation (c)

Post-contrast enhancement may also be heterogeneous on magnetic resonance (MR) images. Computed tomography (CT) scan revealed osteolysis of distal humerus without soft tissue involvement. All of these workups were in favour of GD as most probable diagnosis. After short period and completion of these workups, we decided to reoperate the patient for fixation through previous incision and paratricipital approach by slight shortening, bilateral TBW, and small plate as Persian fixation technique (Figure 4a). In this method, fracture fragments such as articular surface fractures were reconstructed anatomically and stabilized with Kirschner wires (K-wires) (1.2-1.5 mm). K-wires were bent at the medial and/or lateral side. Proximally, away from the fracture site, a reconstruction plate with three or four holes was placed transversely to stabilize the K-wires on the bone. This plate was fixed to the bone with two screws in a divergent manner to prevent iatrogenic fracture. This stabilization prevents pulling the wires out at the elbow motions (7).

Post-operative regular follow-ups were done and early ROM of elbow started. The fracture site was healed 3 months after operation and 4 months later, we decided to remove devices for prevention of stress riser fracture; therefore, he underwent device removal and intraoperative evaluation showed union of fracture site (Figure 4b). Now, after 2 years, the patient is pain free with arc of motion of 30-100 degrees in flexion extension and full supination pronation without any recurrence in X-ray (Figure 4c).

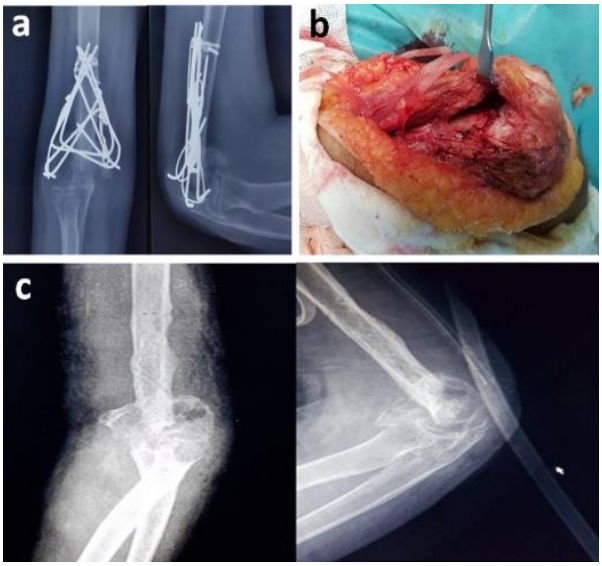

Figure 4. Persian fixation technique (a), union of fracture site (b), union without recurrence in 2 years after operation (c)

Discussion

GD can imitate conditions such as inflammation, infection, endocrine disorders, and malignant bone tumors. The diagnosis should be made after the condition has been ruled out (1). Charcot arthropathy and avascular necrosis should be part of the differential diagnosis in patients with joint involvement (8). GD is common in young adults and adolescents. However, it may occur at any age. In this disease, family history, racial prevalence, and sex are not preferable. Sometimes a history of minor trauma is found in these patients. Spontaneous fractures are common. The lesion may be present for several months

or years before a definitive diagnosis (1). Alkaline phosphatase (ALP) may be elevated but hematological and biochemical tests are often normal. MRI typically demonstrates increased signal on T2-weighted images with features of heterogeneity. Post-contrast enhancement may also be heterogeneous on MR images. Bone scan performed with technetium-99m methylene diphosphonate (Tc-99m MDP) initially shows increased radiotracer uptake; and with ongoing resorption, it becomes negative (9).

In the pathology of this disease, there are no signs of malignancy, neuropathic factors, or infection causing this disease, and only one malformation is seen in the bone tissue (10). GD is self-limiting in rare cases, but in most cases, the disease is progressive. This disease is rarely fatal and course of the disease is not predictable. Current treatment regimen usually indicates the use of medication against osteoclasts such as calcium and vitamin D supplementation and bisphosphonate use, surgery to treat tumor, and radiation therapy to prevent re-growth of tumor by inducing sclerosis. In bones such as the pubic bone, fibula, iliac bone, and clavicle, which are expandable, bone resection without reconstruction is a common treatment. In weight-bearing bones such as the femur and tibia, reconstruction using vascularized fibular graft, autografts, allografts, distraction osteogenesis, and prostheses is required (11). Radiotherapy has good definitive results in the treatment of these patients and has few long-term side effects. The use of sodium fluoride, vitamin D, calcitonin, bisphosphonates, interferon (IFN), androgens, and interleukins (ILs) as a medical treatment is recommended, but there is not enough evidence for their usefulness (1). To our knowledge, although international reports of GD are frequent, there are little case reports from developing countries that makes it an informative addition to already available literature.

Moreover, to our knowledge, GD in distal humerus and pathological joint surface fractures have not been reported so far. The case that we are considering is the first case with primary involvement of the distal humeral articular surface to be reported. Tavakoli et al. reported a case of GD of ulna with secondary involvement of radius and distal humerus with successful treatment by casting (12).

We started treatment of our patient by casting but due to fracture displacement and nonunion, involved part was resected and fixed by lateral plating, medial TBW, and applying fibular allograft, but due to pathology of bone, stress riser fracture above plate occurred; therefore, we reoperated the patient by Persian fixation technique and slight shortening, and then early removal of device for prevention of stress riser fracture was done.

We think that the risk of stress riser fracture was high in GD. Thus, in case of GD that surgeon decides to do surgery for pathologic fracture, we strongly recommend fixation techniques that reduce the risk of stress riser fracture such as long bridging plate or proximal and distal plating with overlapping to cover entire long bone, and in cases of GD of distal humerus with articular involvement, Persian fixation technique is a good option and early device removal after healing of fracture is highly suggested for prevention of stress riser fracture.

Conclusion

In cases of GD of distal humerus and pathologic fracture, Persian fixation is a good option for fixation and we suggest early device removal for prevention of stress riser fracture.

Conflict of Interest

The authors declare no conflict of interest in this study.

Acknowledgements

None.

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