Short Communication

Vanishing bone disease of hemimandible

Abstract

Vanishing bone disease also known as Gorham's disease, massive osteolysis, and phantom bone disease is a rare entity characterized by spontaneous or posttraumatic progressive resorption of bone. It is a rare idiopathic disease leading to extensive loss of bony matrix, which is replaced by proliferating thin-walled vascular channels and fibrous tissue. Most often diagnosis of vanishing bone disease is based on clinical examination, radiologic imaging, and histopathologic examination of the affected area. Very few cases have been reported in Indian literature considering the rarity of the disease occurring in the jaws; we report a rare case of vanishing bone disease of hemimandible in a 65-year-old male patient.

Key words:
Gorham's disease, mandible, osteolysis, vanishing bone disease

Introduction

Vanishing bone disease is a very rare skeletal condition of uncertain etiology, characterized by the uncontrolled proliferation of distended, thin-walled vascular or lymphatic channels within bone, which leads to resorption and replacement of bone with angiomas and/or fibrosis.[1] Various names have been used in the literature to describe this condition such as Gorham's disease, phantom bone disease, massive osteolysis, disappearing bone disease, and acute spontaneous absorption of bone. Gorham and Stout presented the first overview of vanishing bone disease in 1955 and concluded it as a progressive osteolysis associated with angiomatosis of blood or lymphatic vessels, which is now known as Gorham disease.[1,2] Partial or complete resorption of normal appearing bone occurs, which may continue to progress for years. In some cases, resorption stops spontaneously, but regeneration fails to occur.[3] The etiology remains unknown, the prognosis is very much unpredictable, and proper management protocol has not been determined. Incidence of the disease may be due to history of minor trauma, although as many as half of the patients have no history of trauma.[4] However, the disease has been described in patients as young as 1-month to as old as 75 years. The bones of the upper extremity and the maxillofacial region are the predominant osseous locations of the disease. Approximately, around 60% of all cases with vanishing bone disease occur in men.[5] Very few cases have been reported in Indian literature considering the rarity of the disease occurring in the jaws; we report a rare case of vanishing bone disease of hemimandible in a 65-year-old male patient.

Access this article online	
Website: www.ddtjournal.org	Quick Response Code

Case Report

A 65-year-old male patient reported to Department of Oral and Maxillofacial Surgery with a chief complaint of pain and deformed lower jaw since 2 years. He gave a history of progressive loosening of teeth in the right side of the lower jaw 4 years back with no other major illness during that period. There was no history of trauma, infection, neoplasm or previous jaw surgery.

Extraoral examination revealed hypoplastic mandible with significant asymmetry over the right side of the face [Figure 1]. Intraoral examination revealed the absence of teeth with atrophy of the alveolus on the right side of the mandible. Computerized tomography scan was advised

Yadavalli Guruprasad, Chandan Prabhakar[1], O. R. Hemavathy[1], Dinesh Singh Chauhan

Department of Oral and Maxillofacial Surgery, AME'S Dental College Hospital and Research Centre, Raichur, [1]Government Dental College and Research Institute, Bengaluru, Karnataka, India

Address for correspondence:
Dr. Yadavalli Guruprasad,
Department of Oral and Maxillofacial Surgery, AME'S Dental College Hospital and Research Centre, Raichur - 584 103, Karnataka, India.
E-mail: guru_omfs@yahoo.com

to detect any bony lesions, which revealed radiopaque remnants of resorbed bone with extensive osteolysis over right hemimandible. Residual fragments of resorbed mandible were observed over the symphysis, body and ramus of the mandible [Figures 2-6]. There was no bony sequestrum or subperiosteal bone deposition to indicate osteomyelitis. Blood investigation, serum calcium, acid and alkaline phosphatase were normal, and no endocrine abnormalities were identified.

Correlating the history, clinical and radiographic findings, and a provisional diagnosis of vanishing bone disease of hemimandible was made, and differential diagnosis of chronic osteomyelitis, hereditary osteolysis and skeletal angioma was made. Incisional biopsy was done in the region of the right body of the mandible under local anesthesia and subjected to histopathological examination, which revealed extensive osteolysis replaced by fibrous tissue. A final diagnosis vanishing bone disease of hemimandible was made based on clinical, radiographic and histopathology reports. As the patient was not willing for surgery, multidisciplinary approach for resection and reconstruction of the jaw was planned at a later stage.

