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Vanishing Bone (Gorham's) Disease of the Mandible: A Case Report

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Abstract: Vanishing bone disease (VBD) is a rare, chronic condition of massive osteolysis characterized by proliferation of vascular channels, and leading to resorption and progressive disappearance of the affected bone. Since its first description by Jackson in 1838, 150 or more cases have been described in the literature, but only a very few of these are reported in the jaws. The exact cause of the disease is not known and its natural history and prognosis unpredictable. We report here, a case of VBD affecting the mandible of a young Ghanaian man who initially presented with acute severe toothache and Trismus, of sudden onset, that progressed to unusually severe chronic pain similar to neuralgic pain involving the right inferior alveolar nerve.
Keywords: Vanishing, Mandible, Toothache, Neuralgic pain..

INTRODUCTION

Vanishing bone disease (VBD) or Gorham's disease is a very rare, destructive condition of the skeletal system resulting in progressive resorption and disappearance of osseous structures with replacement by vascular and fibrous connective tissue[1]. The condition has many names: Gorham's disease; massive spontaneous, or progressive osteolysis; acute essential bone resorption, disappearing bone disease and phantom bone disease[2]. The disease may or may not be painful and is most commonly detected after fractures or history of injury[3]. When the jaw, tooth sockets, neck, face, and head are affected, possible symptoms include loose teeth, fractures, and pain [3]..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[3]. There has been no previous report of this condition in the present literature in a Ghanaian. The purpose of this report is to remind us of this disease, and to report on its occurrence in the mandible, with an unusual symptom of severe chronic neuralgic pain involving the right inferior alveolar nerve.

CASE REPORT

A 25-year old male presented to us with a two month history of jaw pain in the lower right quadrant after a dental extraction. His symptoms started abruptly as a severe toothache involving tooth number 47 with trismus. He reported to his general dental practitioner shortly after, and had the tooth easily extracted. This was followed by jaw pain; which was constant, disturbed sleep and interfered with mastication. His medical and social histories were unremarkable and there had been no previous any such ailment in his family, hence ruling out hereditary origin. Three different antimicrobials namely, Amoxicillin, Metronidazole, and Clindamycin were prescribed, but had no significant effect. Analgesics such as diclofenac, only gave temporary relief.

There was no swelling nor obvious facial asymmetry. He had trismus and a deviation of the mandible to the right (ipsilateral side) on opening. Palpation of the lower border of the mandible revealed a step deformity on the right. A general examination of his systems revealed no abnormalities. Intra oral examination showed normal appearance of of the oral structures and mucosa except a sore at the extraction site of tooth number 47 which showed signs of healing. The initial impression at the time was fractured mandible as a complication of dental extraction and or a pathological fracture. A panoral radiograph (OPG) however showed loss of the mandibular bone affecting the right condyle to the body, just distal to the roots of the first molar on that side (Fig 1).

Under general anaesthesia, a biopsy was carried out and samples taken from both hard and soft tissues in the region of the suspected pathology. At the same time, intermaxillary fixation (IMF) using eyelet wires was carried out to stabilize the jaw and reduce the severe pain following movement. The results of the biopsy, while suggesting "proliferation of thin-walled

unusually wide capillary-like vessels consistent with chronic inflammatiory changes", ruled out neoplastic disease. Mantoux test was negative for tuberculosis. Chest and pelvic x-rays did not reveal any abnormality.

The patient returned two weeks after the IMF, for follow-up review. He appeared very anxious and still complained of pain in the right mandible that disturbed his sleep. He was further placed on suppository diclofenac, and diazepam. Three weeks subsequently, he returned with a additional complaints of "tingling sensation and feeling of electric shocks" in the lower right lip, prompting the inclusion of central giant cell tumour, brown tumour, and multiple myeloma in the differential diagnosis. Investigations covering serum calcium levels, parathormone, calcitriol, alkaline phosphatase, and Bence Jones proteins were carried out, and the results of these and other blood tests for full blood count, fasting blood glucose, as well as urinanalysis, were all within normal range, and ruled out metabolic disease.

