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內文：

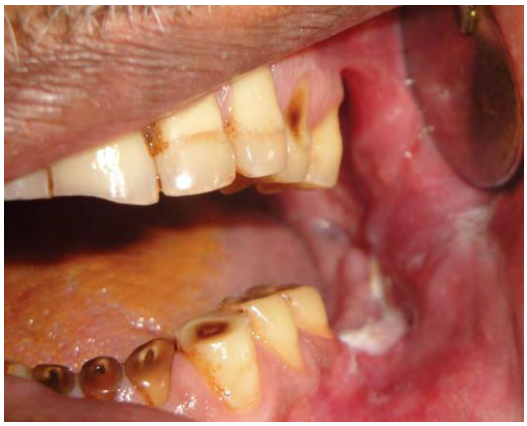
Abstract

1. Gorham's disease is a rare condition characterized by progressive osteolysis of bone with ultimate total disappearance of bone.
2. The etiology is undetermined and may affect any bone of the body, although there is predilection for the pelvis, humerus, axial skeleton and the mandible.
3. Due to the rarity of the disorder it often goes unrecognized and this leads to different treatment modalities(often with limited success)
4. This is a case of Gorham's disease of the mandible in a 62-year-old man and the literature is reviewed with emphasis on etiology and diagnosis management

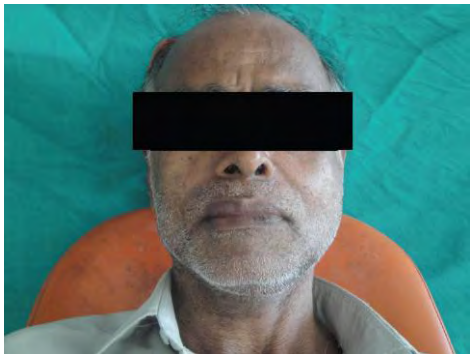
Introduction

1. Gorham's Disease (GD) is a rare disorder characterized by destructive proliferation of vascular channels within the bone and the surrounding soft tissues.
2. The first report of a case of boneless arm was in 1838 and has had different names over the course of history: phantom bone, disappearing bone disease, acute spontaneous absorption of bone, hemangiomatosis, lymphangiomatosis, idiopathic osteolysis, and Gorham's disease
3. The condition may affect any part of the skeleton, but most commonly involves the skull, shoulder, and pelvic girdle. About 50 cases of Gorham's disease involving the maxillofacial region are reported.

Case Report



Intraoral examination – missing molars

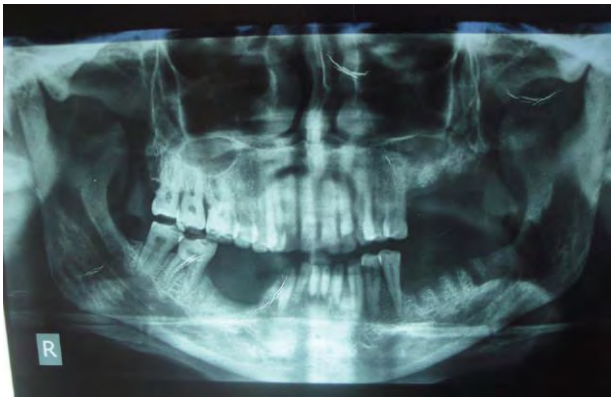


Extraoral exam – mild diffuse swelling of left lower face

1. A 62-year-old male patient visited OPD with chief complaint of mild discomfort and slight swelling in relation to lower left side of face.
2. Patient gave history of mobile tooth which was extracted 1 month previously.
3. Extra oral examination revealed mild diffuse swelling on lower left side of jaw. The overlying skin was normal. Intraoral examination revealed partially

