DISEASES OF BONE

Dept.of Oral Pathology
& Microbiology

Learning Objectives

At the end of the lecture student should be able to describe

- Etiopathogenesis, Clinical features, oral manifestations, radiographic features, histopathological features,& surgical management of *PAGET'S DISEASE*

PAGET'S DISEASE (OSTEITIS DEFORMANS):

- Characterized by excessive & abnormal remodelling of bone
- Named after ***Sir James Paget,*** British surgeon, 1st to describe this condition & its clinical course

- Paget's Disease is characterized by enhanced resorption of bone by giant multinucleated osteoclasts
- Process evolves through various phases of activity, followed by a quiescent stage. Typically, 3 phases:
- Lytic
- Mixed: lytic & blastic
- Sclerotic or burned out.

ETIOLOGY:

- Unknown
- Genetic: Autosomal dominant
- Inflammatory
- Autoimmune, CT disorder, vascular disorder
- Slow virus infection has received considerable attention (unproven)

Slow virus-is one which produces a disease with a prolonged incubation period.

CLINICAL FEATURES:

- Most cases are polyostotic
- Age: Prevalence increases with age
- Most commonly after 50 yrs
- Male: Female ratio- 1:1
- Geographic predilection: England, France, Germany
- Usually asymptomatic

- Most common presenting feature is bone pain: dull, constant aching pain, deep below the soft tissues.
- Pain may exacerbate during night
- Involved bone becomes warm to touch because of the increased vascularity
- Pathologic # may result from weakened pagetic bone

- Other symptoms: non specific head ache, impaired hearing, tinnitus, back pain, neck pain
- Progressive pain, paraesthesia, limb paresis, waddling gait, bowel/ bladder incontinence due to compression on spinal cord/ spinal nerve

- Deformity of skeletal bones: patient's hat size may increase
- Nausea, dizziness, syncope, ataxia, dementia with hydrocephalus, basilar invagination, cerebellar or brainstem compressive syndromes

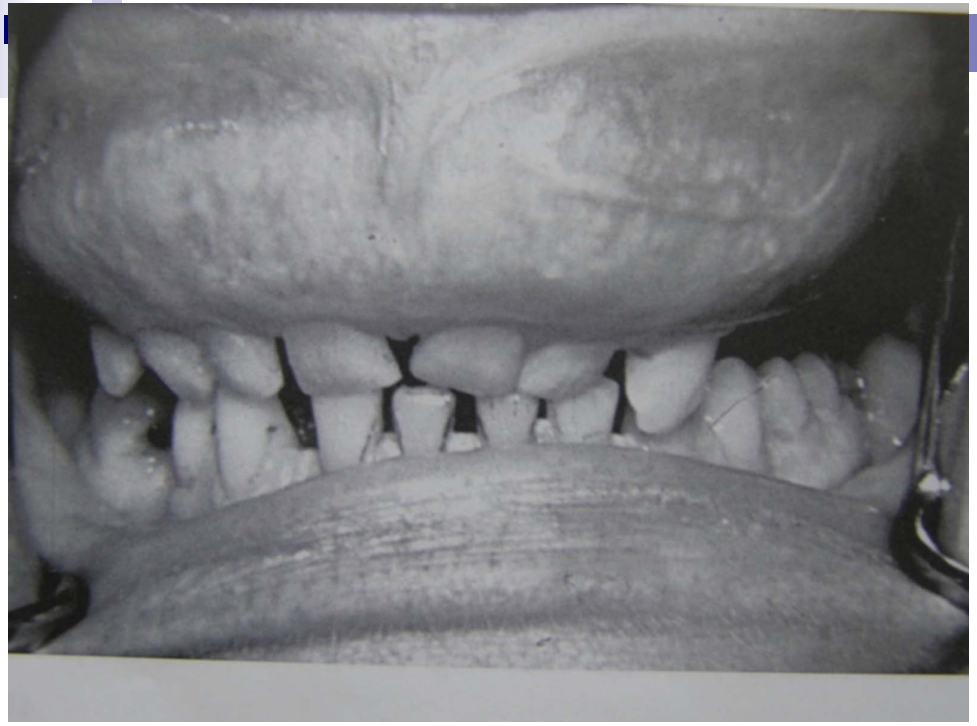
- Uncommonly: cranial nerve palsy. Optic nerve involvement may lead to impaired vision.
- Platybasia: descent of cranium onto cervical spine due to softened bone at base of skull.
- Occassionally, facial bones involved: *Leontiasis Ossea* (lion- like facies)