The patient has since been seen at three monthly intervals in with complaints of severe pain over the area of the mental nerve supply; for which he has intermittently been placed in IMF and maintained on diclofenac and carbamazepine as he appeared to benefit from that. Two years after he was first seen, a second OPG revealed further cortical bone loss, extending further to the region of tooth number 43 (Fig. 2). The bone resorption had not abated and facial asymmetry and deviation of the mandible on opening had become obviously noticeable (Fig. 3 & 4) . His main complaint then continued to be severe pain, especially to touch, in the right lower jaw to the midline region; characteristic of neuralgic pain involving the right inferior alveolar nerve. Considering the clinical and radiographic features and course of the disease, a diagnosis of Gorham's disease of the mandible was made. Emphasis on his treatment shifted and he was started on a course of bisphosphonate, neurobion forte, prednisolone and diclofenac which appeared to offer some initial benefit. At his last visit. Nearly three years following the initial symptoms, though the pain is under control, there is no evidence of cessation of bone resorption.

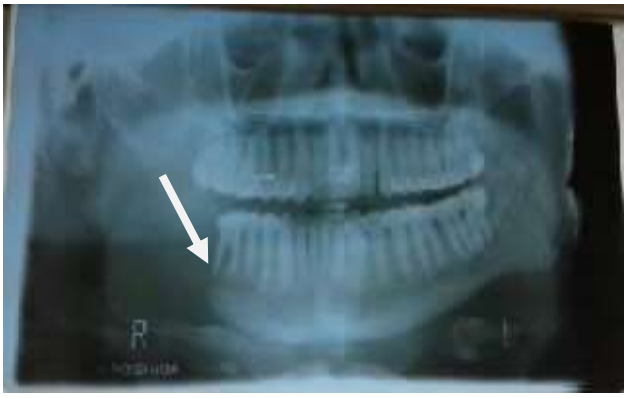

Fig. 1. Orthopantomograph of patient showing loss of mandibular ramus and angle (R)

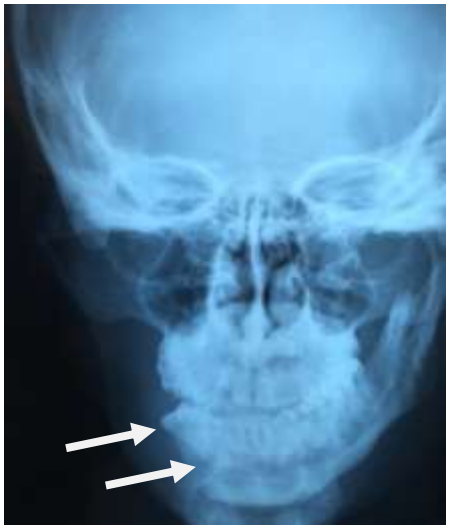

Fig. 2: Digital Skull Xray of patient (Posterior–Anterior view) showing further loss of bone in mandibular body (R)