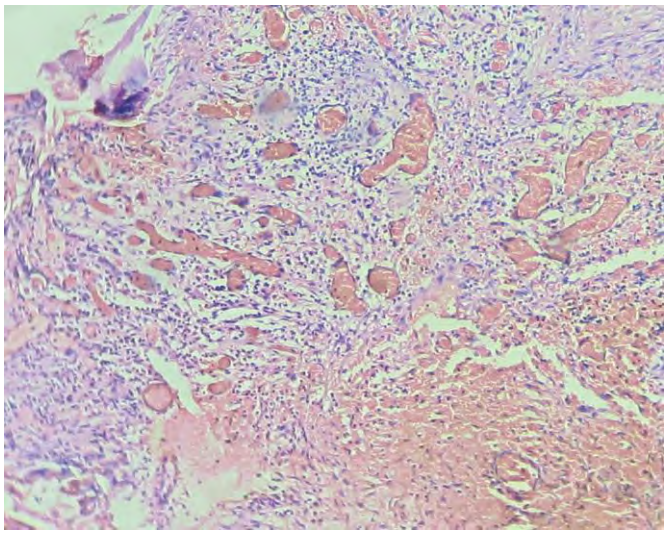
edentulous left mandible with missing molars which was extracted because of mobility.

4. Panoramic X-ray revealed an extensive ill-defined osteolytic lesion of body of mandible involving angle of mandible and extending up the ramus.
5. Provisional diagnosis of intraosseous malignant neoplasm was made. Differential diagnosis included metastatic tumour, metabolic disease, and osteomyelitis.



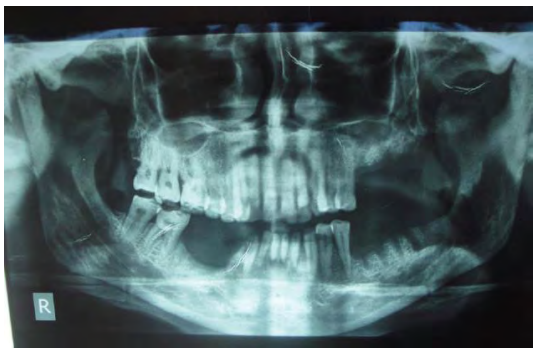
Pano – ill-defined osteolytic region involving body and ascending ramus of mandible

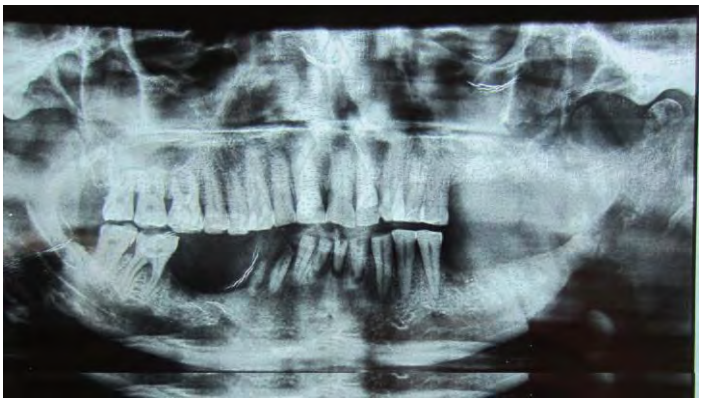
6. The laboratory investigation ruled out metabolic disease as his hematological levels were found to be within normal range.
7. Medical and familial histories were non-contributory
8. Curettage of affected part was undertaken and the intraoperative finding was complete absence of bone. The specimen was sent for histopathological examination.
9. Histopathological examination revealed fibrous connective tissue with numerous thin walled vascular spaces and minimal chronic inflammatory infiltrate with no cellular atypia seen. The histopathological feature was suggestive of an angiomatous lesion



Fibrous connective tissue showing numerous thin walled vascular spaces (H&E 10x).

