Fibro osseous lesions of the jaws

CONTENTS

Replacement of normal bone by a tissue composed of collagen fibers and fibroblasts that contain varying amounts of mineralized substance which may be bone or cementum in appearance

Classification

Waldron 1993 JOMS:51;828-835

I Fibrous dysplasia-Monostotic & Polyostotic

II Reactive (dysplastic) lesions arising in tooth –bearing area which are of periodontal ligament origin

Periapical cemento-osseous dysplasia

Focal cemento-osseous dysplasia

Florid cemento-osseous dysplasia

III Fibro-osseous neoplasms Ossifying fibroma

Juvenile active ossifying fibroma

Fibrous Dysplasia

Lichenstein 1938 - polystotic lesion

Lichenstein & Jaffe - Monostotic form

HAMARTOMATOUS

Developmental tumor like condition that is characterized by replacement of normal bone by an excessive proliferation of cellular fibrous connective tissue intermixed

with irregular bony trabeculae.

Fibrous Dysplasia

Sporadic condition

Post zygotic mutation in GNAS1 gene

Mutation occurring at different stages:

Early embryonic life – polystotic + endocrine

Later stages of embryonic

development - Polystotic

Post natal - Monostotic

Clinical & radiographic features

MONOSTOTIC

- More common
- Jaws & skull
- Other bones

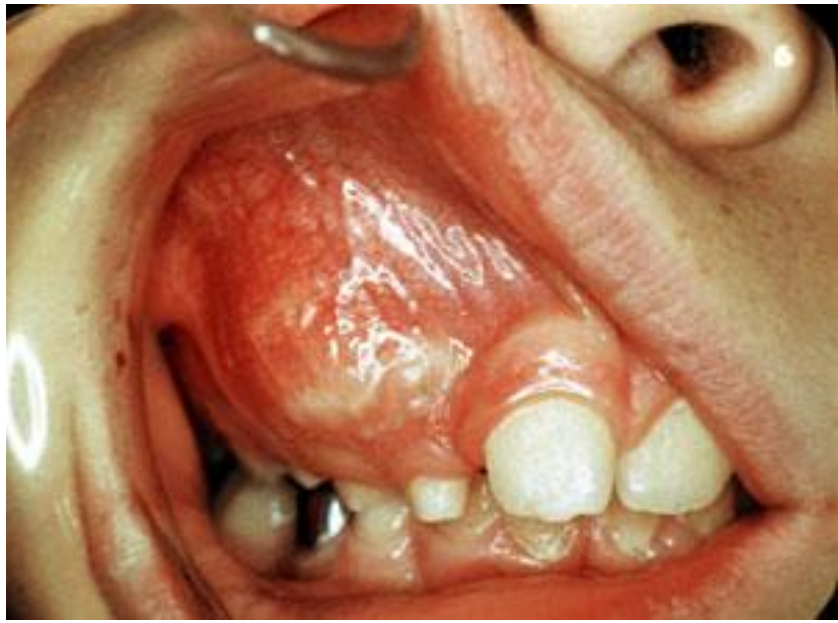
RIBS
FEMUR
TIBIA
MAXILLA

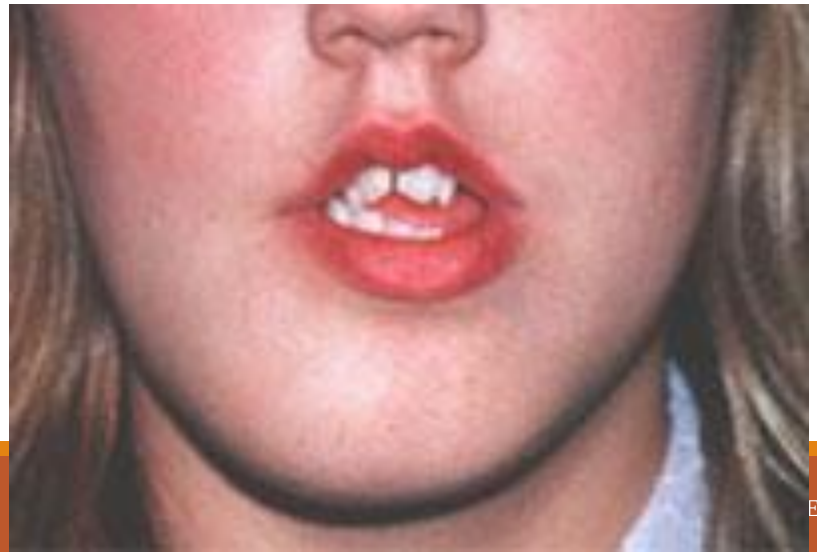
Clinical & radiographic features

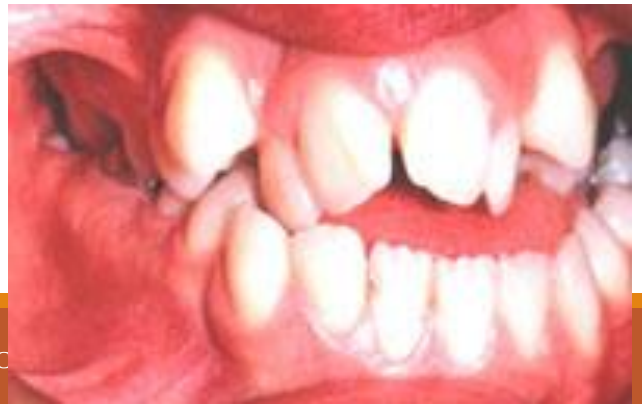
Cranio facial form

M:F::1:1

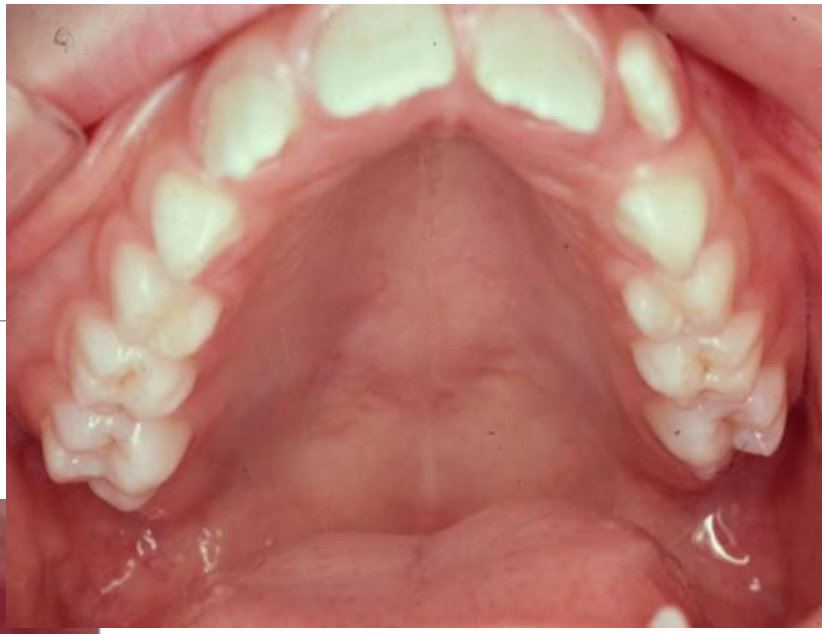
