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# Paget's disease of Mandible: A rare case report.

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***Abstract:***
*Paget disease (PD) of bone is a chronic progressive disease of unknown etiology which is characterized initially by an increase in bone resorption, followed by a disorganized and excessive bone formation, causing pain, fracture and deformities. The disease is often asymptomatic and commonly seen in older individuals. The diagnosis of the disease is mostly based on radiographic examination and biochemical markers of bone turnover. Elevated serum alkaline phosphatase is a constant feature while calcium and phosphate levels are typically within normal limits. PD is being successfully treated by bisphosphonate, a group of anti- resorptive drugs, thereby decreasing the morbidity and mortality associated with the disease. We report a case of an eighty year old female patient who presented with complaints of swelling and pain in the jaw. Panoramic radiography accompanied by laboratory findings confirmed the diagnosis which was crucial for choosing the appropriate treatment.*

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## I. Introduction

PD of bone is a rare disorder of unknown etiology, first described by Sir James Paget as "osteitis deformans" in 1877[1]. It is generally diagnosed in patients older than 40 years of age as an incidental finding. It may be monostotic or polyostotic in nature[2]. The most commonly involved sites are, pelvis femur, spine, tibia, skull and humerus[3]. It is a geriatric disease reported above 5$^{th}$ to 6$^{th}$ decades of life, occurring in both in men and women, with male predominance characterized by increased bone remodelling and abnormal bone architecture which is initiated by an increase in osteoclast-mediated bone resorption, accompanied by a compensatory increase in bone formation. This remodelling results in a disorganized mosaic of woven and lamellar bone which is highly vascular and gradually becomes enlarged and structurally weak[4]. The complications include bowing deformity of the long bones, fracture, and a variety of nerve compression syndromes. Malignant degeneration is rare[5].

In 1923, Moore reported that PD of bone may also affect the jaws. Maxillary involvement being much more common as compared to mandible. Novark and Burket in 1944, and Jocobs in 1945 described that the jaw lesions may be the first to present leading to detection of the disease in other parts of the skull or in other bones[6]. Therefore, it becomes imperative to correctly investigate and diagnose a case of PD involving jaws and formulate an appropriate treatment plan. Since mandibular involvement is a rare presentation of this disease, we hereby report a case of PD with mandibular involvement.

## II. Case Report:

Eighty year old female patient reported to Department of Oral Medicine, Diagnosis and Radiology, with complaints of pain in the extraction site of lower right back tooth region of jaw since 2 weeks and restricted mouth opening since the last 3-4 days. History revealed that patient was apparently alright a month ago when she experienced pain in mandibular right posterior region, following which she visited a local dentist where extraction of the offending tooth was done but patient had no relief. Seven days later, patient re-visited the same dentist when, a root piece was discovered in the same extraction socket which was subsequently removed but patient started experiencing worsening of pain and reduced mouth opening. (Figure 1).

When she reported to us, the nature of pain was dull, aching, intermittent in nature and radiating over right temporal area. Patient was unable to eat solid food stuffs due to restricted mouth opening. A detailed extraoral examination revealed (Figure 2) facial asymmetry; left being more prominent than right and prognathism with mandibular enlargement. The overlying skin was normal. On palpation, the swollen left side

of the mandible was bony hard, non- tender. The lower border was thickened and loss of antegonial notch was seen bilaterally. The local temperature was not elevated. Bilateral reduction in movement of temporomandibular joint with no tenderness, clicking or deviation was observed. No abnormality was detected in eyes, ear, nose, hair and skin of face. Lips were incompetent and a single submandibular lymph node was palpable on right side, which was firm in consistency, mobile and tender in nature. Intraoral examination showed increased size and altered shape of the mandibular arch, and partially edentulous, thickened cortical plate. Trismus secondary to infected socket in 46 and 47 region was noted. The pain was caused by these infected sockets. Oral analgesics and antibiotics were administered and patient was advised to follow up with investigations.