Discussion

Vanishing bone disease is a very rare disorder characterized by uncontrolled, destructive proliferation of vascular or lymphatic capillaries within bone and surrounding soft tissue. Various names have been used in the literature to describe this condition such as Gorham's disease, phantom bone disease, massive osteolysis, disappearing bone disease, acute spontaneous absorption of bone.[5] It is a nonhereditary disease with no sex predilection and are younger than 40 years. More than 150 cases have been documented in the literature until date. Among them, the pelvic and shoulder regions were most frequently involved, although any bone may be affected. The skull is among the least common locations of involvement, and its occurrence

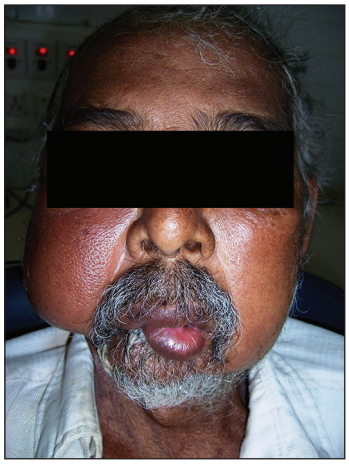

Figure 1: Clinical photograph showing frontal view of the patient with significant asymmetry over right hemimandible

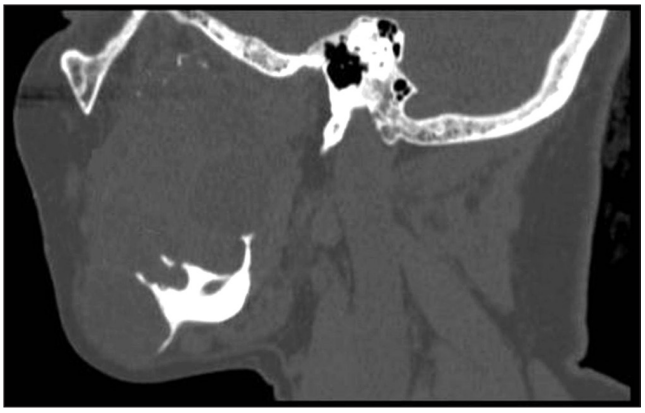

Figure 2: Coronal computerized tomography scan view of right hemimandible shows radiopaque remnants of mandibular bone with extensive osteolysis

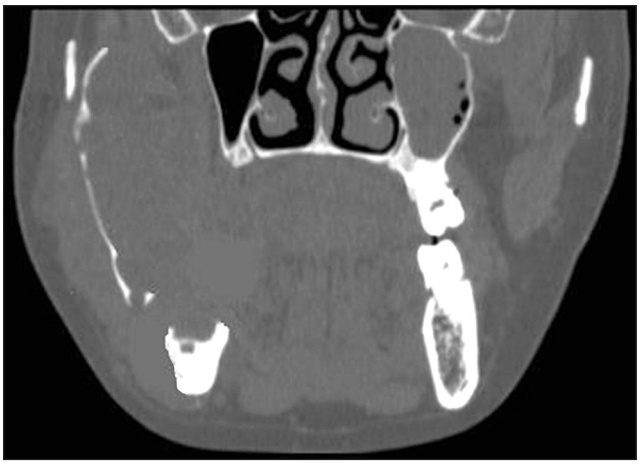

Figure 3: Sagittal computerized tomography scan view of right hemimandible shows radiopaque remnants of mandibular bone with extensive osteolysis

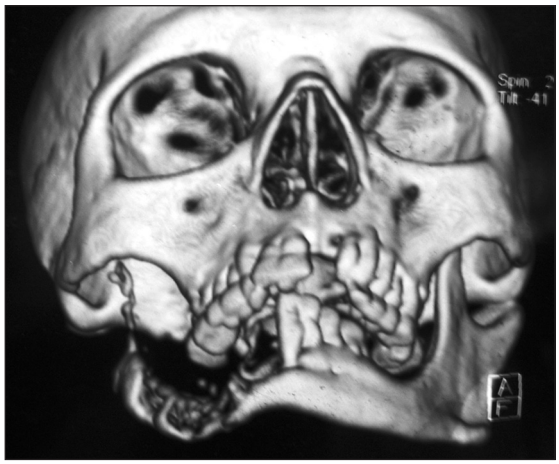

Figure 4: Three-dimensional computerized tomography submental view shows radiopaque remnants of mandibular bone with extensive osteolysis over the right hemimandible

Figure 5: Three-dimensional computerized tomography lateral view shows radiopaque remnants of mandibular bone with extensive osteolysis over the right hemimandible

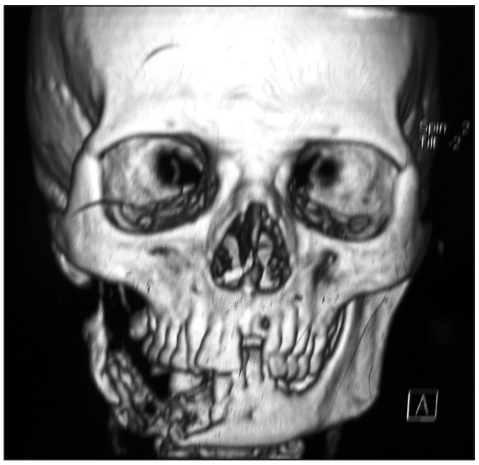

Figure 6: Three-dimensional computerized tomography frontal view shows radiopaque remnants of mandibular bone with extensive osteolysis over the right hemimandible

in the jaws is very rare.[6,7] Although the exact etiology of osteolysis is unclear, it is considered by most to be due to nonmalignant, neoplastic proliferation of hemangiomatous (or lymphangiomatous) tissue.[7]

