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CASE REPORT

Paget's Disease of Maxilla

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ABSTRACT

Paget's disease is an idiopathic, non-neoplastic affliction characterized by disordered bone formation and resorption. It affects about 3% of the population. Paget's disease affects mainly the skull, femur, tibia and pelvic bones. Facial skeleton is involved in about 17% of the cases. We present a case of Paget's disease involving maxillary and mandibular skeleton which developed osteomyelitis following extraction of maxillary right posterior teeth.

Keywords: Paget's disease, Maxilla, Osteomyelitis, Hereditary.

INTRODUCTION

Paget's disease (osteitis deformans) is an idiopathic, non-neoplastic affliction characterized by disordered bone formation and resorption.[1] Nature of the disease is unknown, although evidence suggests that it is a multicentric benign tumor of the osteoclasts. The possibility of an infective viral etiology is suggested by ultrastructural demonstration of intranuclear inclusion bodies in abnormal osteoclasts found in these patients.[2]

Paget's affects about 3% of the population[2-4] in the European countries. There are no epidemiological studies or literature showing incidence of Paget's disease in the Asian population. Its occurrence is more common in males (78.6%) than females (21.4%). It is found to be familial in about 40% of the patients.[5] In all familial cases, the existence of atleast one first degree relative is present. In the following patient, chronic osteomyelitis of the maxilla developed following extraction of right maxillary posterior teeth.

CASE REPORT

A 45-year-old female patient reported to the department of oral and maxillofacial surgery with a chief complaint of halitosis and swelling in the right side of upper jaw. Patient gave a history of extraction of three teeth, 6 months before in a private dental clinic following which she developed halitosis and slight discharge in the right side of maxilla. On examination extra-orally, a diffuse swelling was present in the right maxillary region extending from the infraorbital region superiorly to line joining the tragus to corner of the mouth inferiorly. On palpation, swelling had diffuse borders and was firm in consistency. Intraorally entire maxillary alveolus was enlarged and right maxillary alveolus extending from canine to second molar showed necrotic bone exposed into the oral cavity with inflammation of the surrounding mucosa (Fig. 1).

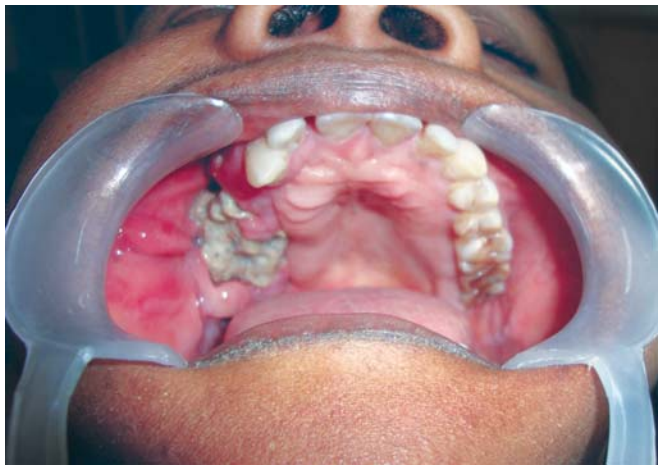

Fig. 1: Preoperative necrotic right maxilla

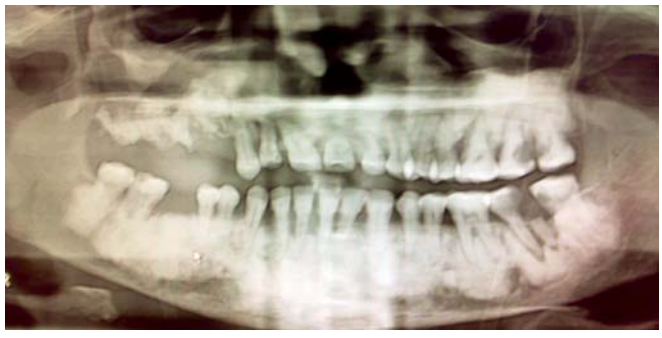

Fig. 2: OPG showing the typical "Cotton Wool" appearance

Orthopantomograph showed cotton wool appearance of upper and lower jaws in the tooth bearing region with loss of lamina dura on all the teeth (Fig. 2). Further investigation with CT scan showed radiolucent and radiopaque areas in the maxilla and mandible with sequestration of the right maxillary alveolus (Figs 3 and 4). Laboratory investigations showed an increase

