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Paget's Disease Of The Jaws: A Contemporary Reappraisal

Jenisha.S[1], Raghuram.M[2], Bhargavi.A[3], Brittilla.K[4], Thamizhchelvan. H[5], Vijayanirmala subramani[6*]

[1]III Year Post graduate, Department of Oral Pathology, Sri Ramachandra Dental College& Hospital, Sri Ramachandra Institute of Higher Education and Research, No 1, Ramachandra Nagar, Porur- Chennai 600116, Tamilnadu, India.
Email: jenisha.sj02@gmail.com,
[2]BDS RESIDENT, Department of Oral Pathology, Sri Ramachandra Dental College& Hospital, Sri Ramachandra Institute of Higher Education and Research, No 1, Ramachandra Nagar, Porur- Chennai 600116, Tamilnadu, India.
Email: raghuramm284@gmail.com,
[3]II Year Post graduate, Department of Oral Pathology, Sri Ramachandra Dental College& Hospital, Sri Ramachandra Institute of Higher Education and Research, No 1, Ramachandra Nagar, Porur- Chennai 600116, Tamilnadu, India.
Email: kavisree8798@gmail.com,
[4]III Year Post graduate, Department of Oral Pathology, Sri Ramachandra Dental College& Hospital, Sri Ramachandra Institute of Higher Education and Research, No 1, Ramachandra Nagar, Porur- Chennai 600116, Tamilnadu, India.
Email: k.brittillasharon@gmail.com,
[5]DEAN,PROF& HOD, Department of Oral Pathology, Sri Ramachandra Dental College& Hospital, Sri Ramachandra Institute of Higher Education and Research, No 1, Ramachandra Nagar, Porur- Chennai 600116, Tamilnadu, India.
Email: hthamizh@gmail.com,
[6*]READER, Department of Oral Pathology, Sri Ramachandra Dental College& Hospital, Sri Ramachandra Institute of Higher Education and Research, No 1, Ramachandra Nagar, Porur- Chennai 600116, Tamilnadu, India.
Email: Subramani.viji3@gmail.com,

***Corresponding Author: Vijayanirmala Subramani**
*Email: subramani.viji3@gmail.com

Abstract:
Paget's disease is a chronic, slowly progressive metabolic disorder of bone. Paget's disease of the head and neck primarily affects the skull and facial bones; the jaws are infrequently involved. An elderly patient is commonly affected. Radiographically, it shows a typical cotton wool appearance and is accompanied by raised serum alkaline phosphatase or urinary hydroxyproline. The histopathological features of Paget's disease resemble fibro-osseous lesions like florid cemento-osseous dysplasia and sequestrum of non-healing sockets. Osteoarthritis, osteosarcoma, and giant cell tumors are complications of Paget's disease. Hypercalcemia can occur in patients with Paget's disease who are immobilized, and high-output cardiac failure can arise as a complication of active disease. Despite there being very limited published literature on the Indian population, it is both underdiagnosed and undertreated, with many patients never coming to medical attention. This review focused on an overview of Paget's disease and the mechanisms underlying expansile osteolysis and associated conditions of the jaws.

Key Words: Osteitis Deformans, Osteolysis, Bone turnover, mandible & Maxilla, Sequestosome 1

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INTRODUCTION: Paget's disease is a rare bone disorder, largely inherited as an autosomal dominant trait with high penetrance by the sixth or seventh decade of life.[1] In 1877, Sir James Paget used the term Osteitis Deformans to characterize Paget's disease (PD).[2] It ranks as the second most prevalent osteodystrophic disorder following osteoporosis. Accelerated skeletal remodeling is the hallmark of this localized disorder, which can impact one bone (monostotic) or several bones (polyostotic), resulting in bone enlargement, cortical thickening, and an atypical bone structure, leading to deformities and increased fragility.[1] The symptoms and signs of PD can vary depending on the affected areas and the rate of bone turnover. While some individuals may be asymptomatic, the primary manifestations of the disease usually include bone pain, deformities, and fractures. The enlarged and misshapen bones may put pressure on nearby nerves and blood vessels, leading to neurological symptoms. Bone alterations are predominantly observed in the pelvis, with the skull and jaws following closely behind. Common clinical features include enlargement of the skull, hearing loss, cardiovascular issues, curvature of the tibia, and a rise in skin temperature above the affected bone.[3,4] New Zealand, England, the US, France, Canada and South Africa have high rates of Paget's disease. The occurrence of this disease in Asian populations, particularly among Indians, is very uncommon.Paget's disease is an age-related condition typically observed in individuals aged 50 and older.[5-8]