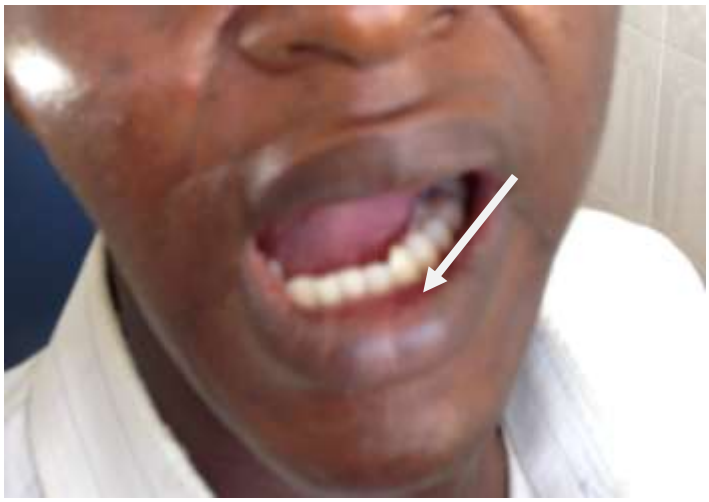

Fig. 3. Photograph of patient showimg deviation of the mandible on opening

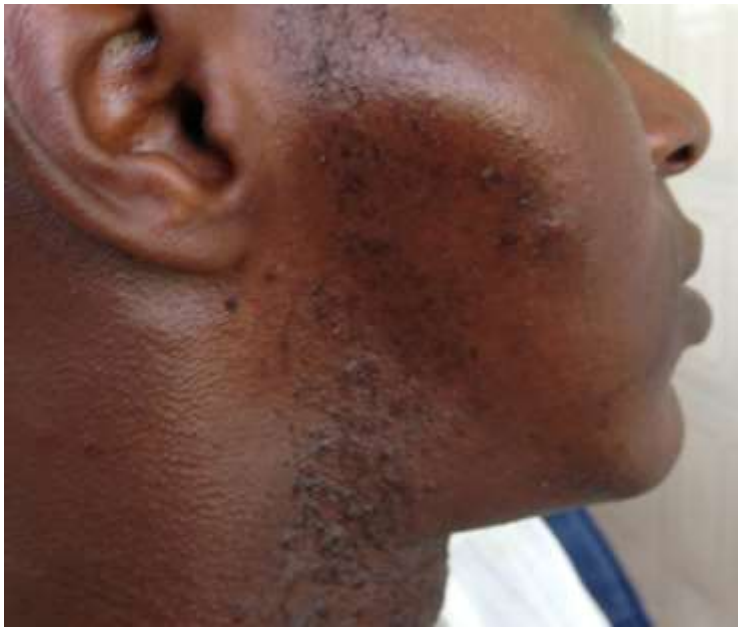

Fig. 4: Photograph of patient showimg loss of bone support in the mandibular ramus and angle region (R)

DISCUSSION

VBD was first described by Jackson in 1838, when he reported the case of a young man with a progressively disappearing humerus [1]. It became known as Gorham-Stout disease in 1955 when Gorham and Stout published a review of 24 cases, with histological features of massive osteolysis [1]. Facial involvement was first reported in 1924 in a 31-year-old woman, and complete mandible lysis in 1933[2].

The process may affect the appendicular or axial skeleton. The bones of the upper extremity and the maxillofacial region are the predominant osseous locations of the disease; with cases being reported in the skull (8 cases), maxillofacial region (42 cases), spine (18 cases), pelvis (18 cases), trunk (including clavicle and ribs) (35 cases), upper extremity (including scapula) (41 cases), and lower extremity (22 cases), in addition to multicentric involvement (11 cases) [3]. Fourteen cases have been reported that involved multiple contiguous bones of the head, which represents a more advanced stage of the disease [4]. The maxilla was never involved alone, whereas the mandible was affected alone by the osteolysis, partially or completely, in 23 cases [3]. This report is yet another case; as so far, there is no evidence of the disease in any other part of the patient except in the mandible. The outcome in this case though, is yet uncertain as the disease process has not halted.

The disorder starts in childhood but has been described in patients ranging in age from 1month to 75 years [2]. In more than one third of the cases, the age at presentation is above 35 years. This patient was twenty

five years when he presented to us, though the condition might have started earlier.

Sexual predilection is not clearly established, however, Huvos et al reported approximately 60% of all cases with vanishing bone disease occur in men [5]. as is the case of the patient presented in this report.

30% of cases with maxillofacial involvement presents with pain, malocclusion, and deformity [6] 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 [5]. In this case, we think that the pain and difficulty to open his mouth, encountered abruptly by the patient, may have been due to a pathological fracture he suffered suddenly; this then revealed the disease that might have been present for some time. However, the patients main complaint as the desease progressed, was that of severe pain, especially to touch, in the right lower jaw to the midline region; characteristic of neuralgic pain involving the right inferior alveolar nerve. We believe this kind of pain is quite significant, as to the best of our knowledge, it has not been previously associated with VBD of the mandible in previous reports.

Thorough history and meticulous physical examination, appropriate blood investigations and radiographic studies are needed to rule out other common underlying causes of osteolysis, such as infection, cancer, and inflammatory or endocrine disorders. The diagnosis of Gorham-Stout syndrome should be suspected or made only after excluding these aforementioned conditions [7]. A biopsy report is required to rule out or confirm aneurysmal bone cyst, and extensive metastatic bone disease, among other diseases that may resemble vanishing bone disease [7]. The patient was investigated thoroughly to rule out any major infectious diseases including tuberculosis, as well as metabolic disease, and all tests turned out negative. A biopsy was done as well, and the result ruled out neoplastic disease.