Second decade

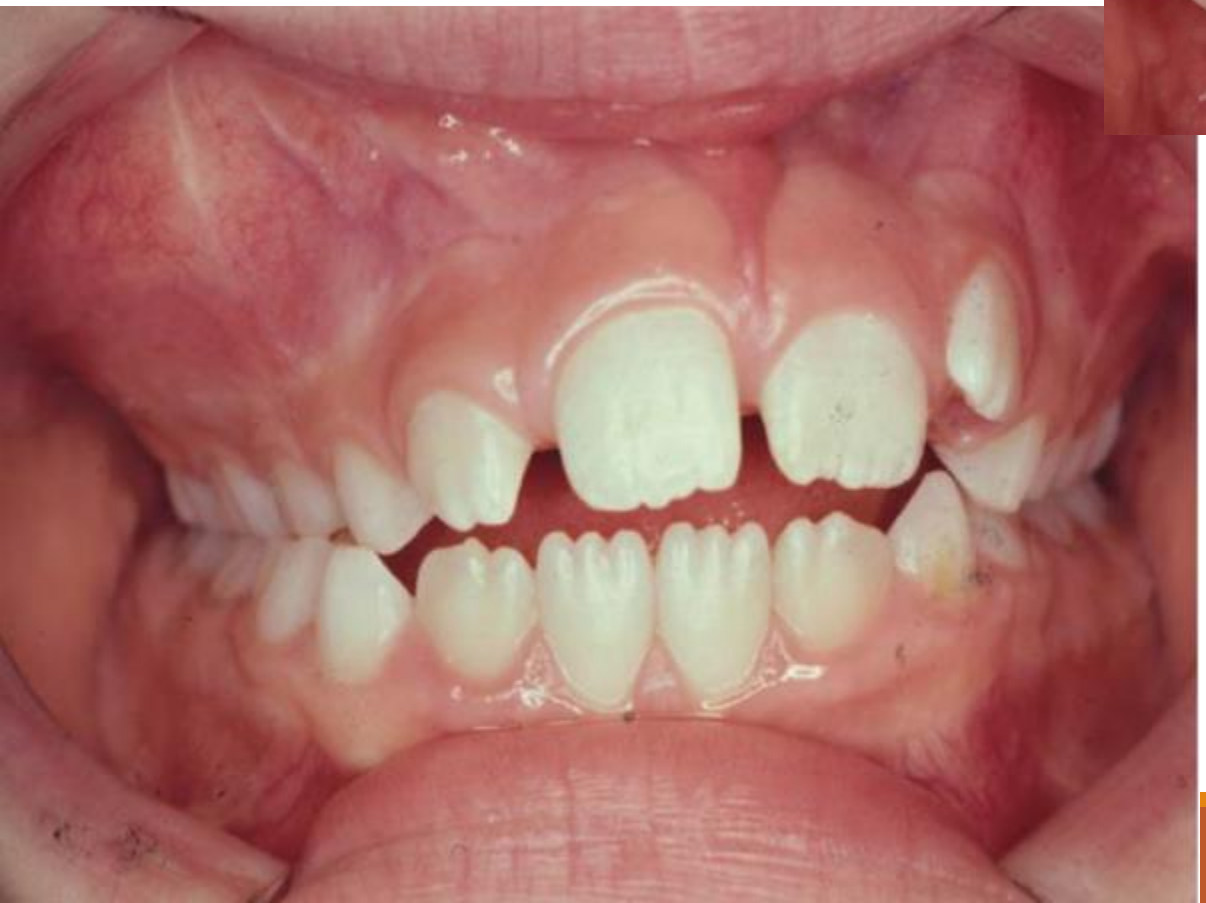






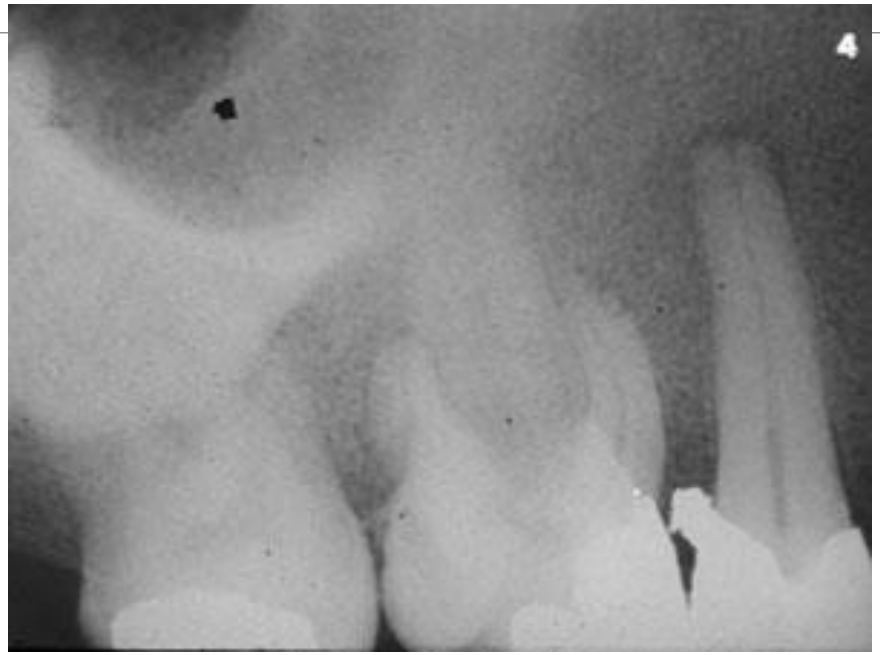
Pain less expansion





Maxilla > Mandible

Clinical & radiographic features



Early stage-unilocular radiolucency

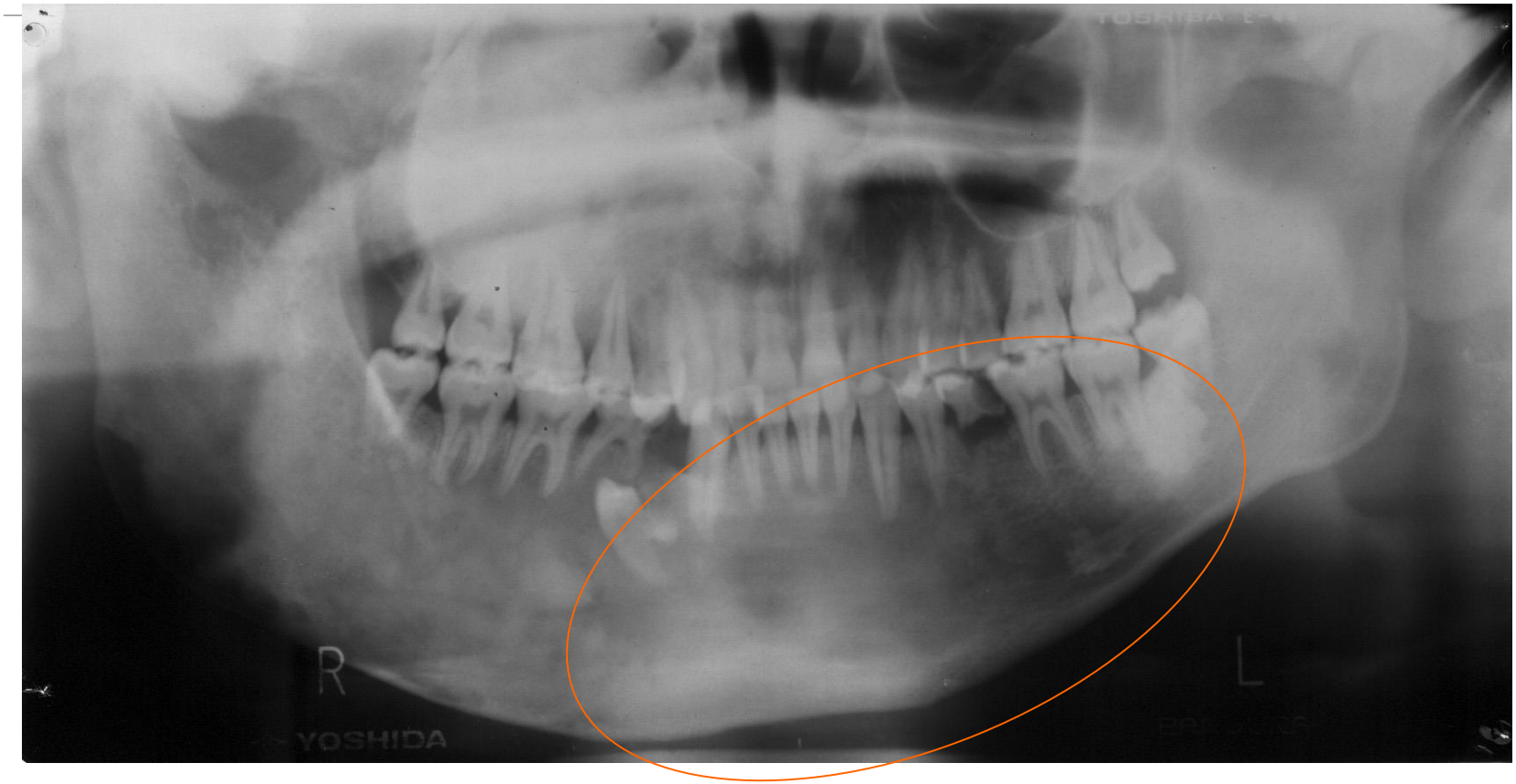
Maturation-Mottled radiolucency

Further Maturation-

GROUND-GLASS
FROSTED GLASS
OR
ORANGE
PEEL

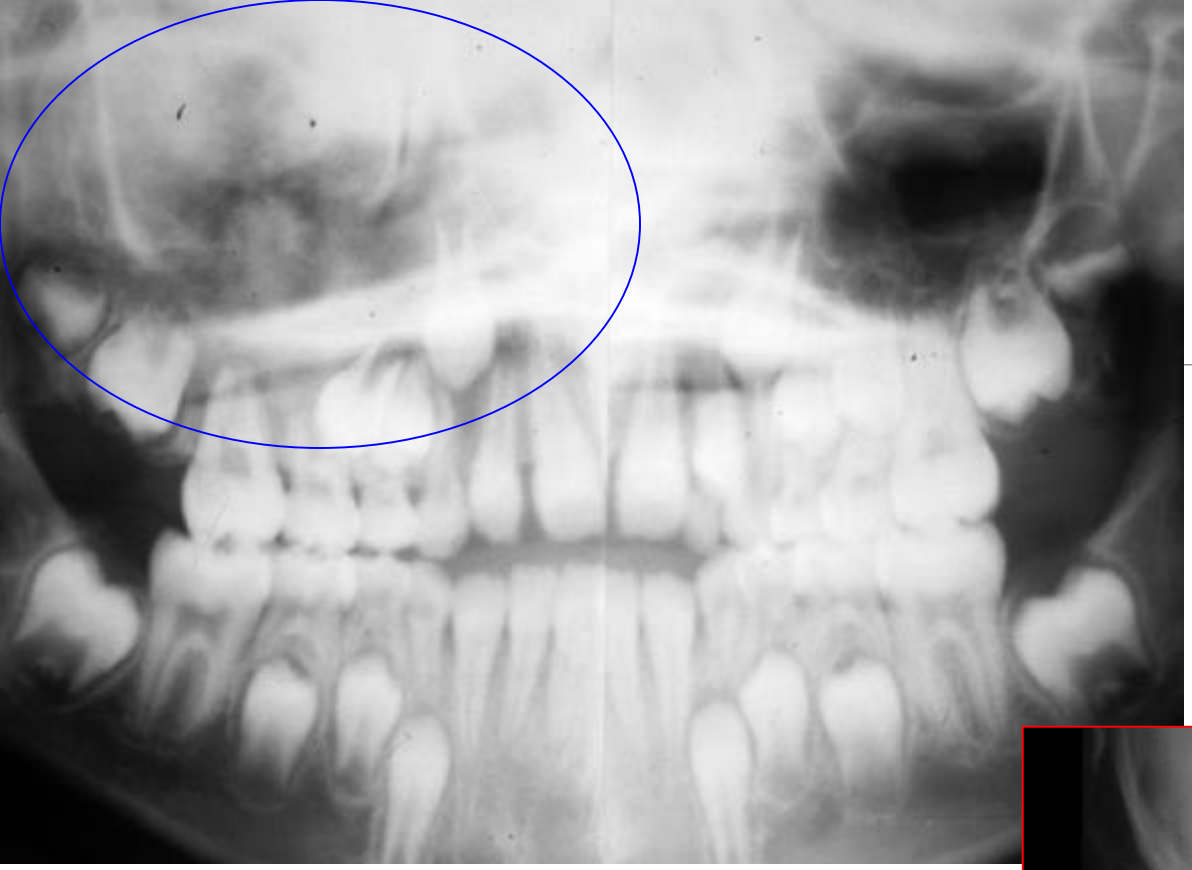
MARGINS ARE NOT WELL DEFINED

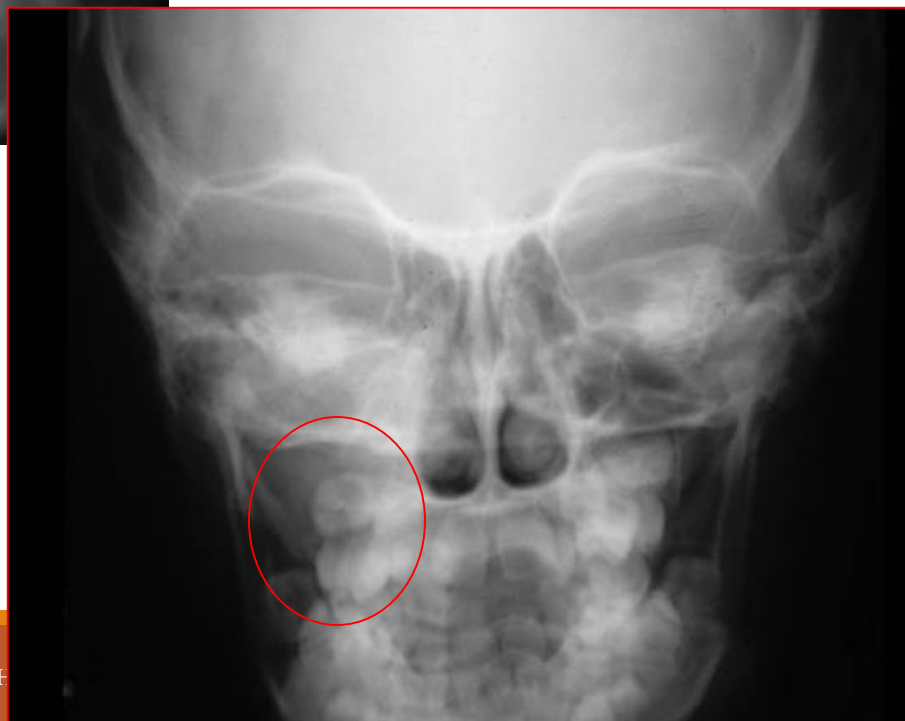
Clinical & radiographic features



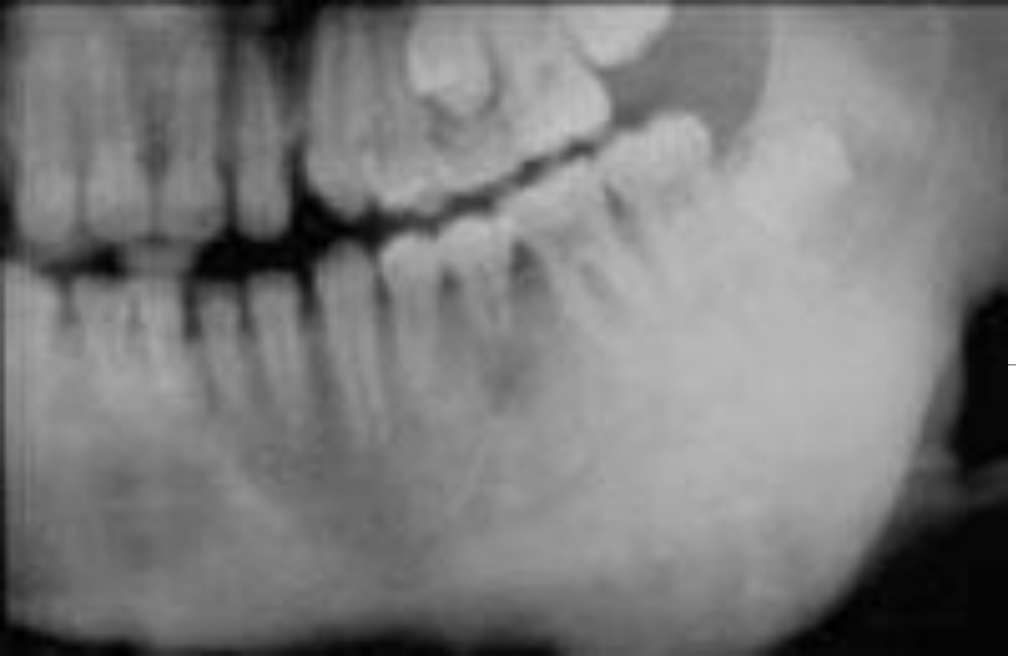
Lack of sharp marginal definition

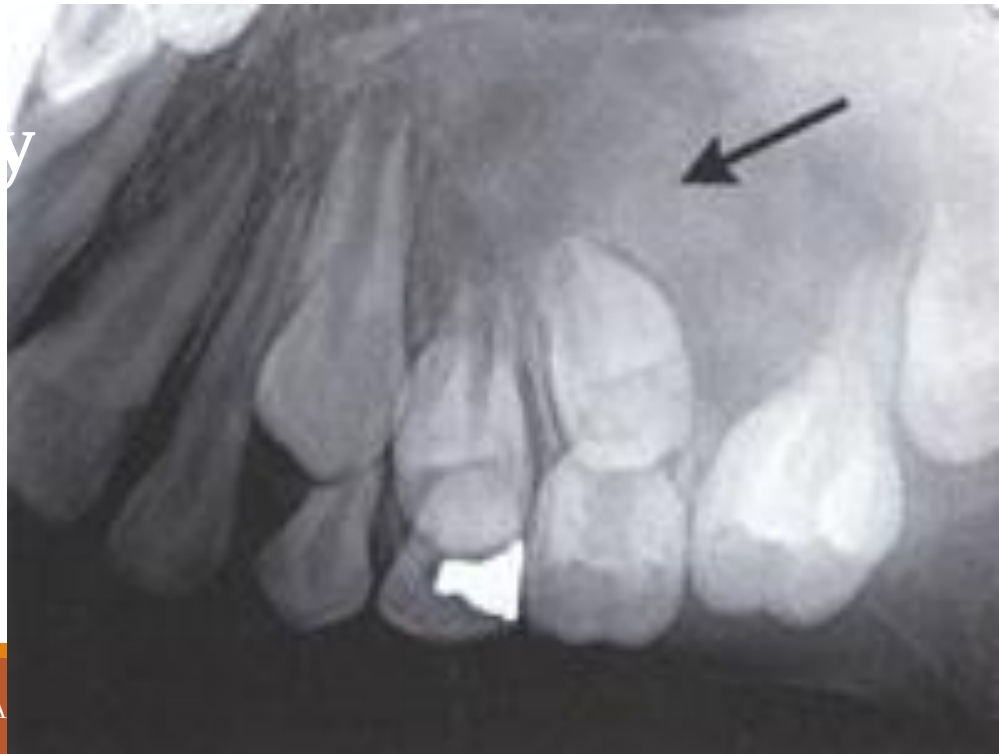
Ill defined lamina dura





Radiograph of right maxillary opacity (fibrous dysplasia)





Clinical & radiographic features

Most characteristic feature of FD

- Increase in density of the base of the skull involving the occiput, sella turcica, roof of the orbit, & frontal bones

Polyostotic FD

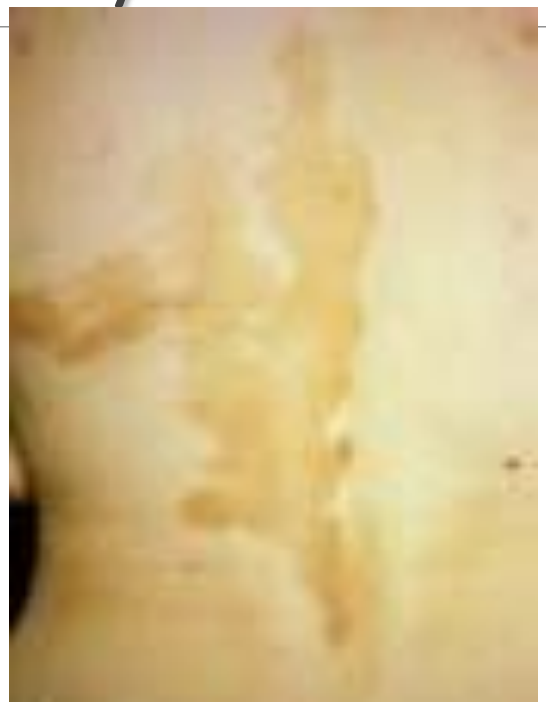
Uncommon

Females

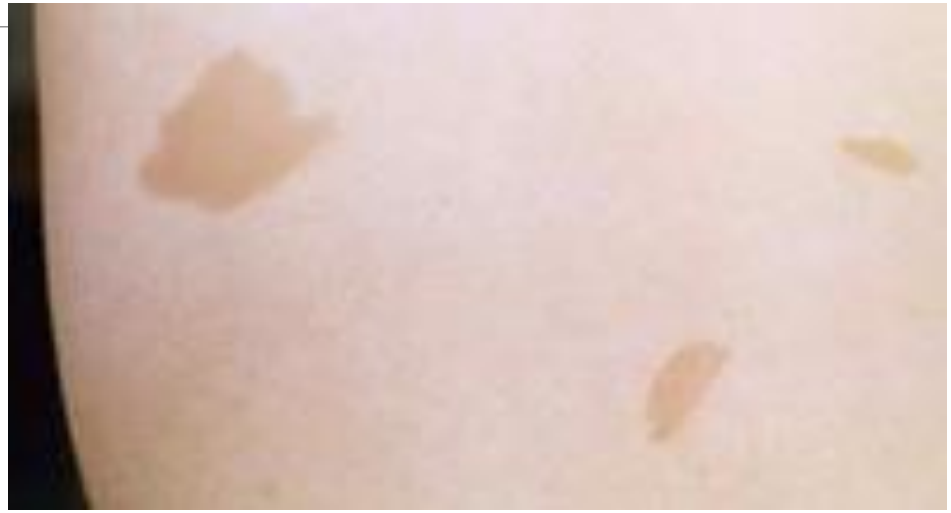
Few bones to 75% of skeleton

Jaffe Lichenstein syndrome

Polyostotic FD



Coastline of Maine



Coastline of California

Jaffe Lichenstein syndrome

Pigmented oral mucosal lesions may be present

Polyostotic FD

Pathologic fracture with pain and deformity

HOCKEY-STICK DEFORMITY-

Leg length discrepancy due to involvement of upper portion of femur

McCune-Albright Syndrome

Polyostotic fibrous dysplasia + café-au-lait pigmentation+multiple Endocrinopathies