It is more frequently observed in males (78.6%) compared to females (21.4%). Genetic factors play a significant role, with approximately 15% of patients having a positive family history. The risk of developing the disease is 7-10 times greater in first-degree relatives of patients than in the general population. However, environmental factors also affect disease onset. Other rare disorders, such as familial expansile osteolysis, expansile skeletal hyperphosphatasia, and early-onset familial Paget's disease, share clinical, histological, and radiographical features with Paget's disease. Potential environmental triggers for Paget's disease include low dietary calcium intake, vitamin D deficiencies, zoonotic infections, viral infections, and occupational exposure to toxins.[9] [10]Additionally, this paper may attempt to provide updated clinical guideline on diagnosis and treatment of the diseases

ETIOLOGY

Paget's disease occurs in a hereditary manner in approximately 40% of affected individuals. In every familial instance, there is at least one first-degree relative involved."Chronic familial hyperphosphatasia," a rare metabolic bone illness that is inherited in an autosomal recessive fashion, is the juvenile form of Paget's disease of bone. It differs from the conventional form of Paget's disease, which usually affects older people, and is characterized by increased bone turnover brought on by increased osteoclastic activity.[9]

There are several genetic theories that suggest the human leukocyte antigen (HLA) on chromosome 6 and a particular gene on chromosome arm 18q play important roles, but studies on HLA have not produced conclusive evidence, and the gene on chromosome arm 18q has not been identified as the main focus in all families studied, suggesting that genetic diversity is likely present. It has been observed that between 1% and 40% of individuals diagnosed with PDB have a first-degree relative affected by the disease.

Four genes that cause classical Paget's disease and related disorders have been shown to have mutations or variants. These genes include Valocin, which encodes p97, Sequestosome 1 (SQSTM1), which encodes p62, TNFRSF11A, which encodes RANK, and TNFRSF11B, which generates Osteoprotegerin (OPG). Since all of these genes are part of the RANK-NF-κB signaling system, it is likely that the mutations make people more vulnerable to PD by interfering with regular signaling, which causes osteoclasts to become activated. Additionally, mutations in a member of the NF-κB modulating protein family are also considered one of the contributing factors for PD. Mutations in SQSTM1, the most important gene for the development of classic Paget's disease, have been identified, with mutations accounting for 20-50% of cases of familial disease and 5-20% of cases of sporadic disease.[10]

In Paget's disease, osteoclast precursors have been found to be overly responsive to RANKL, which is a member of the tumor necrosis factor-α superfamily that facilitates the formation of osteoclasts. One potential explanation is that heightened RANKL expression may play a role in the localized characteristics of the disease. Furthermore, these osteoclast precursors seem to be excessively responsive to 1,25-(OH)2D3 and calcitonin and show increased levels of the c-fos proto-oncogene and the bcl2 antiapoptotic gene. The effectiveness of bisphosphonates in treating Paget's disease may stem from their ability to reduce RANKL-induced bone resorption, leading to lower RANKL levels and enhanced production of OPG[10].

Several investigations have shown that people with PD had higher levels of IL-6 and/or macrophage colony-stimulating factor (M-CSF). Significant levels of IL-6 are released into the conditioned media by osteoclasts produced from bone marrow cultures taken from PD patients; concentrations of IL-6 can exceed 2000 pg/ml. Furthermore, patients with Paget's disease have elevated levels of IL-6 in both their peripheral blood and the bone marrow plasma of the afflicted bones. It is conceivable that IL-6 plays a role in the elevated osteoclast development in PDB since it has been shown to stimulate osteoclast formation. On the other hand, the elevated IL-6 levels observed in PDB patients may merely serve as an indicator of the heightened osteoclast activity. Research has shown that certain hormonal imbalances, such as hypoparathyroidism, can occur alongside PD.[11]

Idiopathic hyperphosphatasia, also known as juvenile Paget's disease, is a recessive disorder characterized by bone turnover abnormalities and widespread deformities that develop during childhood. It is caused by loss of function mutations in the TNFRSF11B gene, which encodes osteoprotegerin, a decoy receptor for RANKL. These mutations impair the ability of osteoprotegerin to inhibit RANKL-induced bone resorption, resulting in an increased bone turnover. The rare syndrome of hereditary inclusion-body myopathy, Paget's disease, and frontotemporal dementia (IBMPFD) is caused by mutations in the VCP gene, which encodes p97, a protein involved in the ubiquitin-proteasome system. The molecular mechanisms causing the development of Paget's disease and other features of this syndrome remain to be elucidated.[9]

The predominant theory is known as the “slow virus theory.” Messenger RNA sequences from the measles virus have been identified in osteoclasts and various mononuclear cells within lesional bones. Additionally, nucleocapsid antigens from the canine distemper virus have been detected in osteoclasts of individuals with PD. However, while the existence of these paramyxovirus-like nuclear inclusions is noted, it does not confirm that they are the cause of these lesions; instead, these inclusions might simply be indicators of the disease itself.

