CASE REPORT

Gorham's Disease [Vanishing Bone Disease] - A case report

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ABSTRACT

Gorham's disease is a very rare disorder characterized by uncontrolled destructive proliferation of vascular or lymphatic capillaries within bone and the surrounding soft tissues. Gorham's disease is characterized by proliferation of vascular channels that result in destruction and resorption of osseous matrix. Since the initial description of the disease, fifty years have elapsed but still the precise etiology of Gorham's disease remains poorly understood and largely unknown.

Key Words: Gorham's disease; massive osteolysis; bone resorption

INTRODUCTION

Gorham's disease is characterized by proliferation of vascular channels that result in destruction and resorption of osseous matrix. There is no evidence of a malignant, neuropathic, or infectious component involved in the causation of this disorder. The mechanism of bone resorption is unclear.[1]

Gorham's disease or vanishing bone disease is an uncommon and unusual disease characterized by progressive osteolysis of bone with ultimate total disappearance of bone. It was first described by L. W Gorham in a patient with massive osteolysis and subsequent resorption of the clavicle.[2]

This disease results total radiological disappearance of the involved bone which helps in distinguishing of the condition. Clinical manifestations are usually related to the area of involvement. When the lower or upper jaw, tooth sockets, or other bones in the face, neck or head are affected, symptoms may include pain, loose teeth, fracture, facial deformity, and recurrent meningitis.[3]

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The clinical presentation of Gorham's disease is variable and depends on the site of involvement. It often takes months or years before the offending lesion is correctly diagnosed. Patients with Gorham's disease may complain of dull aching pain or insidious onset of progressive weakness. Pathological fracture may be a diagnostic feature. Gorham's disease may be progressive in most patients. However, in some cases, the disease may be self limiting. The clinical course is protracted but rarely fatal, with the eventual stabilization of the affected bone being the most common sequence. The medical treatment for Gorham's disease includes radiation therapy, anti-osteoclastic medications (bisphosphonates), and alpha-2b interferon. Surgical treatment options include resection of the lesion and reconstruction using bone grafts and/or prostheses[1].

CASE REPORT

A 17 year old female patient was referred to our Department of Oral Medicine and Radiology with complaint of pain and swelling in the lower left posterior teeth region. The patient stated that she was asymptomatic 6 months back, after which she started feeling pain and complained of swelling in the same region which appeared later and gradually increased to attain the present size.

Extra-oral examination showed facial asymmetry due to deformity in the lower left posterior mandible. There was depression seen on the left pre-auricular region with deviation of the mandible on the left side on opening of the mouth. The inferior border of the left mandible was not palpable. (Parasymphyseal region and angle of mandible).The overlying skin was normal and sub-mental lymph nodes were palpable and enlarged.

Intra-oral examination revealed segmental mobility of the left mandibular 36, 37 and 38 regions. Step-defect was seen between 35 and 36 region. The ascending ramus of the mandible was not palpable.36, 37 and 38 were displaced and found below the occlusal plane. The oral hygiene of the patient was poor with a lot of supra-gingival calculus (Fig 1).

Orthopantomogram showed mesial drifting of 35-38 parallel to the inferior border of mandible. However there was complete resorption of ramus. Root apices of 35, 36, 37, and 38 were in approximation to the inferior border of mandible, 37 and 38 showed root resorption (Fig 4) C T SCAN also revealed

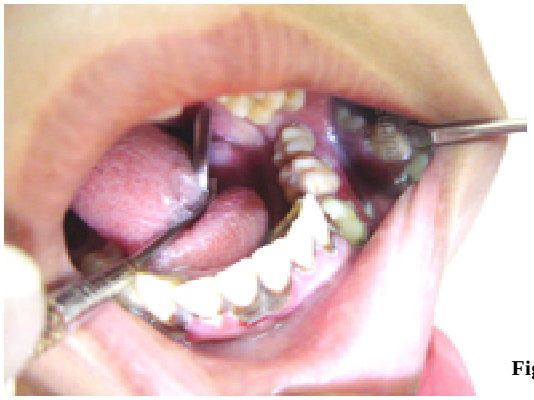
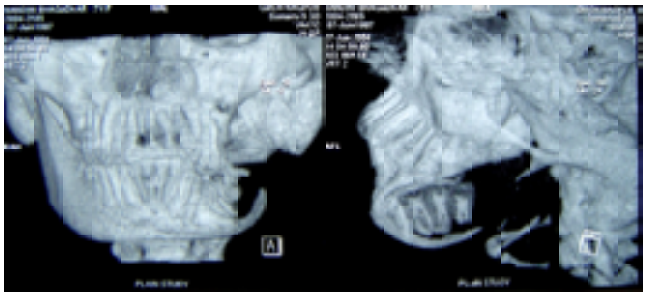