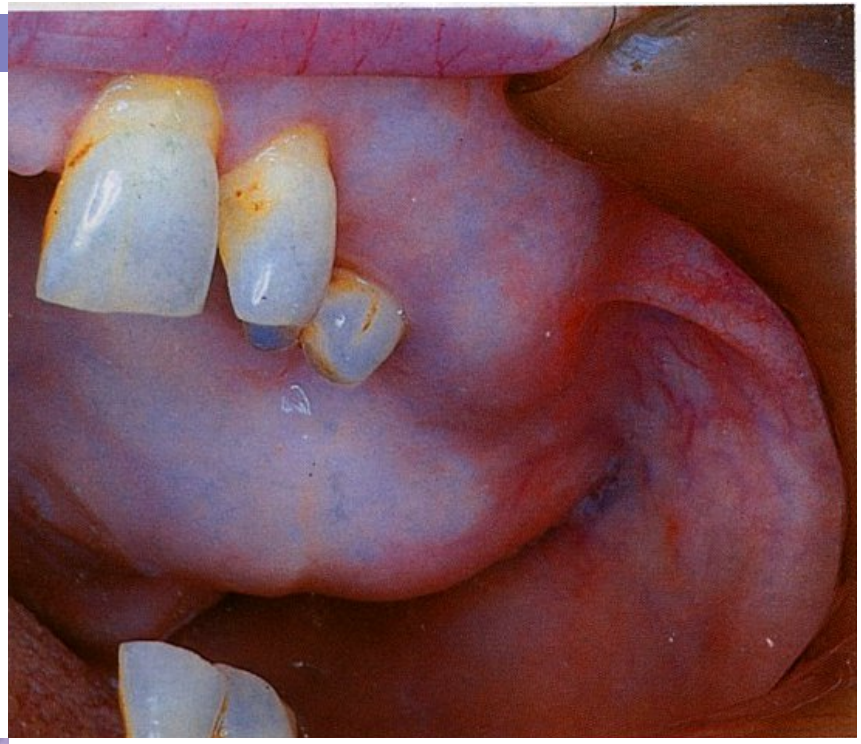
Fig. 8.26 Paget's disease: replacement of both maxillae by pagetoid bone can be completely symmetrical or nearly so, as here, producing a characteristic facies of the leontiasis ossea type.