Accelerated skeletal growth

Cushing's syndrome

Hyperthyroidism

Hyperparathyroidism

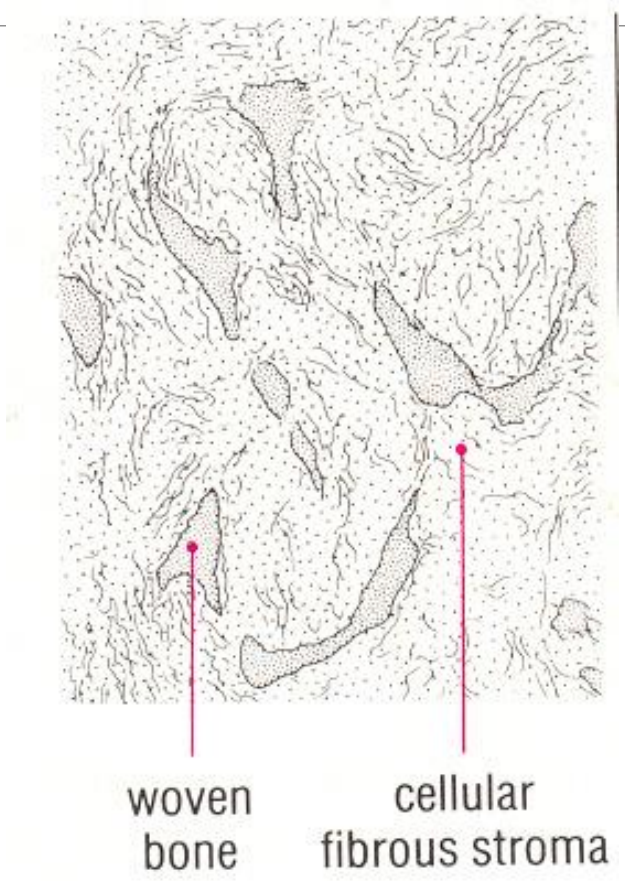
Diabetes mellitus

Sexual precocity in females

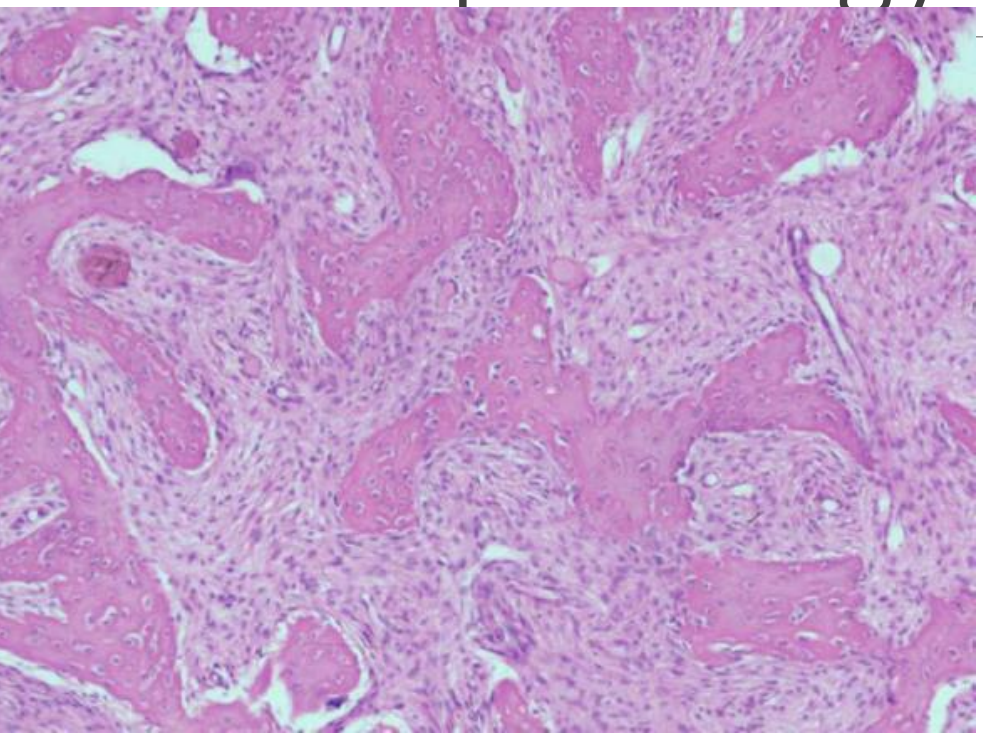
Gynaecomastia

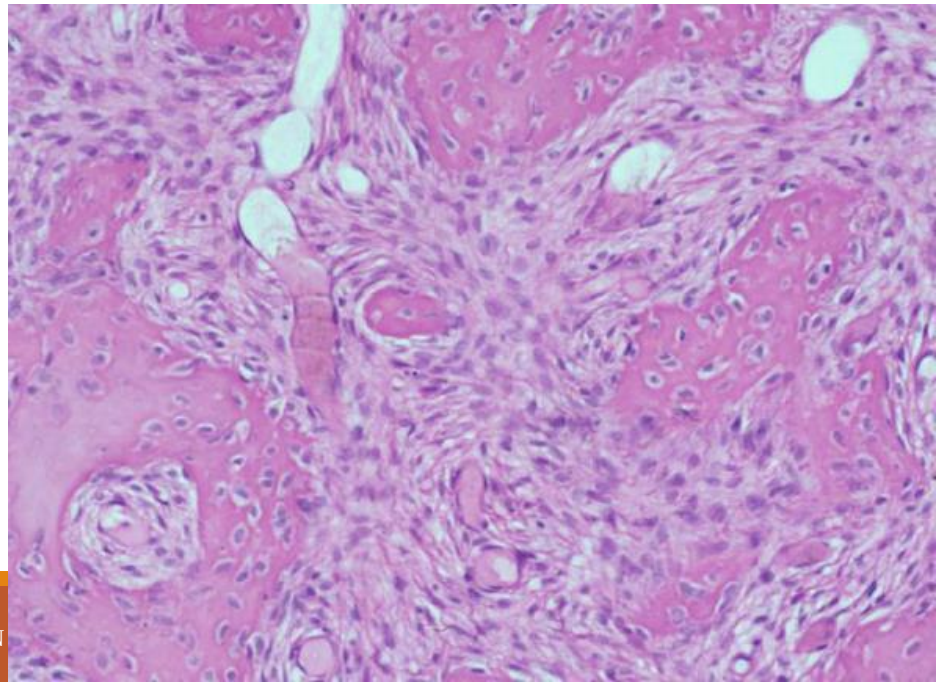
Multiple soft tissue myxomas

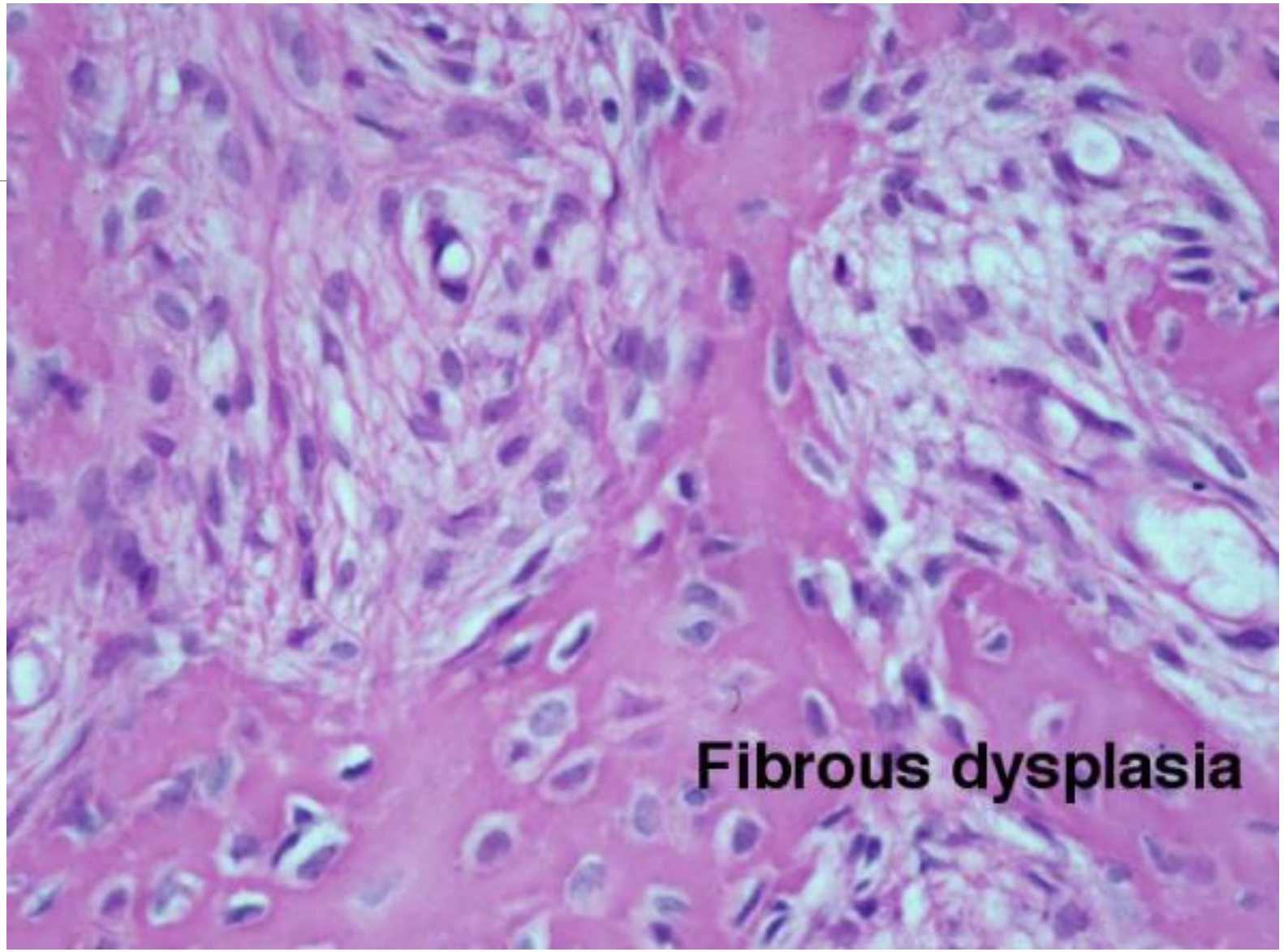
Histopathology

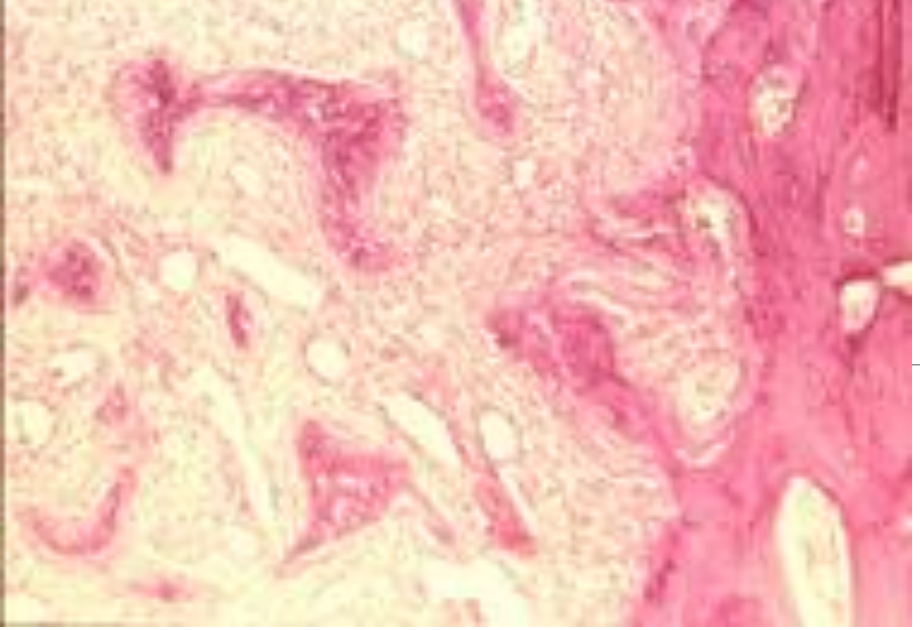


Histopathology





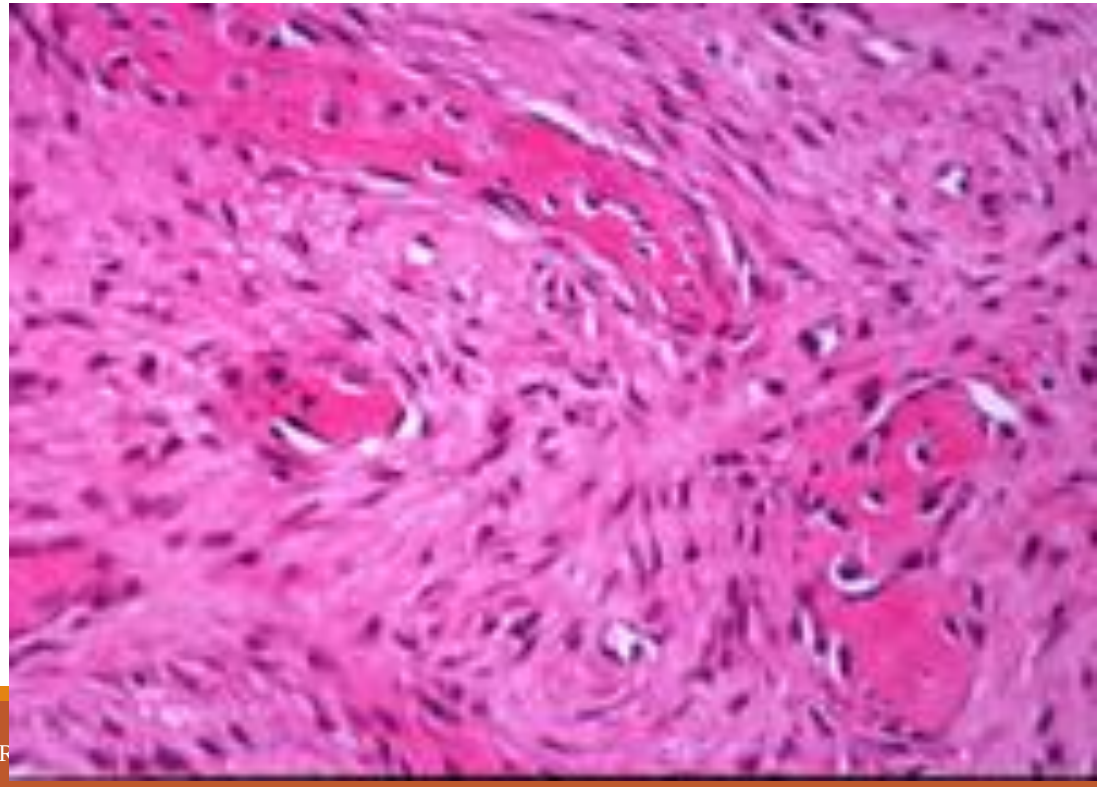

Fibrous dysplasia



No osteoid rim

or

osteoblastic rimming ???



Maxillofacial fibro-osseous lesions: classification and differential diagnosis.
Slootweg PJ.

Semin Diagn Pathol. 1996 May;13(2):104-12.

Fibrous dysplasia shows evenly distributed islands of woven bone that fuse with surrounding bone. *The presence of lamellar bone and osteoblastic rimming does not contradict that diagnosis as they would for lesions occurring outside the maxillofacial bones.*

Calcified spherules may be seen but not numerous

FD displays a monotonous pattern unlike OF

Fusion of lesional bone with normal bone

Fibrous Dysplasia

LABORATORY FINDINGS

- Serum alkaline phosphatase increased
- Premature secretion of pituitary follicle-stimulating hormone
- Elevated BMR

Differential Diagnosis

Ossifying fibromas - demarcated or encapsulated, broad variation in mineralized material (Fibrous dysplasia shows evenly distributed islands of woven bone that fuse with surrounding bone)

Osteosarcoma- osteoid rimming

Differential Diagnosis

Reed viewed fibrous dysplasia as an arrest of bone maturation at the woven stage of development, whereas ossifying fibroma was believed to be a benign neoplasm of bone, in which normal bone architecture was replaced by a tissue composed of collagen fibers, fibroblasts, and various amounts of calcified tissue with the potential for unlimited and destructive growth

Differential Diagnosis

Osteosarcoma- osteoid rimming

Treatment

Smaller lesions may be resected in entirety

Surgical recontouring

25-50% of patients show regrowth after surgical shave-on

Contra-indication-Radiotherapy because of risk of osteosarcoma

Reactive (dysplastic) lesions arising in tooth –bearing area

Most common fibro osseous lesions of the jaws

Periapical cemento-osseous dysplasia

Focal cemento-osseous dysplasia

Florid cemento-osseous dysplasia

Periapical cemento-osseous dysplasia

Apical areas of vital Mandibular incisor teeth

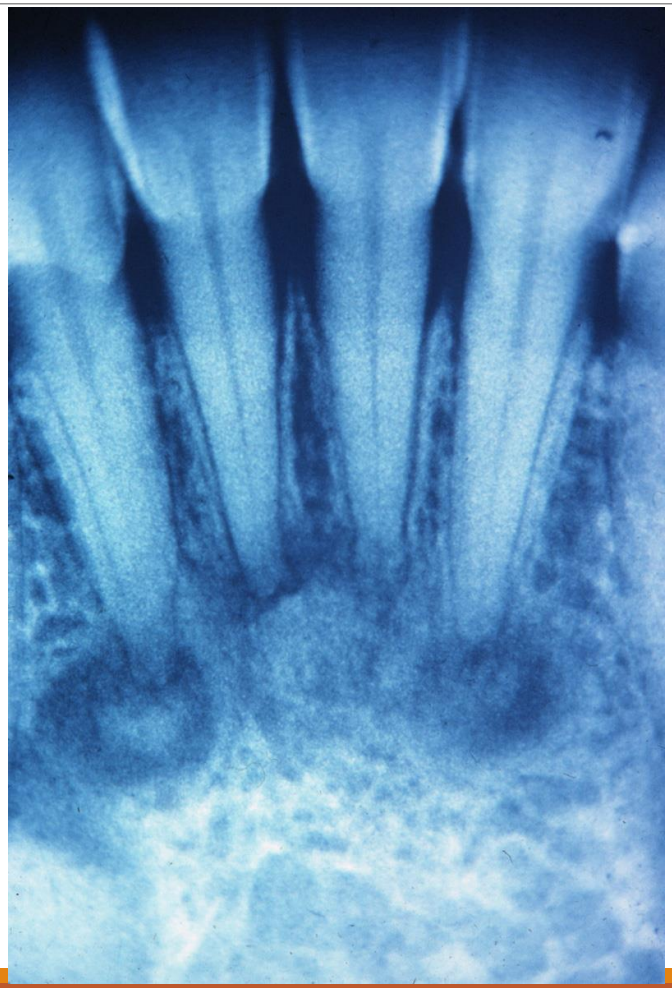
Multiple lesions, rarely exceeds 1 cm in diameter

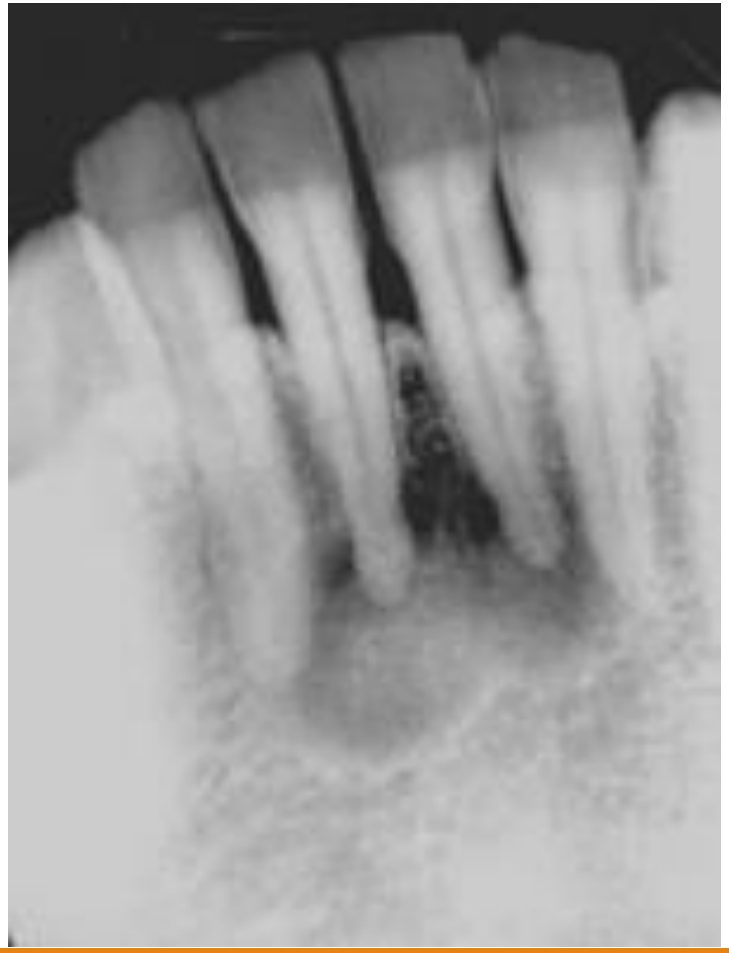
Females & blacks

Over 30 years

Asymptomatic ; on radiographic examination

Periapical cemento-osseous dysplasia





Thin radiolucent rim surrounding lesion

Periapical cemento-osseous dysplasia

Does not require any treatment

Isolated lesion- biopsied

Focal cemento-osseous dysplasia

Did not meet the criteria for FD or OF

"localized fibro-osseous-cemental lesions"

Designated as *osseous dysplasia* reaction of bone to injury

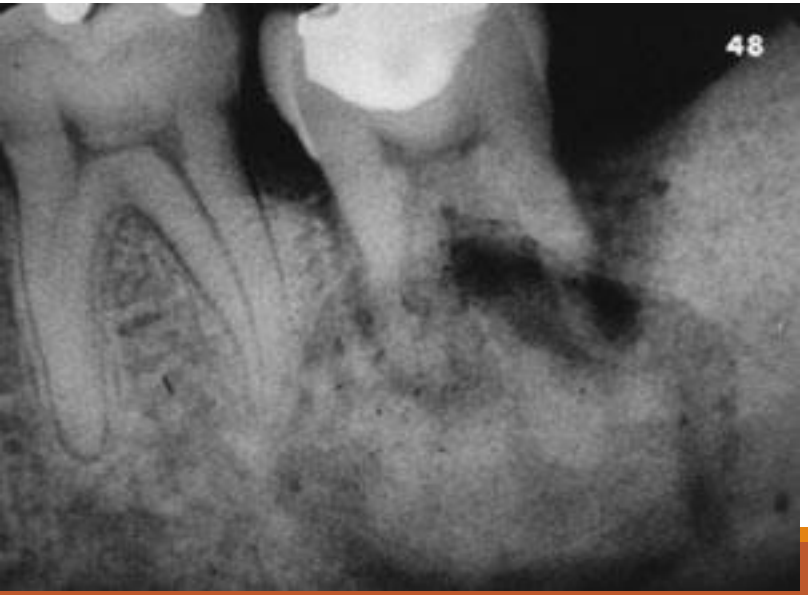
Females ; posterior mandible; edentulous area

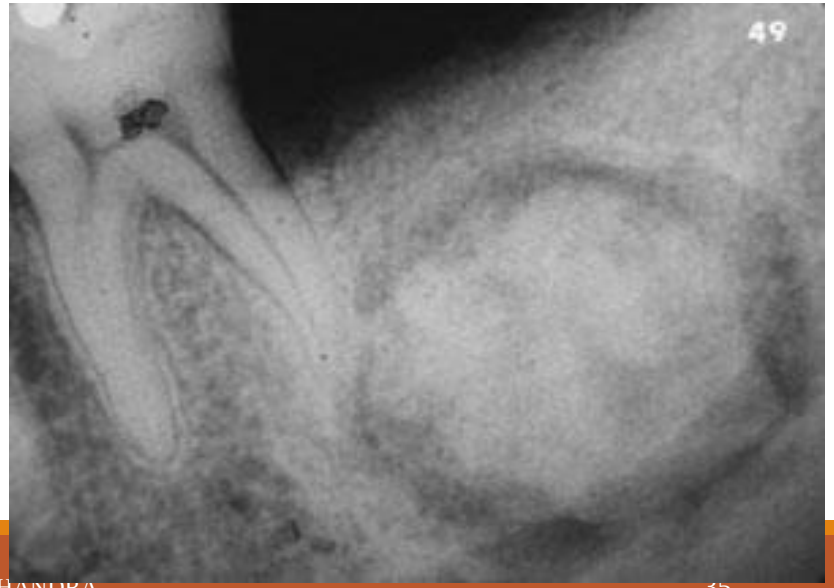
4^{th} to 5^{th} decade

Asymptomatic

Focal cemento-osseous dysplasia

Radiolucent to densely Radiopaque with a thin Peripheral radiolucent rim
Common Is mixed radiolucent-radiopaque stage
No bone expansion





Focal cemento-osseous dysplasia

On surgical exploration the tissue is gritty, hemorrhagic & removed by curettage in small fragments

Simple bone cyst may be associated with FCOD

NO TENDENCY TO ENLARGE
PARTIAL REMOVAL MAY CAUSE REGRESSION ON LESION

Florid Cemento-osseous dysplasia

Middle age to elderly black females

Sclerosing osteitis, multiple enostosis, gigantiform cementoma

Seen only in tooth bearing area

Striking tendency toward bilateral, symmetrical involvement

Florid Cemento-osseous dysplasia

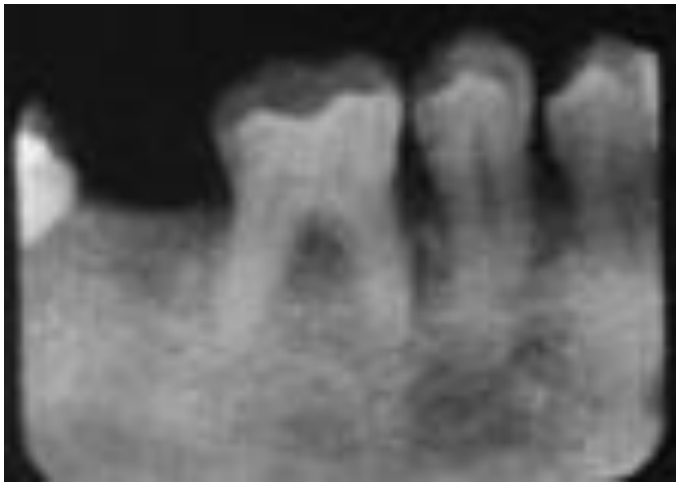
FCOD is defined as "Lobulated masses of dense, highly mineralised, almost acellular cemento-osseous tissue typically occurring in several parts of the jaws".