Fig. 1

complete destruction of body and ramus of left side of mandible only small part of condylar process was spared which was displaced into anterior infra-temporal fossa. CT scan also showed lytic expansile destructive lesion of left hemi-mandible with cortical destruction and associated soft tissue swelling with loose teeth in the matrix of lesion (Fig 2).



The standard laboratory blood tests were usually within normal limits. Serum Calcium, Serum Phosphorus, Serum alkaline phosphatase and Parathyroid hormone levels were within normal range.

Intraoral biopsy revealed a highly vascular connective tissue stroma with numerous dilated and engorged thin walled blood vessels. Dense chronic inflammatory cell infiltrate mainly composed of lymphocytes were seen interspersed between the loosely arranged collagen fiber bundles (Fig 3).

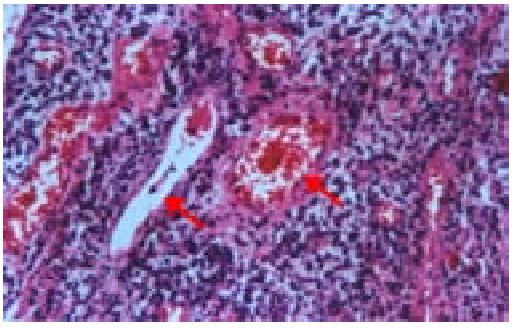


Fig. 3

The patient was referred to Departments of Oral Surgery, Interventional Radiology & Prosthodontics for further treatment. A treatment of surgery combined with the use of Bio- phosphonates was planned to arrest the osteolysis

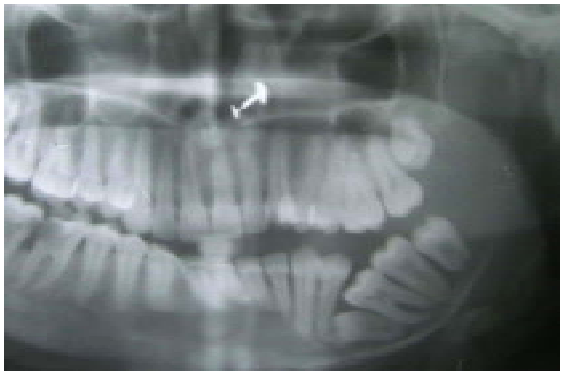


Fig. 4

process. Prosthetic rehabilitation of the left side of mandible was planned thereafter.

DISSCUSSION

Gorham's disease or massive osteolysis is a very rare disorder characterized by spontaneous and progressive osteolysis of one or more skeletal bones. The radiographical findings associated with Gorham's disease are particularly dramatic as in some cases a complete resorption of the involved bone can occur leading to the definition of phantom bone, vanishing bone or disappearing bone disease.[4]

Some of the Eponyms of Gorhams disease are, Gorham's Syndrome, Gorham-Stout Syndrome, Morbus Gorham-Stout Disease, Massive Osteolysis, Idiopathic Massive Osteolysis, Progressive Massive Osteolysis, Massive Gorham Osteolysis, Disappearing Bone Disease, Vanishing Bone Disease, Phantom Bone Disease.[1]

Gorham's disease is relentlessly progressive & involvement of vital structures can occur leading to high morbidity and mortality. Extension of the disease from the scapula, ribs or thoracic vertebra can result in pericardial and pleural effusions and Chylohtorax[1]. Chylothorax and vertebral disease are uncommon but usually fatal in Gorham's disease.[5]

