Case Report
Journal of Pierre Fauchard Academy (India Section), *Vol 36(2), DOI: 10.18311/jpfa/2022/31026, June 2022 p. 71-75*
ISSN (Print) : 0970-2199
ISSN (Online) : 2405-772X

Paget's Disease with Osteomyelitis of Maxilla – A Case Report

Mahalakshmi Loganathan, Thirumal Mohan* and Suman Jhansi Lakshmi

KSR Institute of Dental Science and Research, Tiruchengode – 637215, Tamil Nadu, India; mthirumal089@gmail.com

Abstract

Paget's Disease is a rare bone disease, with altered osteoclastic activity and osteoblastic activity in same area leads to production of poorly mineralized woven bone. PD has an autosomal dominant inheritance pattern. It may be present with concomitant infection and bony inflammation affecting the surrounding periosteum presenting as pagetoid osteomyelitis. Here, we are presenting a unique case of Paget's Disease with associated osteomyelitis. In this article we report an atypical presentation of Paget's Disease with osteomyelitis in maxilla, its biochemical, radiographical and histopathological findings and its effective management by alendronate.

Keywords: Osteoclast, Osteomyelitis, Paget's Disease, Resorption

1. Introduction

In late 19th century, Sir James Paget was the one who firstly coined Paget's Disease (PD) as a bony disorder under the term "osteitis deformans"[1]. Bone is an active living tissue which will frequently undergo modifications. There are some distinct molecular mechanisms which makes progenitor cells to differentiate as osteoblasts and osteoclasts. Cytokines, hormones and repeated mechanical stimuli modifies the bone turnover. These factors decide the amount and character of tissue alterations[2]. It is relatively an uncommon bony disorder characterized by repeated phases of excessive and uncoordinated new bone formation. Finally, it results in formation of large masses of structurally defective bone in different parts of skeleton, which often causes severe distortion and weakening of skeletal tissues as the age advances predominantly in the 5th and 6th decade of life. Its etiology is still unknown but some of predisposing factors are hereditary, Viruses like (Paramyxovirus, Canine distemper virus, respiratory syncytial virus[1]. In this article we report an atypical presentation of Paget's Disease with osteomyelitis in maxilla and its effective management by Alendronate.

2. Case Report

A geriatric male patient, 62 years age came to our institution with swelling and teeth loosening in the right side of upper jaw since past 6 months. History of presenting illness revealed that patient he had undergone extraction of his upper right back tooth six months ago. After few weeks, he noticed a small swelling in the same region which gradually increased in size. Few weeks later, he noticed a small swelling in the upper right back teeth region which was slower in onset, initially smaller in size and gradually progressed to present size. Patient had also given history of intermittent dull aching pain which aggravated on mastication and relieved on medication. He was under analgesics prescribed by a general dentist during previous extraction. He had a habit of betel chewing, three times a day and discontinued 3 months ago. He had no relevant familial history on diabetes mellitus, hemophilia, hypertension and any malignancies.

Extraorally, a diffuse swelling on the right side of face was noticed (middle third region) measuring about 3 x 5 cms approximately and extending 2 cm below the inferior eyelid inferiorly till the line joining the right corner of the mouth and right ear lobe. Mediolaterally one cm from the

*Author for correspondence



Figure 1. **A**. Extraoral image showing diffuse swelling seen on the right middle third of face (Blue arrow head); B. Intraoral image showing diffuse swelling seen on the right hard palate (Black arrow head) and missing 14, 15, 16.

lateral margin of ala of nose to 3 cm in front of the tragus of the auricle (Figure 1A). The swelling was found to be obliterating the nasolabial fold on the right side but with normal overlying skin in appearance. On palpation of the swollen region, it was found painful, hard but slightly pinchable.

Intraoral examination revealed that the swelling was located on the right maxillary alveolar ridge in relation to edentulous region of tooth #14, #15, #16 and the dentulous region of tooth #17, #18 and till the tuberosity measuring 6 x 3 cms approximately (Figure 1B). It was extending buccally causing obliteration of the upper buccal sulcus and palatally till 2 cm from the midline. The surface of the swelling appeared erythematous and smooth without any secondary changes. On palpating, the consistency of the swelling was hard andtender on mild pressure. The associated tooth #17 was also tender on percussion. The buccal aspect dentoalveolar region of tooth #16 showed evidence of discharge of pus.

