

Metabolic conditions of the bone

Bone cells & their precursors react to the influences of circulating hormones & other regulating substances. Thus the bones are particularly sensitive to physiologic & genetic changes that can result in alterations to their structure & mineral content.

1- Paget's disease (Osteitis deformans)

Is a focal alteration in the histology & morphology of bone, result from uncoordinated increase in the osteoclastic & osteoblastic activity of the bone cells. Confined to one bone or multiple bones throughout the body.

Initially the process is dominated by bone resorption followed by excessive bone formation, result in increase in volume but a decrease in strength.

This uncoordinated function of osteoclast & osteoblast activity characterized by numerous reversal lines that give the bone a characteristic *mosaic* pattern under the microscope.

The etiology is still unknown, but intranuclear viral inclusion bodies have been found within some osteoclast cells.

Clinical features

Asymptomatic, seldom found in patients younger than 40 years, & being more common in the western countries. The disease may involved one bone (Monostotic) or multiple bones (Polyostotic), when multiple bones are involved, there will be some degree of incapacity, pain, & joint disease.

When involve the skull, a frequent symptom is a gradual increase in hat size caused by increased cranial circumference. But the serious concern is the involvement of the base of skull, in this case diminish in the various foramens can result in compression of the spinal cord & cranial nerves lead to paralysis & loss of hearing & sight.

In the jaws, the maxilla is more commonly involved than the mandible, the first sign is spaces between the teeth, & the denture may become tight & uncomfortable within the time.

The most important investigation is alkaline phosphates level, which give information of osteoblastic activity. Normal level = 63 IU/ L, but in Paget's disease may reach 1000-5000 IU/L in polystotic & 200-500 IU/L in monostotic one. Elevation of urinary hydroxyproline level indicate the increase bone resorption (osteoclast activity).

Although considerable Ca^{++} loss from the bone occurs, serum Ca^{++} usually remain within normal levels. The complications, include, bone fracture, heart failure & sarcoma



Radiographic appearance

Varies extensively, in the early osteolytic stage the disease exhibit a diffuse radiolucency, later on there is a diffuse radiopacity.

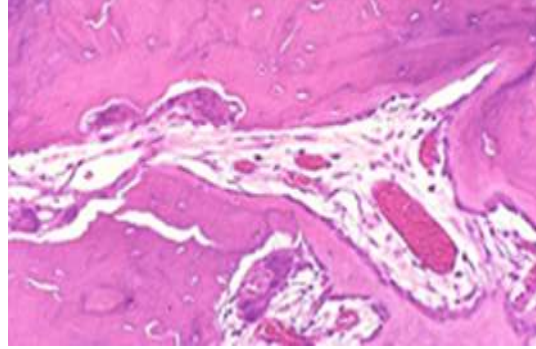
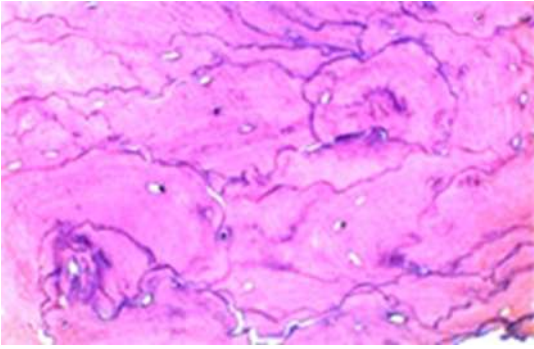
The most common picture is a mixed radiolucent / radiopaque resembling “cotton wool” in a diffusely radiolucent area.

The maxilla & mandible are often enlarged, with loss of the lamina dura & hypercementosis of the roots of the teeth in the area.



histopathology

Consist of an increase in osteoclasts, osteoblasts, & blood vessels & a replacement of the normal bone by a less dense bone having a *mosaic* appearance.



Treatment

The most common effective therapeutic agents are Calcitonin & Diphosphonates such as Disodium etidronate. These compound inhibit bone resorption, this provides the bone forming cells to produce stronger, less fragile bone. However, the treatment does not stop the lesion but does slow it. Surgery is avoided because the diseased bone is prone to hemorrhage & prone to a difficult to manage osteomyelitis.

Surgery is used only to relive sever pain caused by pressure on nerves & spinal cord.

2- Hyperparathyroidism

Loss of bone mineralization (osteoporosis) due to increase in the PTH secretion (primary) or increased demand for serum Ca^{++} (secondary) mostly in renal disease.

The disease lead to multiple systemic complications, loss of alveolar bone architecture, & occasionally giant cell tumor (brown tumor).

Clinical features

Mostly in patients over 60 years of age & female to male ratio 2-1. The disease is mostly asymptomatic.