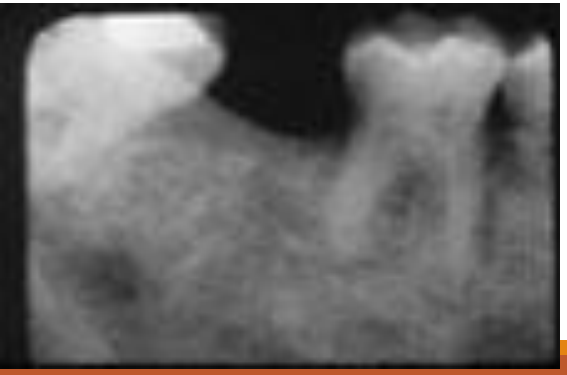
Florid Cemento-osseous dysplasia

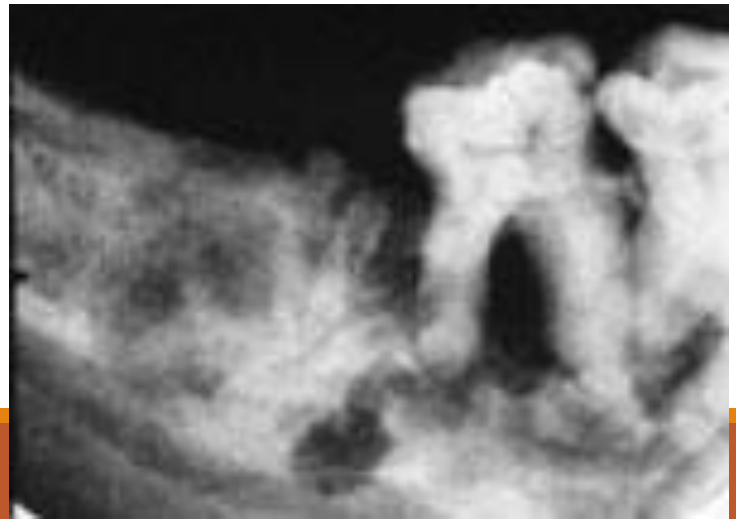
Small- asymtomatic

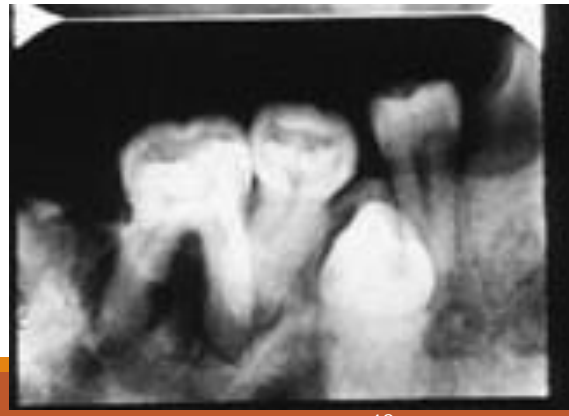
Large-Symptoms are usually associated with exposure of sclerotic cemental masses to oral environment from alveolar atrophy, tooth extraction and biopsy

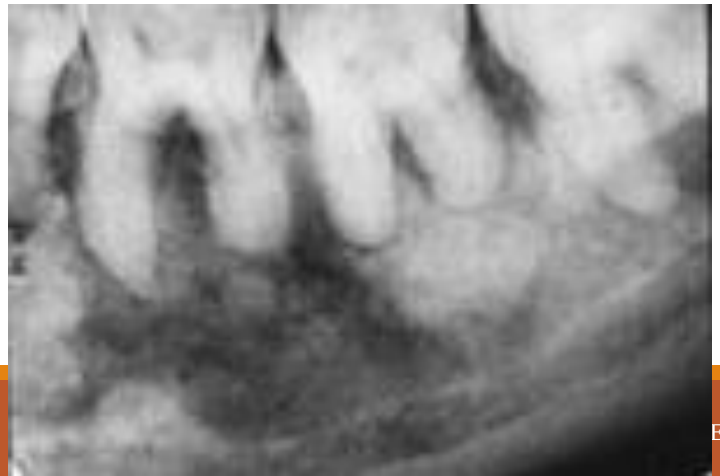
Florid Cemento-osseous dysplasia

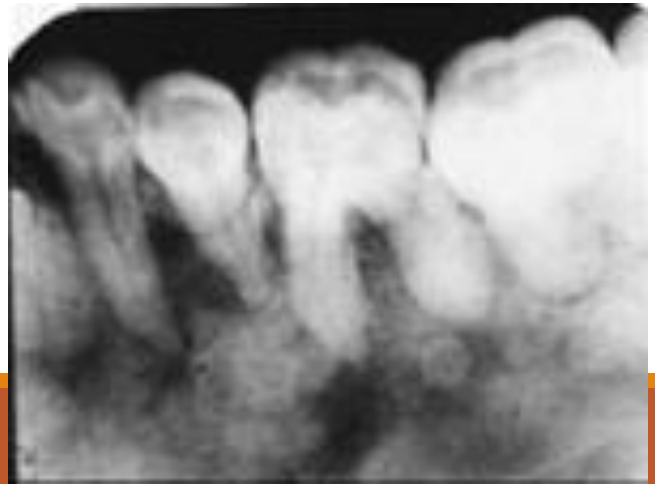




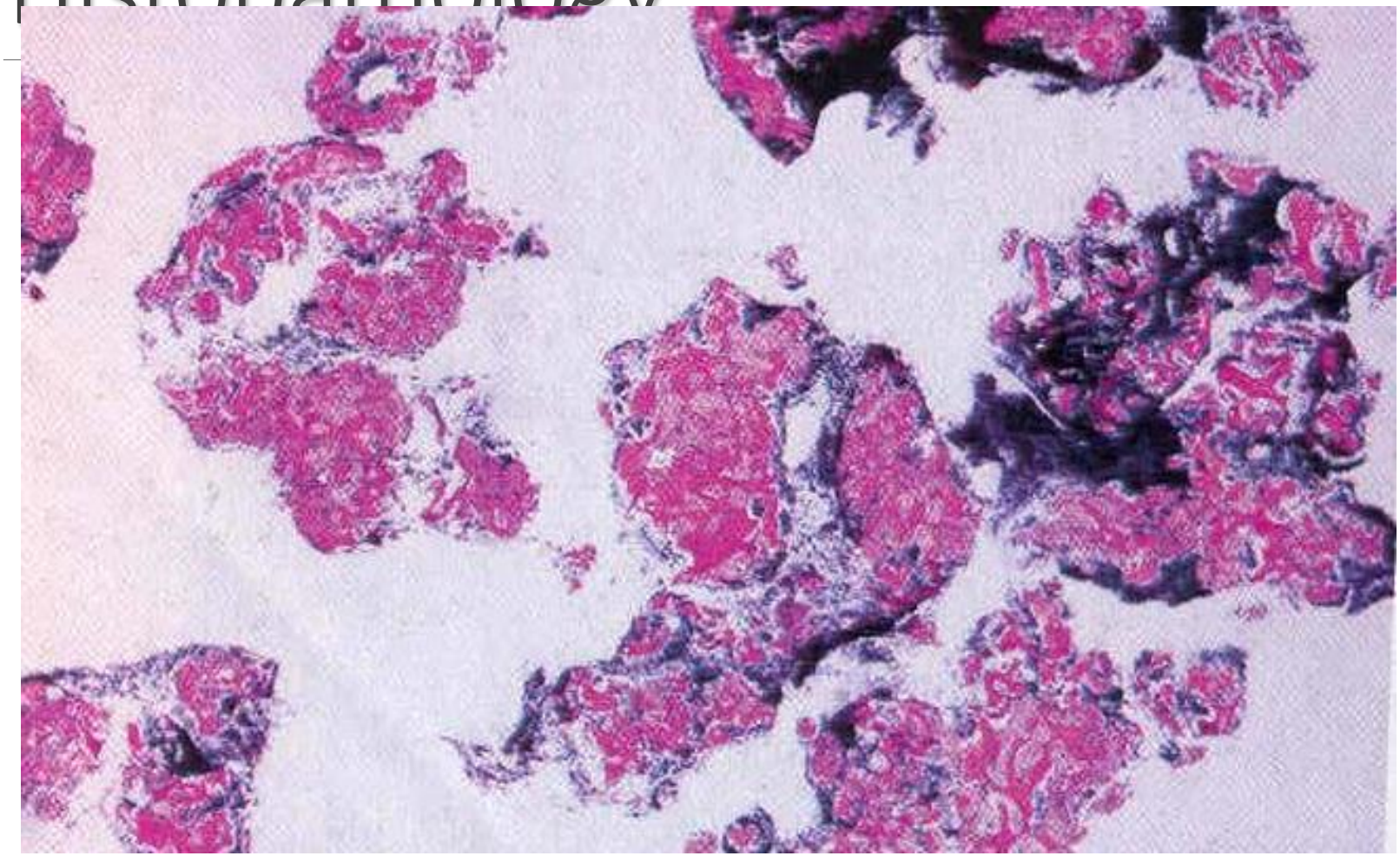






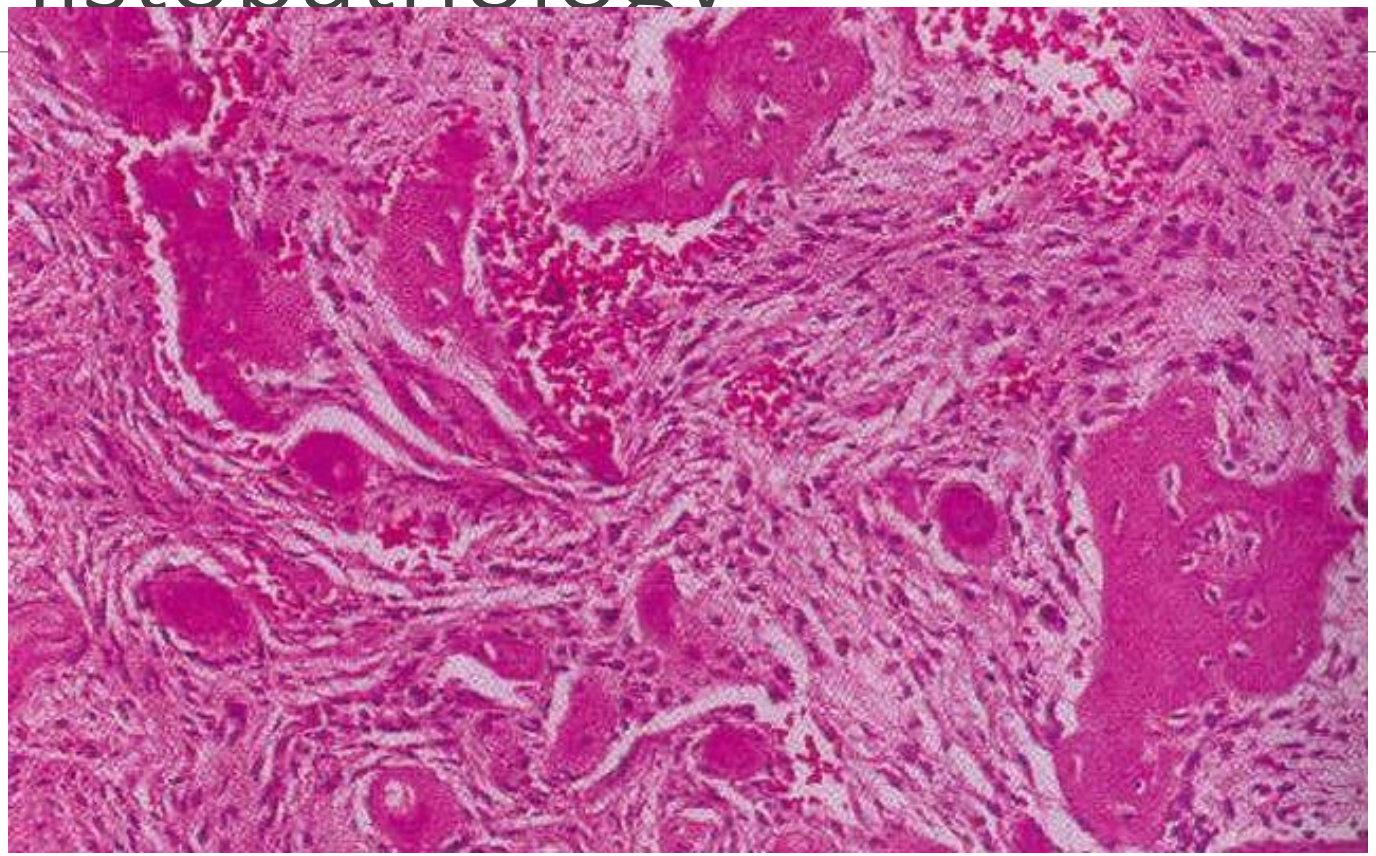


Histopathology



Fragments of cellular fibrous connective tissue containing scattered trabeculae of bone

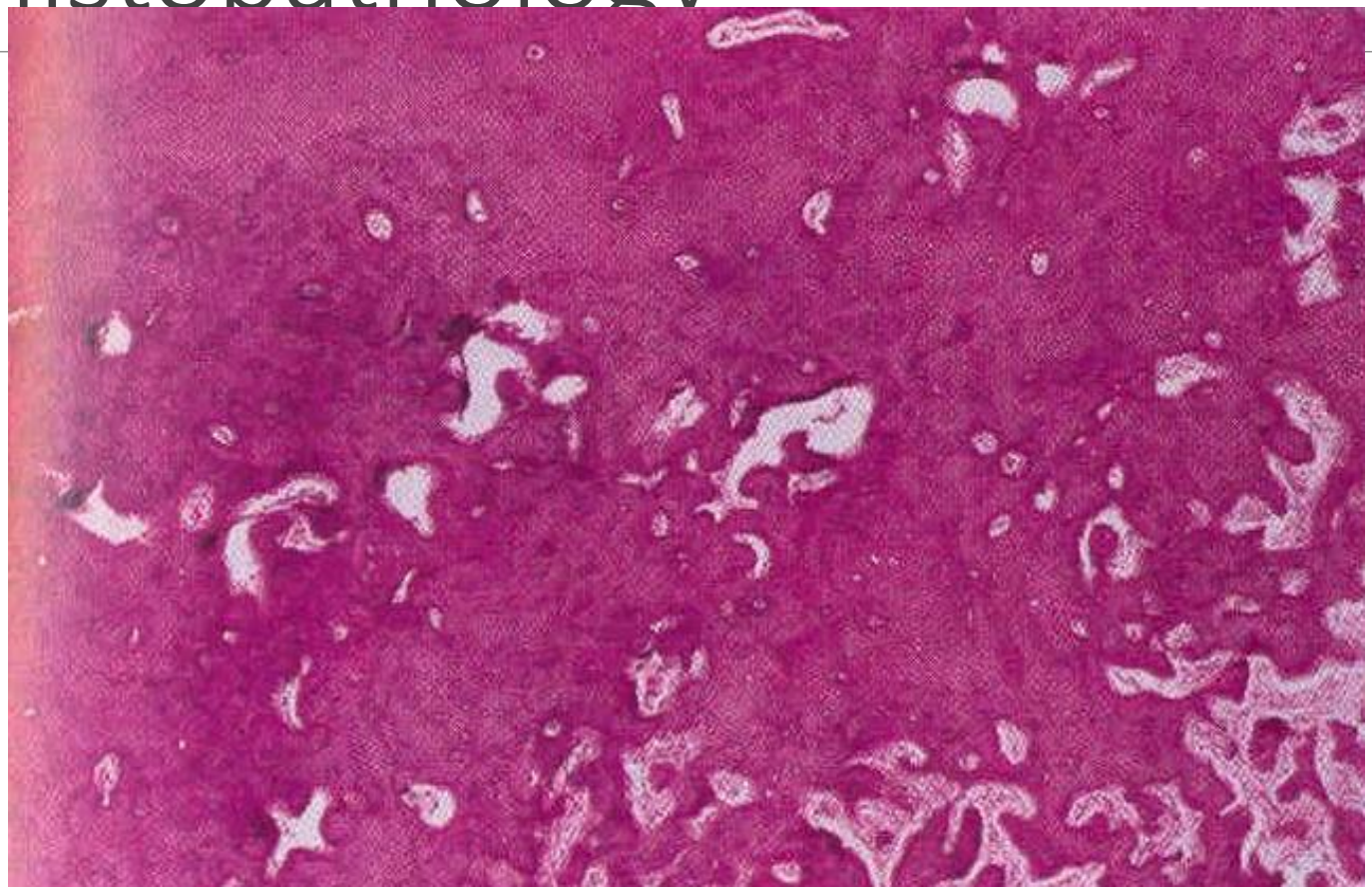
Histopathology



Spicules of bone and cementum like hard tissue with moderately cellular fibrous connective tissue.

