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Case Report

Disappearing bone disease of the humerus and the cervico-thoracic spine: a case report with 42-year follow-up

Anne Kathleen Ganal-Antonio, MD[a,b,c], Dino Samartzis, DSc[a,*], Cora Bow, MCMSc, BHS[a], Kenneth M.C. Cheung, MBBS, MD, FRCS, FHKCOS[a], Keith D.K. Luk, MCh(Orth), FRCSE, FRCSG, FRACS, FHKAM[a], Yat-Wa Wong, MBBS, FRCSE FHKCOS, FHKAM[a,*]

[a]*Department of Orthopaedics and Traumatology, The University of Hong Kong, Professorial Block, 5th Floor, 102 Pokfulam Rd, Pokfulam, Hong Kong, SAR, China*

[b]*Ateneo School of Medicine and Public Health, Don Eugenio Lopez Sr. Medical Complex, Ortigas Avenue, Pasig City, 1604, Philippines*

[c]*Department of Orthopaedics, L1 CP Manahan Annex, Makati Medical Center, 2 Amorsolo Street, Legaspi Village, Makati City, 1229, Philippines*

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Abstract

BACKGROUND CONTEXT: Disappearing bone disease (DBD) is a rare idiopathic musculoskeletal disorder that is distinguished by bone resorption without bone formation, vascular or lymphatic vessel proliferation, and soft-tissue swelling. Long-term follow-up of a patient with DBD has rarely been reported in the literature.

PURPOSE: The following is a case report of a female patient with DBD of the humerus and the spine who was followed for 42 years, documenting the progression of the disease and outcomes.

STUDY DESIGN: Case report.

METHODS: A review of the medical records since the time of initial hospital admission throughout follow-up was performed.

RESULTS: A female patient was first seen at our institution at the age of 14. She later developed DBD of the humerus and the spine. The initial difficulty encountered was reaching the diagnosis, and later on with management of the patient as the disease progressed. The case was complicated by syrinx and arachnoid cyst formation, which caused neurologic changes leading to tetraplegia and shunt infection. The patient's inability to form a solid fusion mass led to repeated implant loosening and progressive deformity despite efforts made to stabilize both the humerus and the spine. The treatment modalities used were oral bisphosphonates, rhBMP, repeated surgeries, and instrumentation with adjunct bone graft and substitutes. At the age of 56 years, the patient died because of septicemia secondary to urinary tract infection from tetraplegia.

CONCLUSIONS: To our knowledge, this is the first report documenting a 42-year follow-up of a patient with DBD of the humerus and the spine. Our report showed that DBD greatly affects the quality of life of the patient. Close follow-up, a multidisciplinary approach, and supportive care are stressed when managing patients with DBD. © 2015 Elsevier Inc. All rights reserved.

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FDA device/drug status: Not applicable.

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The disclosure key can be found on the Table of Contents and at www.TheSpineJournalOnline.com.

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* Corresponding author. Department of Orthopaedics and Traumatology, The University of Hong Kong, Professorial Block, 5th Floor, 102 Pokfulam Rd, Pokfulam, Hong Kong, SAR, China. Tel.: (+852) 2255 4254; fax: (+852) 2817 4392.

E-mail address: yatwa@hku.hk (Y.-W. Wong); dsamartzis@msn.com (D. Samartzis)

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Introduction

Disappearing bone disease (DBD) was first defined by Gorham and Stout in 1955 [1–3]. Its etiology remains unknown and its pathophysiology is not fully understood. Thus, the diagnosis is made out of exclusion [4,5]. There is no gender or race predilection [6,7]. Both axial and appendicular skeleton can be affected, with the skull, clavicle, scapula, humerus, ribs, and pelvis being the most frequently involved sites [3,4,7,8].

Disappearing bone disease should be differentiated from other forms of osteolysis [3,5,7,9]. Angiomatosis is similar to DBD in that it is also angiomatous in origin with unknown etiologies. Furthermore, angiomatosis and DBD are similar in pattern of spread, course, and prognosis. However, DBD is non-familial and is usually monocentric (ie, affecting one region) [9,10]. In DBD, bone is replaced with fibrous tissue, with non-healing fractures and eventual bone resorption during active disease states [9]. Spontaneous resolution has been reported, although the clinical course is unpredictable, depending on the site and extent of involvement [4,6,7,9].

