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Case Report

Floating teeth appearance: A radiographic dilemma

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الملخص

مرض غورهام هو اضطراب نادر للغاية، يتميز بانحلال عظمي كبير للعظم المتضرر غير معروف السبب، ولا يمكن التنبؤ بمصيره. إضافة إلى ذلك، لا يوجد علاج قياسي متاح لمرض غورهام. هذه المقالة تصف تقرير لحالة مريضة من الملايو عمرها ٦١ عاما تم تشخيصها بمرض غورهام في الفك السفلي الأمامي دون تاريخ سابق لأي إصابة. حضرت المريضة وهي تعاني من ألم وتحرك في الجزء السنخي السني من ٣ أشهر. أظهر التصوير الشعاعي "أسنان عائمة" وتوسع مساحة رباط اللثة ومنطقة محدودة بها تدمير عظمي. أظهر فحص الأنسجة، انتشار العديد من القنوات المبطنة البطانية داخل الأنسجة بين الخلايا. تم استبدال الظهور الليفي للعظم بالنسيج الضام الليفي. تم علاج المريضة بإزالة بسيطة للعظم المصاب تحت التخدير الموضعي، أعقبه إعادة تأهيل تقويم الأسنان. تم شفاء مكان الموقع الجراحي وعدم ظهوره مجددا أثناء المتابعة لمدة ٣ أعوام.

الكلمات المفتاحية: انحلال العظم؛ مرض غورهام؛ الفك السفلي؛ الأسنان العائمة؛ إعادة تأهيل تقويم الأسنان

Abstract

Gorham disease (GD) is an extremely rare disorder that is characterised by massive osteolysis of the affected bone with unknown aetiology and an unpredictable prognosis. Additionally, no standard treatment is available for GD. This article describes a case report of a 61-year-old Malay woman who was diagnosed with GD of the anterior mandible without a prior history of trauma. She presented with pain and mobility of the dentoalveolar segment for 3 months. The radiographic findings showed "floating teeth" with widening of the periodontal ligament space and localized area of bony destruction. Histopathologically, there was proliferation of numerous dilated endothelial-lined channels within the intertrabecular tissue. Some areas of bone were replaced by fibrous connective tissue giving rise to the appearance of a benign fibroosseous lesion. The patient was managed with simple removal of the affected bone segment under local anaesthesia, followed by prosthodontic rehabilitation. Healing of the surgical site was uneventful, and no recurrence was reported at the 3-year follow-up.

Keywords: Floating teeth; Gorham disease; Mandible; Osteolysis; Histopathology

Introduction

Gorham disease (GD) is an extremely rare disorder mainly characterized by massive osteolysis of the affected bone with unknown aetiology.[1] Although it was defined as a specific pathological entity by Gorham and Stout in 1954, a case of vanishing bone disease was first described by Jackson in 1838.[1,2] Around 200 cases of GD have been reported

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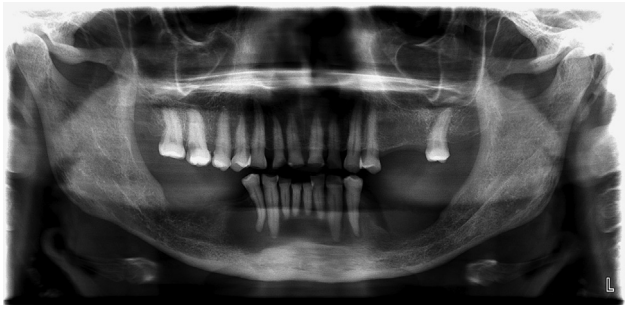

Figure 1: Panoramic radiograph showing presence of an ill defined radiolucency confined to the anterior segment of mandible with a floating appearance of teeth. Widening of periodontal ligament space noted at the periapical area of tooth 33 and 34.