Orthopentamagram (OPG) showed generalised mandibular enlargement, cortical thickening of the body of the mandible, and mixed radiolucent and radio opaque areas on both sides of the mandible, Cotton wool appearance, elongated trabeculae in reticular manner, loss of lamina dura and layered sclerotic inferior border of mandible. Inferior dental canal was not evident bilaterally (Figure 3). Cross sectional mandibular occlusal radiograph showed thickening of body of mandible on the affected side (Figure 4)

Intraoral periapical radiograph (IOPA) of maxillary anterior region revealed hypercementosis (Figure 5). Whole-body X-rays and scans along with Posteroanterior view of skull were done to rule out any extra mandibular involvement. To confirm the diagnosis, patient was advised to get her previous photographs and mandibular radiographs if any.

Biochemical analysis showed elevated alkaline phosphatase (ALP) which was 270 IU/L (normal range 37-147 IU/L). Serum calcium was normal with value 9.8 mg/dl (normal range 8.2-10.4 mg/dl) (Figure 6). Bone biopsy showed increased osteoblastic activity and irregular cortical bones, numerous irregular bony trabeculae interconnected at places with presence of basophilic reversal line giving the characteristic mosaic pattern. Marrow spaces were filled with fibro vascular connective tissue, confirming osteitis deformans. Figure (8)

Based on the clinical, radiographic, histopathological, and biochemical findings, a diagnosis of mandibular PD was made and patient was counselled. Oral prophylaxis was performed on the patient, Root piece were extracted with respect to 22, 23. Conservative and endodontic treatment was advised for 31, 32, 33,34 ,41, 42,43,44. Prosthetic replacement of missing teeth along with surgical contouring of mandible was advised. Calcitonin and Bisphosphonates were administered.

## III. Discussion:

Prevalence of PD is reported to be between 0.01% and 3% in patients above the age of 40 (Barnett & Elfenbein, 1985) increasing to 10% in patients above the age of 70 (Monteiro & Rout, 2007). It may have jaw bone changes similar to those seen in Fibrous Dysplasia (FD), Florid Osseus dysplasia(FOD) and other bone disorders. Since our case showed mixed radiopaque radiolucent lesion in the radiograph, it became difficult to differentiate it from FD; a bone disorder which also causes expansion of jaws. The patients age, absence of ground glass appearance along with localised enlargement helped in differentiating our case. [7] Cotton wool appearance, mixed radiopaque radiolucent lesion seen in patients older than 30 years, are some of characteristics which were observed in our patient common to those seen in FOD. But, absence of localised enlargement of jaw and radio opacities rimmed with radiolucent borders helped us differentiate it from the same.[8]

We advised the patient to get some previous photographs where mild facial asymmetry was observed since 2008 when patient was 74 years old. This asymmetry has gradually progressed to the present state. When the patient reported to us, she complaint of pain in extraction site of lower right back tooth region of jaw for 15 days and restricted mouth opening for 3-4 days. On examination, right submandibular lymph node was found palpable which was attributed to infection in the extraction site. Thus, the pain was due to the infected socket and the swelling on the opposite side when investigated revealed PD, making it an incidental finding. Bone biopsy was done to confirm the diagnosis of PD before starting treatment to exclude other pathologies such as FD, FOD and osteomyelitis.

Treatment of PD includes medication to help regulate bone remodelling, relieve pain, physical therapy and surgery. Bisphosphonates are the first-choice treatment to regulate bone growth. They work by controlling the cells that absorb old bone, which means the bone regeneration process should return to normal. A type of bisphosphonate called risedronate has proved to be effective in treating Paget's disease and is usually recommended.