Note hemorrhage around bony trabeculae

Histopathology



Late stage lesion showing a sclerotic mass of cemento-osseous material

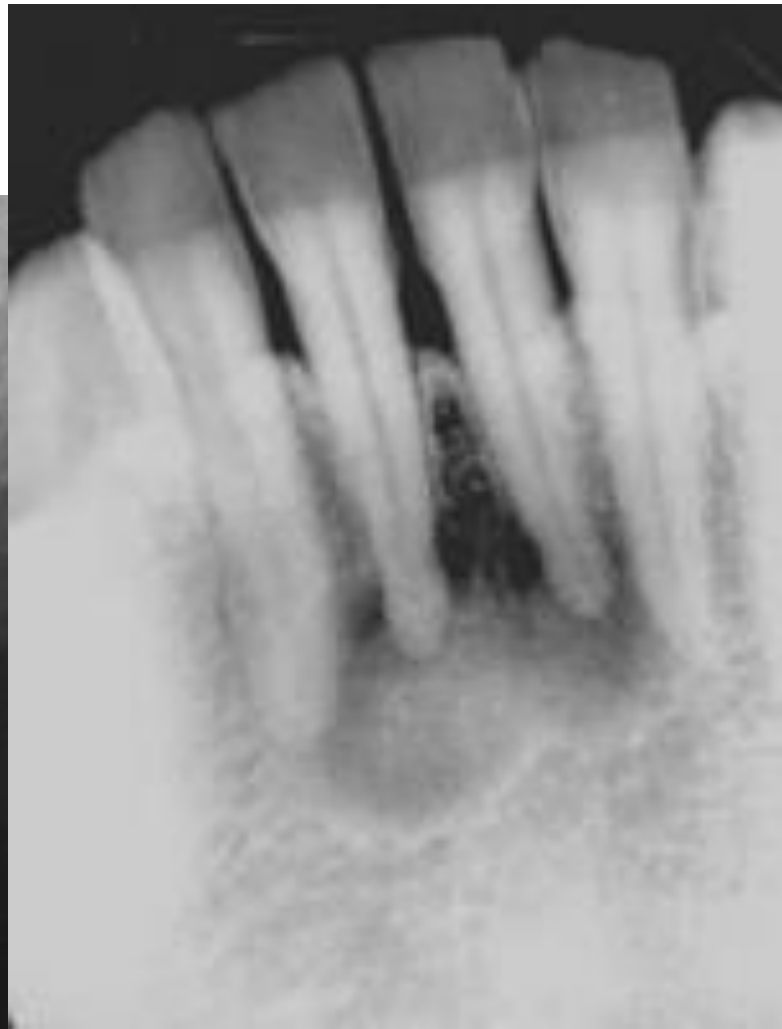
Differential Diagnosis

FD or OF CAN BE DISTINGUISHED ON THE BASIS OF CLINICAL PRESENTATION

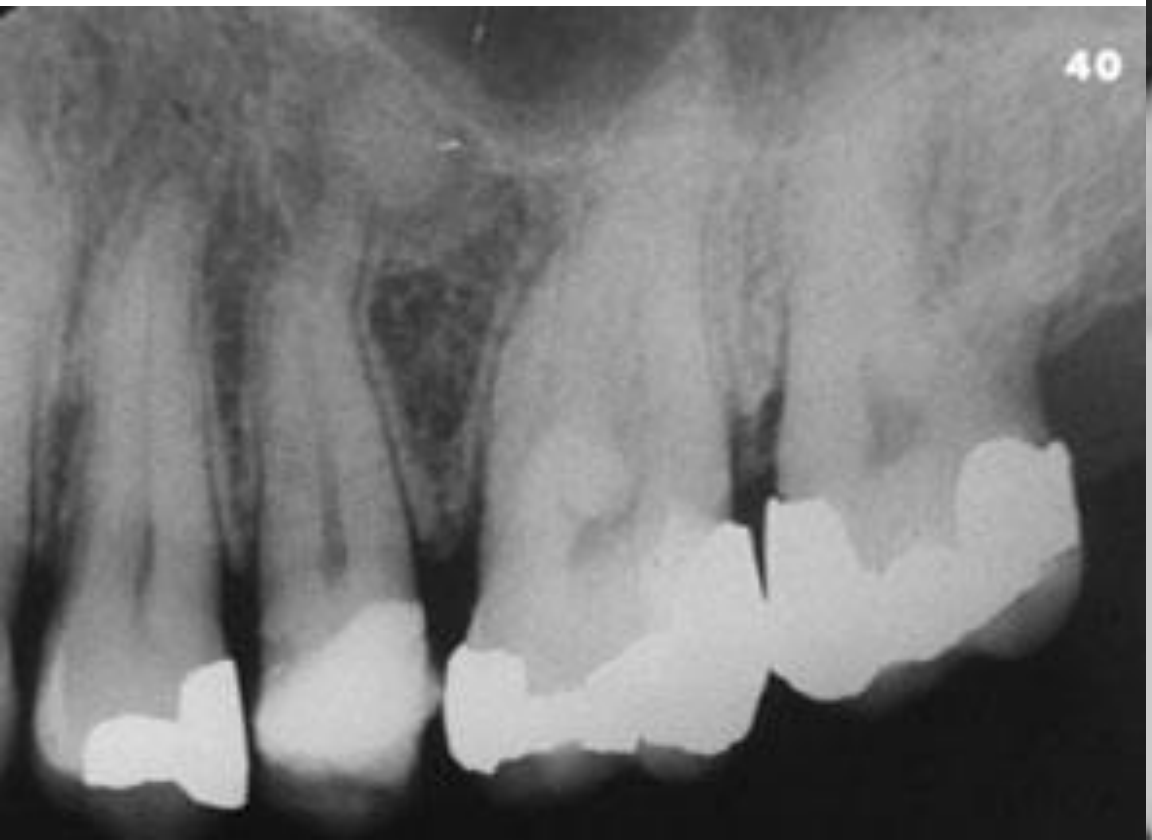
-BONY EXPANSION NOT SEEN
-CEMENTUM LIKE CALCIFICATION ARE SEEN
-GROSSLY GRITTY & HEMORRHAGE (UNLIKE *OF*)
-Periapical cemental dysplasia can be found in the tooth-bearing jaw area and are similar to ossifying fibroma but without demarcation.

Differential Diagnosis

PCD



OF



Differential Diagnosis

Periapical cemental dysplasia should be distinguished from cementoblastoma, a lesion similar to osteoblastoma but connected with tooth apices

Treatment

Management is often difficult & not satisfactory

Asymptomatic patient-wise to keep under observation

Symptomatic patient- antibiotic should be administered

Sequestration of cementum

like masses will occur slowly

followed by healing

Fibro osseous neoplasms

Ossifying fibroma

Juvenile active ossifying fibroma

Ossifying fibroma

1972 WHO – CF:odontogenic

1992 WHO – CF & OF variants

Defined as demarcated & occasionally encapsulated lesion consisting of fibrous tissue containing variable amounts of mineralized material resembling bone and /or cementum

Ossifying fibroma

Hamner *et al* - periodontal origin of ossifying fibroma.

capable of producing cementum and osteoid.

Krausen *et al* and Spjut *et al*, postulated that primitive mesenchymal cells in areas such as the ethmoid bone and long bones may produce cementum at sites distant from odontogenic tissue. They discredited the notion that these tumors arise from ectopic periodontal tissue in these locations.

