Journal of Oral Science, Vol. 52, No. 3, 513-516, 2010

Case Report

Vanishing mandible

Atul Sharma, Nageshwar Iyer, Amit Mittal, Debdutta Das and Shalinder Sharma

Department of Oral and Maxillofacial Surgery, MM College of Dental Sciences and Research, Mullana (Ambala), Haryana, India

(Received 4 March and accepted 2 July 2010)

Abstract: Massive osteolysis is a rare, insidious, chronic disease characterized by progressive resorption of contiguous osseous structures. In 1838 Jackson first described a case of disappearing humerus. More than 150 cases have since been described in the international literature, with fewer then 35 involving a maxillofacial site, usually the mandible. The exact cause of this disease remains unknown, but ongoing clinical research attempts to better understand the etiology. This case report is to our knowledge the second case in the international literature (after that reported by Thoma in 1933) to report complete resorption of the mandible. (J Oral Sci 52, 513-516, 2010)

Keywords: Gorham's disease; massive osteolysis; disappearing bone disease; phantom bone disease; vanishing bone disease; mandible.

Introduction

Massive osteolysis is a rare, insidious, chronic disease characterized by progressive resorption of contiguous osseous structures (1). First described in 1838 by Jackson, who reported the case of a young man with a progressively disappearing humerus, the syndrome became known as Gorham-Stout disease in 1955 when Gorham and Stout published a review of 24 cases (2). Facial involvement was first reported by Romer in 1924 (3), and complete mandible lysis was reported by Thoma in 1933 (1). The condition has many names: Gorham's disease; massive, spontaneous, or progressive osteolysis; acute essential bone resorption; and disappearing, phantom, or vanishing bone disease (4).

In most patients, the disease develops in the fourth decade of life, although it has been described in patients ranging in age from 18 months to 72 years. There is no genetic basis for the transmission of the disease, no sex or racial predilection, and no associated endocrine, metabolic, or neurologic disturbance (5). Any bone can be affected, although there is a predilection for the pelvis, humeral head, humeral shaft, axial skeleton, and mandible. To date more than 150 cases have been described in the international literature, with fewer then 35 involving a maxillofacial site, usually the mandible. The condition may or may not be painful and is most commonly detected after fractures or history of injury. When the jaw, tooth sockets, neck, face, and head are affected, possible symptoms include loose teeth, fractures, and pain. Because early diagnosis is crucial to prevent major morbidity and in some cases even mortality, dental surgeons should consider including Gorham's disease among the differential diagnosis of pathological entities leading to tooth mobility and subsequent osteolysis.

Correspondence to Dr. Atul Sharma, 296, Sector 13, OHB, Karnal, Haryana, India, Pin: 132001
Tel: +91-9896032239
Fax: +91-1842206066
E-mail: maxfacesurgeon@gmail.com

Case Report

A 32-year-old man presented to the outpatient department of MM College of Dental Sciences & Research, Mullana, Ambala, India, with a chief complaint of inability to chew food due to absence of the lower teeth. The patient gave a history of a fall from height ten years previously leading to abrasions on the face for which no medical attention was sought. He subsequently noticed progressive loosening of all his mandibular teeth to the point where he eventually lost all his teeth four years previously. He did not have any systemic complaints. There was no past history of any major illness and no other family member had a similar disorder.

On examination, the patient had a typical "bird face"

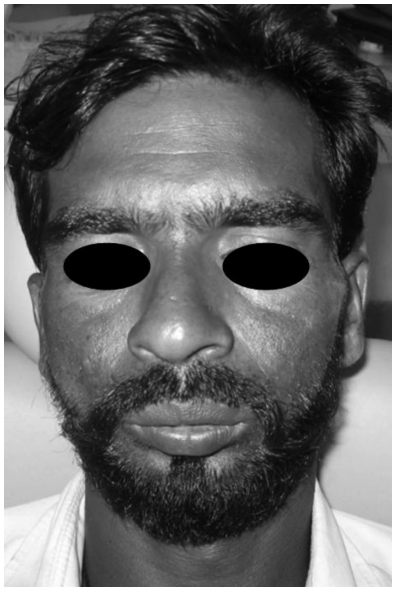

Fig. 1 Frontal view of the patient.