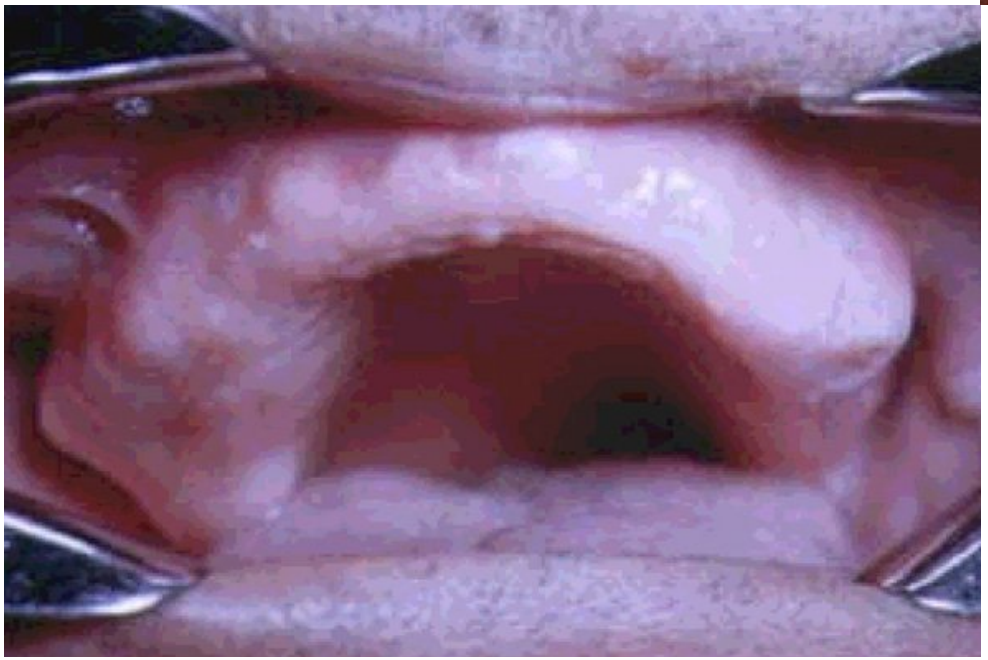
ORAL MANIFESTATIONS:

- Jaw involvement is common
- Predilection for maxilla involvement (ratio mx: mn:: 2.3: 1)
- Maxilla exhibits progressive enlargement, alveolar ridge becomes widened, palate is flattened. If present, teeth become loose & migrate, producing spacing.

- Mandibular involvement not as severe, but similar C/F
- Denture wearing patients complain of tightness of dentures & inability to wear appliances because of increase in size of jaws.