Laboratories struggle to detect viruses in Paget's disease of bone due to insufficient sensitivity of techniques. Some argue that positive results may be due to PCR contamination or non-specific binding of probes and antibodies. A recent study found no evidence of paramyxovirus transcripts in samples using the most sensitive methods available.[11]

PATHOPHYSIOLOGY: Paget bone lesions show increased osteoclastic bone resorption, marrow fibrosis, increased vascularity of bone, and disorganized bone formation. These abnormalities can give rise to a mosaic appearance with a mix of woven and lamellar bone. Osteoclasts in Paget bone lesions are increased in number and size, containing more nuclei than normal. These osteoclasts also contain characteristic nuclear inclusion bodies, which are not specific for Paget osteoclasts.[11] The rapid rate of bone turnover in Paget's disease leads to disorganized bone with reduced mechanical strength, increasing the risk of deformities and fractures. Evidence suggests involvement of mesenchymal cells and osteoblasts in the pathogenesis of the disease. Osteoblasts cultured from Paget bone lesions show increased expression of genes, contributing to focal abnormalities of bone turnover.[12]

CLINICAL FEATURES: Paget's disease is a rare bone disorder that presents with bone pain and deformity, often presenting at rest, at night, and upon use of an affected limb. It is often difficult to distinguish from secondary osteoarthritis or co-existing musculoskeletal disorders.[13]Clinical features favor a Paget origin, such as localization over an affected site with biochemical or scintigraphic evidence of continuing metabolic activity. Pseudo fractures should be suspected in patients with deformity of weight-bearing limbs who develop localized bone pain of acute onset. Paget's disease can be associated with several complications, including deafness, pathological fracture, secondary osteoarthritis, cranial nerve compression syndromes, and spinal stenosis. Osteosarcoma is a rare complication of Paget's disease, with a combination of increased bone pain, local swelling, and in some cases a pathological fracture. The true incidence of osteosarcoma in Paget's disease is unclear. [14]

Paget's disease can be either monostotic or polyostotic, with the former being the more prevalent variant, primarily affecting the axial skeleton. The disease presents with significant musculoskeletal challenges, along with potential neurological and cardiac complications. Involvement of the facial bones can occur occasionally, which is referred to as leontiasis ossia. Clinical manifestations include pain and deformity, and it may result in fractures of the involved bone, although the disease may start off asymptomatically. Paget's disease is a skeletal condition marked by excessive and irregular reconstruction of bone, leading to deformation and fragility of the bones involved.[15]

ORAL MANIFESTATION: Jaw involvement is frequently observed in PD, with an approximate incidence of 17%, as indicated by various studies in Table1.[16-28]

TABLE :1 shows occurrence in the jaws.

Authors	YEAR	SITE
Sekar et al[16]	2010	Mandible
GC Rajkumar et al[17]	2011	Maxilla
Karunakaran K et al [18]	2012	Mandible
Shankar UK et al[19]	2013	Maxilla
Dineshwarran et al[20]	2014	Maxilla
Polisetti N et al[21]	2014	Maxilla
Chakravarthi et al [22]	2015	Mandible
Rai NP et al [23]	2016	Mandible
Jayachandra et al [24]	2017	Mandible
Vanjari GK et al [25]	2021	Mandible
Loganathan M et al [26]	2022	Maxilla
Arti saluja et al [27]	2023	Maxilla and mandible
Nunez gil et al [28]	2024	Maxilla

More frequently than not, the maxilla is impacted when the jaw is involved. It is possible for the afflicted jaw to gradually expand while the alveolar ridge widens. If there are teeth, they may break free and move about, creating interdental space. The inverted triangle appearance of facial features is caused by the size difference between the maxilla and mandible. Too much cementum builds up on tooth roots, causing hypercementosis, which makes dental extractions more difficult. Localized osteitis is often seen, and the healing of extraction sites is typically delayed. An infection at the extraction sites may result in osteomyelitis in a significant proportion of individuals. Dystrophic calcifications with mosaic patterns can be identified within the pulp, along with signs of internal resorption of dentin. Edentulous patients with dentures often report challenges in wearing their appliances due to alterations in arch size. Additionally, there is an increased occurrence of salivary calculi in patients with PD.[29]

