INTRODUCTION

Paget’s disease (PD) of bone was first described, in 1877, by Sir James Paget under the term “osteitis deformans” and then was later given his name. He credited the first use of the term osteitis deformans to Czerny who described an acute inflammation of the tibia in a young man in 1873. However, Wtany in 1867 and Wilks in 1869 described the disease several years before Paget (Barry 1969).

The aetiology of Paget’s disease is, strictly speaking, unknown. Paget first described this disorder in 1877 as a chronic, non-infectious inflammatory disease that affects bone. Later, however, circulatory disturbances and viral infections were also found to be associated with this disease. However, by and large, genetic and environmental factors have been implicated in the pathogenesis of this disease. It is an osseous dysplasia seen in the middle-aged and elderly and is the second commonest osteodystrophic condition. The reported overall prevalence of Paget’s disease in population surveys ranges from 0.5% to 10% of individuals over 40 years of age, and increases in incidence with age (by 0.3% per annum after 55 years). Paget’s disease is rare in patients under 40 years, and has a male: female ratio of approximately 3:1:2.

The primary early feature of this disease is the increase in bone resorption and replacement with poorly mineralized, less dense, fibrotic bone with disorganized trabecular patterns. The disease process overcompensates with bulk for the replaced bone, and most commonly affects the weight bearing bones, especially the sacrum, spine, femur, and tibia. In some cases, Paget’s disease of bone may affect the jaws which has been known since Moore’s (1923) report of a case in which there was a bony tumour of the maxilla together with osteoporosis circumscripta of the frontal region, though until quite recently the disease in this site has been considered rare. The maxilla is much more often involved than the mandible.

PDB can be monostotic or polyostotic, the former being the more common form of the disease, affecting the axial skeleton. All bones of the craniofacial complex however, may be affected to varying degrees. It is characterized by rapid bone resorption and deposition, resulting in formation of numerous reversal lines, which give rise to a mosaic pattern in the lamellar bone with profuse local vascularity and fibrous tissue in the marrow. In the initial phase of PD, there is excessive bone resorption followed by increased deposition. However, both may occur simultaneously, resulting in osteoporosis and sclerotic bone. Within the sclerotic bone there is reduced vascularity (localized vascularity) and the ability for normal healing is compromised, hence...
it can lead to osteonecrosis.\textsuperscript{10}

\textbf{CASE REPORT}

An 84-year-old male reported to a village camp in the outskirts of Greater Noida, Uttar Pradesh, India with the chief complaint of pain and growth on the right side of the upper jaw since the past 4-5 months. The patient gave history of a traumatic extraction of his posterior teeth he got 6 months ago following which he started noticing a painful growth inside his mouth along with a swelling on the face which had increased with time (Figure 1). He also complained of halitosis. The medical history revealed that patient had been suffering from pain in his legs since the past 10 years for which he had not seen a qualified doctor, but instead had seen a healer at his village who referred him to a nearby hospital for blood examination. He used a crutch to walk around. A blood report shown by him, however showed increased serum alkaline phosphatase level of 246 U/L. The calcium and phosphorus levels were within normal limits.

Vital signs were within normal limits. On examination extraorally, a diffuse swelling was present in the left 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, the left maxillary alveolus extending from canine to second molar showed necrotic bone exposed into the oral cavity (Figure 2). Similar kind of deposition was also seen on the right side and in the maxillary rugae area but the extent of involvement was small as compared to the left side (Figure 3). The mucosa surrounding that area was pale pink in color and was tender on palpation.

The patient was called for further examination and investigations to the institute but due to some reason he could not show up. However, on the camp, he was advised to get a few investigations done in the nearby hospital in case he is not able to get to our hospital. These included an OPG, biochemical tests for serum calcium, phosphorus and alkaline phosphatase. The hospital at the village did not have the facilities for doing an OPG. On the other hand, the biochemical tests revealed normal serum calcium and phosphorus (9.0 mg/dL and 3.8 mg/dl respectively), but markedly raised serum alkaline phosphatase (SAP) upto 1326 IU.

Considering the examination done and the results revealed in the examination, it was diagnosed as a case of Paget's disease.

\textbf{DISCUSSION}

Both the eponym "Paget's disease of bone" and "osteitis deformans" are being recently used but a variety of other titles had been given to this disease. For example, Weinman & Sicher (1947) used the term "osteitis hyperplastica" and Schuller (1926) suggested the term "osteoporosis circumscripta" to decumscripta" to describe characteristic changes seen in skull radiographs. Virchow (1862) was the first to use the nonspecific term "leontiasis ossea" to describe the enlargement of the facial bones sometimes seen in Paget's disease (Drury 1962).\textsuperscript{4}

The incidence of Paget's disease as jaw lesions has been uncertain, as most of the published reports concern single cases or relatively small series. The prevalence of Paget disease shows wide geographical variation, common in Western Europe, Americas, and Australia, but rare in Asia and Africa. However, recent studies have reported an unexplained reduction in both prevalence and disease severity. This disease is relatively common in older people, occurs in approximately 3-4\% of the population aged over 50 years with a slight male gender predilection.\textsuperscript{9} Shankar YU\textsuperscript{9} and Rajkumar GC\textsuperscript{6} published an intensive case report on Paget's disease of bone involving the dentofacial region in the year 2013 and 2011 respectively. Another case was reported by Venkatesh R\textsuperscript{7} in the year 2011. These three cases were reported in the recent times in southern parts of India namely, Karnataka and Odisha. As an exception to small case series and reports, a report by Stafne and Austin (1938), who found 23 cases with mandibular or maxillary involvement out of 138 cases in which the skull was affected.\textsuperscript{3}

