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Gorham-Stout syndrome: a case report

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Abstract. – Gorham-Stout syndrome, is an extremely rare disease of the bone, characterized for vascular and lymphatic channels proliferation in bony segments and consequent osseous resorption. There are around 200 cases reported around the world. Although bisphosphonates are used for symptoms relief, there is no standardized treatment established. We present a case that was diagnosed in our centre secondary to a resistant epistaxis and a literature review of this condition.

Key Words:
Gorham Stout, Vanishing bone, Disappearing bone disease.

Introduction

First reported by Jackson in 1838 and then described by Gorham and Stout in 1955, also known as Vanishing or Phantome bone disease[1]. There is a spontaneous progressive resorption of bone with (no malignant) vascular proliferation structures. The diagnosis is one of exclusion and it is based on combined histological, radiological, and clinical features[2]. Its etiology and molecular mechanism is still unknown. Bone matrix is destroyed as vascular and/or lymphatic vessels grow in, the bone marrow became fatty and the trabeculae became thinner. X-rays imaging demonstrates progressive osteolysis with resorption and cortical loss[3]. Patients with Gorham's disease may complain of dull aching pain or insidious onset of progressive weakness. In some cases, pathologic fracture often leads to its discovery[4]. It is a progressive disease but rarely fatal, the most common sequelae are those from fractures; chylothorax is probably the most serious complication that can lead to death.

Case Report

A fifteen year old girl, with no previous symptoms, is admitted in our centre in hemorrhagic shock due to massive epistaxis. Blood tests results were normal apart from a low red cell count, hemorrhagic diathesis as clotting defects were ruled out. Bleeding was stopped with an anterior and posterior nasal packing. Imaging studies demonstrated multiple osteolytic lesions in skull (Figure 1), long bones and spine (Figures 2, 3), but no vascular malformations susceptible to embolization were found. There was no evidence from pleural effusions or mediastinum congestion (Figure 4). Orthopaedic and spine division were consulted in order to take a bone biopsy and evaluate vertebral lesions. The bone samples were taken from tibia and fibula with a Jamshidi type needle under general inhalator anaesthesia. Spine was considered stable and with no fracture risk at the moment. Anatomical pathology results defined diminished bone matrix with typical vascular structures, patterns compatible to Gorham stout disease. After 2 years of follow-up with no treatment at all no further complications were registered.

Discussion

The Gorham disease is a non hereditary condition with equal prevalence in males and females, characterized for venous and lymphatic malformations in skin, mediastinum and bones[5]. The presence of abundant, leaky systemic lymphatic vessels is often accompanied by chylous ascites. There is no standardized treatment available for Gorham's disease, and its molecular mechanisms remain unclear[6,7]. Epistaxis is a complication due to affected bones in the skull base or face. The medical treatment for Gorham's disease includes radiation therapy, anti-osteoclastic medications (bisphosphonates), and alpha-2b interferon[2,7].

Conclusions

Gorham stout syndrome is a rare condition in wich abnormal venous and lymphatic channels

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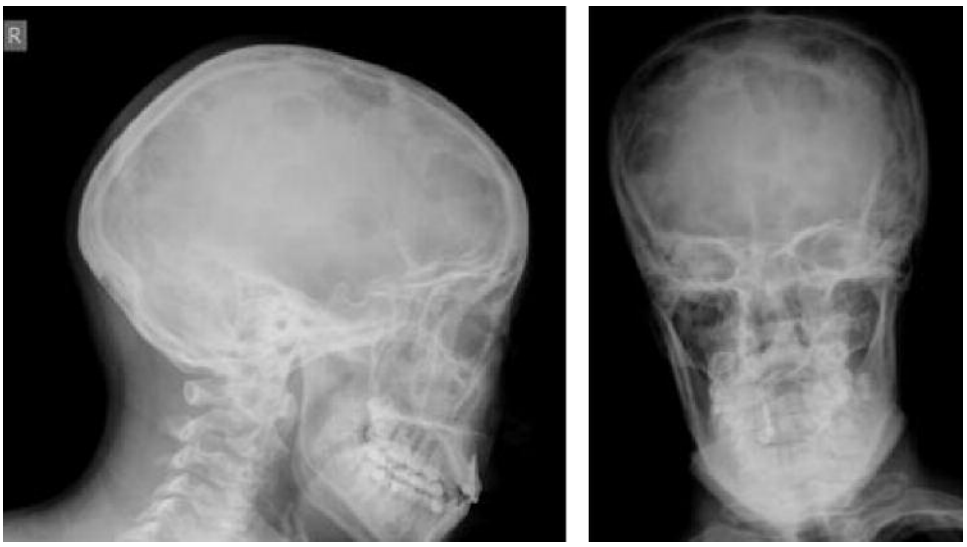

Figure 1. Patient skull X-Rays with the typical osteolysis images.