Ossifying fibroma

3rd to 4th decades of life

Posterior mandible may extend into the ascending ramus

70-80% in premolar and molar area

Female predilection

Large tumors –slowly progressing enlargement of jaws

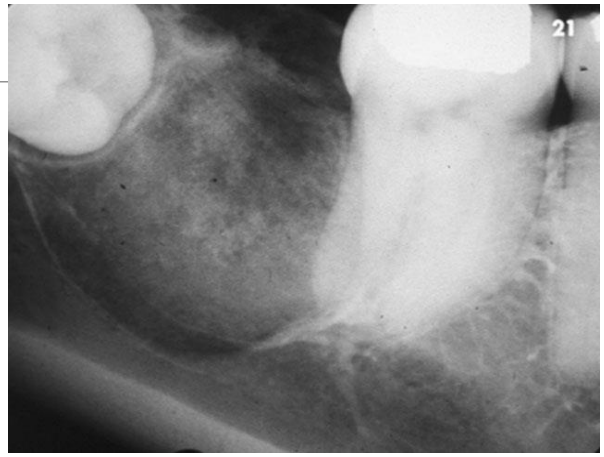
Painless swelling of involved bone

Facial asymmetry

Ossifying fibroma

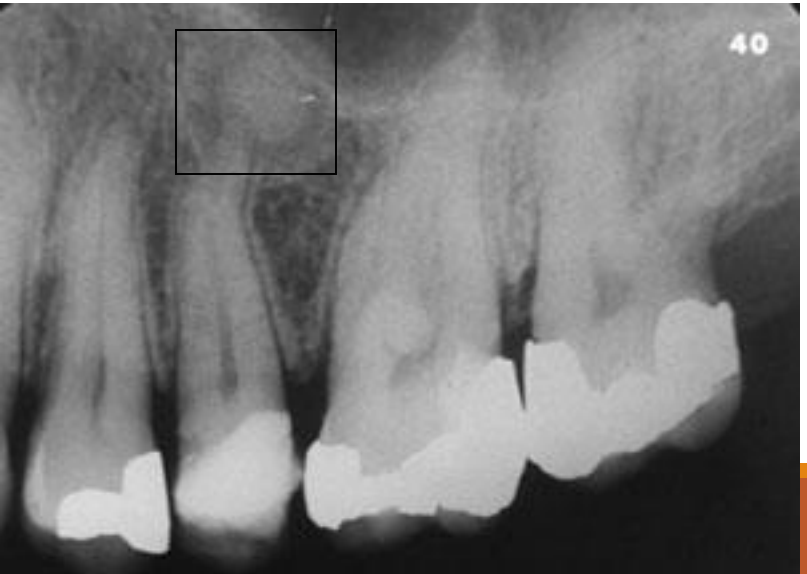
Unilocular lesion

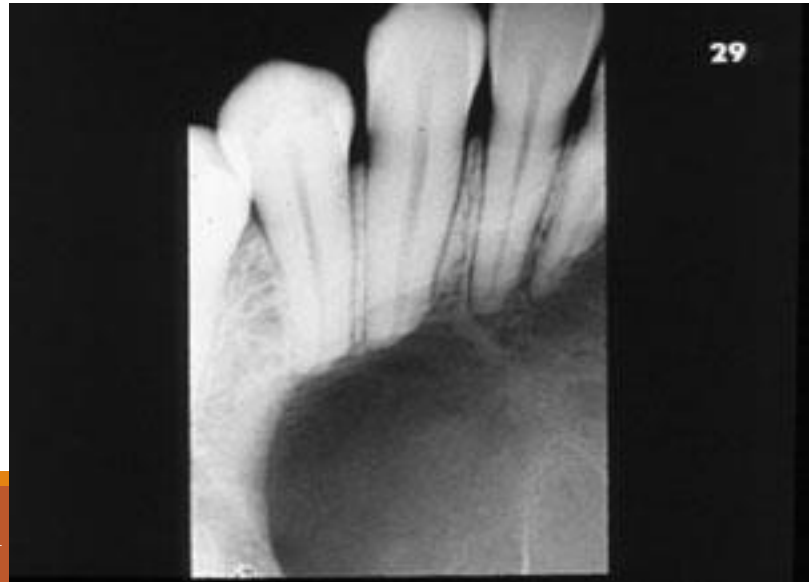
Root resorption may/may not be seen



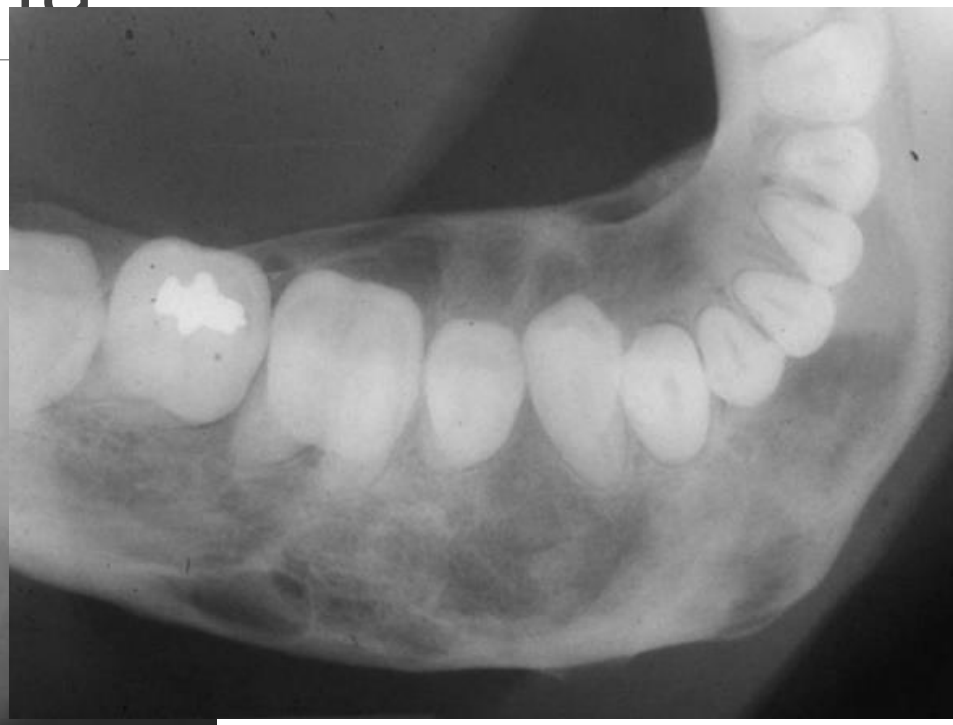
Radiolucent

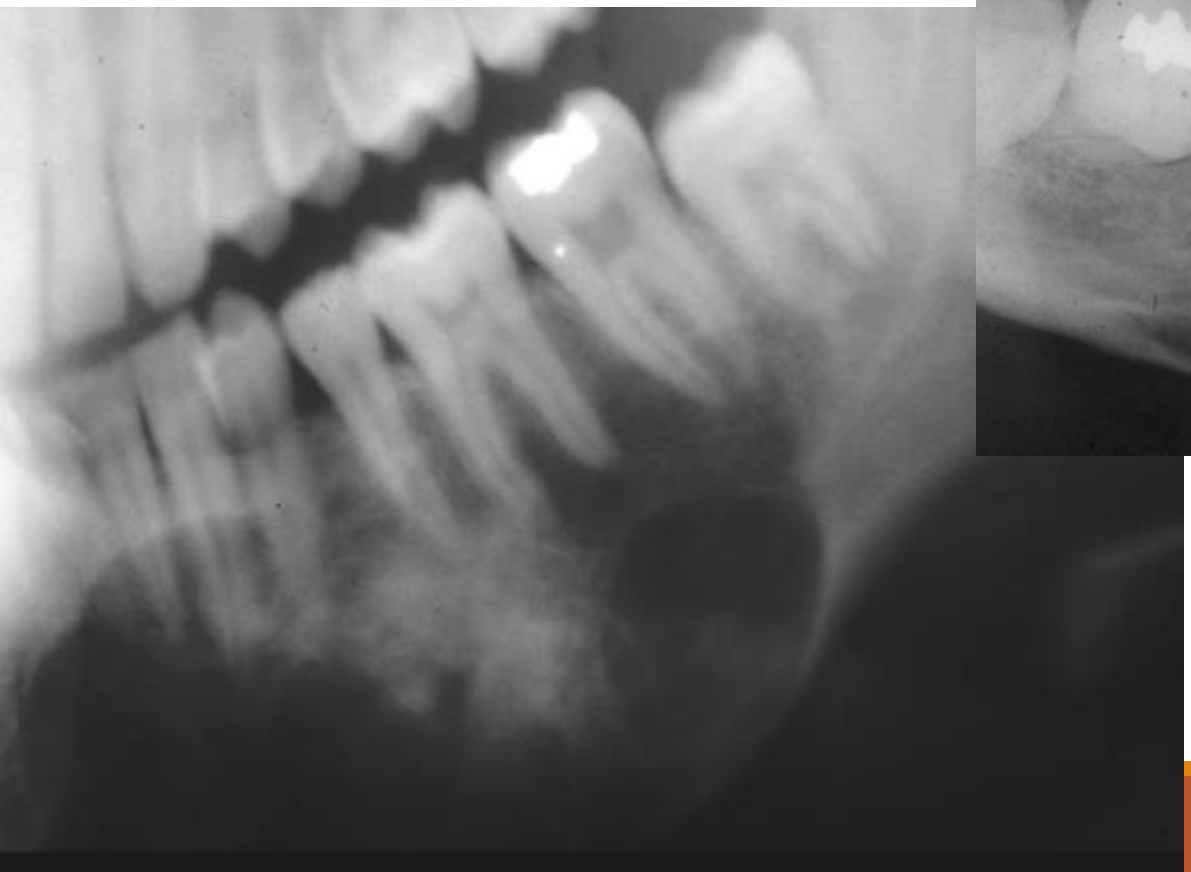
Radiopaque



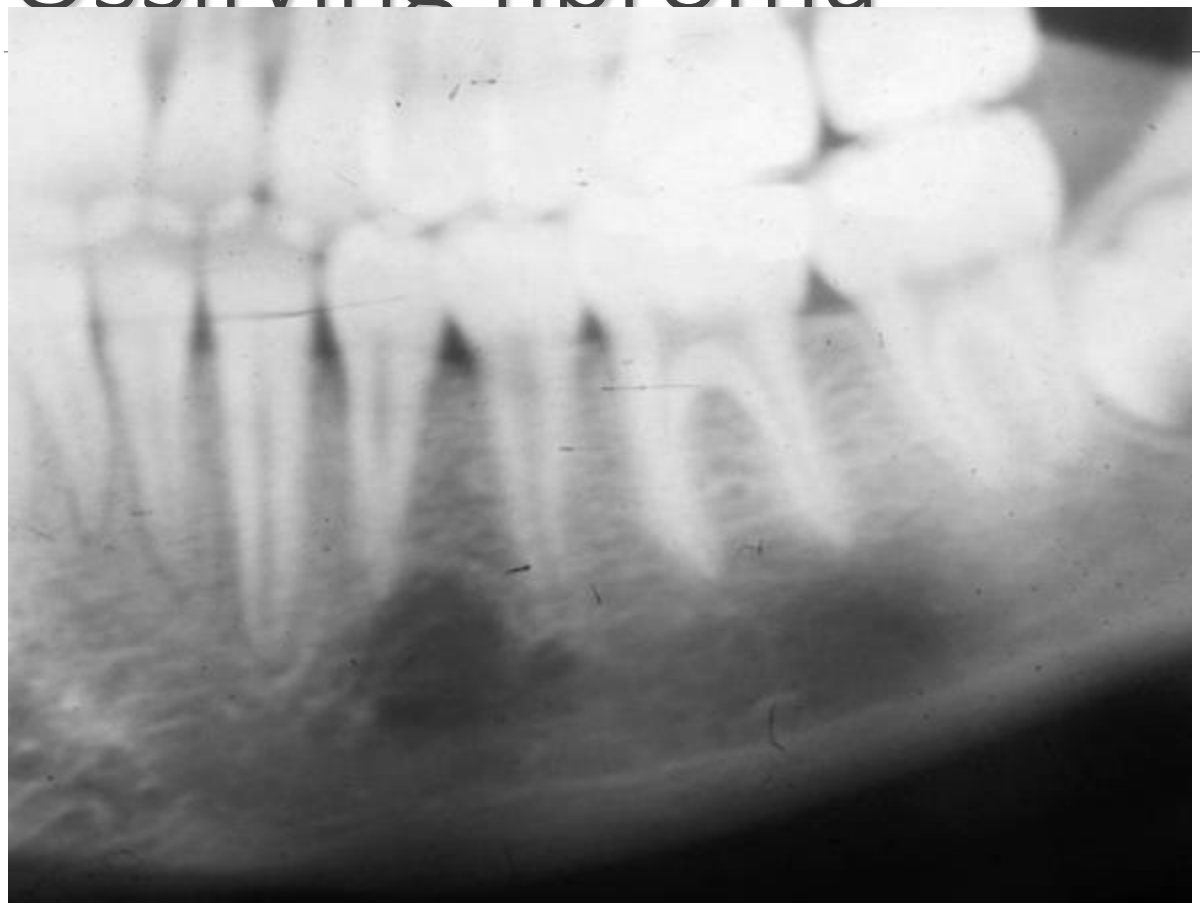


Ossifying fibroma



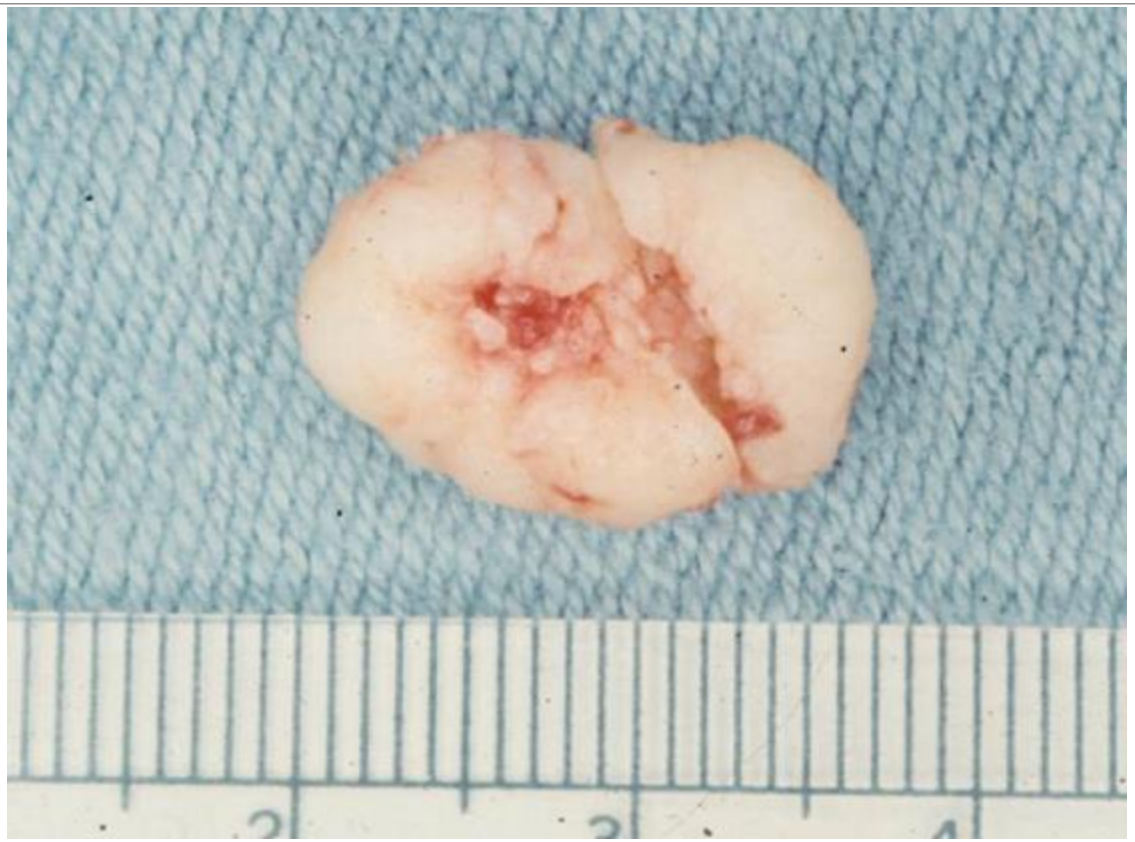


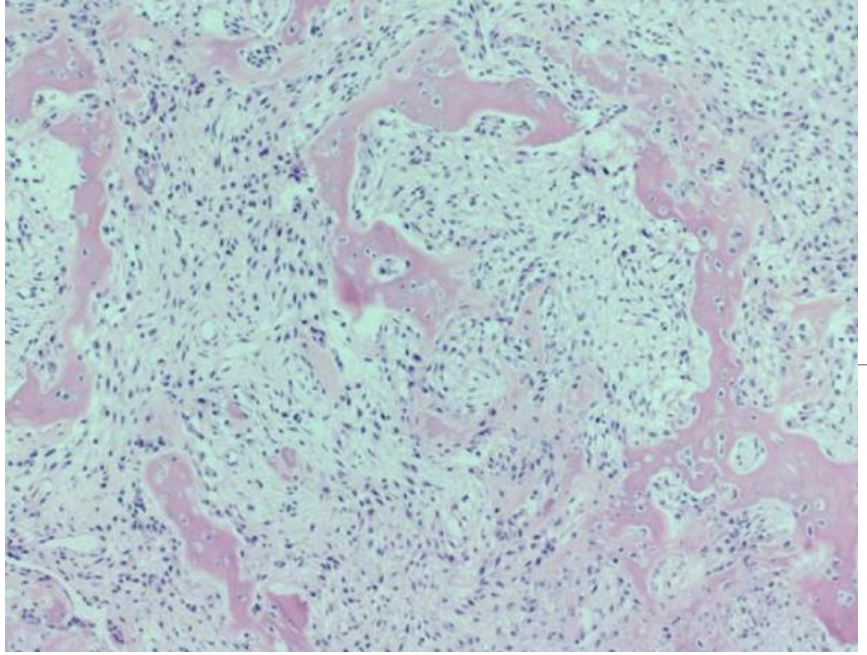
Ossifying fibroma



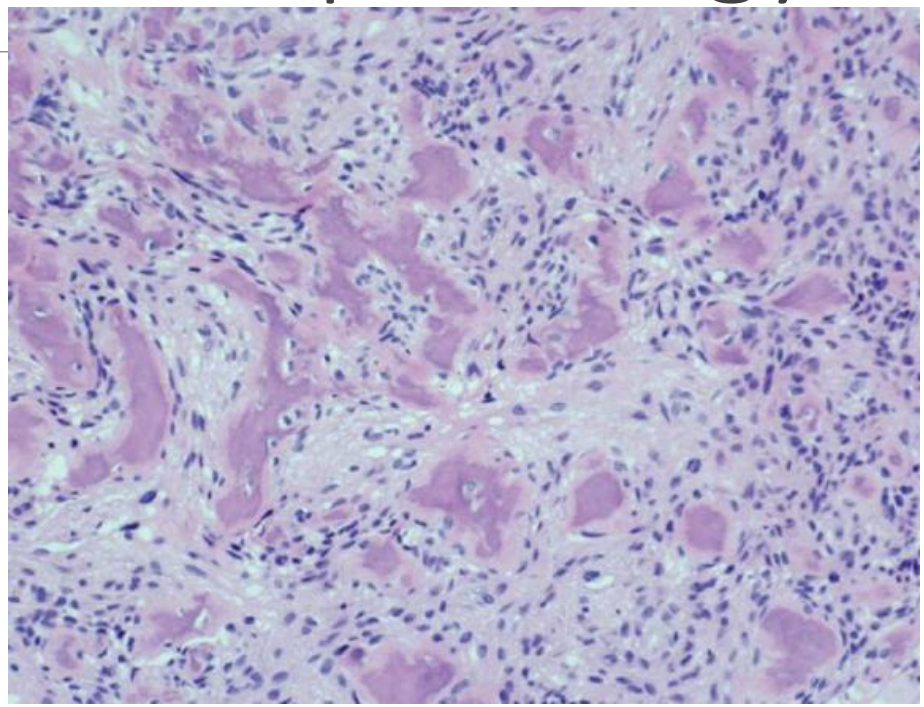
Radiograph of well-defined radiolucency in the body of the mandible (ossifying fibroma).

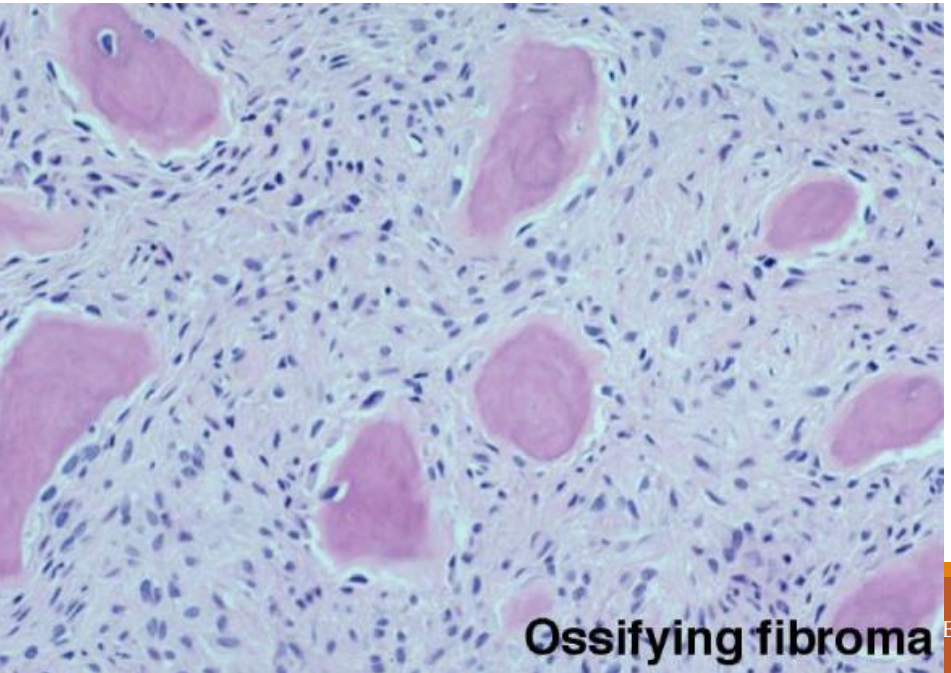
Ossifying fibroma



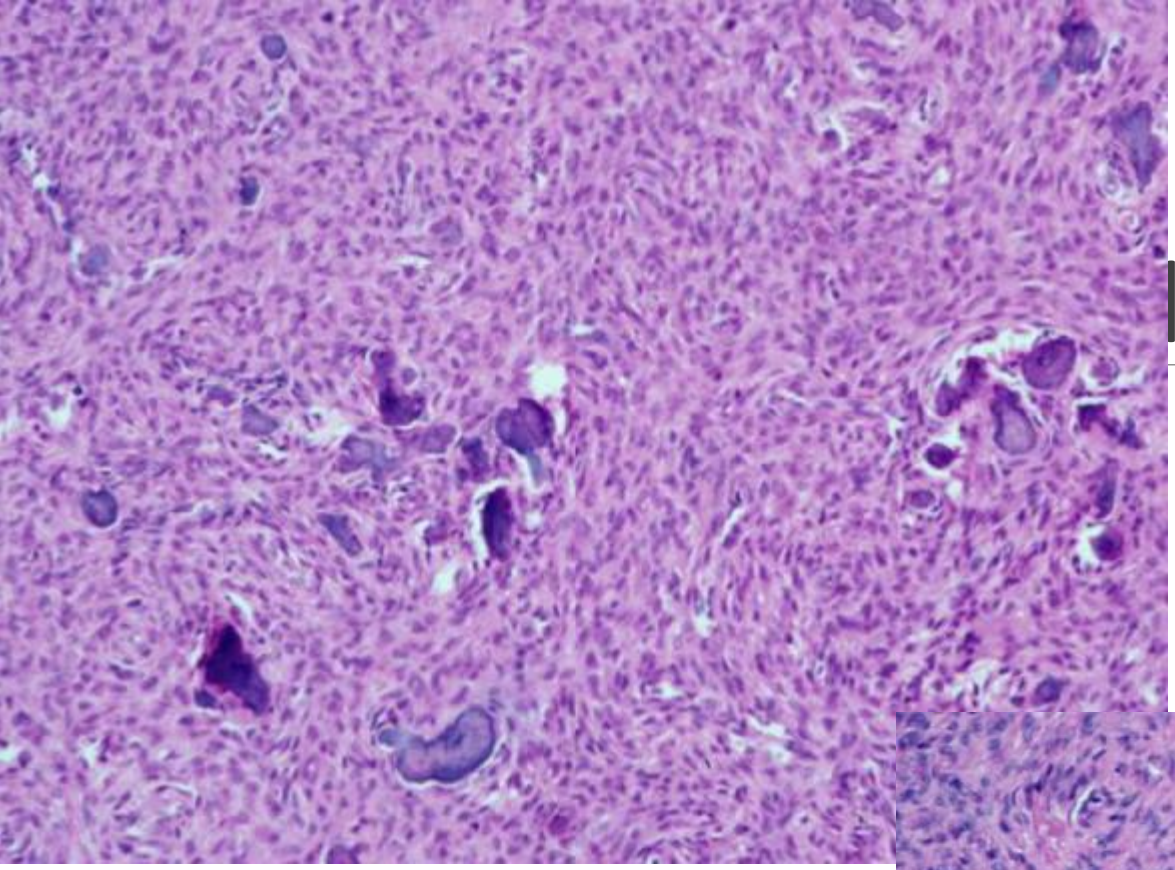


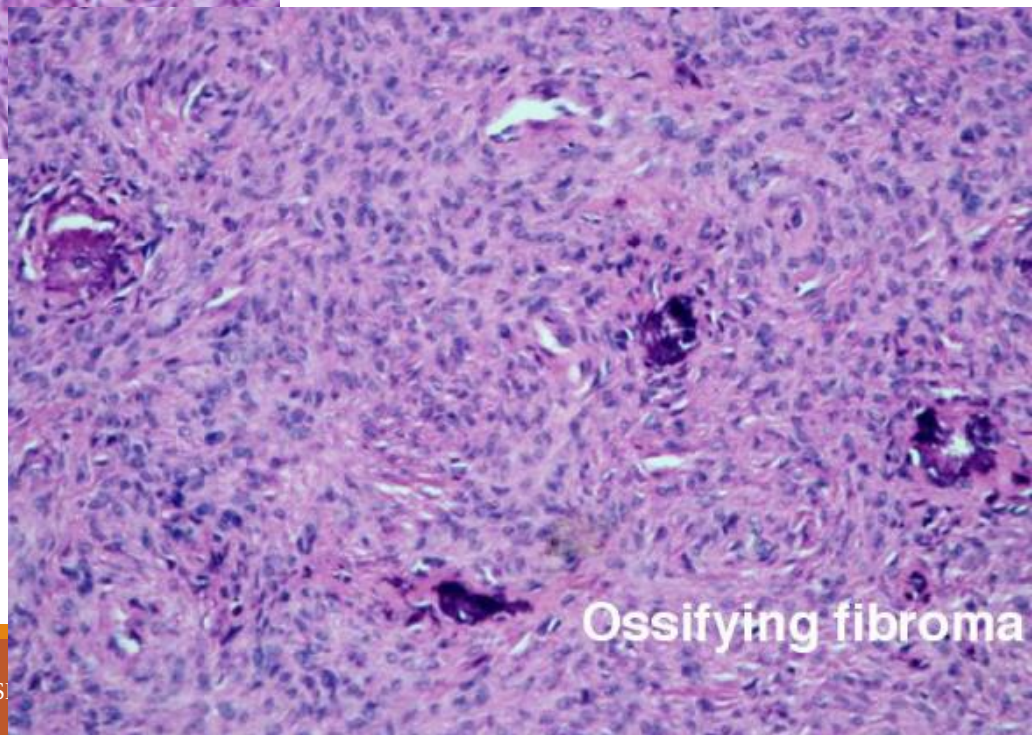
Histopathology





listopathology





Ossifying fibroma

Growth rate varies : histopathology cannot predict growth rate

Difficult to distinguish FD &OF Histopathologically

OF generally excised in one piece ; large lesions may require local resection and subsequent bone grafting