RADIOLOGICAL FEATURES: Dental X-rays can show the characteristic cotton-wool texture of the bone, along with regions exhibiting hypercementosis or a bulbous appearance at various root tips. As part of the preliminary diagnostic evaluation, a radioisotope bone scan may be recommended for all patients in order to detect the spread of the disease, particularly in places such the long bones, spine, and base of the skull that may cause difficulties. [30,31]

Significant involvement of the mandible may sometimes show radiotracer uptake from one condyle to the other, leading to what is known as the “Lincoln sign” or “Black beard” sign. In cases involving spinal stenosis, cauda equina syndrome, compression fractures, or suspected malignancy, computed tomography or magnetic resonance imaging may be helpful.[26]
Paget disease starts with the lytic phase, characterized by heightened bone resorption and an increase in the population of osteoclasts located in a region known as Howship’s lacunae, which is found at the affected bony area. This marked escalation in bone resorption triggers a second phase (referred to as the mixed phase), during which there is a rapid rise in bone formation accompanied by a notable increase in osteoblasts, which, while increased in number, retain their normal morphology. In the final phase of Paget disease, called the sclerotic phase, bone formation prevails, resulting in newly formed bone exhibiting a disorganized structure (woven bone) that is weaker than typical adult bone.[23]

HISTOPATHOLOGICAL FEATURES: In Paget’s disease, histopathological findings reveal osteolysis, which is subsequently followed by a compensatory rise in bone formation due to the recruitment of osteoblasts to the affected area. This process leads to an increased deposition of lamellar bone in a chaotic manner. The resorbed bone is replaced, and excess fibrous connective tissue fills the marrow spaces, accompanied by a notable increase in blood vessels, resulting in hypervascular bone. The abnormally enlarged trabeculae display a mosaic pattern of reversal lines, which arise from ongoing bone deposition occurring alongside disordered bone resorption. The presence of short, fragmented reversal lines is characteristic of Paget’s disease. [25]The primary cell impacted by PD is the osteoclast. In PD, osteoclasts exhibit both structural and functional abnormalities. In terms of structure, osteoclasts found in PD are both larger and more numerous than normal osteoclasts, possessing up to 100 nuclei compared to the typical 3 to 20 nuclei seen in healthy osteoclasts. Another distinguishing feature of osteoclasts in Paget’s disease is the presence of unique nuclear inclusions, which resemble para-crystalline structures akin to the nucleocapsids of paramyxoviruses. Functionally, osteoclast precursors in PD show heightened sensitivity to various osteoclastogenic factors, such as 1,25-(OH)2D3 and RANKL. While osteoblasts are also found in increased numbers at the affected sites, they remain morphologically unchanged and are not considered a primary contributor to the pathophysiology of PDB.

DIFFERENTIAL DIAGNOSIS: Cemento-osseous dysplasia, chronic sclerosing osteomyelitis, and medication-related osteonecrosis of the jaws are the primary differential diagnoses for Paget's disease of the jaws.[24 25] Various biochemical markers associated with bone remodeling that show increased levels in PD are valuable for diagnosing the condition. Serum deoxypyridinoline crosslinks of type I collagen, serum N telopeptide of type I collagen, serum C telopeptide of type I collagen, and urine hydroxyproline are all elevated markers of accelerated bone resorption. Serum bone-specific alkaline phosphatase, SAP, osteocalcin, and the N-terminal propeptide of type I collagen are all upregulated indicators of bone formation.[32] According to Selby PL et al., serum total alkaline phosphatase is the most frequently utilized approach for tracking disease activity, and it is recommended to assess it every three months during the initial six months following treatment, and then every six months afterward.[33]According to the Systemic review and meta-analysis by Al Nofal et al, P1NP levels are used to track PD activity. However, urine N-terminal telopeptide, bone-specific alkaline phosphatase, and total alkaline phosphatase are also excellent substitutes for PD patients who are not receiving treatment.[12]