Orthopantomogram (OPG) of this patient revealed that there was a mixed radiolucent and radio opaque lesion seen in the right maxilla marking its boundary from distal aspect of permanent right canine to the tuberosity region. Diffuse area of mixed radiodensity marking its boundary from the crest of alveoli to the right maxillary antrum. The radiopacity was heterogeneous in nature (Figure 2). Resultant images of Computed Tomography of upper jaw (plain and contrast of Maxilla) revealed osteolytic lesion in the alveolar process of right maxilla in relation to the level of molars and premolars and extends till the soft tissue structures of right cheek region. No abnormalities were detected in right maxillary air sinus and it appears normal (Figure 3).

After all these investigations, the provisional diagnosis of "Fibro osseous lesion of maxilla" was given. To diagnose the type of fibro osseous lesion an incisional biopsy was performed under local anesthesia in the right maxillary alveolar ridge for histopathological evaluation. Microscopic examination of H and E stained decalcified sections showed fibro-cellular connective tissue stroma along with numerous irregular bony trabeculae showing prominent reversal lines, osteoblastic rimming and osteoclasts. Bony trabeculae without osteocytes suggestive of non-vital bone were evident in few areas. The connective tissue stroma was made up of dense inflammatory cell infiltrate and blood vessels of varying sizes (Figure 4A to 4D). Correlating all the clinical, radiological, biochemical and histopathological findings, a final diagnosis of Paget's Disease of maxilla with osteomyelitiswas given.

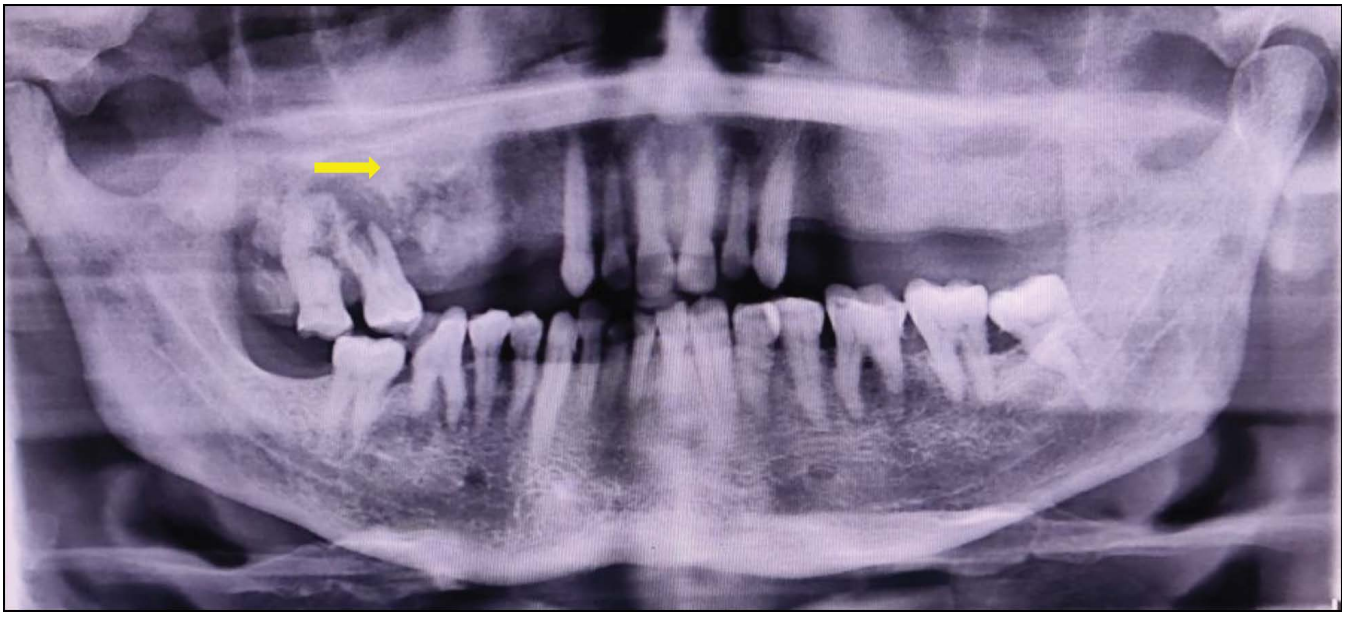


Figure 2. Orthopantomogram shows the lesion with mixed radiolucent-radio-opaque in the right maxillary region.