Prognosis is excellent ; no recurrence; no malignant change

Juvenile Aggressive Ossifying Fibrom

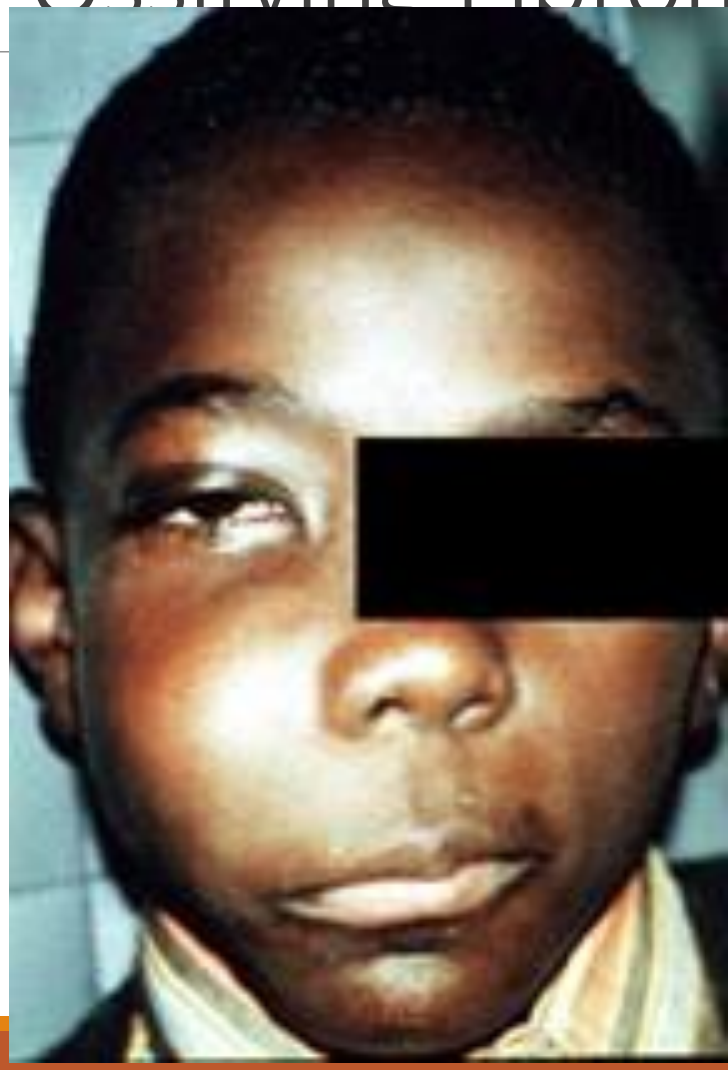
Trabecular variety

Psammomatoid variety

No sex predilection

5 To 15years(60%-70%)

Cortical expansion may result in clinically detectable facial enlargement



Juvenile Aggressive Ossifying Fibron

Maxilla, Para nasal sinuses & orbital & frontoethmoidal bone

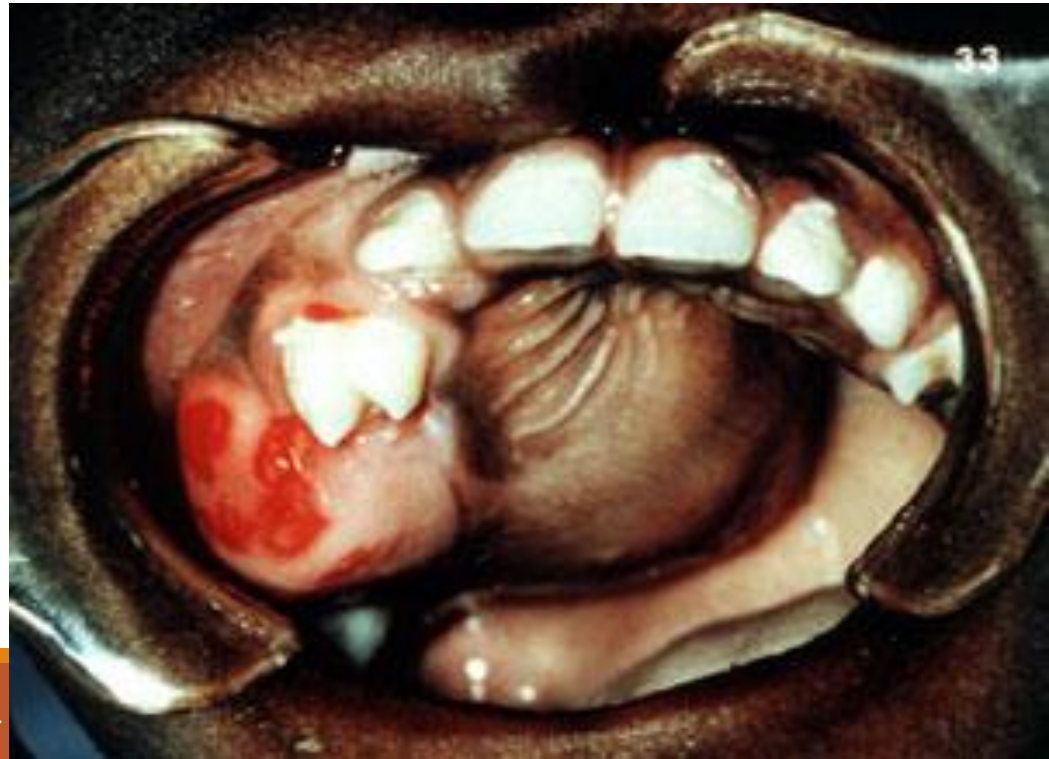
NASAL OBSTRUCTION

EXOPTHALMOS

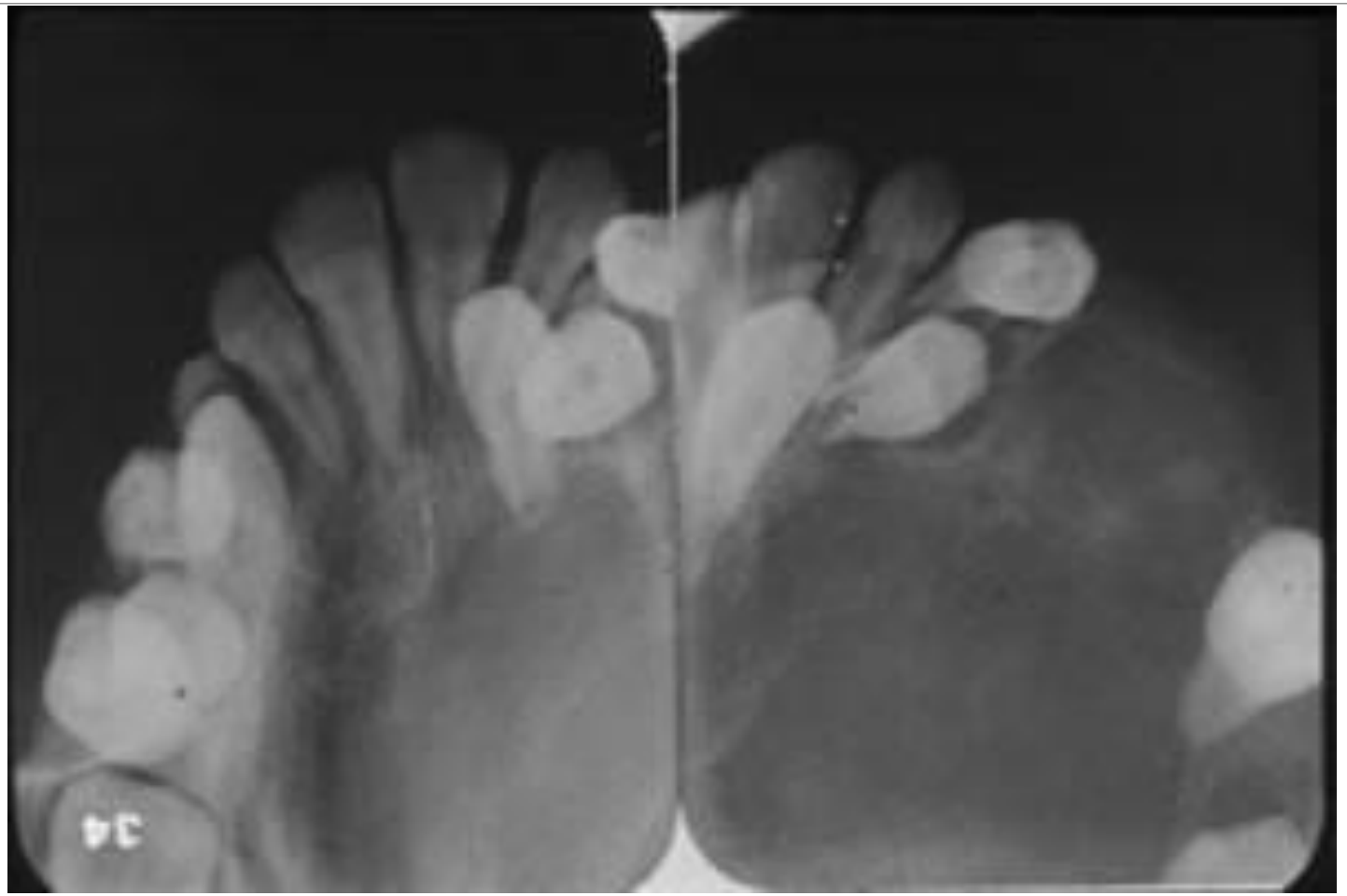
PROPTOSIS

BLINDNESS

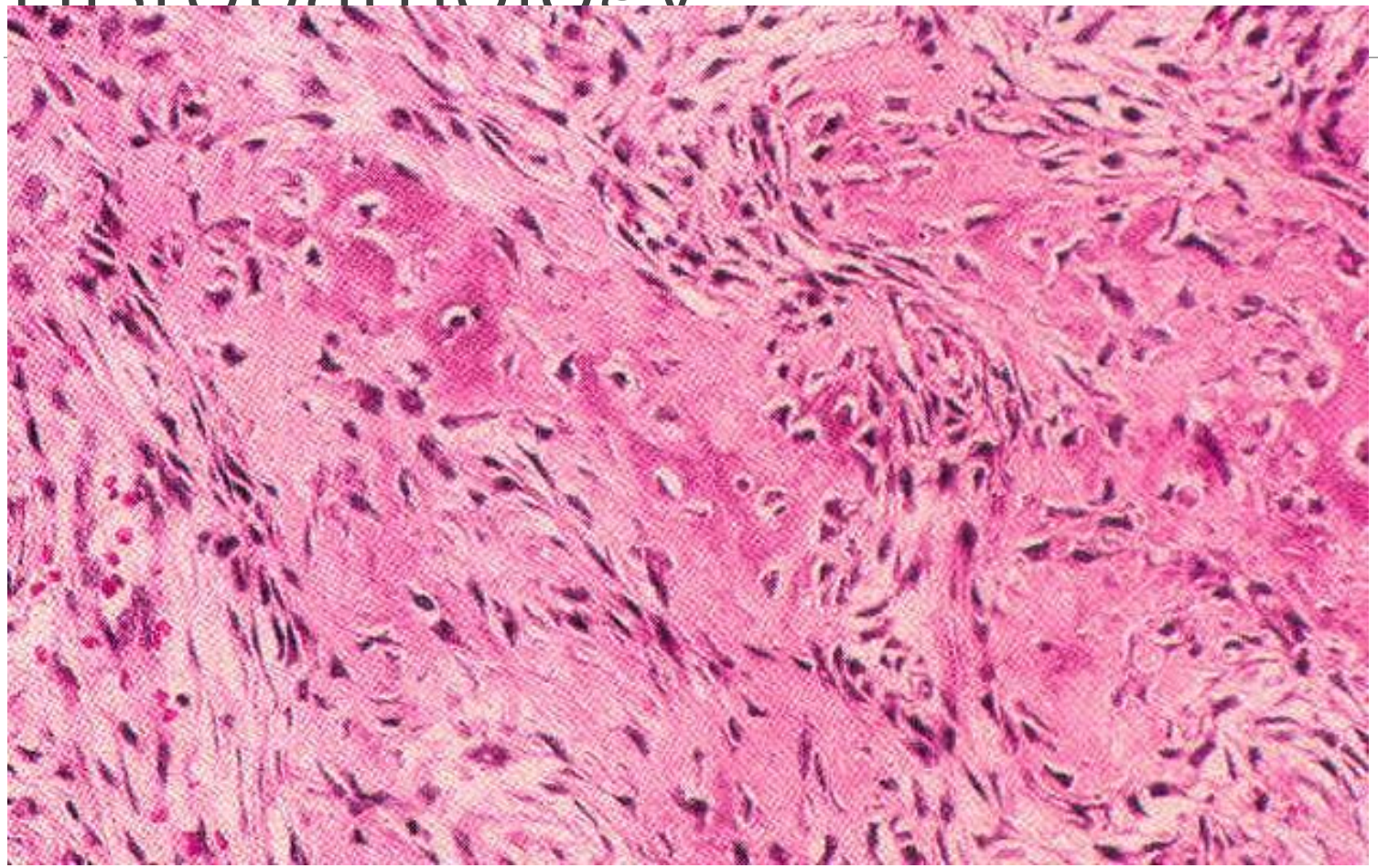
MENINGITIS



Juvenile Aggressive Ossifying Fibron

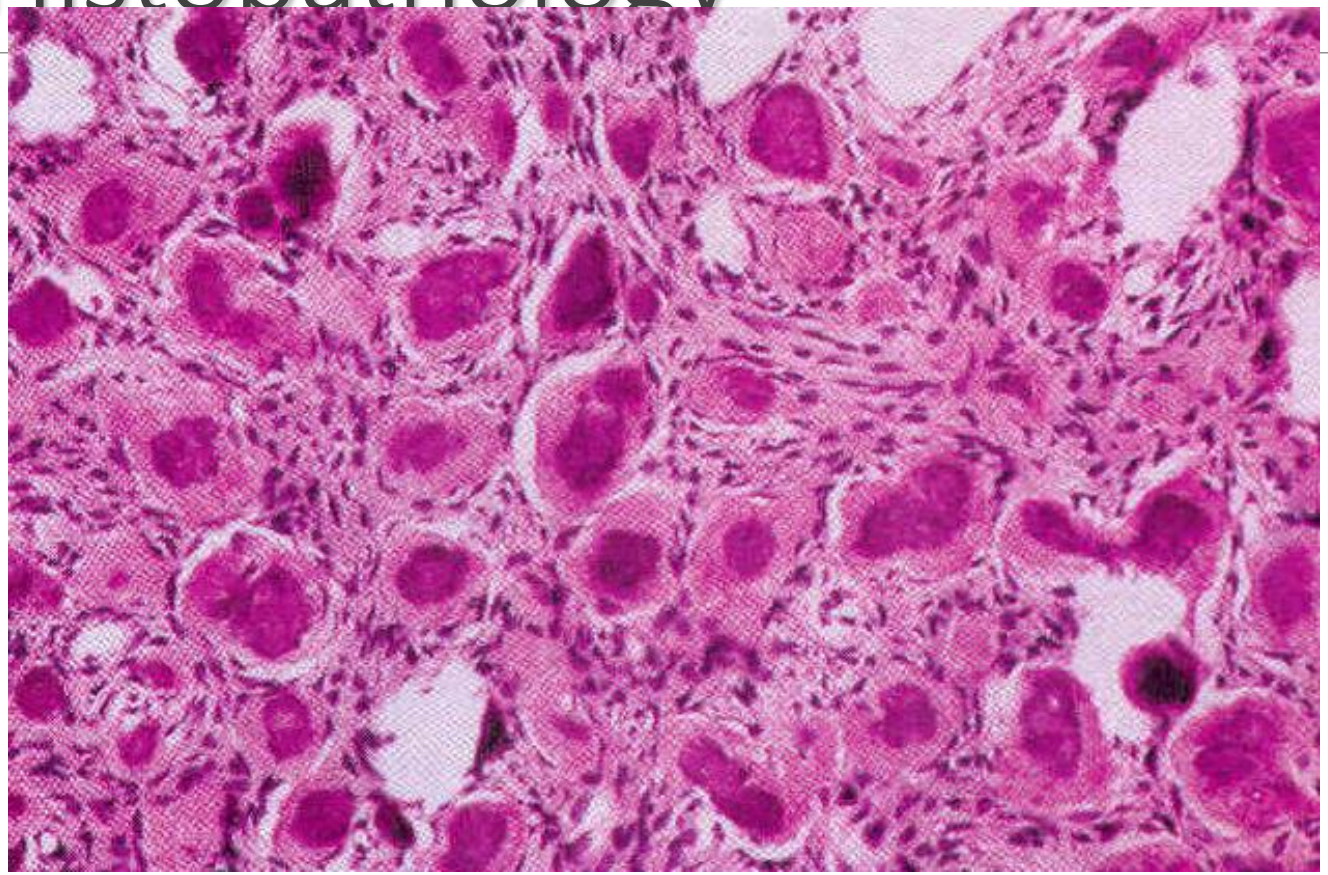


Histopathology



Trabeculae of cellular woven bone present in a cellular fibrous stroma

Histopathology



Cellular fibrous connective tissue containing spherical ossicles with basophilic centers and peripheral Eosinophilic rims

Distinction from OF

NON ENCAPSULATED

THE TUMOR IS MORE CELLULAR THAN *OF*

AREAS OF HEMORRHAGE; MULTINUCLEATED GIANT CELLS

Treatment & prognosis

Clinical management is uncertain,

Some show rapid growth & are aggressive mostly seen in infants and young children

30-58% recur

No malignant transformation rate

Familial Gigantiform Cementoma

Disorder of gnathic bone that leads to formation of massive sclerotic masses of disorganized mineral material

Autosomal dominant disorder

Caucasians and African blacks

Equal in males and females

Limited to jaws

Facial asymmetry,impaction,malposition and malocclusion of involved dentition

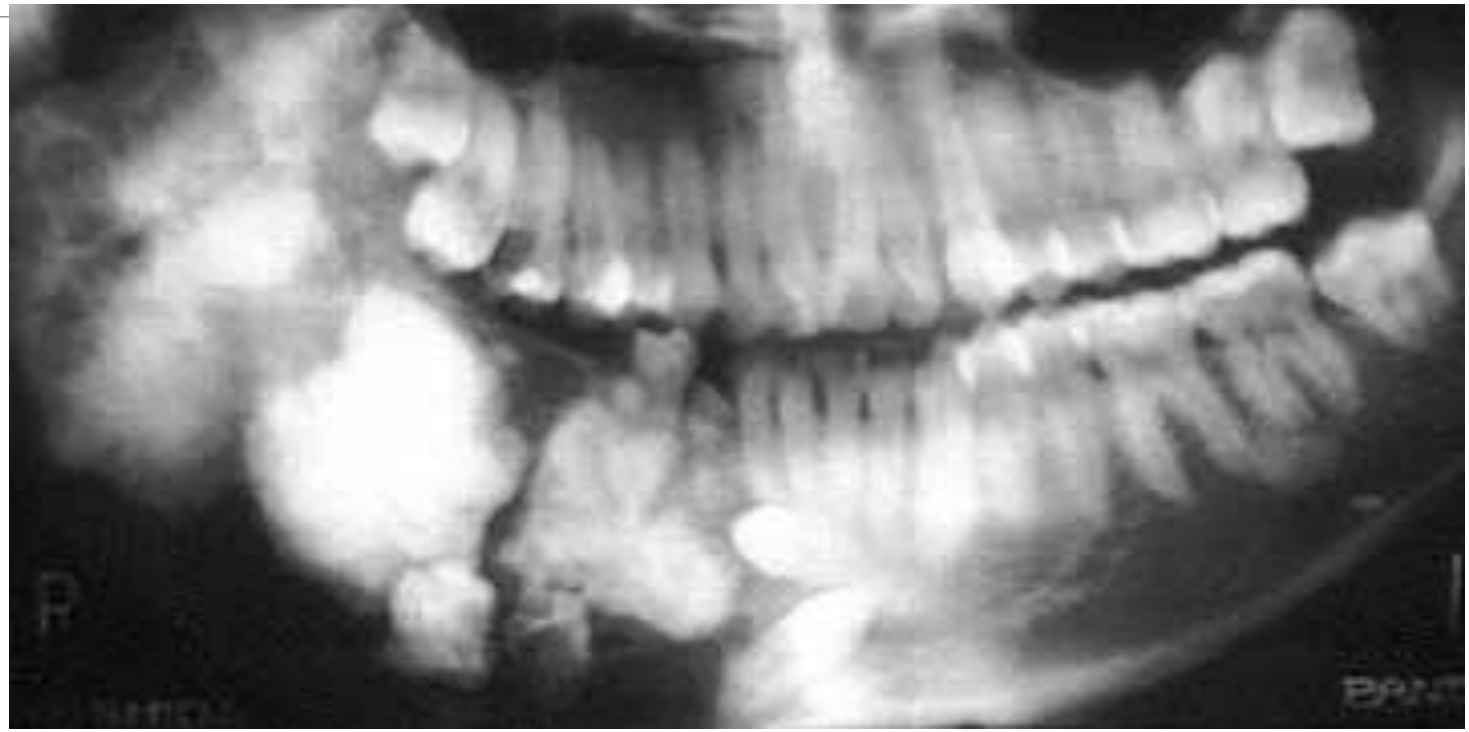
Familial Gigantiform Cementoma

Radiographic alterations seen during first decade of life

Clinically obvious swelling by adolescence, followed by a rapid thickening & expansive growth pattern

If not treated enlargement usually ceases during *fifth decade* of life

Familial Gigantiform Cementoma



Radiopaque with radiolucent thin rim

Familial Gigantiform Cementoma

Elevated serum alkaline phosphatase levels

Anemia in females

Histopathology is similar to florid cemento osseous dysplasia

Treatment & prognosis

Dysplastic bone rapidly regrows if removed before sclerotic stage

Once the lesion is radio opaque, partial removal causes sequestration

Paget's disease

Abnormal and anarchic resorption and deposition of bone , resulting in distortion and weakening of affected bone

Discovered by Sir James Paget

in 1876

Etiology

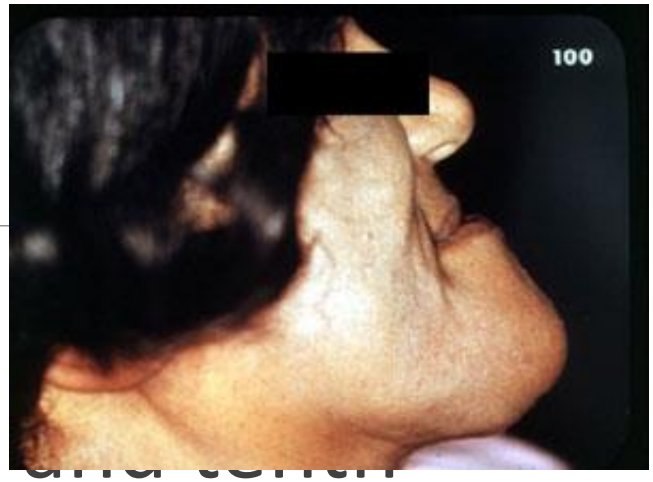
Slow virus infection,e.g. nucleocapsids from a ***paramyxovirus*** has been detected in osteoclasts in patients with pagets disease

Circulatory disturbance

Protracted clinical course,absence of acute ***inflammatory process*** and geographic and familial clustering favors a **viral** cause

15-30% positive ***family*** history

Clinical features



Men

incidence over 40 years-3% and tenth decade is 10%

Monostotic and polyostotic

LINCOLN' S SIGN/BLACK BEARD SIGN

Clinical features

Bone pain- may be quite severe, is a common complaint

Lumbar vertebrae> pelvis> skull> femur

Bones become thickened, enlarged, bowing deformity (monkey like stance)