since.[3] The disease can be monostotic or polyostotic, affecting any bone, although it has a predilection for the pelvis, humerus, and axial skeleton.[4] Although the exact aetiology is unknown, it has been linked to a history of trauma, as demonstrated by elevated interleukin 6 levels and evidence of osteoclast activity, with the proposed biomarkers being PDGF BB, sRANKL, and osteoprotegerin.[5–8] In addition, increased levels of local vascular endothelial growth factor (VEGF A and C) that are associated with lymphangiogenesis have been proposed to drive the abnormal proliferation of endothelial-lined channels seen in GD.[8] Involvement of the maxillofacial bones was first described by Romer in 1924[9] and approximately 50 cases have been reported.[10–12] The disease is characterised by initial progressive bone destruction with discomfort, followed by an asymptomatic latent period.[13] The clinical features may include pain, deformity, malocclusion, fracture, and tooth mobility. It may mimic malignancy in a clinical setting, but the underlying bone loss and clinical outcome run a benign course.

Case presentation

A 61-year-old woman visited the dental clinic of Hospital Universiti Sains Malaysia with a chief complaint of pain and mobility of her lower front teeth for 3 months duration. The pain was dull in nature, triggered upon biting, and gradually eased upon release. There was no previous history of trauma or infection involving the jaw area. She had underlying hypertension or osteoarthritis and had been compliant with medication. There was no history of allergy to medication or food. She was a retiree and a mother of five children.

Upon examination, the face was symmetrical with no apparent abnormality noted. No clicking or tenderness was elicited over the temporomandibular joint area, and she presented with adequate mouth opening. There was no numbness or abnormal sensation over the face and jaw areas. The maxilla was firm upon palpation. The dentoalveolar

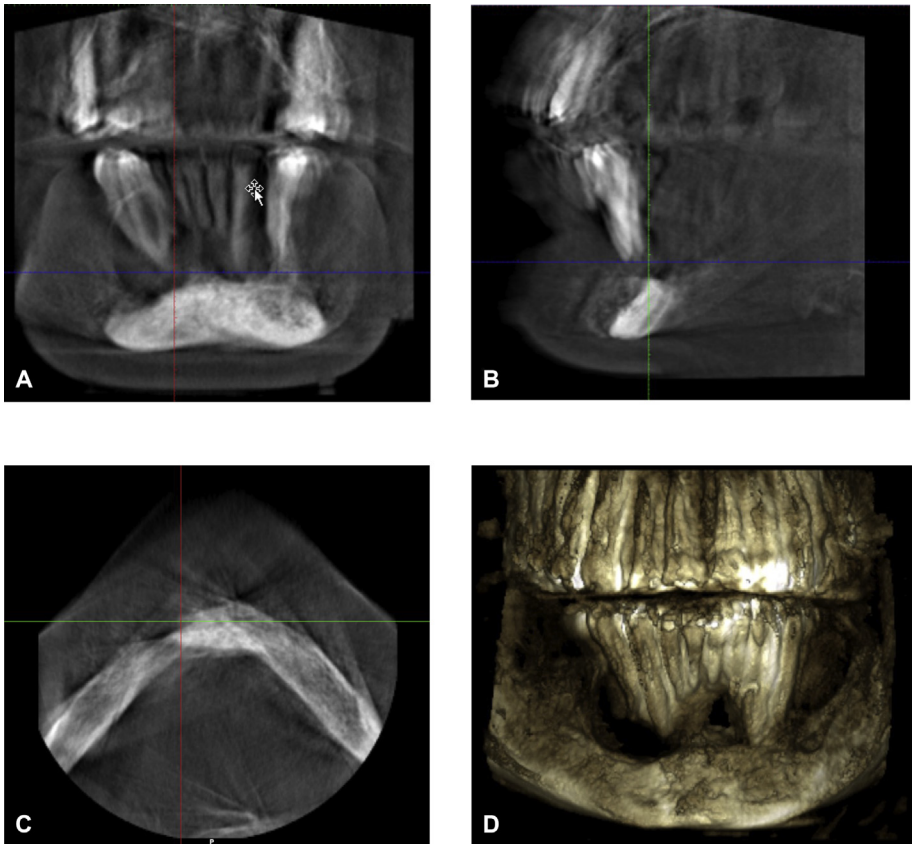

Figure 2: Coronal (A), sagittal (B), axial (C) and reconstructed 3D cone beam computed tomography (D) images showing presence of osteolytic lesion localized to the anterior mandible, respectively.