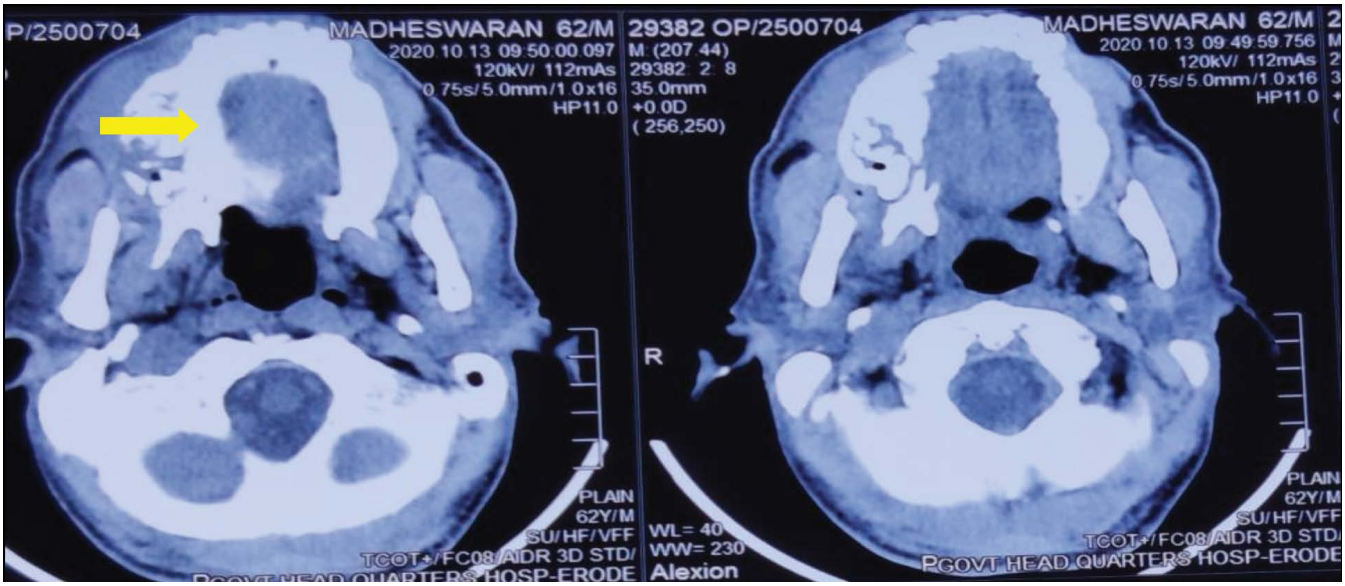


Figure 3. Computed tomography image shows mixed radiolucency and radiopacity in the right maxilla.

3. Discussion

"Osteitishyperplastica" term was the coined in 1947 by Weinman and Sicher. In 1877, Sir James Paget, first named the condition as "Paget's Disease of bone or Osteitis Deformans"[4]. PD is a geriatric pathology and affects both the gender, with male predominance of approximately 3:2 ratio[5].

Etiology of PD is unknown, but there is a hypothesis that vitamin D deficiency in children may express later as PD. Wide variations in prevalence are not completely understood. Studies have suggested that it was found to be associated with Virus (Paramyxovirus, respiratory syncytial virus and canine distempervirus)[1,6]. It is due to defective function (inactivation and mutation) in the Osteoprotegrin/TNFRSF11A or TNFRSF11B or B-RANKL/RANK pathway, the molecular regulators of osteoclastogenesis. Mutations in SQSTM1 (p62 are often encountered in classic PD. The pathology involves disordered bone formation that results in overall

