Paget's Disease (of The Jaw Bones)

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Overview

Paget's disease, or Osteitis Deformans, is a chronic bone disorder characterized by the simultaneous occurrence of excessive bone production and resorption. This condition occurs in the axial skeletal system, giving rise to thick, soft, and porous bones that can lead to potential fractures and deformities, especially in the skull, spine, pelvis, and legs.

Etiology

The etiology of Paget's disease is unknown. Three potential factors that may cause the disease include genetic predisposition, mutation of the SQSTM1 gene affecting osteoclast-regulating proteins, or viral infections that induce dysfunction in osteoclasts (Wang et. al, 2020). The disease is composed of three phases: the early phase characterized by osteoclastic activity, the middle phase made of osteoblast-osteoclast action, and the late phase dominated by osteoblastic movement (Gennari et. al, 2019). Osteoclasts rapidly resorb bone in the early phase. The loss of bone stimulates osteoblasts to increase bone formation during the middle phase. Eventually, the osteoclasts stop, leaving overactive osteoblasts to increase alkaline phosphatase (ALP) levels (Reid, 2019).

Clinical Presentation

Some objective clinical signs of Paget's disease are hearing loss, loose teeth, and bone deformities. Thickening of the skull impinges on the cranial nerves, leading to hearing loss. Patients may present bowing of long bones, enlargement of the skull, changes in posture and

gait. A larger skull would cause an increase in hat size. Progressive enlargement of jaw bones, particularly the maxillary arch, may result in migration, loosening, and spacing of teeth (Gupta et. al, 2020). An extreme bone deformity in the face can illustrate a lion-like feature known as Leontiasis Ossea (Gennari et. al, 2019). Chronic progression of the disease may lead to osteosarcoma, which is a bone tumor of malignant osteoblasts laying malignant bone throughout the body. Radiographically, Paget's disease appears as a radiopaque "sunburst" pattern. Some subjective signs of Paget's disease are bone pain, swelling of the joints, tenderness, and redness of the skin over affected areas. The greater the osteoblastic activity, the greater the amount of bone is laid that can impact on local nerves and cause tremendous pain.

Demographic

Paget's disease primarily affects Caucasian women who are older than 50 years old. It is less observed in Blacks and Asian descents.

Biopsy / Histology / Radiographs

Diagnostic methods involve an x-ray, blood test, or bone scan. A blood test would examine for elevated amounts of ALP; patients diagnosed with Paget's disease often have raised ALP activity. A biopsy may also be used to confirm the disease; a pattern of unfused bone would be seen. Histologically, there are osteoblastic and osteoclastic activities in the same region. Radiographically, there is a "cotton wool" radiopaque appearance.

Differential Diagnosis

The remodeling activity of osteoclasts and osteoblasts can cause bone pain and swelling, which can be misdiagnosed for diffuse sclerosing osteomyelitis (DSO). DSO manifests similarly but primarily affects the mandibular arch. Clinical swelling in the orofacial region may resemble

Paget's disease. Radiographically, there is a mixture of sclerotic and resorbing bone (Antao et. al, 2019).

Treatment

While there is no cure for Paget's disease, treatment options range from no treatment, NSAID prescriptions, and anti-resorptive medications like bisphosphonates. NSAIDs alleviate swelling, inflammation, and pain while bisphosphonates slow down disease progression by inhibiting osteoclasts (Reid, 2019). These interventions help the patient manage the disease to make it more tolerable.

Prognosis

With treatment, patients may experience reduced pain, discomfort, and swelling. Without treatment, the disease may worsen, leading to exacerbated symptoms and potential development of osteosarcoma.

Professional Relevance

Paget's disease holds professional relevance in the dental field, especially in the oral cavity. It can cause enlargements and fractures of the jaws (osteonecrosis), alveolar bone loss, tooth loss, or malocclusion. Edentulous patients require periodic examinations due to alveolar bone expansion, which can lead to fractures in dentures. Ill-fitted dentures can cause ulcers or bacterial infections of the gingiva and palate. Inflammation of the bone can worsen periodontal disease. As a future dental hygienist, it is essential to understand the underlying cause of these oral manifestations. Early recognition of the disease allow for better dental services, accurate diagnosis, and proper modifications to the recommended treatment plan to improve the overall health of patients.

Citations

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