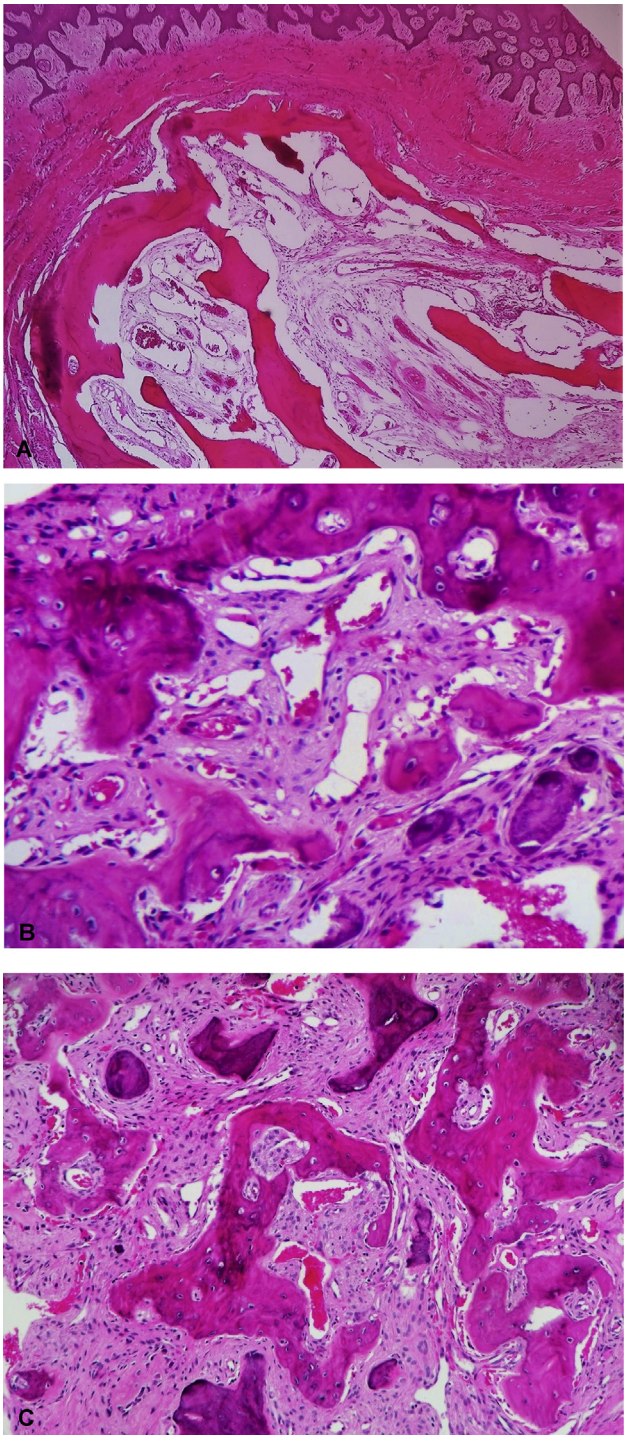

Figure 3: Histopathological examination of the haematoxylin-eosin stained specimen. Residual cortical bone below the surface oral mucosa with loose fibrous connective tissue background bearing numerous dilated endothelial-lined channels (Original magnification ×40) (A); Irregular trabeculae of woven bone exhibiting reversal lines, within a vascular fibrous connective tissue background (Original magnification ×400), (B); Curvilinear trabeculae of woven bone with moderately cellular stroma resembling benign fibroosseous lesion appearance. (Original magnification ×200), (C).