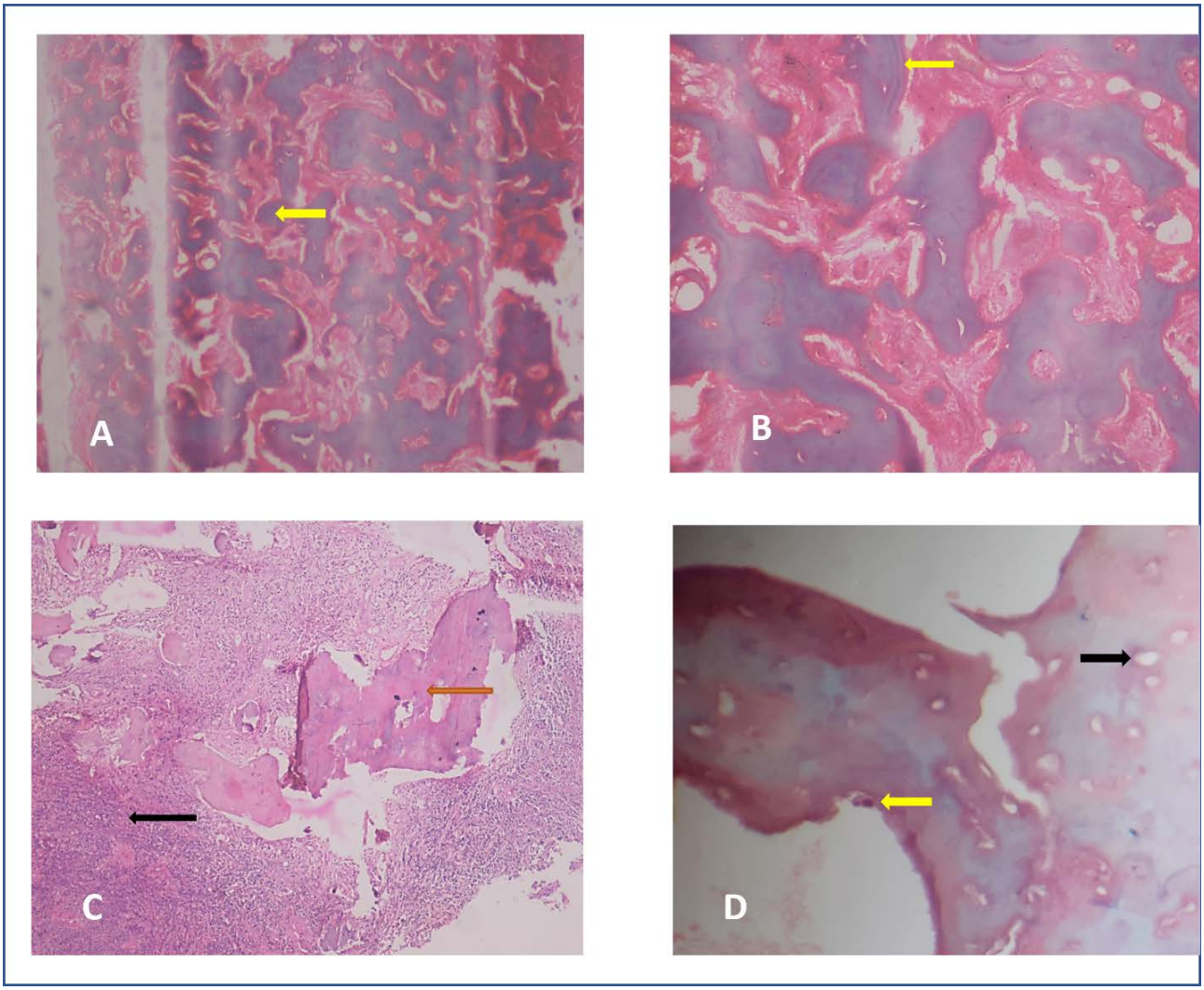


Figure 4. Histopathological images depicts **A)** Numerous irregular bony trabaculae (Yellow arrow head); **B)** Irregular bony trabaculae with prominent reversal lines (Yellow arrow head); **C)** Osteoblastic rimming (brown arrow head) and fibrocellular connective tissue stroma (black arrow head); **D)** Osteoclast (Yellow arrow head) and bony trabaculae with empty lacunae (Black arrow head).

abnormal bone remodeling and is the key primary event in PD.

PD occurs usually in three major stages:

- Lytic stage, it is characterized by predominant osteoclastic resorption.
- Mixed stage, involves irregular fashion of both osteoclastic and osteoblastic events with predominant osteoblastic activity.
- Osteoblastic stage, a late inactive one and has gradually decreased osteoclastic activity and the osteoblastic phase becomes dominant, resulting in excessive bone formation which is fibrous and coarse.

These unorganized bone remodeling process results in formation of disorganized new immature or woven bone (also called as mosaic pattern) and it may involve any skeletal bone[7,8].

Paget's Disease with involvement of multiple bones (Polyostotic) is more common than involvement of single bone (Monostotic)[9]. Usually, the clinical signs and symptoms varies based of predominant stage of PD as all these three phases can be present in single PD patient at different sites on same time. As PD is chronic disease the clinical symptoms will develop slowly and patients with PD may experience pain in bone, severe headaches, dizziness, ear deafness, visual disturbances, facial paralysis,

mental and visual disturbances. PD patients may remain asymptomaticand is usually diagnosed accidentally during radiographs and laboratory investigations.

