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內文：

Introduction

I. **Vanishing bone disease**, or **Gorham disease**, was first defined as a specific entity by **Gorham and Stout** in **1955**; it is a rare disorder characterized by proliferation of vascular channels, which results in destruction and resorption of osseous matrix.

II. Only a few cases have been reported in the jaws. **Vanishing bone disease** or **massive osteolysis** of the lower jaw will **initially** affect the **mandibular basal and alveolar bone**, which **subsequently** involves the **rami and the condyles**.

III. The etiology remains unknown, the prognosis is unpredictable, and effective therapy has still not been determined.

IV. Numerous names have been used in the literature to describe this condition, such as **phantom bone**, **disappearing** or **vanishing bone** disease, **acute spontaneous absorption of bone**, **hemangiomatosis**, **lymphangiomatosis**, **idiopathic osteolysis**, and **Gorham disease**.

V. **Idiopathic osteolysis** is characterized by the spontaneous and progressive destruction of one or more of the skeletal bones.

VI. Idiopathic osteolysis was described first in ***1838*** and again in ***1872*** by ***Jackson***, who reported a case of a ***"boneless arm."*** ***Romer*** reported the first case in the ***jaws in 1924, in a 31-year-old woman***.

VII. In 1954, Gorham reported on 2 patients with **massive osteolysis** of the bone. One was a *boy, aged 16 years*, with right clavicle and scapula involvement. ***Chylothorax*** eventually developed, and the patient ***died***. The other patient was a *man, aged 44 years*, who also had involvement of the right clavicle and scapula.

VIII. Gorham disease is usually associated with **angiomatosis** of blood

vessels and sometimes of **lymphatic** vessels.

IX. Any bone can be affected, although there is a predilection for the pelvis, humerus, axial skeleton, and mandible.

Etiopathology

- Wildly proliferating neovascular tissue causes massive bone loss, but non-neoplastic vascular tissue, similar to a hemangioma or lymphangioma.
- **In early stage**, bone is replaced by an abundance of **thin-walled capillary-sized** vascular channels and, at a **later stage**, by **fibrous connective tissue**.
- One main structural feature of the lesion is the **presence of unusually wide capillary-like vessels**, and therefore it is likely that the **blood flow** through these vessels is **slow**. It has been suggested that the slow circulation produces ***local hypoxia and lowering of the pH***, favoring the activity of various hydrolytic enzymes.
- ***Gorham and Stout*** reported that active hyperemia, changes in local pH, and *mechanical forces* promote bone resorption. They hypothesized that trauma may trigger the process by stimulating the production of vascular granulation tissue and that "***osteoclastosis" is not necessary***.
- In contrast, ***Devlin*** have suggested that bone resorption in Gorham disease is due to enhanced osteoclast activity and IL-6 may play a role.
- ***Moller*** reported 6 cases of Gorham-Stout syndrome with histopathologic findings and presented evidence that osteolysis is due to an increased number of ***stimulated osteoclasts***.
- ***Hirayama*** has also been suggested ***thyroid C cells and calcitonin*** may play an important role in the pathogenesis of Gorham disease
- The disease can be monostotic or polyostotic, although multicentric involvement is exceptional.
- **No ethnic or gender** predilection has been noted.
- The disease appears to be **nonhereditary** and is **most common in children and young adults**.
- In 30% of cases maxillofacial involvement is seen with pain, malocclusion, and deformity.

Clinical Features

- Most cases occur in children or in adults aged less than 40 years.
- However, the disease has been described in patients aged as young as 1 month to as old as 75 years.
- Approximately 60% of all cases occur in men.
- More than 200 cases have been reported in the literature. The process may affect the appendicular or axial skeleton. Cases have been reported in the skull (8 cases), maxillofacial region (42 cases), spine (18 cases), pelvis (18 cases), trunk (including clavicle and ribs) (35 cases), upper extremity (including scapula) (41 cases), and lower extremity (22 cases), in addition to multicentric involvement (11 cases).
- The mandible was affected alone by the osteolysis, partially or completely, in 23 cases, whereas the maxilla was never involved alone.
- Some patients present with a relatively abrupt onset of pain and swelling in the affected area, whereas others present with a history of *insidious onset of pain* in the involved jaw.
- The medical, personal, and family histories are usually noncontributory.

Investigations

- The standard laboratory blood tests are usually within normal limits and are not helpful. The serum *alkaline phosphatase* level may be slightly elevated.
- Plain radiographs, radioisotope bone scans, CT, and MRI must been used.
- There will be resorption and decreased vertical height in panoramic radiograph.