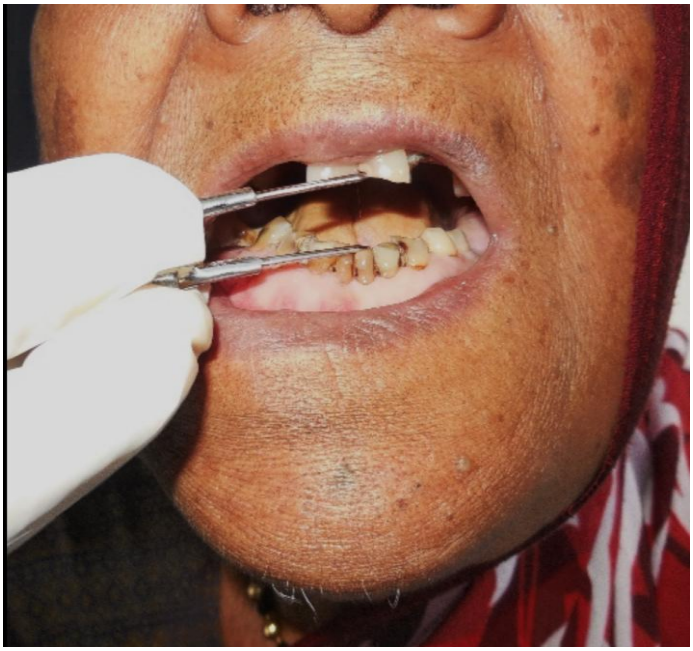

Figure 1 Showing reduced mouth opening

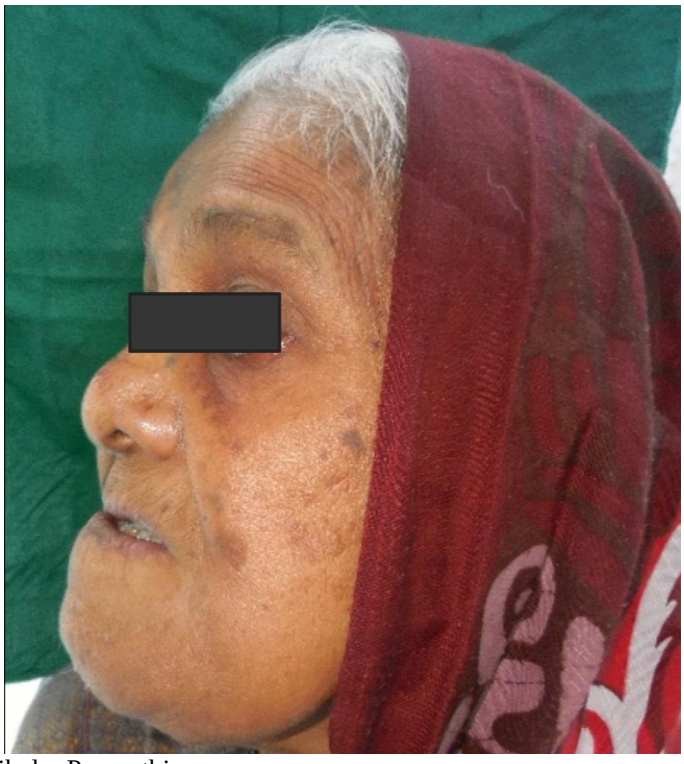

Figure 2 Showing Mandibular Prognathism

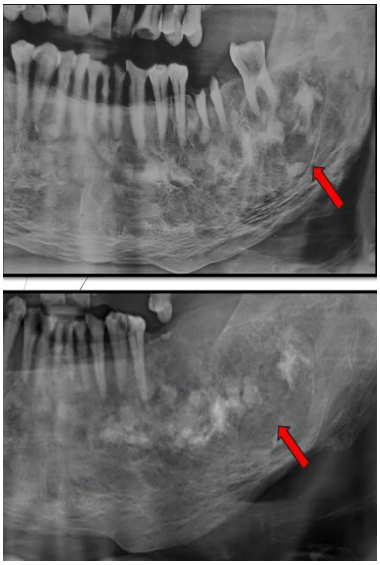

Figure 3 Orthopantomograph

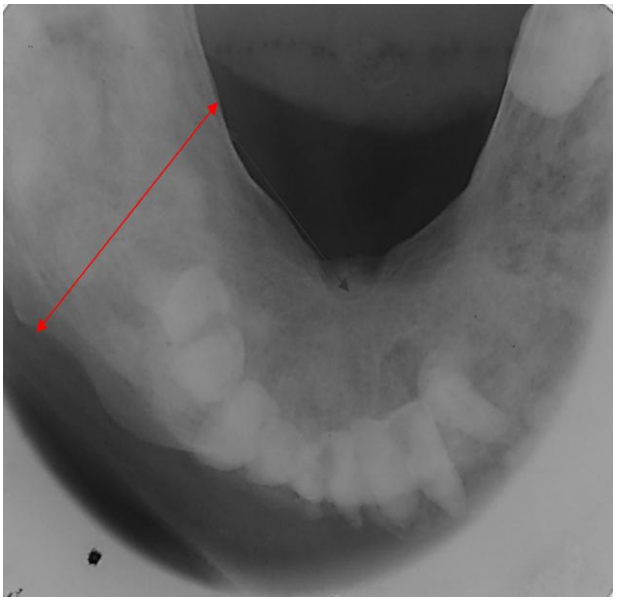

Figure 4 Cross sectional mandibular occlusal radiograph showed thickening of body of mandible on the affected side