The proposed etiopathogenesis of vanishing bone disease is nonmalignant, neoplastic proliferation of hemangiomatous or lymphangiomatous tissue replacing the resorbed bone.[7,8] Some authors have suggested local hypoxia and acidic environment, and some hydrolytic enzymes such as acid phosphatase and leucine aminopeptidase can cause the bone destruction. Others have also postulated the role of mechanical forces may promote bone resorption and that trauma might trigger the process. Johnson and McClure suggested that there are two stages of the disease.[9,10] The first stage of hemangiomatosis characterized by vascular proliferation in connective tissue. This explains some of the pathology reports of Gorham disease as "skeletal hemangioma." Second is the stage of fibrosis that replaces the absorbed bone. Whether osteoclasts are involved in the mechanism of bone destruction remains controversial. Most authors have not observed osteoclasts in the areas of excessive bone resorption by microscopy.[9,10]

Clinical manifestations include pain, swelling, or a pathologic fracture, whereas others may be asymptomatic or have an insidious onset of soft tissue atrophy and there are no significant specific laboratory findings. Preoperative Gorham's or vanishing bone disease must be distinguished from osteolysis secondary to other pathologic processes, including the hereditary, metabolic, neoplastic, infectious, and immunologic etiologies. Common differential diagnosis includes hereditary multicentric osteolysis, essential osteolysis with nephropathy, metastasis, osteomyelitis, and rheumatoid arthritis.[10,11]

Different treatment modalities, surgical and nonsurgical methods have been used in the management of vanishing disease of bone. The nonsurgical options include radiation therapy, antiosteoclastic medication (bisphosphonates), and alpha-2b interferon. The surgical options include surgical resection, reconstruction using a bone graft, or prosthesis. Most surgeons, based on their personal experience have observed that the bone graft undergoes resorption. In recent years, most patients have been treated with surgery and/or radiation therapy.[12]

Conclusion

Vanishing bone disease of the jaws is very rare condition occurring in the maxillofacial region. Diagnosis is often delayed in most cases as laboratory studies are usually within normal limits. Clinical diagnosis together with characteristic radiographic and histopathological findings is helpful for making an early accurate diagnosis. Proper diagnosis is very important to avoid mishaps in management, and a more cautious multidisciplinary approach is required to treat such type of lesions. It is a surgical challenge for reconstruction and rehabilitation for the patient postoperatively.

References

1. Gorham LW, Stout AP. Massive osteolysis (acute spontaneous absorption of bone, phantom bone, disappearing bone); its relation to hemangiomatosis. J Bone Joint Surg Am 1955;37-A:985-1004.
2. Gorham LW, Wright AW, Shultz HH, Maxon FC Jr. Disappearing bones: A rare form of massive osteolysis; report of two cases, one with autopsy findings. Am J Med 1954;17:674-82.
3. Ohya T, Shibata S, Takeda Y. Massive osteolysis of the maxillofacial bones. Report of two cases. Oral Surg Oral Med Oral Pathol 1990;70:698-703.
4. Bouloux GF, Walker DM, McKellar G. Massive osteolysis of the mandible: Report of a case with multifocal bone loss. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1999;87:357-61.

5. Holroyd I, Dillon M, Roberts GJ. Gorham's disease: A case (including dental presentation) of vanishing bone disease. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2000;89:125-9.
6. Benhalima H, Lazrak A, Boulaich M, Mezahi M, Ourlai A, Kzadri M, *et al*. Massive osteolysis of the maxillo-facial bones: Case report and review of the literature. Odontostomatol Trop 2001;24:35-40.
7. Motamedi MH, Homauni SM, Behnia H. Massive osteolysis of the mandible: A case report. J Oral Maxillofac Surg 2003;61:957-63.
8. Raghuveer HP, Jayalekshmy R. Gorham's massive osteolysis of the mandible- a progressive radiographic presentation. Dentomaxillofac Radiol 2009;38:292-5.
9. Skidmore TB, Lee CM, Abbott TM, Wiggins RH 3rd, Anderson GE, Tward JD, *et al*. Gorham disease of the mandible: Radiographic findings and radiotherapy response. Ear Nose Throat J 2008;87:E4-7.
10. Gomez-Diez SG, López LG, Escobar ML, Gutiérrez LJ, Oliva NP. Progressive facial hemiatrophy with associated osseous lesions. Med Oral Patol Oral Cir Bucal 2007;12:E602-4.
11. Sharma A, Iyer N, Mittal A, Das D, Sharma S. Vanishing mandible. J Oral Sci 2010;52:513-6.
12. Deng R, Tang E, Lu M. Massive osteolysis of the mandible. J Craniofac Surg 2011;22:2386-8.

How to cite this article: Guruprasad Y, Prabhakar C, Hemavathy OR, Chauhan DS. Vanishing bone disease of hemimandible. Drug Dev Ther 2014;5:168-71.

Source of Support: Nil. **Conflict of Interest:** None declared