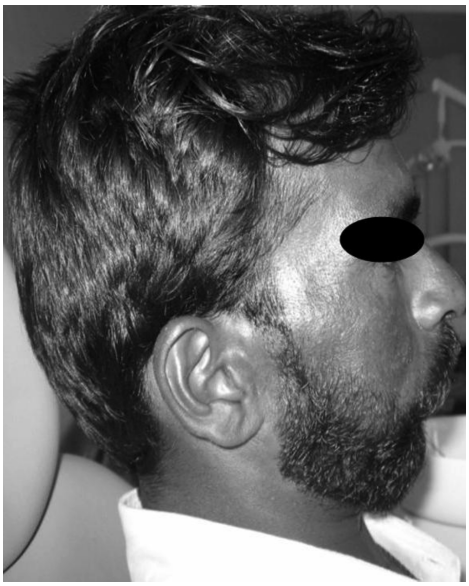

Fig. 2 Right lateral view of the patient.

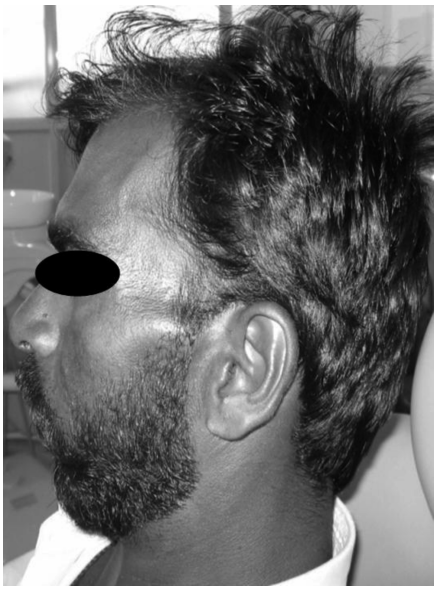

Fig. 3 Left lateral view of the patient.

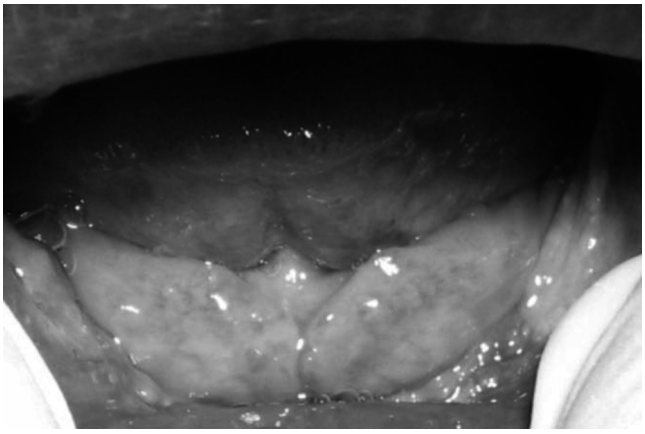

Fig. 4 Intraoral view.

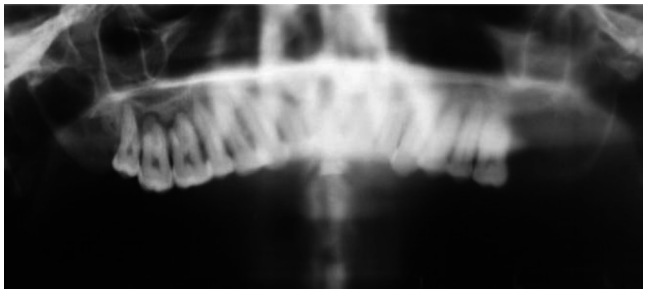

Fig. 5 Panoramic radiograph showing complete absence of the mandible.

appearance with a deep mentolabial furrow (Figs. 1-3), and on intraoral examination the lower jaw was edentulous (Fig. 4). On palpation, the entire mandible was missing with no tenderness or lymphadenopathy. There were no surgical scars on the face but there was massive attrition of the upper teeth suggestive of longstanding presence of their lower counterparts. The patient underwent routine hematological and biochemical tests (haemoglobin, total leucocyte count, differential leucocyte count, random blood glucose, and renal and liver function tests) but all were within normal limits. The patient declined a biopsy of the region.

Panoramic radiography was performed to evaluate the extent of mandibular loss; it showed complete absence of the mandible (Fig. 5). To better evaluate the extent of the resorptive process, we performed axial computed tomography (CT) of the maxillofacial region along with three-dimensional reconstruction. CT sections showed residual parts of the resorbed mandible in the symphysis, ramus, and condyle areas. (Figs. 6-8)

The clinical and radiographic features were compatible with the diagnosis of Gorham's disease of the mandible, without any other skeletal involvement. Aplasia of the mandible was ruled out because the patient gave a history of presence of the lower teeth (and this was supported by the finding of attrition of the upper teeth) and because CT showed residual parts of the resorbed mandible. Moreover, Frederiksen et al. (5) found that the focal lucencies seen in Gorham's disease of tubular bones are generally not seen in the mandible. This apparent difference may result from either the type of bone involved or the stage of disease at initial presentation.