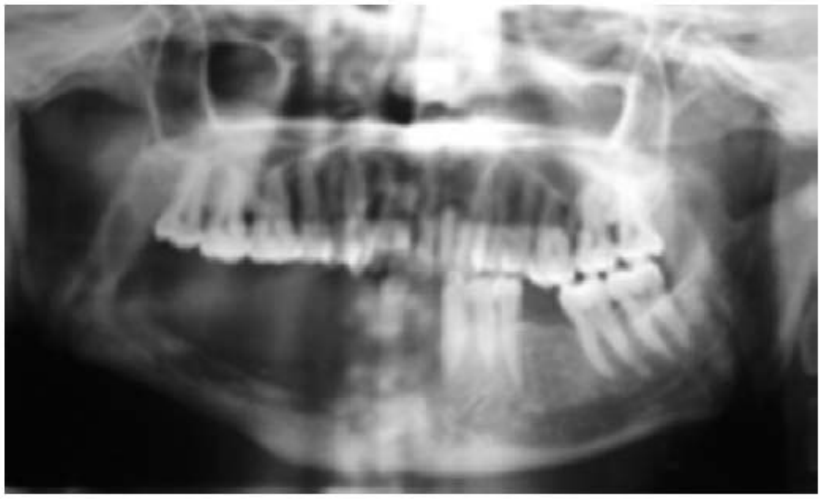


FIGURE 1. Panoramic radiograph showing missing teeth in right quadrant, resorption, and decreased vertical height of right mandibular body with bone resorption extending to mesial aspect of left canine.[31]

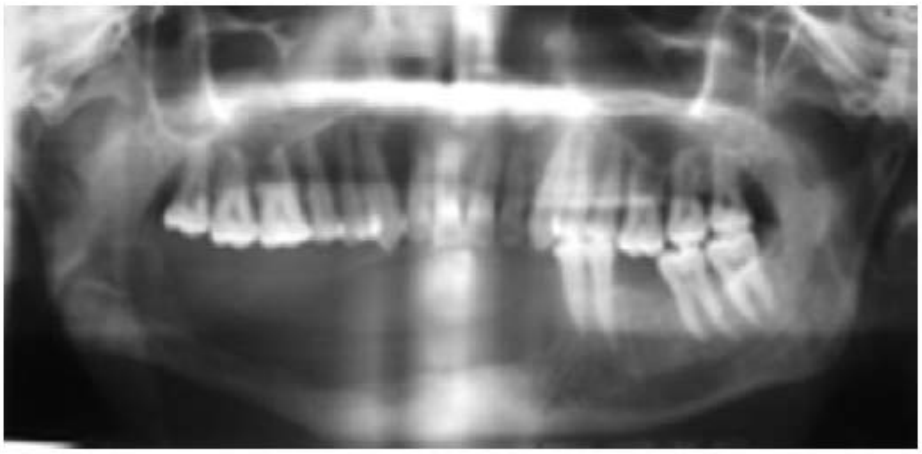


FIGURE 2. Panoramic radiograph taken 3 months after initial panoramic radiograph showing progressive resorption of right mandibular body and loss of bone, encroaching on mandibular canal space.[31]

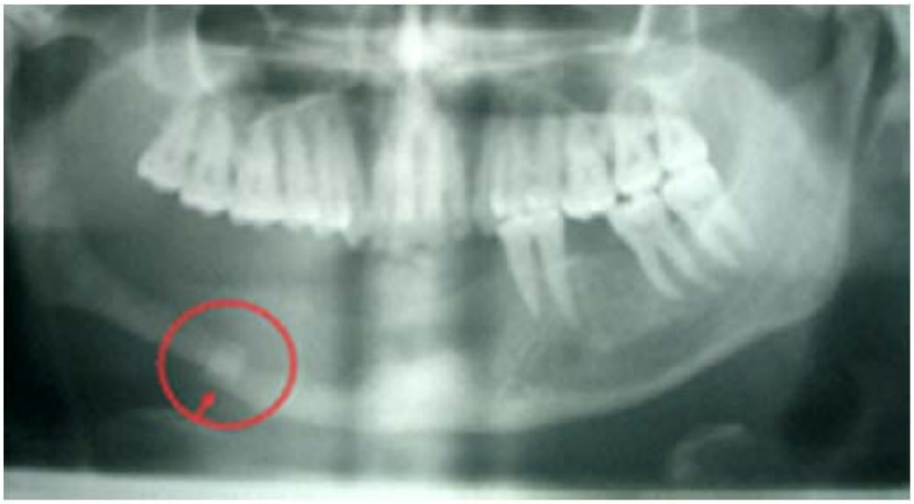
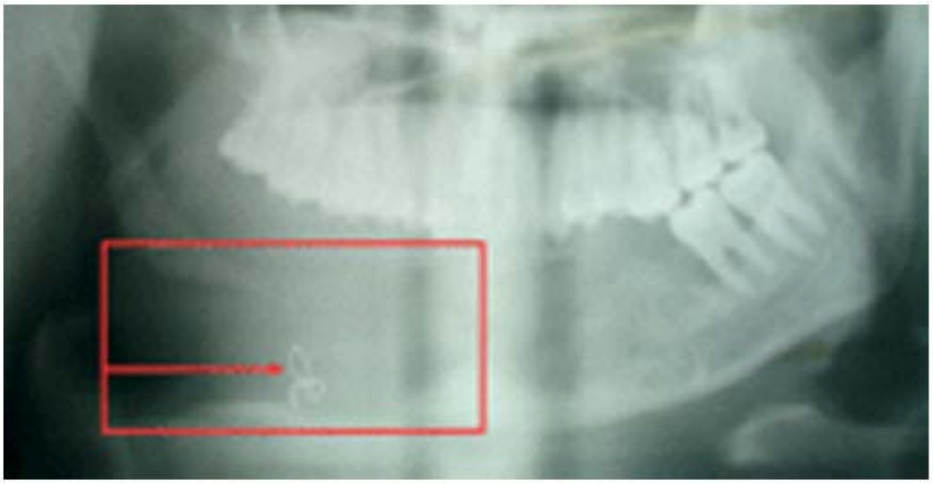