10. Patient was lost to follow-up for 3 months and returned with complaint of mild discomfort and another Panoramic X-ray was advised





3 Month follow-up pano – osteolysis extending to involve condyle

11. Segmental mandibulectomy was undertaken and the histopathological examination of biopsied sample revealed an angiomatous lesion. Based on patient's history, clinical behavior, histopathological and radiological features diagnosis of Gorham's disease made as per Heffez et al. criteria.

Discussion

1. Gorham's disease is an extremely rare disorder that may affect any bone, but has a predilection for the pelvis, humerus, axial skeleton, and mandible.
2. Over 150 cases are described in international literature with more than 50 cases reported involving the maxillofacial region.
3. Most cases reported were under 40 years of age with a mean age of 18 months – 70 years.
4. The clinical manifestation depends on the site of involvement. Some patients present with abrupt onset of pain and swelling or pathological fracture on the affected site.
5. Gorham's disease of lower jaw will initially affect basal and alveolar bone, which subsequently involves rami and condyle Involvement of temporomandibular joint can be mistaken for tempromandibular joint dysfunction.
6. The clinical course is unpredictable; bone resorption may arrest spontaneously after variable number of years in some cases. Despite the extensive regional loss of bone with resultant deforming atrophy, these patients have only mild disability

Etiopathogenesis

1. The exact etiology is unknown but the pathological process involves replacement of normal bone by nonneoplastic vascular tissue.
2. Gorham and Stout reported that active hyperemia changes in local PH, and mechanical forces promote bone resorption
3. Some believe the disorder has to do with osteoclast activity and interleukin-6, while some believe the increased osteoclast formation is due to an increase in sensitivity in circulatory precursors to humoral factors, and recently it has been suggested that cells of monocyte-macrophage lineage may play a role in pathogenesis

Histopathological Features

1. The typical histopathological finding is the replacement of bone by connective tissue containingmany thin walled blood vessels. It does not represent a hemangioma of bone which is a localised legion.
2. Most believe that the disease is not due to increased osteoclastic activity, although osteoclasts may be found in the tissues.
3. Heffez et al. described the criteria for diagnosis of massive osteolysis as:
   (1) positive biopsy for angiomatous tissue;
   (2) absence of cellular atypia;
   (3) minimal or no osteoblastic response and absence of dystrophic calcification;
   (4) evidence of local progressive osseous resorption;
   (5) nonexpansile, nonulcerative lesion;
   (6) absence of visceral involvement;
   (7) osteolytic radiographic pattern;
   (8) negative hereditary, metabolic, neoplastic, immunologic, or infectious etiology.
4. The diagnosis of Gorham's disease is difficult. It must be differentiated from osteomyelitis, hyper parathyroidism, intraosseous malignancies, metastatic disease, eosinophilic granuloma, and osteolysis associated with disease of CNS such as tabes dorsalis, syringomyelia, leprosy or myelodysplasia.

Management

1. Different forms of therapy include: medical treatment with oestrogen, calcium, vitamin D, bisphosphonates, and calcitonin, alpha 2B interferon.
2. Surgical treatment options include resection of the lesion, bone grafting, and prosthetic implants, the success of these treatments is very limited
3. Low dose radiation therapy is controversial and is not accepted, as there is chance of radiation induced malignancies

Conclusion

Gorham's disease is a rare musculoskeletal disorder. The etiology is undetermined, and currently, there is no effective treatment; therefore, it is important for dental surgeons to be aware of its existence as a rare case of osteolysis in maxillofacial skeleton.

題號	題目
1	以下哪種治療 Gorham's syndrome 方式為非： (A) Resection (B) Radiation therapy (C) Alpha 2B interferon (D) Biphosphonates
答 案 (B)	出處：Oral Pathology – Clinical Pathological Correlations 6th edition
題號	題目
2	以下不是 Gorham's syndrome 特徵之一？ (A) Angiomatous tissue (B) Negative hereditary, metabolic, neoplastic, or infectious etiology (C) Progressive osseous resorption (D) Increased osteoclastic activity
答 案 (D)	出處：