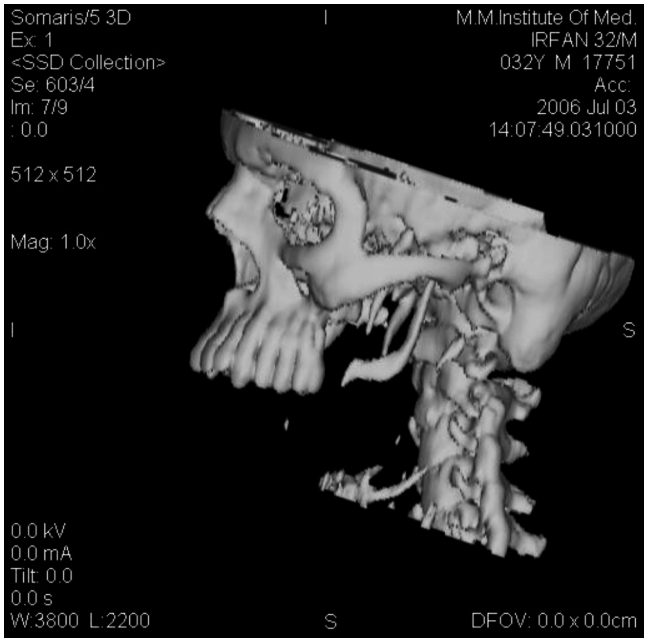

Fig. 6 Three-dimensional CT image showing remnants of resorbed mandibular condyle.

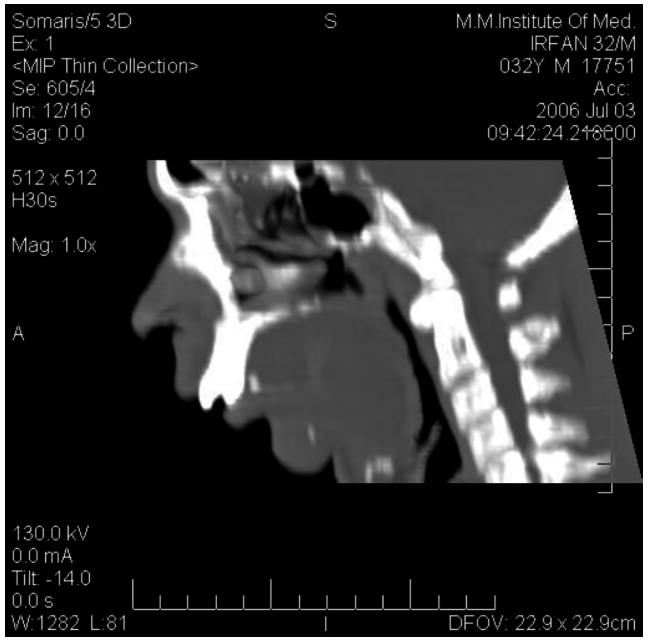

Fig. 7 CT image showing remnants of mandible in the symphyseal region.

As Gorham's disease is rare there is no accepted treatment protocol, although most treatment modalities focus on stopping osteoclastic resorption and vascular proliferation. Thus we advised the present patient to undergo radiotherapy or medical treatment with bisphosphonates to stop the ongoing osteolytic process and allow reconstruction of the

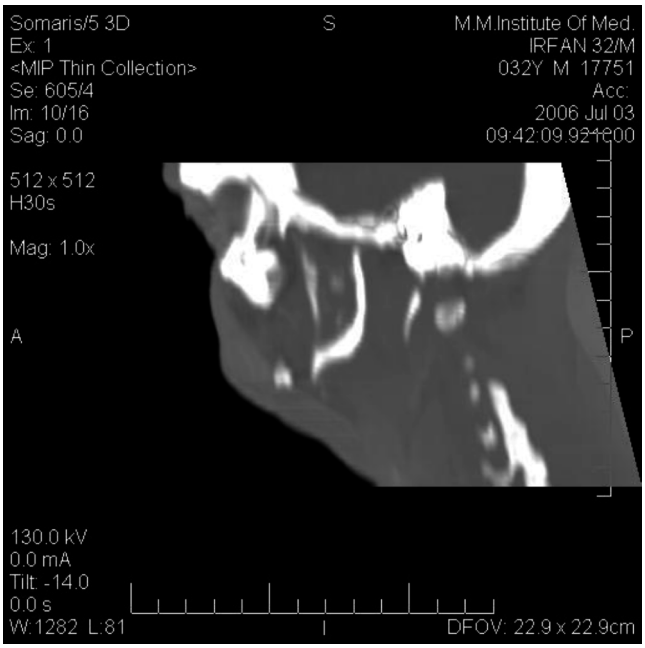

Fig. 8 CT image showing remnants of resorbed mandibular condyle.

mandible at a later stage. However, the patient was not willing because of potential side effects of both approaches.