Hypercalcemia is the most common manifestation & the disease diagnosed when serum Ca^{++} is above the normal range of 8.6-10.4 mg/100 ml, associated with decrease serum phosphates level.

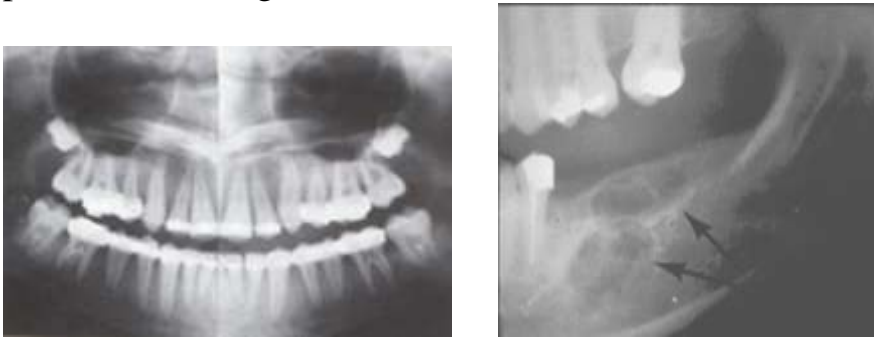
Serum alkaline phosphates & urinary hydroxyproline are normal, but urinary phosphate may be elevated.

Radiographic appearance

Generalized reduced bone density (osteoporosis) or mottled areas of radiolucency.

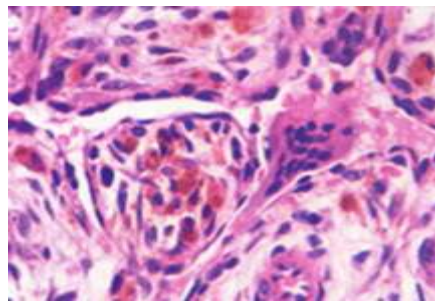
In the mandible & maxilla, the normal trabecular pattern may be lost, with

lack of distinctness of lamina dura in late stage. A large radiolucency may be present, indicate giant cell tumor.



Histopathology

Non specific, However, the lesion consisting of increased osteoclastic activity, thinning of trabecular bone, & wide zones of osteoid with activated osteoblast. Also some replacement of marrow with loose C.T. may occur.



Treatment

Treat the cause & dietary phosphate supplements, & Vit. D supplements have been beneficial.

3-Osteopetrosis

Is a generalized hereditary condition in which the bones become more denser than normal due to excessive bone mineralization, resulting in altered stature, frequent fracture, lack of bone marrow hematopoietic function, & a tendency for sever Osteomyelitis of the jaws.

Clinically

Early symptoms in infancy include difficulties in breathing & hearing due to

oversized facial & mastoid bones.

Followed by functional defects in the ocular & trigeminal nerves as a result of compressed by sclerosis of the foramina of the base of skull.

Delayed eruption of teeth is a common oral finding, & healing is difficult after extraction.

Depletion of platelets, leukocytes, & erythrocytes, make a tendency for spontaneous hematomas, multiple infections, & anemia.

In sever cases the patient die from complications of marrow depletion before 10 years of age.

radiographically

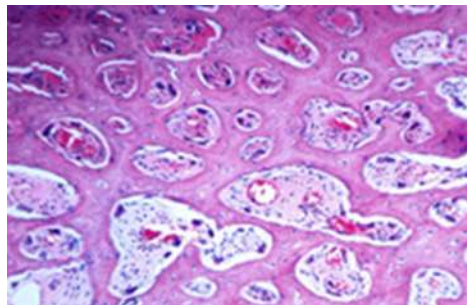
The disease characterized by a generalized increase in bone density with obliteration of normal internal architecture.



histopathology

The bone is dense & sclerotic, with most of the marrow spaces replaced with bone or fibrous tissue.

The bone is somewhat avascular & in some cases islands of calcified cartilage are found, which normally resorbed during development.



Treatment

Depends on the condition. Either by bone marrow transplantation, or by oral Calcitriol combined with a calcium deficient diet.

4- Osteogenic imperfecta

is a bone disease characterized by defective matrix formation & lack of

mineralization, resulting in multiple broken bone, blue sclera of the eye, & associated dentinogenesis imperfecta

Clinically

The neonatal form is lethal, the patients suffering multiple fractures during gestation & delivery. Patients with moderate type, have blue sclera, & dentinogenesis imperfecta.



X-Ray

show large areas of cyst like radiolucency, multiple fractures & healed fractures are present. The teeth exhibit bulbous crowns with shortened roots.

Histopathology

The bone show markedly thinned bone cortices composed of immature woven bone

Treatment

No therapy is capable of altering the course of the disease.