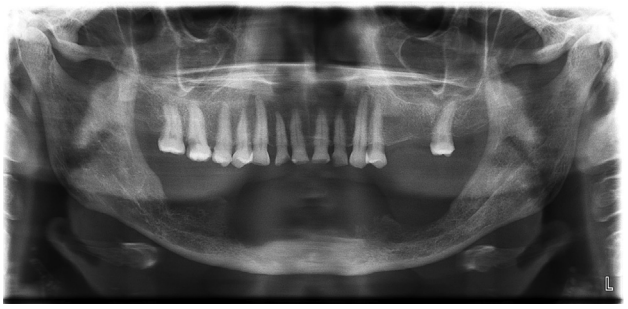

Figure 4: Panoramic radiograph showing presence of an area of radiolucency confined to the anterior segment of mandible at 4 months post-surgical intervention.

segment from tooth 34 to 44 was mobile when palpated against the firm inferior border of the mandible. There was no evidence of bone expansion, with a normal appearance of overlying mucosa. The affected teeth were non-carious, with an absence of periodontal pockets, and responded to pulp sensibility testing. Marked attrition was observed on the anterior teeth on the upper and lower jaw.

A panoramic radiograph was taken to evaluate the alveolar bone condition. Widening of the periodontal ligament space indicating loss of lamina dura was noted on the periapical area of teeth 33 and 34. Overall, an ill-defined radiolucency appeared to be localized to the dentoalveolar segment of teeth 34 to 44, producing an appearance of floating teeth (Figure 1). Further evaluation with a cone beam computed tomography (CBCT) scan revealed evidence of alveolar bone destruction localized to the anterior mandible (Figure 2).

Surgical removal of the mobile dentoalveolar segment was performed under local anaesthesia. Histopathological examination showed irregular trabeculae of lamellar and woven bone exhibiting reversal lines, supported by loose to dense fibrous connective tissue. Numerous dilated channels lined by flattened endothelial cells that were largely capillary and tortuous were observed within the inter-trabecular tissue and close to the bone trabeculae surface. Mild to moderate chronic inflammatory cell infiltrate was present (Figures 3A and 3B). Some areas showing replacement of the osseous component by moderately cellular dense fibrous connective tissue were reminiscent of a benign fibroosseous lesion (Figure 3C). The microscopic diagnosis of GD was made. At a 4-month follow-up, the affected areas appeared fully healed. Radiographic findings showed a localized area of radiolucency (Figure 4) with intact remaining bone surface (Figure 5). The patient was referred to a prosthodontist for denture construction. There were no signs of disease recurrence at the 3-year follow-up.

Discussion

GD or phantom bone disease is an extremely rare osteolytic condition that involves extensive, locally aggressive resorption of bone and is, in most cases, a diagnosis of exclusion. The disease shows no clear inheritance pattern, and most cases have involved adults below the age of 40 and children aged as young as 3 years.[12,14–16] The present case,

however, occurred in a much older patient in her 60s and affected the mandibular bone, which is the most common site reported in cases involving the jaw.[17]

GD generally lacks characteristic clinical features other than tooth mobility accompanied by mild dull pain, while other local manifestations such as bone swelling and pathological fracture are infrequently encountered.[17] As it is a locally aggressive disease, systemic signs and symptoms are rather unlikely, although fever was recently reported in a male patient aged 34 years.[11] The radiographic findings of GD are also nonspecific, with common characteristics including ill-defined radiolucencies, widening of the periodontal ligament space of involved teeth, and localized bone destruction with the "floating teeth" appearance.[18,19] Given the clinical presentation and radiographic features of the present case, several pathologic entities of either inflammatory or neoplastic origin may be considered.

A number of bone lesions such as chronic periodontitis, chronic osteomyelitis, plasmacytoma, primary intraosseous carcinoma, and osteosarcoma may clinically present with tooth mobility and bone destruction. Chronic periodontitis is included in the differential diagnosis because it is the most common disease affecting the periodontium in the adult population. Its main clinical presentation of teeth loosening and dull pain is associated with attachment and alveolar bone loss, affecting multiple teeth in a quadrant or with generalized involvement.[20–22] The present case exhibited a pattern of horizontal bone loss, with destruction of more than two thirds of the alveolar bone level. However, neither signs of periodontal pocketing nor receding gingiva were observed. The structural mobility of the tooth in this patient also appeared segmental in nature, as it moved as a unit upon palpation, as opposed to the mobility of an individual tooth typically seen in cases of chronic periodontitis.