FIGURE 3. Panoramic radiograph taken 4 months after initial panoramic radiograph, with an *arrow* pointing toward fracture in right mandibular body with overriding fragments and osteolysis extending onto angle and ramus with resorption of coronoid process.[31]



- T1-weighted spin echo MRI shows uniformly low signal intensity in the involved bones, whereas an increased signal intensity generally is observed on T2-weighted spin echo images.

Differential Diagnosis

- Differential Diagnosis is needed to rule out other common underlying causes of osteolysis, such as infection, cancer, and inflammatory or endocrine disorders.
- D.D：(can be confirmed with biopsy)
  - ☑ Aneurysmal bone cyst
  - ☑ Extensive metastatic bone disease due to carcinomaof the breast
  - ☑ Osteosarcoma

Treatment

- There is no standard therapy.
- Non-OP ： radiation therapy, anti-osteoclastic medication (bisphosphonates), and interferon alfa-2b
  OP：surgical resection and reconstruction by use of a bone graft or prosthesis.
  ➔ The success rate after the use of a bone graft is low.(dissolution)
- A moderate doses (40-45 Gy in 2-Gy fractions) appears to result in a good clinical outcome
- In children and adolescents who receive high-dose RT,exists：
  - ☑ Potential for secondary malignancy
  - ☑ Growth restriction
- The prognosis of Gorham disease is generally good unless vital structures are involved.

Discussion

- The term used by Gorham and Stout was “haemangiomatosis,” implying:
  - ■ This connective tissue contains many thin-walled vessels, sometimes with red blood cells. In other areas, freely anastomosing vascular spaces lined by endothelial cells can be seen. The fatty marrow also contains some dilated blood vessels.

- (1958)Johnson and McClure: Frequency as follows →
  - ☑ clavicle, scapula, proximal end of humerus, ribs, iliac bone, ischium, and sacrum.
  - ☑ The disease has not been observed in the calvaria or in the distal bones of the extremities.
- In recent years, most patients have been treated with surgery and/or radiation therapy
- Heffez suggested 8 criteria for definitive diagnosis of massive osteolysis:

1. Positive biopsy findings in terms of angiomatous tissue presence
2. Absence of cellular atypia
3. Minimal or no osteoclastic response and absence of dystrophic calcifications
4. Evidence of local bone progressive resorption
5. Non-expansive, nonulcerative lesion
6. Absence of visceral involvement
7. Osteolytic radiographic pattern
8. Negative hereditary, metabolic, neoplastic, immunologic, and infectious etiology

題號	題目
1	Massive osteolysis is primarily related to a proliferation of blood or lymphatic vessels and has been termed _____ of bone (A) Resorption (B) Destruction (C) Angiomatosis (D) Osteosclerosis
答案 (C)	出處： Oral and Maxillofacial Pathology , 3rd edition , p622
題號	題目
2	What kind of treatment is the most successful and widely accepted mode of therapy? (A) Radiation therapy (B) Surgical resection (C) Chemotherapy (D) No further treatment, but keep follow up closely
答案 (A)	出處：Oral and Maxillofacial Pathology , 3rd edition , p623