TREATMENT: Paget's disease of bone is primarily treated for bone pain due to increased metabolic activity. Treatments include bisphosphonates and calcitonin, which inhibit osteoclastic bone resorption. Bisphosphonate therapy reduces bone turnover, improves pain, promotes healing, and restores normal bone histology. However, no adequately powered studies have been conducted to determine its effectiveness in preventing Paget's disease complications. The advent of potent bisphosphonates like zoledronic acid may improve long-term metabolic control. Bisphosphonates are anecdotally effective in improving paraplegia and symptoms of spinal stenosis, but a comparison with other management strategies has not been conducted. Hypercalcaemia is considered a definitive indication for bisphosphonate therapy.
Calcitonin was used in the past because it provides prompt pain relief and decreases bone resorption. However, it frequently resulted in unpleasant side effects such nausea, vomiting, and flushing. Second-generation nitrogen-containing bisphosphonates (disodium pamidronate, alendronate, and risedronate) are currently the main treatment for PD. These drugs are efficient inhibitors of bone resorption and cause a longer-lasting remission of the condition while also significantly lowering bone turnover markers when compared to calcitonin.
Dietary deficiencies in calcium and vitamin D are common in elderly patients with Paget's disease, and correcting this before starting bisphosphonate therapy is crucial to avoid complications like hypocalcaemia. Focal osteomalacia has been reported in patients treated with etidronate, and this defect in mineralisation cannot be prevented by active vitamin D treatment. Other rare side effects include uveitis, skin rashes, renal impairment, and osteonecrosis of the jaw. Calcitonin is now rarely used for Paget's disease due to its short duration of action and weak antiresorptive effects, but is effective at controlling bone pain.Analgesic drugs and anti-inflammatory drugs are often needed for symptom control. Non-pharmacological approaches like acupuncture, physiotherapy, hydrotherapy, and electrical nerve stimulation can also help. Aids and devices can improve quality of life.
Paget's disease often necessitates surgical intervention, including joint replacement, fracture fixation, osteotomy, spinal stenosis correction, and prophylactic surgery. The treatment can be challenging due to bone enlargement, deformity, and increased vascularity. Orthopaedic surgery is necessary for osteosarcoma patients, but the prognosis is poor, with a 5-year

survival rate of 6%. Bisphosphonate treatment is often used to reduce operative complications. Treatment with these newer medications encourages the formation of new bone with a more normal lamellar structure, and mineralization issues are rarely, if ever, observed, leading to a more proactive management approach where both symptomatic disease and asymptomatic cases at high risk of progression and anticipated complications serve as clear grounds for starting treatment.[32]

COMPLICATIONS & Malignant Transformation : Osteosarcoma (71%), fibrosarcoma (14%), chondrosarcoma (3%), malignant fibrous histiocytoma (1%), malignant giant cell tumor, and lymphosarcoma (0.5%) are the most common complications that have been seen in some patients.[14]Malignant transformation represents the most severe complication associated with PD, with osteosarcoma being the most prevalent cancer linked to it. A diagnosis of sarcoma should be considered in patients who report a significant increase in bone pain intensity or a substantial rise in serum alkaline phosphatase levels. Those with Paget's disease who are over 40 years old face a 30-fold higher risk of developing osteogenic sarcoma compared to healthy individuals. In contrast to osteosarcomas that occur in children, these tumors carry a notably grim prognosis. With the introduction of neoadjuvant chemotherapy, the five-year survival rate for pediatric patients has soared to almost 80%, while for individuals with Paget sarcoma, it remains around 10%. Pagetic osteosarcomas are most frequently observed in affected bone during the mixed lytic/sclerotic stage of the disease. Additionally, fibrosarcomas and giant cell tumors may develop from long-standing Paget's disease.[31]

CONCLUSION: Familial expansile osteolysis is a rare autosomal-dominant bone dysplasia that in its early stage may resemble Paget disease. Patients present in the second through fourth decades of life. Progressive osteoclastic resorption is accompanied by medullary expansion of bone, producing both generalized and local bony changes that unlike Paget disease are rare in the axial skeleton. Pain, pathologic fracture of disintegrated bone, skeletal deformity, loss of dentition, and deafness are features of the disease. Total serum alkaline phosphatase concentrations are used to assess Paget's disease of bone activity and monitor antiresorptive treatment.[33] Other markers are more sensitive but have higher costs. Advanced treatment options such as bisphosphonates have been shown to be effective in enhancing patient care. Some clinicians routinely administer antiresorptive therapy when alkaline phosphatase concentrations rise, while others only treat patients with Paget's disease. symptoms. A comprehensive diagnostic approach that includes clinical characteristics, radiological evidence, biochemical tests, and histopathological evaluations is vital for managing the disease.

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