Case Report

Vanishing the Existence of the Mandible?

Abstract

Phantom bone disease, also known as Gorham's disease, is a rare disorder characterized by the progressive destruction of one or more skeletal bones. Commonly involved bones are the upper and lower extremities. Very few cases have been reported in the maxillofacial region with unilateral mandible being commonly involved. This case, to our knowledge, is the third case of Gorham's disease involving the whole length of the mandible in a 55-year-old female with a brief discussion on its clinical, radiographic, and histopathological presentation and treatment options.

Keywords: *Gorham's disease, mandible, oral pathology, osteolysis, phantom bone disease, vanishing bone disease*

Narayan Dutt Pandey[1], Shrikant Dusane[1], Bhushan Ramdas Bhagat[2], Shubhangi Bhushan Bhagat[3]

[1]Department of Oral and Maxillofacial Surgery, Sinhgad Dental College and Hospital, Pune, Maharashtra, India, [2]Department of Oral and Maxillofacial Surgery, Dr. D. Y. Patil Dental College and Hospital, Dr. D. Y. Patil Vidyapeeth, Pune, Maharashtra, India, [3]Department of Pharmaceutics, Dr. D. Y. Patil College of Pharmacy, Pune, Maharashtra, India

Introduction

Phantom bone disease (PBD), also known as Gorham's disease, is a rare disease characterized by the progressive replacement of bony framework by the proliferation of endothelial-lined lymphatic vessels.[1] It has been known by various names including massive osteolysis, vanishing bone disease, disappearing bone disease, acute essential bone resorption, spontaneous or progressive or idiopathic osteolysis, hemangiomatosis, lymphangiomatosis, and progressive osteolysis.[1,2] Although the exact etiology of this disease remains unknown.[3] The first case of this disease in the maxillofacial region was reported by Romer in 1924 in a middle-aged woman.[4] Till date, around 200 cases have been reported in the literature, out of which, 30% of the cases were found in the maxillofacial region with the unilateral mandible being the most commonly affected site.[1] In the present article, we report a unique case of PBD in a middle-aged female involving full length of the mandible.

Case Report

A 55-year-old female reported to the department of oral medicine with the chief complaint of pain in the mandible and difficulty in sleep for the past few weeks. History revealed a continuous slow reduction in the lower jaw size for the past 2 years which drastically changed her appearance [Figure 1a and b]. The patient gave a history of mild-to-moderate pain in the lower jaw for the past few weeks which was accompanied with reduced masticatory efficiency. Furthermore, difficulty in breathing while sleeping was reported for the past few days. There was no history of trauma, infection, neoplasm, or previous jaw surgery. The patient's past medical, social, and family history was noncontributory. Past dental history revealed exfoliation of some of the mandibular teeth and extraction of the remaining mandibular teeth by a local dentist progressively over a period of 1 year primarily because of pain and mobility, except for the left mandibular premolars and right mandibular first molar. On clinical examination, the patient appeared undernourished with the presence of a typical bird face appearance. Other clinical findings included incompetent lips, everted lower lip, retruded mandible, Grade III mobile mandibular left first and second premolars, and atrophy of the mandibular alveolar ridge, whereas the overlying mucosa appeared normal with no signs of inflammation or trauma. Computed tomography (CT) scan was advised which revealed diffuse loss of bony structures involving the whole length of the mandible, giving rise to floating teeth appearance, extending posteriorly and superiorly to involve both the condyles and coronoid processes which may have resulted in the pathological fracture of

How to cite this article: Pandey ND, Dusane S, Bhagat BR, Bhagat SB. Vanishing the existence of the mandible? Contemp Clin Dent 2023;14:176-8.

Submitted : 15-Nov-2022
Revised : 16-Apr-2023
Accepted : 15-May-2023
Published : 30-Jun-2023

Address for correspondence:
Dr. Bhushan Ramdas Bhagat, Department of Oral and Maxillofacial Surgery, Dr. D. Y. Patil Dental College and Hospital, Dr. D. Y. Patil Vidyapeeth, Sant Tukaram Nagar, Pimpri, Pune - 411 018, Maharashtra, India.
E-mail: bhagatbhushan10@gmail.com

Access this article online

Website: https://journals.lww.com/cocd

DOI: 10.4103/ccd.ccd_545_22

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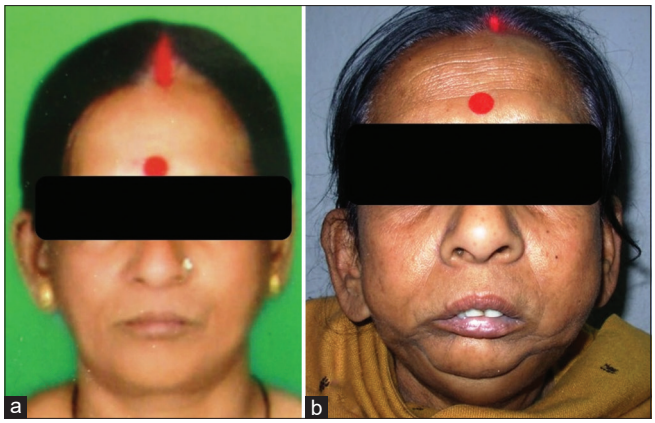