Chronic osteomyelitis, on the other hand, can manifest as segmental alveolar bone mobility due to the presence of a large bony sequestrum. Radiographic findings of ill-defined radiolucency involving a segment of alveolar bone that appears detached from the surrounding vital bone is a common finding.[23] However, given the 3-month duration of the problem, a periosteal inflammatory reaction is expected to have taken place as part of the bone remodelling process in its attempt to heal. This is typically represented by a sclerotic bony margin, which was not observed in this case.[23] Additionally, the absence of a common initiating factor for osteomyelitis such as a history of trauma to the jaw, either by fracture or surgical procedure, makes this clinical diagnosis rather unlikely. The lack of inflammatory signs and symptoms such as redness, swelling, or tenderness and the absence of a contributary immunocompromising state of systemic illnesses further ruled out this possibility.

Plasmacytoma is a malignant neoplasm of plasma cells that can affect single or multiple bones.[24] It is prevalent in adults between the ages of 40 and 82 years, with a mean age of 55 years.[24,25] The mandible is more frequently affected than the maxilla, but it is considered uncommon

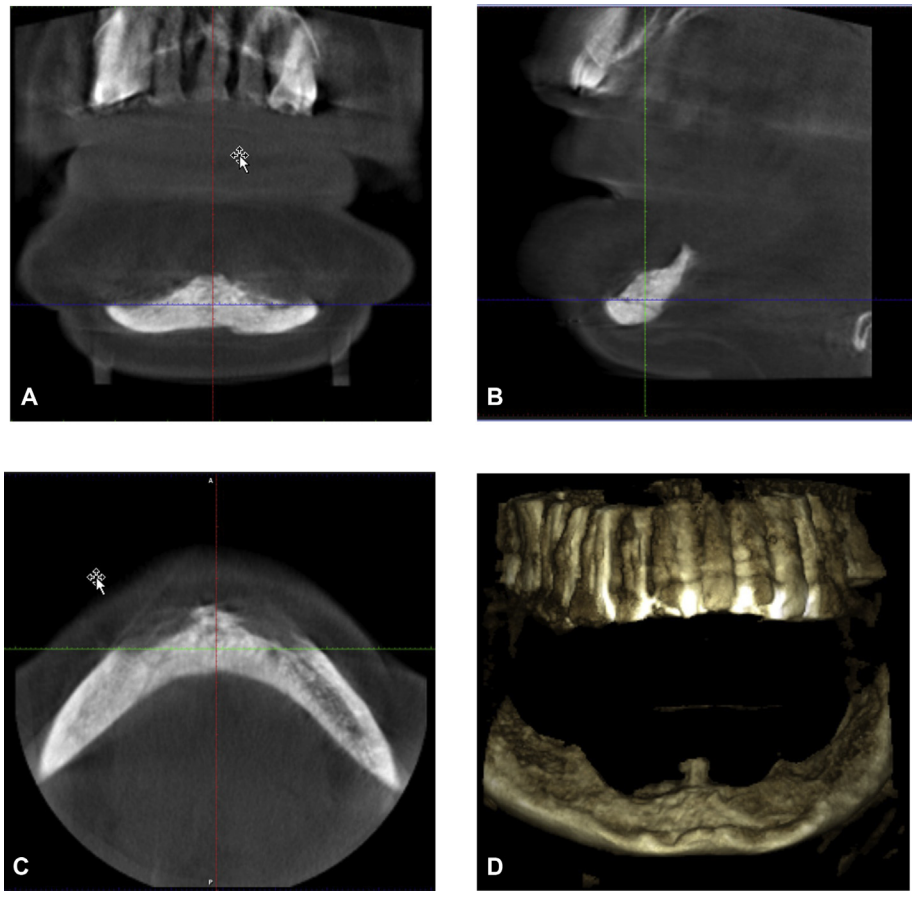

Figure 5: Coronal (A), sagittal (B), axial (C) and reconstructed 3D cone beam computed tomography (D) images revealed intact bone surface of the remaining mandibular cortex after 4 months post surgery, respectively.