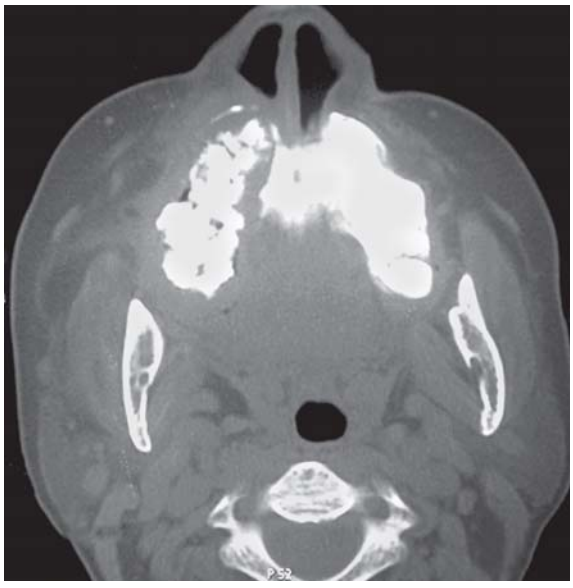

Fig. 3: Axial CT scan of the maxilla

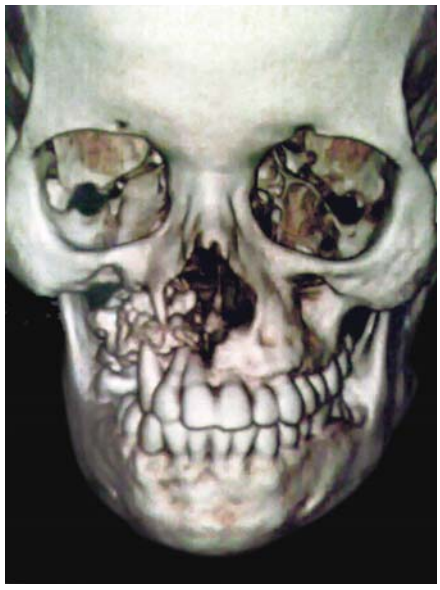

Fig. 4: 3D CT reconstruction showing the necrotic right maxilla

in serum alkaline phosphatase level 242 U/L. Calcium and phosphorous levels were within normal limit. Bone scan of the entire skeleton showed increased mitotic activity only in maxilla and mandible with no other involvement of the skeleton. After clinical and radiographic examination, the case was diagnosed as osteomyelitis of right maxilla due to underlying Paget's disease.

Under local anesthesia, a buccal mucoperiosteal flap was raised extending from the canine to second molar region on right side, necrotic bony sequestrum in the region was removed, curettage of the remaining bone was done. Hemostasis was achieved and a primary closure was done. Patient was prescribed antibiotics and analgesics. Postoperatively, the wound healed uneventfully (Fig. 5). Based on biochemical investigations, it was decided not to put the patient any medication to inhibit.