To date, the etiology and pathophysiology of this poorly understood disease remains undetermined.[1,3,4,6] The pathological process is the replacement of normal bone by an aggressively expanding but non-neoplastic vascular tissue, similar to a hemangioma or lymphangioma. Wildly proliferating neovascular tissue causes massive bone loss. In the early stage of the lesion, the bone undergoes resorption, and is replaced by hypervascular fibrous connective tissue and angiomatous tissue. Histologically, involved bones show a non-malignant proliferation of thin-walled vessels; the proliferative vessels may be capillary, sinusoidal or cavernous.The stimulus that generates this change is unknown.[1]

It is accomplished; the progressive osteolysis is always

associated with an angiomatosis of blood and sometimes of lymphatic vessels, which are seemingly responsible for it.

A biopsy from the involved area, laboratory investigations, as well as imaging techniques should be performed to find a potential cause of the osteolytic process. Accordingly, definitive diagnosis of Gorham's disease is a diagnosis of exclusion, after having failed in detecting serum biochemical abnormalities, and with microscopic findings showing a non-specific vascular fibrous connective tissue containing foci of lymphocytes, plasma cells and active fibroblasts.[3]

The mandible is one of the most common bones to be involved and its invasion undergoes complete or partial dissolution and spreads across the joints to contiguous bones.[7] Mandibular involvement is considered a potential high-risk disease location, as severe progression from mandible to maxilla, skull, and spine has been reported (Jackson 1838), thus leading to poor prognosis.

Gorham's Syndrome is a combined clinical, radiological and histological entity. Radiologic findings are especially important in the diagnosis of Gorham's Syndrome.[8]

Physicians must take a thorough history and perform a complete physical examination for all patients who present with osteolysis of the shoulder or pelvic girdle, long bones, or vertebrae. 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. The diagnosis of Gorham's disease should be made only after carefully eliminating the aforementioned causes of osteolysis.[1]

DIFFERENTIAL DIAGNOSIS OF OSTEOLYSIS SYNDROMES[1]

Acro-osteolysis of Hajdu and Cheney	Idiopathic multicentric osteolysis (carpal-tarsal osteolysis)
Multicentric osteolysis with nephropathy	Hereditary multicentric osteolysis
Neurogenic osteolysis	Acro-osteolysis of Joseph
Acro-osteolysis of Shinz	Farber's disease
Winchester's syndrome	Massive osteolysis (Gorham's disease)

The medical treatment for Gorham's disease includes radiation therapy anti-osteoclastic medications (lipophosphates) and Alpha 2B Interferon. Surgical treatment, options include resection of the lesion and reconstruction using bone grafts and prosthesis. Various treatment options including pleurectomypleroclasis, thoracic duct ligation, radiation therapy, interferon therapy & bleomycin have been used for the management of patients with Gorham's disease presenting with Chylothorax. The treatment of either Gorham–Stout syndrome or chylothorax is still a dilemma. In general no single treatment modality has proven effective in arresting the disease.[1]

Moller *et al.*[6] reported six cases of massive osteolysis and found histologically increased number of stimulated osteoclasts, they suggested early potent antiresorptive therapy such as with calcitonin or biophosphonates may prevent local progressive osteolysis. The use fo Zoledronic acid, a nitrogen containing high potency biophosphonate was effectively used by Mignona M D.

The principal treatment modalities are surgery and radiation therapy. Surgical options include resection of the lesion, and reconstruction using bone grafts and/or prostheses. The prognosis for patients with Gorham's disease is generally good unless vital structures are involved.[1] Early use of radiation therapy may arrest endothelial cell proliferation and thereby limit the spread of the disease and may avoid relatively complicated surgeries later on.[4]

The role of oral physician is extremely important in diagnosing promptly the disorder and preventing the functional and esthetic consequences of advanced and extensive bone loss. Gorham's disease should be included among the pathological entities mimicking periodontal disease on radiographs, such as inflammatory disease (e.g. osteomyelitis), endocrine disease e.g. hyperparathyroidism), intra-osseous malignancies or (eosinophilic granuloma, histiocytosis X, metastases etc), infective processes (e.g. tuberculosis).

Because of the rarity of the disease, uncertain origin and number of diseases having similar investigatory findings it is the need of Oral physicians to include Gorham's disease as pathologic entity.

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