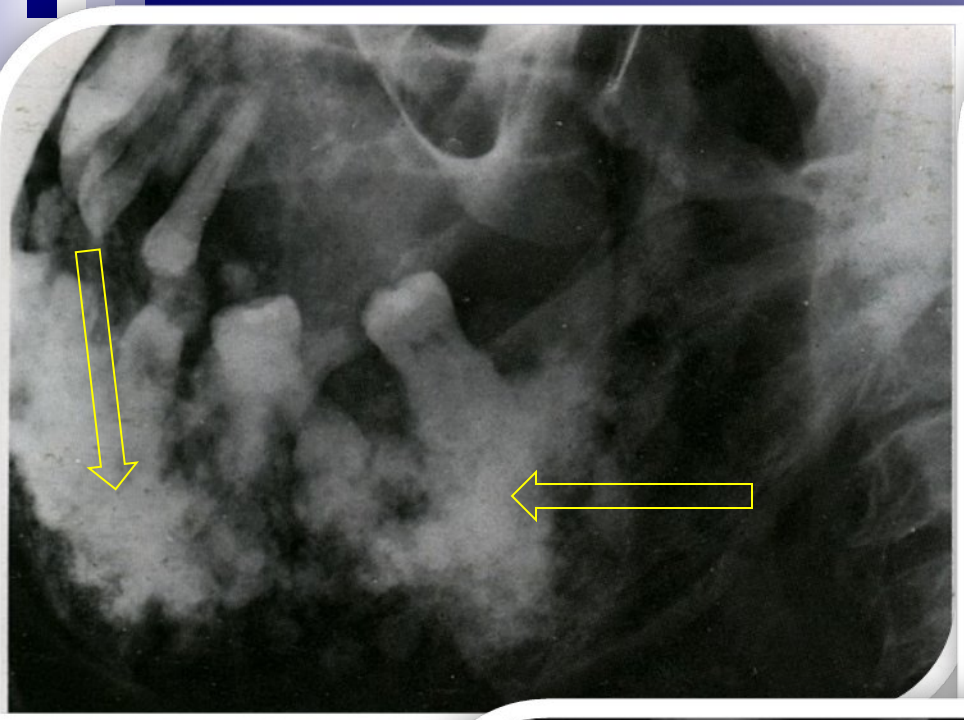


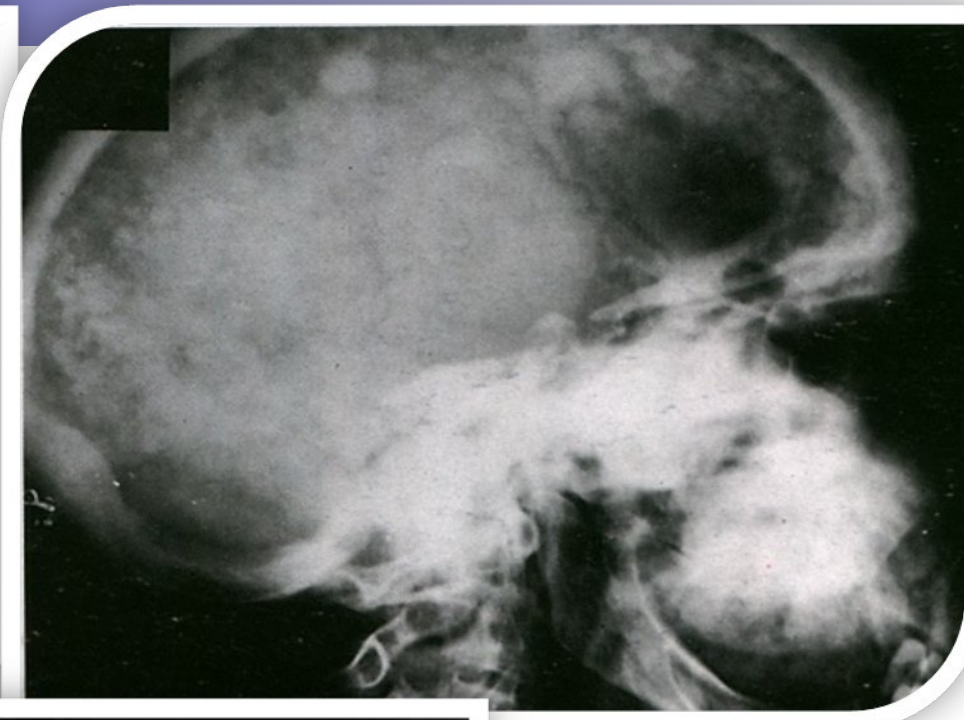
RADIOGRAPHIC FEATURES:

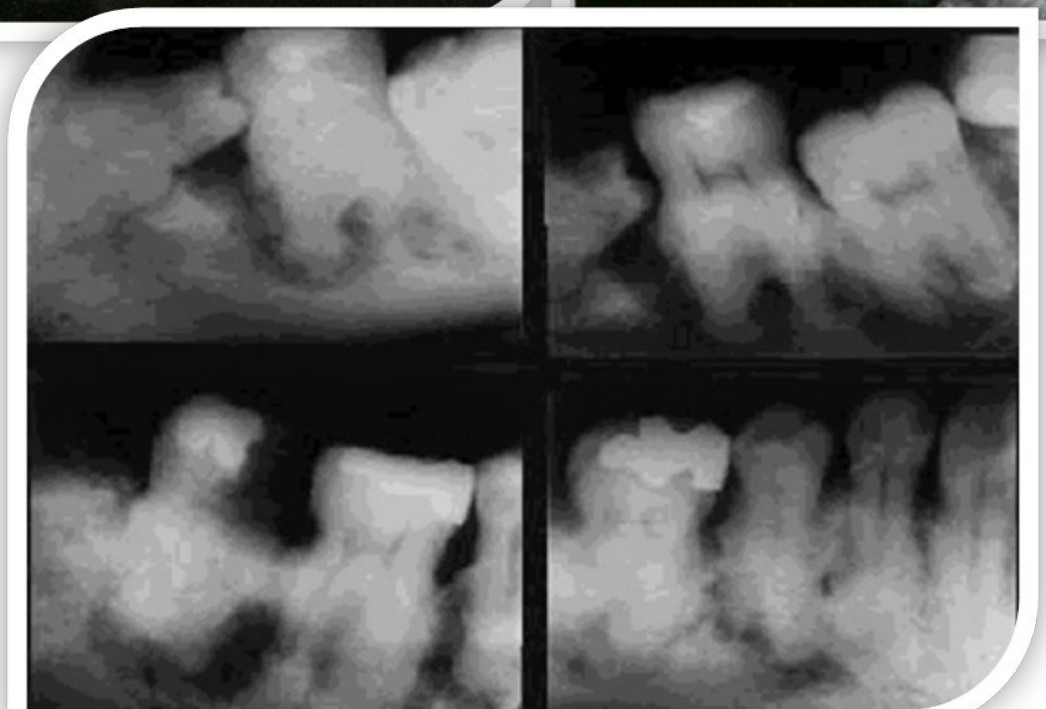
- Varied
- Since disease is described as disorder characterized by an initial phase of de-ossification & softening, followed by bizzare dysplatic type of re-osssification not related to functional requirements, two processes take place simultaneously or alternately