A variety of deformities commonly observed include kyphosis; shortened or bowed limbs, leonine facies; frontal bossing of the forehead; dental abnormalities such as spacing between teeth leading to malocclusion and the loss of teeth and in aggressive cases, because of the cranial enlargement it may be difficult to hold erect[4,10].

The patient with PD of jaw often presents with progressive enlargement of maxilla rather than mandible which result in increased dental arch size with teeth spacing out. Eventually, dentures cease to fit properly. The inverted triangular facial contour is due to the discrepancy of both upper and lower jaws. Teeth will become mobile or tends to migrate pathologically due to widened alveolar ridge[4]. Biochemical markers of bone turnover and radiological examinations plays a vital role in diagnosing PD. Calcium phosphates will remain normal but the Serum Alkaline Phosphatase (SAP) will be elevated for PD patients.

As like clinical findings, the radiographic findings will be varied from phase to phase for this disease. The resorptive stage exhibits as radiolucent lesion and depositional stage exhibits as radio paque lesion. The earliest lesions are most appreciable in skull because it is osteolytic in nature. Radiograph shows typical cotton wool appearance as a result of increased bone formation in same areas and its patchy distribution. Root apices of numerous teeth reveals hypercementosis or club like appearance. Sometimes all the lower teeth or all the upper teeth will show loss of lamina dura, but never in both. Occasionally, lamina dura will be absent in one or more teeth in either or both jaws[2].

Provisional diagnosis of PD will include fibrous dysplasia, ossifying fibroma, condensing osteitis, osteosarcoma and osteoblastoma as clinically and radiographically. However, Paget's Disease can be easily differentiated from the above mentioned lesions on proper histopathological evaluation.

4. Conclusion

Paget's Disease with osteomyelitis is not very common in English language literature. Even though the condition of Paget's Disease can be symptomless for a long period of time, when symptoms do occur, they may include severe bone deformities and pain in the affected area. Hence, careful and earlier examination of same with clinical, radiographical and histopathological findings will help us to treat the patient for better prognosis.

5. References

1. Karunakaran K, Murugesan P, Rajeshwar G, Babu S. Paget's Disease of the mandible. J Oral Maxillofac Pathol. 2012; 16(1):107–9. PMid: 22434946 PMCid: PMC3303501. https://doi.org/10.4103/0973-029X.92984
2. Gupta S, Jawanda MK. Paget's Disease of bone with special reference to dentistry: An insight. Int J Oral Health Sci. 2020; 10:19–25.
3. Kravets I. Paget's Disease of bone: Diagnosis and treatment. Am J Med. 2018; 131(11):1298–303. PMid: 29752905. https://doi.org/10.1016/j.amjmed.2018.04.028
4. Chaudhary M, Prabhakar I, Seth N, Kushwaha S, Sehrawat P. Paget's Disease of Maxilla: A Case Report. IHRJ. 2017; 10(4):15–9.
5. Ripp GA. A complication after extraction in a patient with advanced Paget's Disease. Oral Surg Oral Med Oral Pathol.1972; 33:35–40. https://doi.org/10.1016/0030-4220(72)90205-8
6. Hansen T, *et al.* Osteonecrosis of the jaws in patients treated with bisphosphonates - histomorphologic analysis in comparison with infected osteoradionecrosis. J Oral Pathol Med. 2006; 35(3):155–60. PMid: 16454811. https://doi.org/10.1111/j.1600-0714.2006.00391.x
7. Theodorou DJ, *et al.* Imaging of Paget Disease of bone and its musculoskeletal complications: Review. AJR Am J Roentgenol. 2011; 196: S64–S75. PMid: 21606236. https://doi.org/10.2214/AJR.10.7222
8. Eversole R, *et al.* Benign fibro-osseous lesions of the craniofacial complex. A review. Head Neck Pathol. 2008; 2(3):177–202. PMid: 20614314 PMCid: PMC2807558. https://doi.org/10.1007/s12105-008-0057-2
9. Sadaksharam J, *et al.* A typical presentation of Paget's Disease with secondary osteomyelitis of mandible. J Indian Acad Oral Med Radiol. 2017; 29(3):227–30. https://doi.org/10.4103/jiaomr.jiaomr_54_17
10. Paul Tuck S, *et al.* Adult Paget's disease of bone: A review. Rheumatology (Oxford). 2017; 56(12):2050–9. PMid: 28339664. https://doi.org/10.1093/rheumatology/kew430