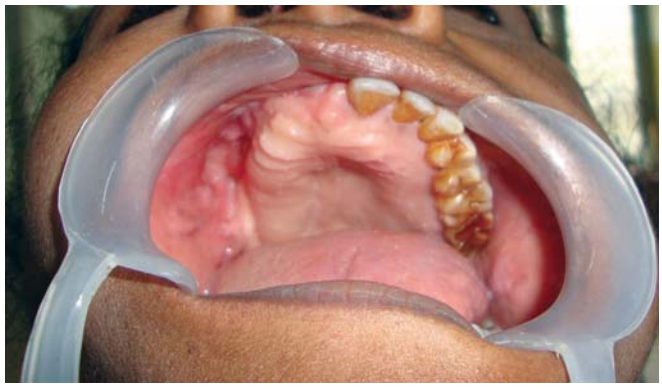

Fig. 5: The right maxilla after curettage 4 weeks postoperative

DISCUSSION

Paget's disease of the bone is a chronic disease of adult skeleton characterized by focal areas of excessive bone resorption followed by bone formation. Radiographically, one sees predominantly osteolytic areas; as the disease progresses there is compensatory almost exuberant overgrowth of bone. The rapidly laid-down bone is soft and extremely vascular responding poorly to stress and resulting deformity, especially in the weight bearing bones. Finally, the disease progresses to the "burned out" stage, where the enlarged bones become well mineralized with decreased vascularity.[6, 7]

Significant observations made in our patient are that she has a family history of Paget's disease with patient's father and a sibling suffering with similar condition. It is stated by Morales et al that in familial cases, age at diagnosis is earlier than sporadic cases as our patient was 45-year-old at the time of diagnosis and already in the burned out stage, sporadic cases generally appear in later decades of life. Radiograph, CT scan and bone scan reports show involvement of only the maxilla and mandible indicating milder form of the disease, which is confirmed by her blood investigations showing mild increase in alkaline phosphatase level. Patient is in the burned out stage of the disease resulting in increased density and decreased vascularity, which has led to fracture of alveolus during extraction. Decreased vascularity has further led to impaired healing of the extraction wound and necrosis.

It has been stated by many authors that bone involved in Paget's disease is highly susceptible to infection when exposed to the oral flora.[3] When there is a necessity for extraction, surgical removal of teeth is recommended along with adequate suturing of the mucosa, followed by antibiotic therapy. In Paget's disease, medical line of treatment mainly includes bone resorption inhibitors, such as calcitonin and bisphosphonates that alter the bone metabolism and biomechanical pattern. Surgical treatment is limited to nerve decompression, arthroplasty and correction of deformities.[8]

REFERENCES

1. Kasper, Braunwald, Fauci. Harrison's Principals of Internal Medicine (16th ed), McGraw Hill Book Company Inc. New York 1966;2279-81.
2. Wood NK, Goaz PW. Differential diagnosis of oral lesions (5th ed). St Louis: Mosby Year Book, 1997:407-08,439-40.
3. Martin S Greenberg, Micheal Glick. Burket's Oral Medicine Diagnosis and Treatment (10th ed). BC Becker Inc, Hamilton Ontorio, Canada.
4. Wen Chen Wang, Yi Shing Lisa Cheng, Chung Ho Chen. Paget's disease of bone in a Chinese patient: A case report and review of literature. Oral Surg Oral Med Oral Oral Pathol Oral Radiol Endo 2005;99:727-33.
5. Morales-Piga AA, Rey-Rey JS, Corres Gonzalez J. Frequency and characteristics of familial aggregation of Paget's Disease of bone. Journal of Bone and Mineral Research 1995;10(4).
6. Howard I Feig, Walter R Edmunds, Richard beaubien. Chronic osteomyelitis of the maxilla secondary to Paget's disease: A complication following dental extraction. J of Oral Surg Oral Med and Oral Pathology Sep 1969;28(3):320-25.
7. Woo TS, Schwartz HC. Unusual presentation of Paget's disease of the maxilla. British Journal of Oral and Maxillofacial Surgery Apr 1995;33(2):98-100.
8. Juan L Martinez, Fernando Almeida, Manue Picon. Maxillomandibular monoblock removal, reshaping, and reinsertion in Paget's disease: 15-year follow-up. J Oral Maxillofac Surg 2005;63:1680-85.