Approximately 200 cases of DBD have been reported in the literature; however, the precise estimate is challenging to determine because this condition has been called by other names, such as Gorham-Stout disease, Gorham syndrome, progressive massive osteolysis, phantom bone, hemangiomatosis, and lymphangiomatosis [5–7]. Nonetheless, all these names pertain to an idiopathic and rare disease that has spontaneous and progressive bone resorption, which is not followed by bone production and without malignant proliferation of vascular structures [6,7].

Long-term follow-up with multicentric involvement of DBD has rarely been reported. As such, we report a case with multicentric DBD involving the humerus and cervico-thoracic spine, documenting the 42-year follow-up, natural course, and outcome of the patient.

Case report

The following is a case report of a 56-year-old female who was first seen at our institution (Department of Orthopaedics and Traumatology, The University of Hong Kong) at the age of 14 years for right upper limb shortening, recurrent swelling, and pain. She had limited shoulder range but had good upper limb function, mostly compensated by the elbow (Fig. 1). Her medical history started at the age of 5, when she sustained a closed injury on her right humerus. The initial impression was chronic osteomyelitis, which was treated with radial drilling with a 3.5-cm shortening of her right arm. Biopsy results yielded sclerotic bone and culture results were negative.

Nine years later at the age of 25, the patient complained of right arm pain. Laboratory parameters all remained normal, although she had instances of mildly elevated alkaline phosphatase. Radiographs showed a pathologic fracture of the right humerus, and the patient was treated by iliac crest bone grafting and a shoulder spica. Grossly, no signs of infection were seen, but instead a small fibrous reaction was observed between the fracture edges. Cultures were negative, and histopathology showed cortical lamellar bone, covered with dense fibrous tissue with marrow spaces showing fibrosis and increased vascularity, and abundant collagenous tissue showing vascular proliferation. No malignancy or fibrous dysplasia was noted. On monitoring, the fracture edges remained atrophic. At 26 years old, the patient received an artificial bone graft with a custom-made hydroxyapatite cylinder to bridge the gap, and subsequently underwent iliac crest bone grafting. Gradual resorption of the bone graft led to pseudo-arthrosis (Fig. 2). Histopathology showed cortical and spongy bone with dense fibrous tissue, highly vascular tissue, and abundant capillaries without inflammatory infiltrates or dead bone, consistent with DBD. Additional imaging showed T10–T12 and L2 involvement.

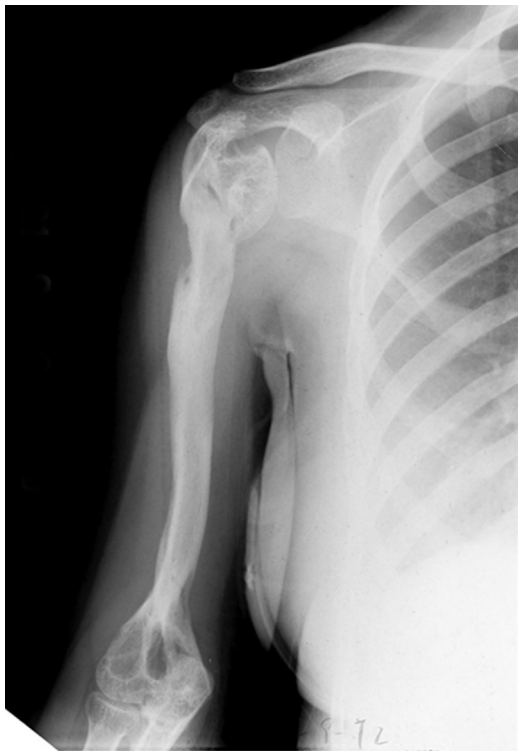

Fig. 1. Anteroposterior plain radiograph of the right humerus at initial presentation of the patient at age 14 years.

At 28 years old, the patient had a vascularized fibular graft placed over the humeral bone gap. There was gradual graft resorption, despite reexploration of the arterial anastomosis and scintigraphy showing a viable graft. In 2 years, the fibular graft was completely resorbed, with the whole length of the humerus remaining dystrophic and thin (Fig. 3). The patient lost all shoulder movement; however, elbow motion compensated quite well. At this point, she was able to adapt to her activities and was able to work as a clerk. She remained right-handed and was able to write, but used

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