The aetiology of Paget's disease, strictly speaking, has been unknown. It has been frequently associated with infection (especially viral infections), erroneous connective tissue biosynthesis, autoimmunity etc.\textsuperscript{4} Genetic factors,
although, have clearly been an important component of the etiology of PDB since 15-40% of affected patients have a first-degree relative with PDB.\textsuperscript{10}

The primary early feature of this disease is the increase in bone resorption and replacement with poorly mineralized, less dense, fibrotic bone with disorganized trabecular patterns. The disease may occur in 3 stages:

1. An early osteoclastic stage,
2. An intermediate stage which demonstrates both osteoclastic and osteoblastic activity, and
3. A mature stage, in which osteoblastic activity predominates.\textsuperscript{5}

In the early phase, termed "osteolytic phase", bone resorption predominates and there is a concomitant increased vascularity of involved bones. Commonly, the excessive resorption of pagetic bone by osteoclasts is followed closely by formation of new bone. During this second phase of the disease ("osteoblastic phase"), the new bone that is made is structurally abnormal, presumably because of the accelerated nature of the remodeling process. Newly deposited collagen fibers are laid down in a disorganized rather than a linear manner, creating the so called "woven bone". With time, the hypercellularity at the affected bone may diminish leading to development of a sclerotic, less vascular Pagetic mosaic bone without any evidence of active bone turnover. This is the so-called "sclerotic" or "burned-out" phase of PDB. Typically, all these three phases of the disease can be seen at the same time at different sites in a single pagetic patient.\textsuperscript{9} Clinical symptoms develop slowly due to the chronicity of the disease. Symptoms most commonly encountered are bone pain, severe headaches, dizziness, deafness, visual disturbances, facial paralysis, weakness, and mental disturbances. The signs of the disease are not always obvious until it has become relatively far advanced.\textsuperscript{9} Asymptomatic patients are usually diagnosed accidentally on radiographs and during laboratory investigations. Patients with bone pain caused by Paget’s disease usually describe the pain as continuous which increases with rest, and at night. 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 severe cases, an enlarged cranium that may be difficult to hold erect.\textsuperscript{9}

The patient with Paget’s disease affecting the jaw complains of progressive enlargement of the maxilla or, much less frequently, the mandible. As a result of the increase in size in the dental arch the teeth become spaced, and if dentures are worn these cease to fit properly. The disproportion in size between the maxilla and the mandible gives rise to the inverted triangle type of facial contour. The alveolar ridges may become widened and the palate flattened. Teeth, when present may become loose or migrate.\textsuperscript{4,8,9}

The radiographic features vary with the stage of the disease.\textsuperscript{4,5} The most common radiographic manifestation of Paget’s disease is a replacement of normal trabecular bone by dense, granular bone which presents as radiopaque areas. This may be accompanied by generalised or focal enlargement of the jaw. Another radiographic appearance of Paget’s disease of the jaws is focal areas of radiolucency. These sometimes have irregular margins and simulate the appearance of metastatic tumour. When such areas of radiolucency involve the roots of teeth the lamina dura is lost.\textsuperscript{4,12}

When bone collagen is resorbed the amino acids hydroxyproline and hydroxylysine are not recycled for new collagen production. Thus, hydroxyproline released during bone degradation is either metabolised or excreted in the urine. The amount excreted in the urine in Paget’s disease may be considerably increased during the osteolytic phase of the disease (e.g., from normal level of 40 mg/24 h to 1 g/24 h). The serum alkaline phosphatase level is raised during osteoblastic phases of the disease, sometimes up to 100 Bodansky units per 100 ml in the polyostotic form.\textsuperscript{13}

The most frequent complications of Paget’s disease of the jaws are associated with dental extractions. Hypercementosis and ankylosis may necessitate surgical extraction of teeth. This may be complicated by excessive bleeding in the highly vascular lytic phase of the disease, or post-operatively by poor healing and infection in the avascular phase. Antibiotics are therefore
indicated in these patients, as is careful surgical technique.²

As the aetiology of Paget’s disease is uncertain, treatment has been mainly symptomatic and empirical. Enlarged jaws may need to be reduced surgically for cosmetic reasons. For relieving pain, aspirin has been used and injections of phenol into the sympathetic chain and xylocaine into the epidural space have been used for extreme cases. Also restoration of negative calcium balance to normal levels after oral administration of sodium fluoride has been recommended. The exact mode of action was unknown but it was thought to be related to replacement of hydroxyl and carbonate ions in the bone by fluoride ions. The response to radiotherapy is variable and is usually not recommended in view of the intrinsic potential for the lesion to undergo malignant change. The only therapeutic agents that have had any consistent success are calcitonin, diphosphonates and mithramycin.⁷

CONCLUSION
Paget’s disease presents itself as an asymptomatic lesion until it progresses into an advanced stage. In our report, we based our diagnosis on intraoral examination and blood investigations, although radiographic examination forms a very crucial part of the diagnosis especially diseases which involve the bone. Traumatic extraction seems to be the most likely cause of this disease in this case as similar observations have been reported in other reports. Early diagnosis and appropriate treatment depending on the extent of the lesion can help prevent further complications.

REFERENCES
Paget’s Disease of Maxilla: A Case Report


Source of support: Nil, Conflict of interest: None declared

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LEGENDS

Figure 1. Frontal profile of the patient showing diffused swelling on the left side of the face.

Figure 2. Left maxillary alveolus showing necrotic bone deposition.

Figure 3. Involvement of right maxillary alveolus and rugae area.