The patient requested a complete denture for his lower jaw and was therefore advised to undergo a surgical procedure for complete replacement of his mandible with a prosthetic metallic implant after stabilization of the disease. The patient is currently being followed while awaiting disease stabilization.

Discussion

The exact cause of Gorham's disease remains unknown, but ongoing clinical research attempts to better understand the etiology of this bone disorder. As most general dentists are not aware of this disorder the majority of cases remain unreported. Even in our dental college most of the postgraduate teaching staff and postgraduate trainees were unaware of this disease. The provisional diagnosis made by house surgeons was edentulous lower jaw. Even after discussing the full case history with the postgraduate trainees they added a differential diagnosis of osteomyelitis of the lower jaw.

The standard laboratory blood tests are usually within normal limits in Gorham's disease and are therefore not helpful in making the diagnosis. Moreover, radiographically in the initial stages it resembles periodontal disease. Hence the condition is frequently misdiagnosed. Gorham's disease should accordingly be suspected in any case with a long history of inexplicable non response to periodontal

treatment, and biopsy should be performed (as this can only be done in the initial stages). Histopathologically, replacement of bone by connective tissue containing thin-walled blood vessels or anastomosing vascular spaces lined by endothelial cells is seen in most cases (1).

Radiation therapy and chemotherapy with interferon alfa-2b, which prevent angiogenesis, are the most widespread forms of treatment, with various degrees of success. Surgical options are resection in the active stage of disease followed by bone grafting. However, bone grafting is only performed after complete stabilization of the osteolytic process because resorption of bone grafts can occur these are implanted during the active phase of disease. Medical treatment with calcitonin, calcium, estrogen, vitamin D, fluoride, and biphosphonates, all of which promote bone regeneration and decrease bone metabolism, has been attempted with variable results (6,7).

Gorham's disease is an intriguing condition. To our knowledge this is the first case of its kind, as in the previous case where complete lysis of mandible was reported (Thoma, 1933) CT was not available and the diagnosis was reached from plain radiographs. In conclusion, we stress that dental surgeons should include Gorham's disease in their differential diagnosis to help researchers elucidate the pathogenesis and treatment of this poorly understood disease. We believe that this disease is not as rare as it seems but rather remains undiagnosed or misdiagnosed.

Acknowledgments

We would like to record our gratitude to Dr. V. K. Sharma, Director Health Services (Retd), Govt. of Haryana, for his inspiring guidance, cooperation and constant encouragement.

References

1. Heffez L, Doku HC, Carter BL, Feeney JE (1983) Perspectives on massive osteolysis. Report of a case and review of the literature. Oral Surg Oral Med Oral Pathol 55, 331-343.
2. Bruch-Gerharz D, Gerharz CD, Stege H, Krutmann J, Pohl M, Koester R, Ruzicka T (2003) Cutaneous vascular malformations in disappearing bone (Gorham-Stout) disease. JAMA 289, 1479-1480.
3. Holroyd I, Dillon M, Roberts GJ (2000) Gorham's disease: a case (including dental presentation) of vanishing bone disease. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 89, 125-129.
4. Ricalde P, Ord RA, Sun CC (2003) Vanishing bone disease in a five year old: report of a case and review of the literature. Int J Oral Maxillofac Surg 32, 222-226.
5. Frederiksen NL, Wesley RK, Sciubba JJ, Helfrick J (1983) Massive osteolysis of the maxillofacial skeleton: a clinical, radiographic, histologic, and ultrastructural study. Oral Surg Oral Med Oral Pathol 55, 470-480.
6. Tsang WM, Tong AC, Chow LT, Ng IO (2004) Massive osteolysis (Gorham disease) of the maxillofacial skeleton: report of 2 cases. J Oral Maxillofac Surg 62, 225-230.
7. Hagberg H, Lamberg K, Aström G (1997) Alpha-2b interferon and oral clodronate for Gorham's disease. Lancet 350, 1822-1823.