- Thus, osteolytic areas are commonly associated with osteoblastic areas
- In early stages – skull might show large circumscribed areas of radiolucency (***osteoporosis circumscripta)***

- During later stages of (osteoblastic) disease there is haphazardly formed patchy areas of radio-opacities (bone formation) which eventually become confluent, but show variation in radio density-giving it a ***"Cotton wool"*** appearance.
- Dental changes : Hypercementosis of roots,Loss of lamina dura,Obliteration of PDL space,Resorption of roots





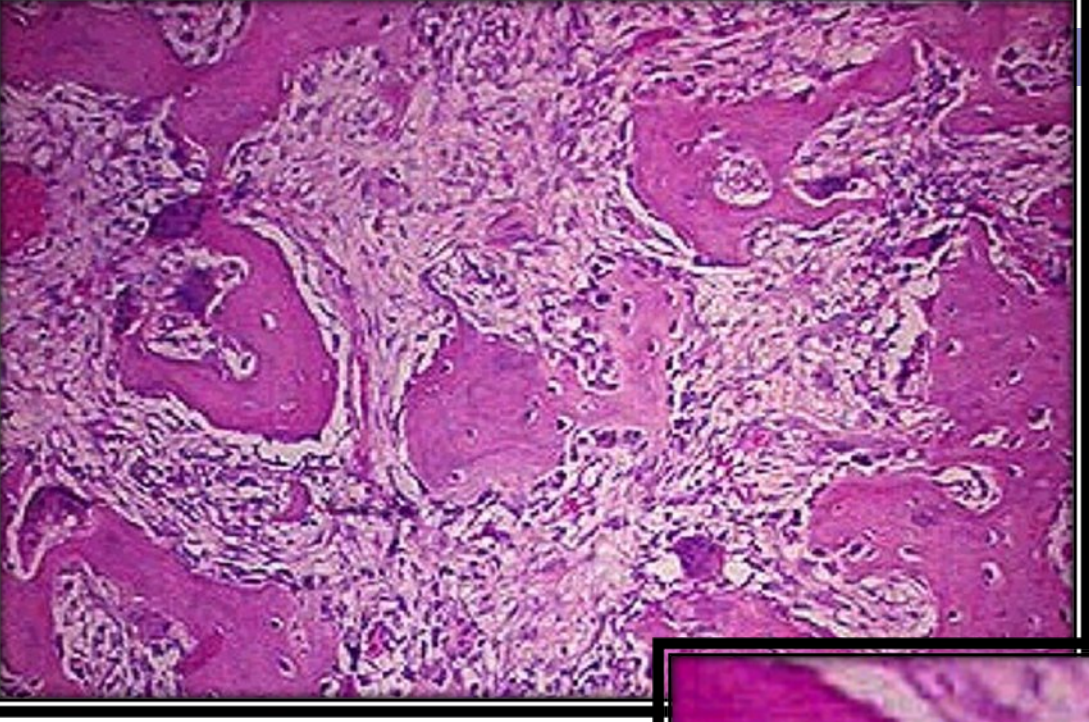


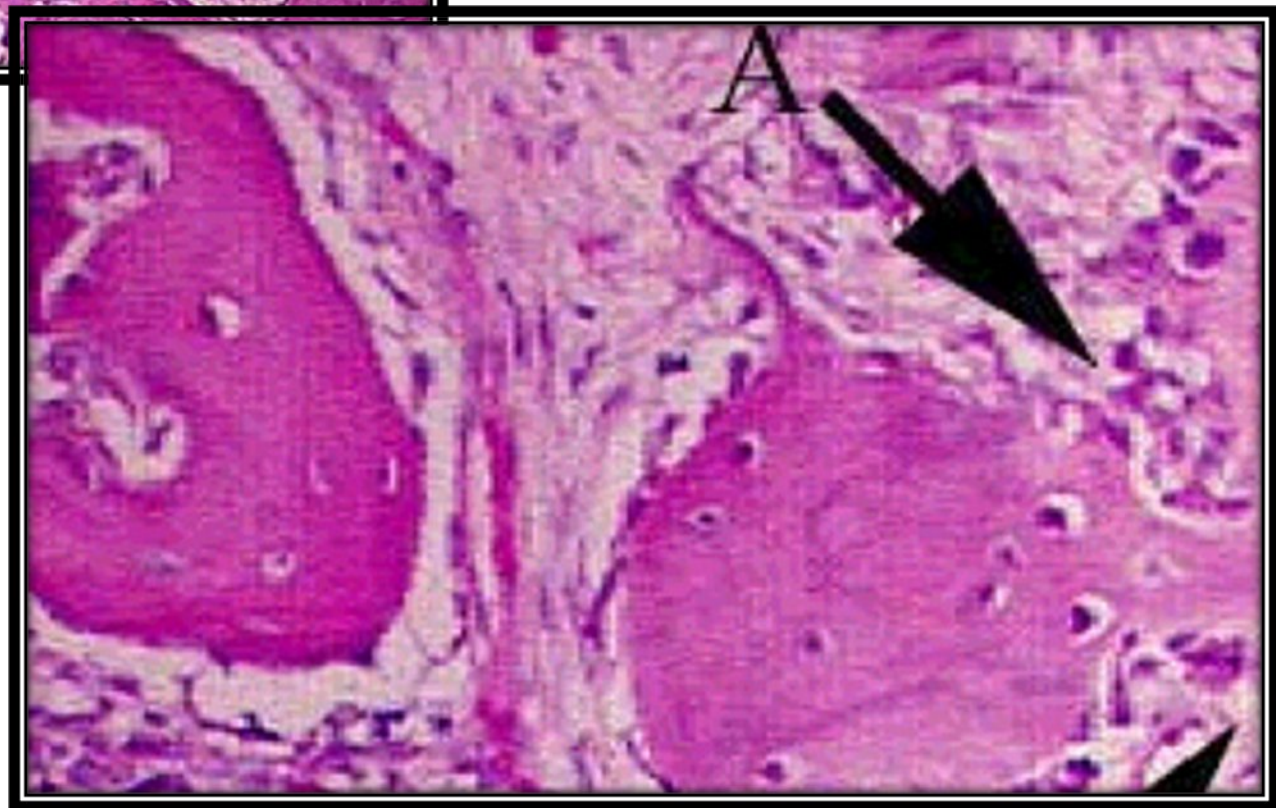
LABORATORY FINDINGS:

- Important information is provided
- Serum Calcium & Phosphate levels are within normal limits
- Serum **Alkaline phosphatase** levels are markedly **elevated**
- Normal value in adult: 1.5 – 5.0 (Bodansky units)
- In Paget's disease : 50 – 250 (Bodansky units)
- Other units for measuring Alkaline phosphatase
- King-Armstrong – Normal value = 5 -10 units
- Phenol units -- Normal value = 1.0 – 3.5 units