Criteria for diagnosis has been suggested by Heffez *et al*. [8] and is as follows: Evidence of local progressive osseous resorption; Minimal or no osteoblastic response and an absence of dystrophic calcification; Non-expansile; non-ulcerative lesion; Absence of visceral involvement; Osteolytic radiographic pattern; Negative findings for a hereditary; metabolic, neoplastic, immunologic or infectious origin.

In the case reported here, the following were present: Evidence of local progressive osseous resorption; Minimal or no osteoblastic response and an absence of dystrophic calcification; Non-expansile, non-ulcerative lesion; Absence of visceral involvement; Osteolytic radiographic pattern; Negative findings for a hereditary, metabolic, neoplastic, immunologic or infectious origin.

Radiographic findings in patients with Gorham disease were described by Resnick [9]. Plain radiographs, radioisotope bone scans, computed tomography, and magnetic resonance imaging (MRI) have all been used [9]. During the initial stage of the lesion, radiolucent foci appear in the intramedullary or subcortical regions, resembling findings seen in patchy osteoporosis. Subsequently, slowly progressive atrophy, dissolution, fracture, fragmentation, and disappearance of a portion of the bone occur with tapering or "pointing" of the remaining osseous tissue and atrophy of soft tissues [9]. According to Kiran and Anupama, there will be resorption and decreased vertical height of the mandibular body with the resorption extended toward the basal bone [10]. Infact, the radiographic features in our case were quite consistent with that described above. At the time of first presentation, there was already a pathological fracture with loss of the right mandibular ramus and bone resorption extending toward the basal bone; and after over two years since first reporting the disease process, in the patient presented here, continues without any sign of regression.

There is no standard therapy available for VBD though, various treatment regimes have been suggested [10]. These include radiation therapy, anti-osteoclastic medication (bisphosphonates), and alfa-2b interferon. The principal treatment modalities according to them are surgery and radiation therapy. Surgical options include resection of the lesion and reconstruction by use of bone grafts and/or prostheses; and this was suggested to our patient without a favourable response. The approach to treatment in our case was largely symptomatic resulting in some limited success.The prognosis for patients with Gorham disease is generally good unless vital structures are involved [10], though 16% fatality has been reported [1].

CONCLUSON

Vanishing Bone Disease (VBD) is a serious disease process that can pose a great challenge to the clinician in terms of both diagnosis and treatment. It may as in this case, present as severe toothache, and subsequently as a pathological fracture of the mandible and severe persistent neuralgia. It is important that clinicians bear in mind this condition, when patients present with any of these symptoms.

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(5): 985-1004.
2. Sharma A, Iyer N, Mittal A, Das D, Sharma S; Vanishing mandible. J Oral Sci., 2010;52(3): 513-516.
3. Papadakis SA, Khadi L; Vanishing bone disease: Review and case reports. Orthopedics, 2008; 31(3): 278.
4. Bouloux GF, Walker M, 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(3): 357-361.
5. Huvos AG; Bone Tumors; Diagnosis, Treatment and Prognosis. 2nd edition, Philadelphia, PA, Saunders, 1991.
6. Anavi Y, Sabes WR, Mintz S; Gorham's disease affecting the maxillofacial skeleton. Head Neck, 1989;11(6): 550.
7. Raghuveer HP, Jayalekshmy R; Gorham's massive osteolysis ofthe mandible—A progressive radiographic presentation. Dentomaxillofac Radiol., 2009; 38(5): 292-295.
8. Heffez L, Doku HC, Carter BL, Feeney JE; Perspectives on massive osteolysis. Report of a case and review of the literature. Oral Surg Oral Med Oral Pathol., 1983; 55(4): 331-343.
9. Resnick D; Osteolysis and chondrolysis. In Resnick D editors; Diagnosis of Bone and Joint Disorders. 4th edition,. Philadelphia, PA, Saunders, 2002: 4920-4944.
10. Kiran DN, Anupama A; Vanishing Bone Disease: A Review. J Oral Maxillofac Surg., 2011; 69:199-203.