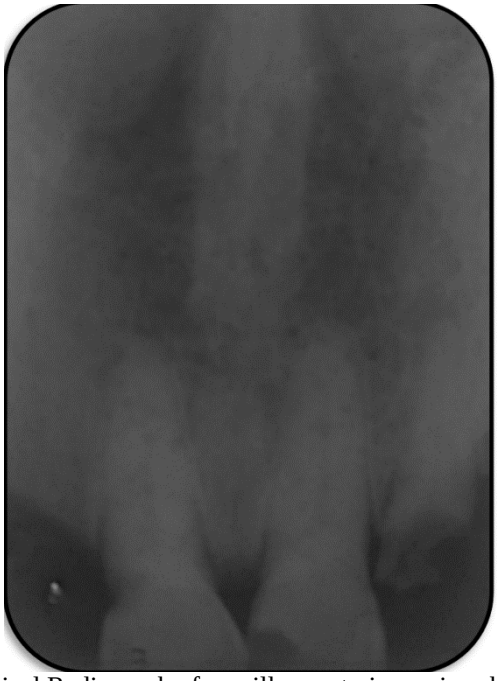

Figure 5 Intraoral Periapical Radiograph of maxillary anterior region showing Hypercementosis

PATIENT'S NAME : CHANDRABHAGA SOMWANSHI SEX : F

REF. BY. DR : DR. ATUL P. DATE : 12/9/14

EXAMINATION OF BLOOD

| TEST | RESULT | NORMAL |
|---|---|---|
| S. CALCIUM | : 9.8 MG/DL | 8.2 – 10.4 MG/DL |
| S. ALKALINE PHOSPHATASE | : 270 IU/L | 37 – 147 IU/L |

THANKS

Neha

DR. MRS. NEHA M. DESHMUKH
M.D. (PATH)

Figure 6: Biochemical analysis showing elevated serum alkaline phosphatase level

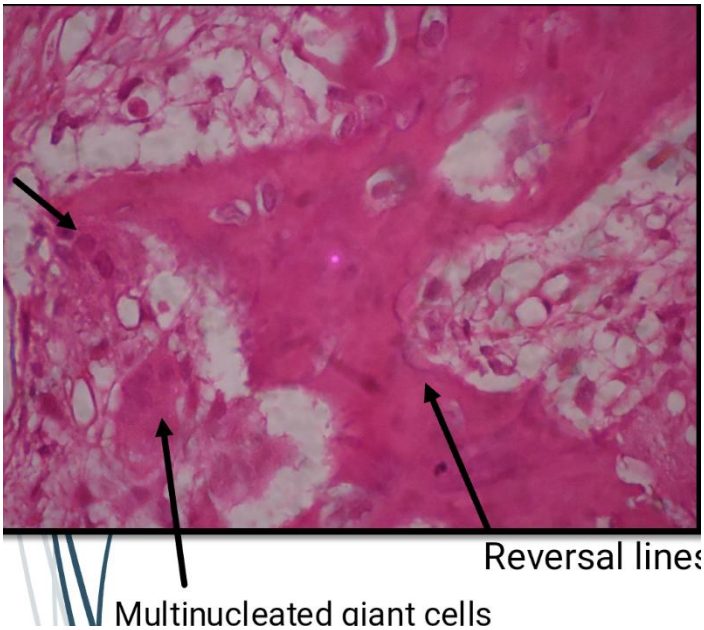


Figure 7 Bone biopsy showing numerous irregular bony trabeculae interconnected at places with presence of basophilic reversal line and multinucleated giant cells giving the characteristic mosaic pattern.

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