HISTOPATHOLOGY:

- Depends on the stage of the disease (osteoclastic or osteoblastic)
- In the active resorptive stages, numerous Osteoclasts surround the bone trabeculae
- Simultaneously, osteoblastic activity is seen with formation of osteoid rims around bone trabeculae

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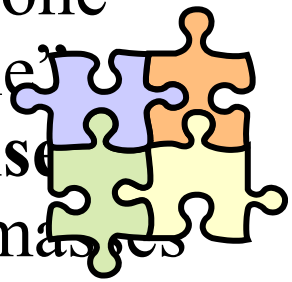
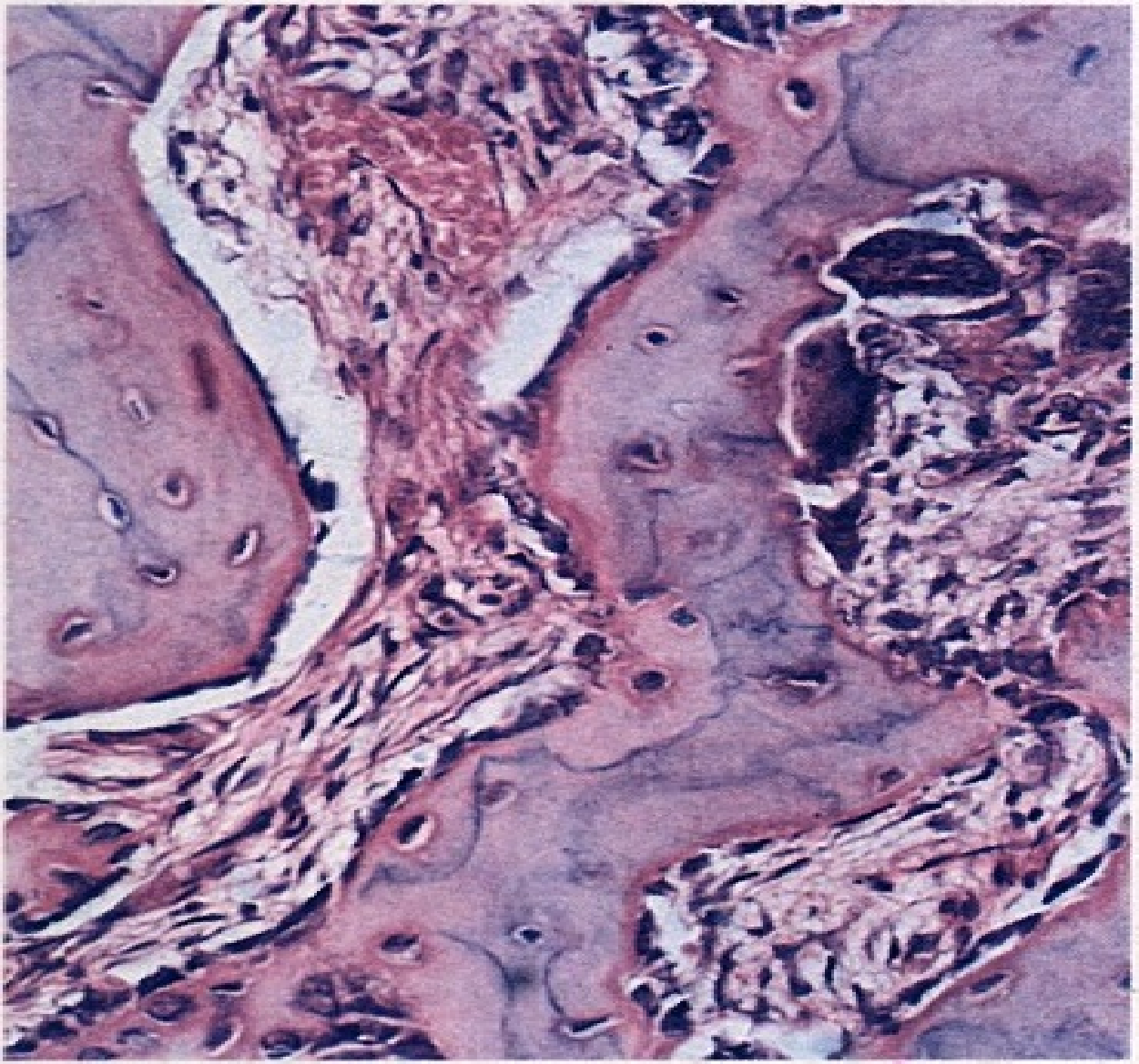
- The marrow is replaced by fibrous C.T.
- Characteristic picture of PD is presence of basophilic reversal lines. These lines indicate the junction between resorptive & formative phases of the bone and results in a “jigsaw puzzle” or “mosaic bone” appearance-**Histologic hallmark of Paget’s Dise**
- Few cases show multiple small acellular bony masses that fuse as the disease progresses.