in the jaws. The patient may present with tooth mobility, dental pain, haemorrhage, swelling, and paraesthesia, although some lesions are occasionally asymptomatic.[24–26] Radiographically, the margin of the lesion is well defined and lacking in periosteal bone reaction. In most cases, the appearance is more ragged and infiltrative, imparting a "punched out" appearance.[27] For solitary plasmacytoma of the jaw bone, consistent local features include swelling, haemorrhage, and paraesthesia. In the case of multiple bone involvement including the jaw bone, there should be accompanying constitutional symptoms of fatigue, weight loss, fever, and typical low back pain, which were not present in our patient.[24,27]

Primary intraosseous carcinoma is a squamous cell carcinoma that arises solely from within jawbones without any involvement of the overlying oral mucosa. It is rare and insidious, as most present as asymptomatic incidental findings, and it typically affects adults between the mean ages of 55–60 years, with tooth mobility being one of its main features, apart from a non-healing extraction socket and slowly progressive jaw swelling.[28] Pain is apparent once it reaches a considerable size. The lesion appears as ill-defined irregular radiolucencies, with a lack of periosteal bone reaction and gradual effacement of the lamina dura. Nevertheless, it has a male predilection and affects the molar region of the mandible rather than the anterior part of the jaw.[27]

Osteosarcoma of the jaws, albeit rare, commonly presents in patients within the 50–60 years age group, which is a decade later than its occurrence in its long bones counterpart[29]. While tooth loosening is a frequent feature, in most cases it is accompanied by a rapid increase in jaw swelling along with pain and tenderness, all of which were absent in the present case. Its radiographic findings include nonspecific features of ill-defined radiolucencies and widening of the periodontal ligament space of the involved teeth, with a more typical "sunray" appearance representing periosteal bone involvement.[27]

The only defining features of GD can be observed through microscopic evaluation. The characteristic histologic abnormalities include a proliferation of thin-walled capillary-sized vascular channels with evidence of a substantial loss of bony matrix and areas exhibiting replacement by fibrous connective tissue,[17,30,31] all of which were observed in this case.

The disease progression is usually halted with surgical resection of the affected jaw, and most cases showed improvement with time. From the authors' viewpoint, other forms of therapy, particularly bisphosphonates and radiation therapy, should be reserved for the management of persistent cases, as an adjunct to failed surgical therapy, given that the complication involving the development of jaw osteonecrosis and osteoradionecrosis respectively, might outweigh the benefits of the disease outcome.[17,32–34] In rare instances, varying degrees of deformity may develop and encroachment on vital structures may be fatal[35,36].

Conclusion

Floating teeth appearance is a common phenomenon observed on panoramic imaging in the clinical setting, and the underlying alveolar bone destruction may be due to a variety of causes. This case report emphasised the clinicoradiological correlation approach in the formulation of the diagnosis. It also discussed GD as a diagnosis of exclusion, which should always be considered in cases that present with this specific radiographic finding.

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Conflict of interest

There is no conflict of interest.

Ethical approval

Ethical approval was obtained from the Human Research Ethics Committee of Universiti Sains Malaysia (USM/JEPeM/17040222).

Consent

Informed consent was obtained prior to the preparation of the case report, and the author/s endeavoured to ensure anonymity.

Authors contributions

NSAM collected the relevant clinical data including radiographic images. SAR was directly involved in the case management. NAR conceived the idea, performed the histopathological examination of the specimen, and wrote the initial and final drafts of the article. MHH revised the article for important intellectual content. All authors have critically reviewed and approved the final draft and are responsible for the content and similarity index of the manuscript.

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