Figure 1: Clinical picture of patient (a) 2 years ago; (b) at the time of presentation

the condylar region [Figure 2]. Furthermore, the saggital section of the CT scan revealed soft-tissue impingement in the airway space leading to compromised airway space which might be the reason for sleep apnea [Figure 3]. To rule out long bone involvement, which is more commonly affected by Gorham's disease, radiographic examination of long bones was done which revealed normal bone structures without any similar pathology. Routine blood investigations, serum calcium, acid, and alkaline phosphatase were also within normal range, and no endocrinal abnormalities were detected. Biopsy was taken from the mandibular left posterior alveolar ridge distal to the premolars, and the specimen was sent for histopathological examination. The histopathological examination showed chronic inflammatory cell infiltrate and abundant thin-walled vascular channels with some active fibroblasts. There was no evidence of malignancy. Based on the history, clinical, and radiographic examination, as well as histopathological assessment, a diagnosis of Gorham's disease was made. The restoration of a defect that involves the entire mandible is rare and challenging for surgeons. In the present case, the treatment plan involved mandibular reconstruction with vascularized-free fibula graft and securing the tongue muscles so as to relieve sleep apnea. The patient was explained about the disease condition and treatment plan. However, the patient was not willing for the treatment due to financial reasons. The patient was on regular follow-up for the next 2 months, but she failed to report back thereafter.

Discussion

Gorham's disease or PBD is an extremely rare benign condition which is marked by the demolition of bone matrix and the proliferation of vascular structures causing massive osteolysis of the affected bone.[5] In the present case, the patient was in her 6th decade of life which is unusual.[5] Gorham's disease has no obvious gender, race, or geographic predilection.[5]

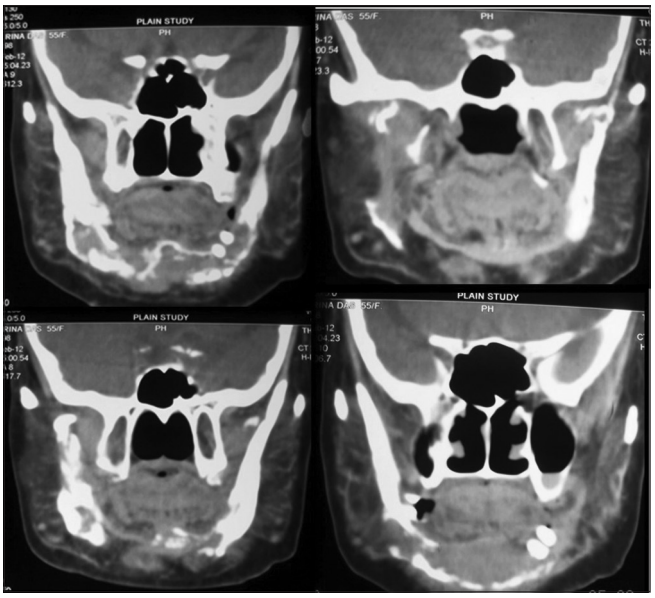

Figure 2: Coronal sections of CT scan showing the disease process affecting the full length of the mandible. CT: Computed tomography

This disease was first reported by Jackson, who found gradual "vanishing" of humerus "A boneless arm" in a young male patient following fracture.[6] However, its first detailed description was given in 1954 by Gorham and in 1955 by Gorham and Stout.[7] In the present case, pain in the mandible was the presenting complaint which was accompanied by sleep apnea which is rarely seen in this condition. Till date, approximately 200 cases of Gorham's disease have been reported in literature,[1] out of which only 53 cases involved the maxillofacial region. The most commonly involved bone in the maxillofacial region was the unilateral mandible, except for the two cases reported by Thoma in 1933 and Sharma *et al*. in 2010, wherein the entire mandible was affected.[7] Thus, involvement of the entire mandible is extremely rare. To the best of our knowledge, the present case report is the third case reported in the international literature thus making it unique. In 1983, Heffez *et al*.[6] established the following diagnostic criteria for PBD. Our case meets all eight diagnostic criteria established by Heffez *et al*.

Radiographic presentation in Gorham's disease was first described in 1958 by Johnson and McClure.[8] CT plays a vital role in evaluating the involvement of maxillofacial bones and helps in determining the extension into the skull.[3] Hence, in the present case, a CT scan was advised which revealed the involvement of the full length of the mandible as well as the compromised airway space.

Gorham's disease is an extremely rare and relatively silent disease with potentially fatal complications. Since the clinical symptoms are not characteristic of the disease, radiographic investigations play a vital role in early diagnosis of this condition. None of the treatment modalities have been proved to be very effective in

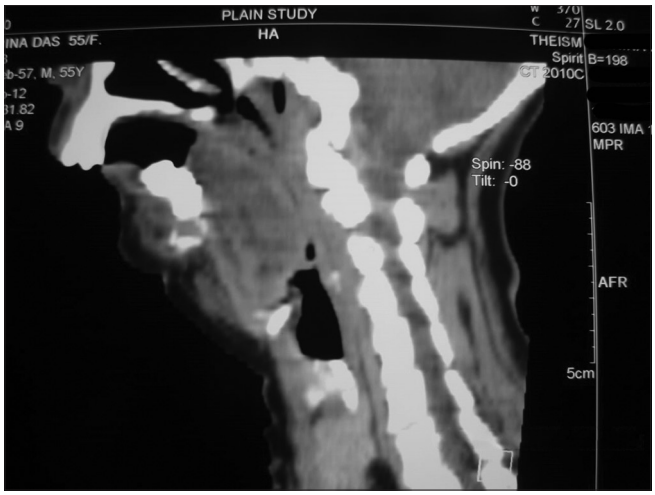

Figure 3: Saggital section of CT scan showing compromised airway due to soft-tissue overlapping. CT: Computed tomography

the long run. Hence, it is important for dentists to be knowledgeable about the existence of this disease as one of the rare causes of massive osteolysis involving the maxillofacial skeleton.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

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