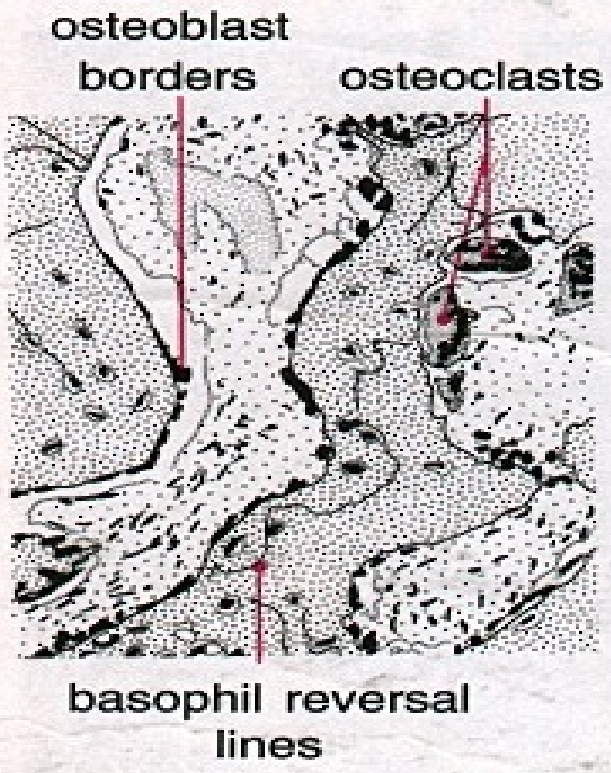


Fig. 8.30 Paget's disease: early stage. There are many osteoblasts and osteoclasts associated with active resorption and formation of new bone, producing the characteristic jigsaw puzzle (mosaic) pattern of bluish reversal lines within the bone.

- As disease progresses, osteoblastic phase predominates, excessive bone formation occurs, causing more compact & dense bone
- Marrow spaces filled with loose highly vascularized CT
- Hypervascular bone combined with increased vasodilation causes increased skin temp

- Normal trabecular pattern distorted with Pagetoid bone formation
- Pagetoid bone shows no tendency to form Haversian system
- Bone is very hard and dense.
- Proliferation of bone & concomitant hypercementosis sometimes result in obliteration of PDL.

- Eventually, osteoblatic phase diminishes, osteoporotic/ burned – out phase predominates: new bone is disordered, poorly mineralized, lacks structural integrity.
- **Complications:**
  - Development of malignant tumor, usually an ***osteosarcoma*** is reported.
  - Other neurological complications.

Treatment:

- No specific treatment : Vitamin, hormone therapy
- Use of parathyroid hormone antagonist, such as calcitonin or biophosphonates can reduce bone turnover
- Dental: - difficulty in extraction is encountered (hypercementosis)
- Edentulous patients may require periodic new & larger dentures

Summary

- Etiopathogenesis, Clinical features, oral manifestations,Radiographic features, histopathological features,& surgical management of Pagets disease

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Thank You