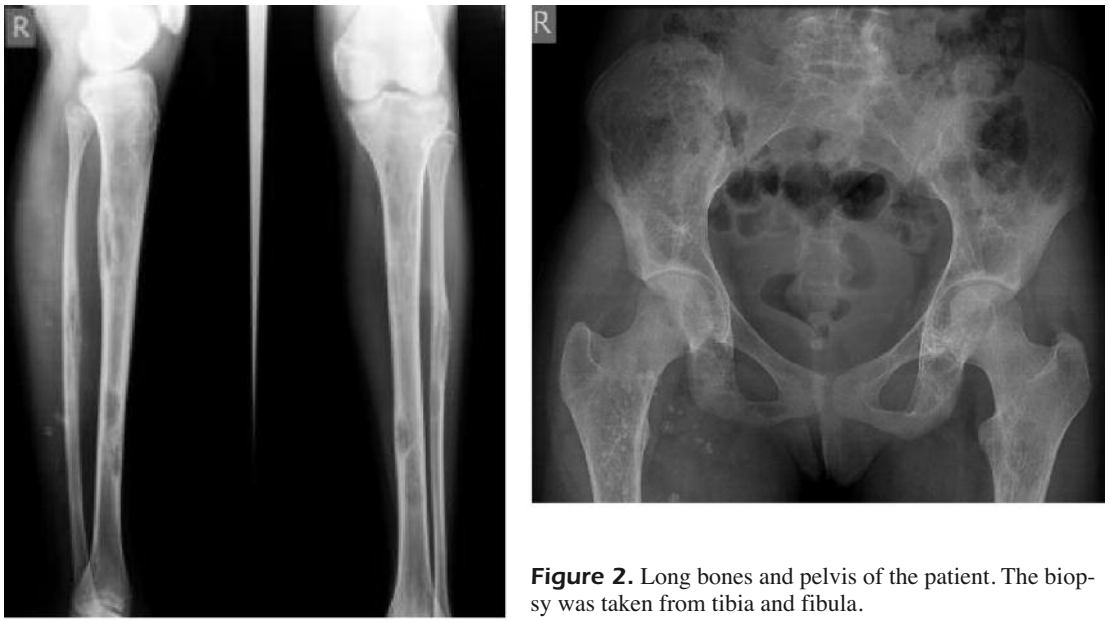

Figure 2. Long bones and pelvis of the patient. The biopsy was taken from tibia and fibula.

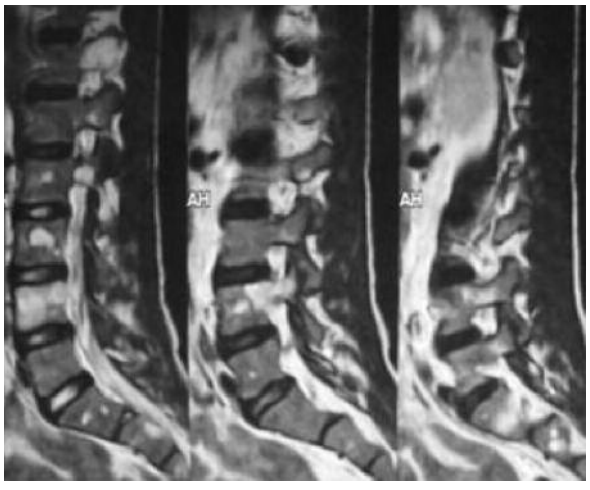

Figure 3. MRI of the spine demonstrates extended lesions, but no instability or compression of the neural elements were found.

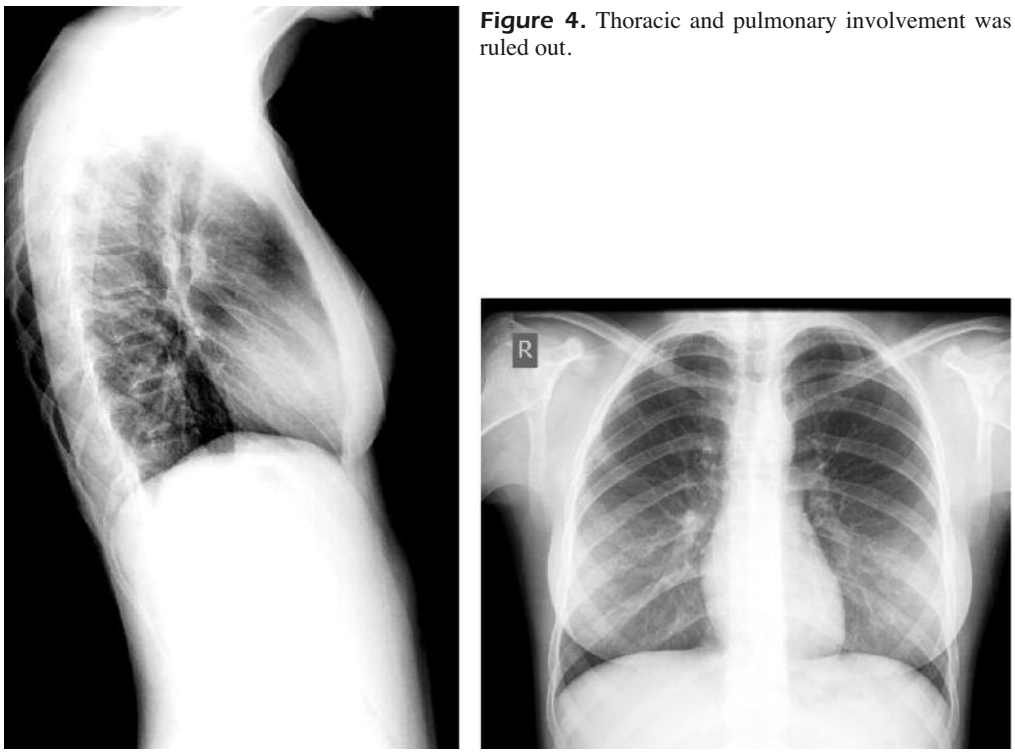

Figure 4. Thoracic and pulmonary involvement was ruled out.

grow into the bone making it disappear, and in some cases causing leakage of blood, lymphatic liquid and even cerebrospinal fluid[8] into cavities. Because its etiology and mechanism remains unclear, treatment is symptomatic and surgical options must be reserved for fractures or unstable spine, bone graft tend to be reabsorb in this affection. Chylotorax in some cases requires ligature of the thoracic duct. Further series must be analyzed to establish treatment guidelines on prevention and complications[9,10].

Conflict of Interest

All authors have disclosed that they have no financial relationships with, or financial interests in, any commercial or pharmaceutical companies pertaining to this educational activity.

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