Skull- increase in circumference

Clinical features

Jaw-17%

Maxilla>mandible

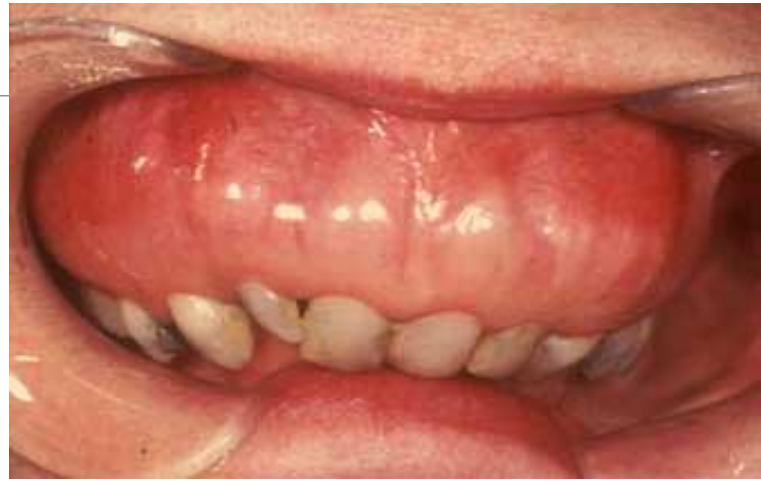
Enlargement of middle

third of face

Lion like facies

Alveolar ridge

symmetric enlargement



Tight fitting Dentures
Spacing of Teeth

Radiographic features

Early stage reveals decreased Radiopacity of bone and alteration of trabeculae

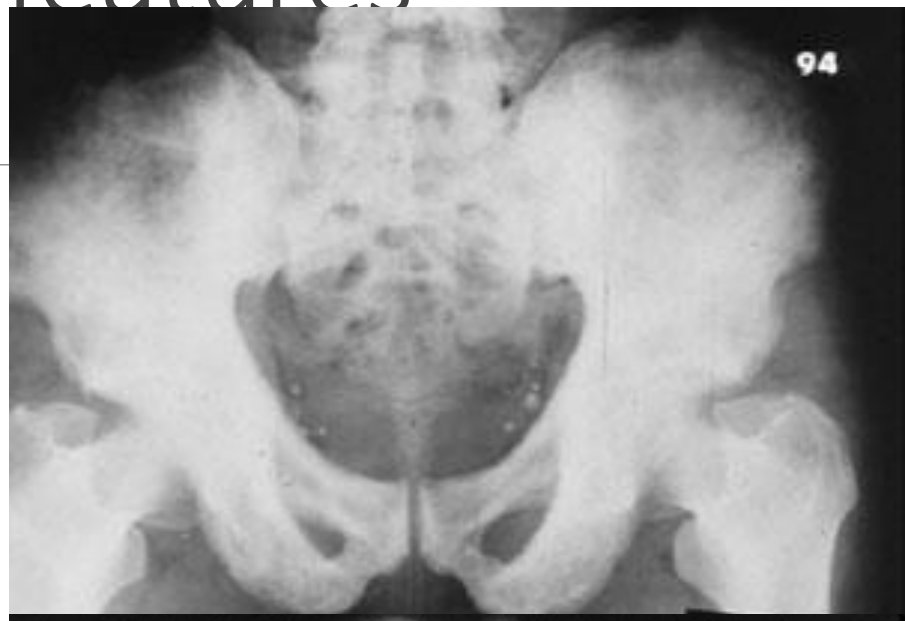
Loss of Trabeculae produces a GROUND GLASS appearance

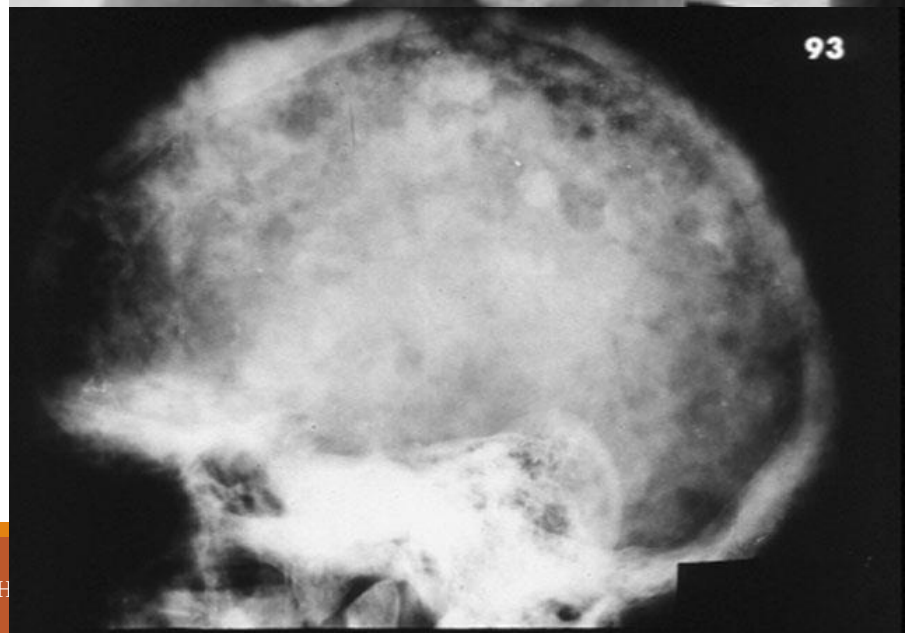
Osteoporosis circumscripta

Patchy areas of sclerotic bone are formed

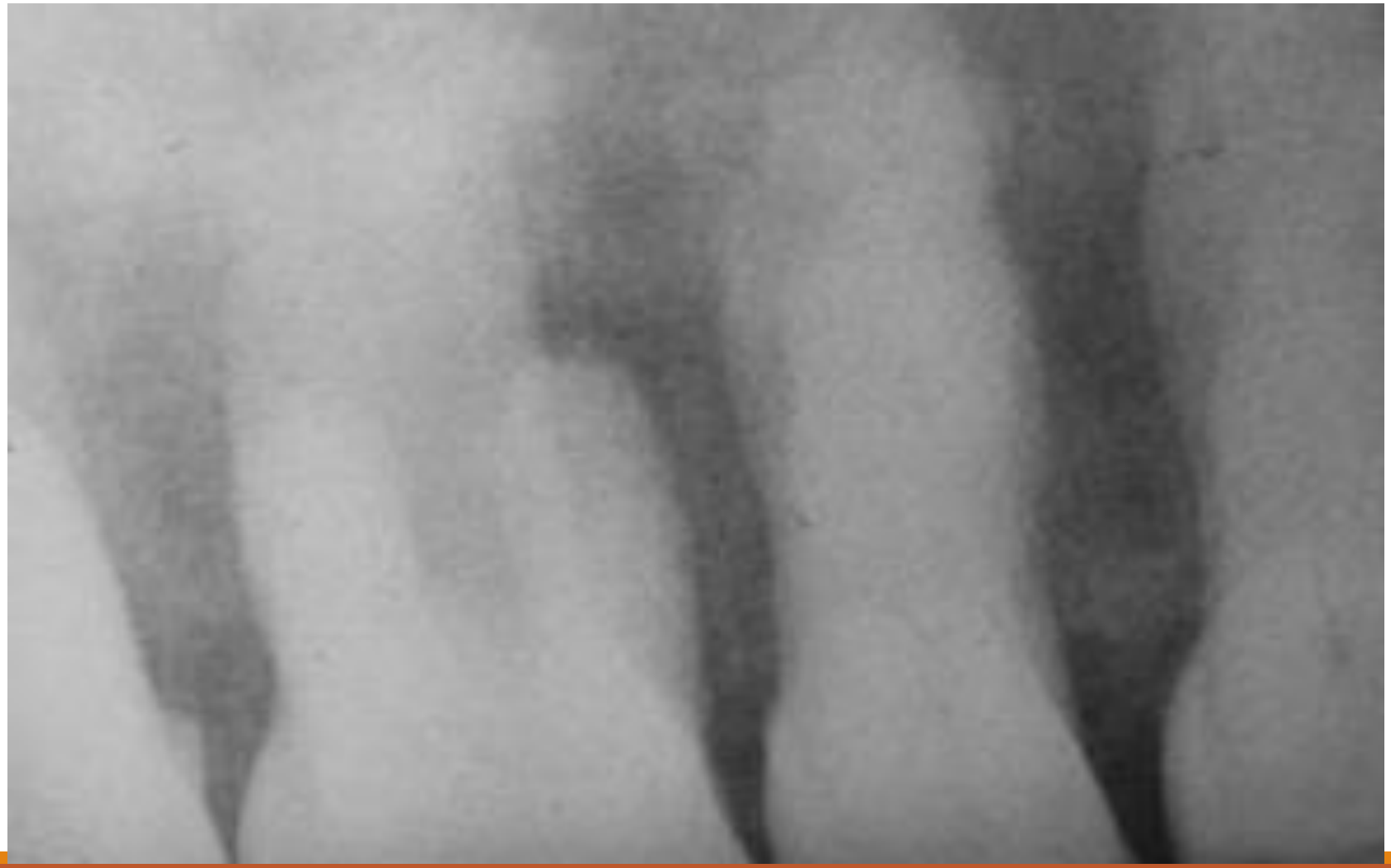
COTTON-WOOL appearance

Radiographic features





Radiographic features



Biochemical findings

Increased serum Alkaline phosphatase

Increased urine Hydroxyproline

Normal serum calcium and phosphorous

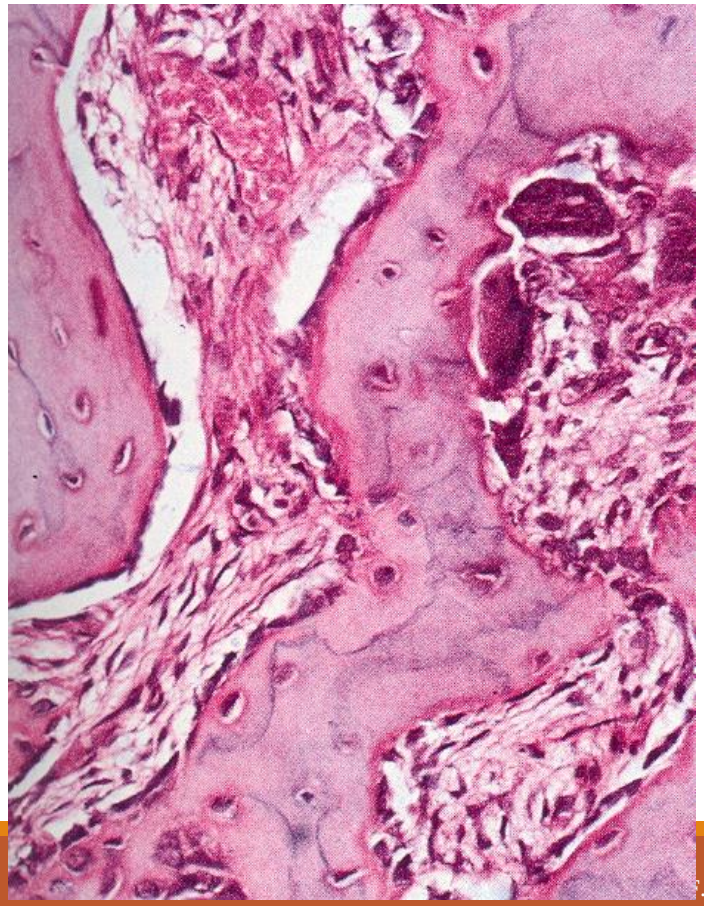
Complications

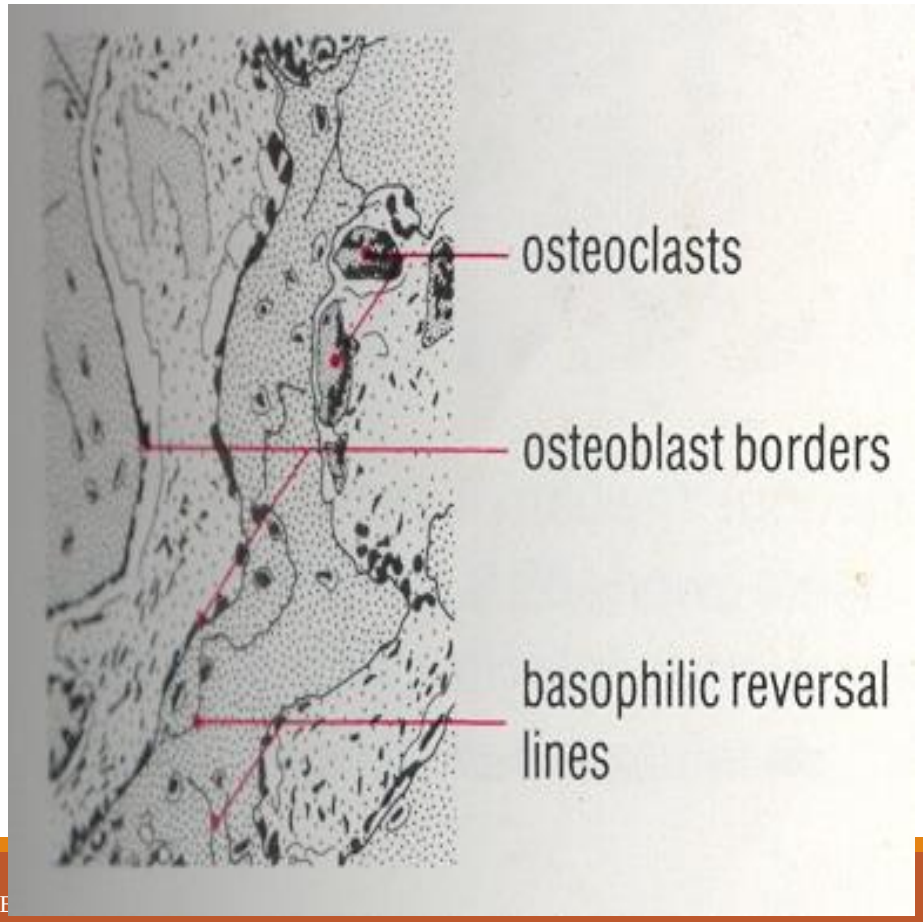
Pathologic fracture

- Osteosarcoma
- Osteomyelitis
- Bleeding
- Deafness
- Blindness

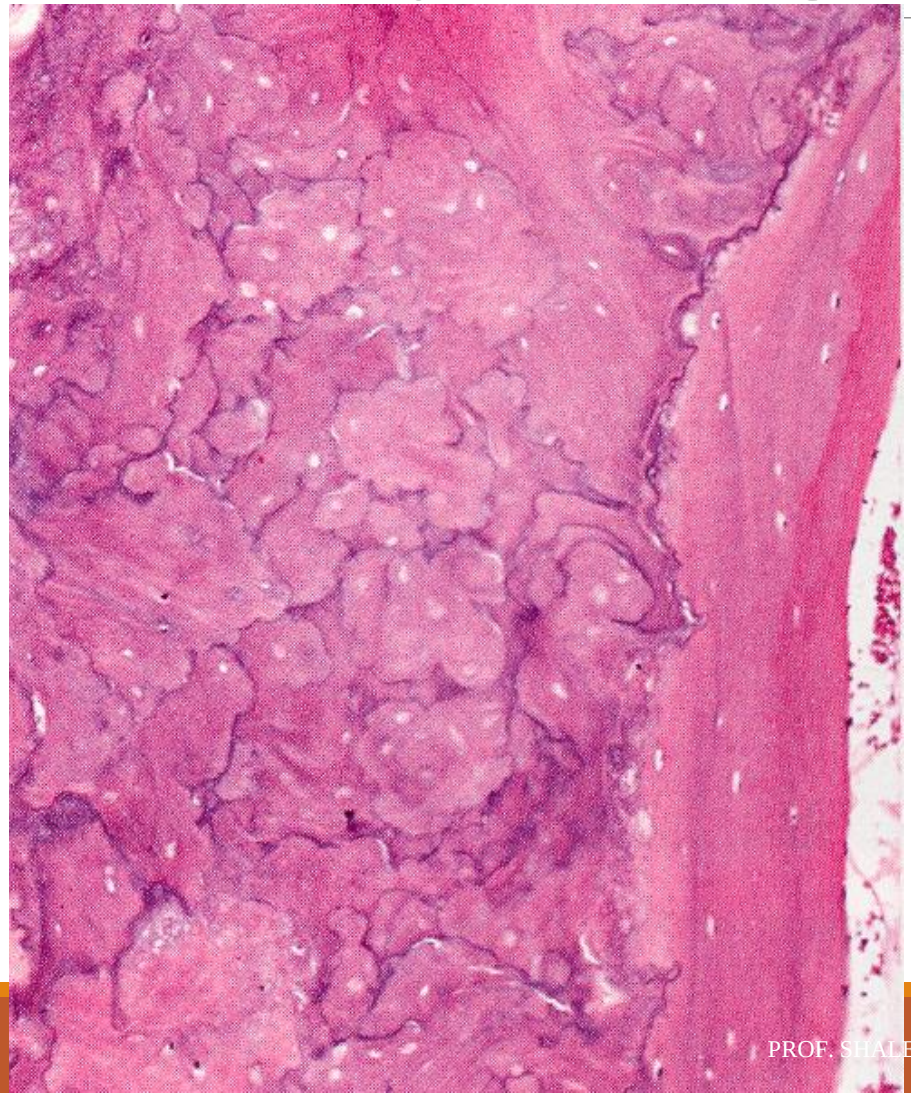
Histopathologic findings

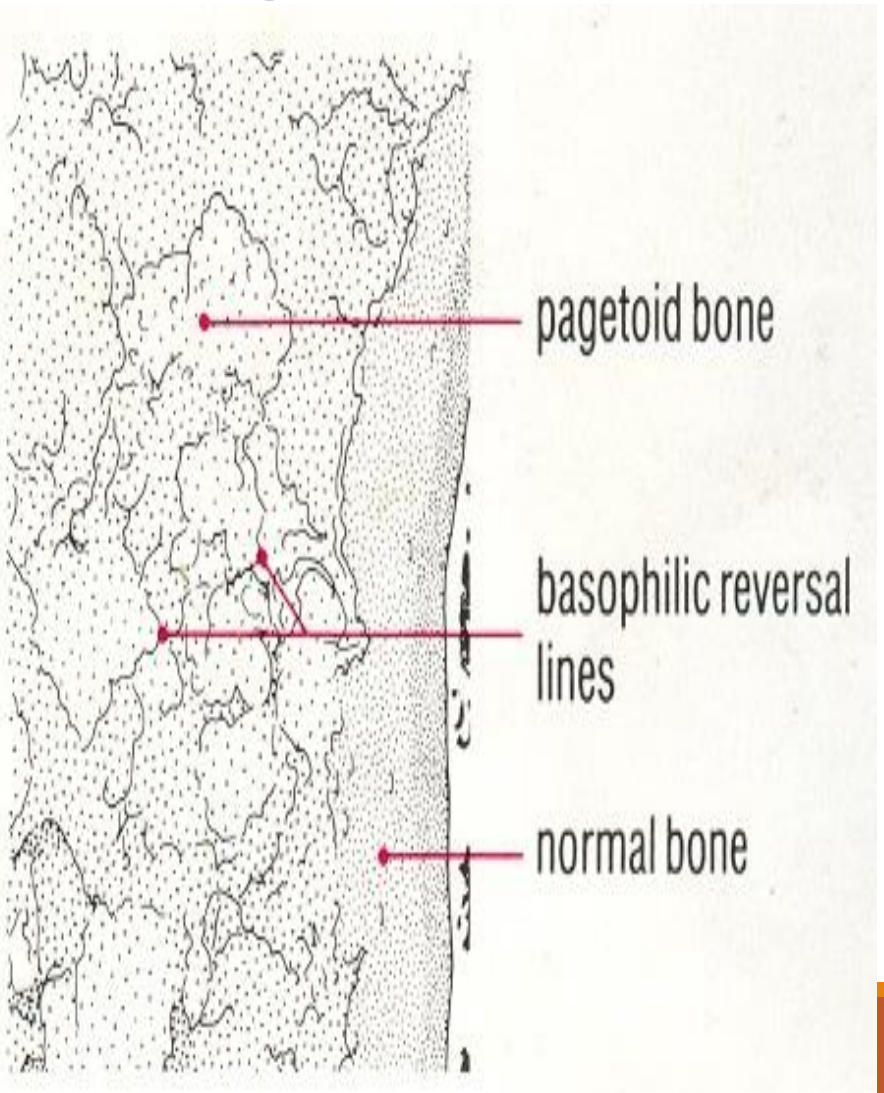
Jigsaw /mosaic appearance





Histopathologic findings





COMPLICATION

OSTEOSARCOMA

TREATMENT

Bone pain is controlled by Aspirin

use of Bisphosponates

Plicamycin given only in severe cases

Antibiotics prior to extraction

Dentures to be continually remade

CHERUBISM

Familial fibrous dysplasia of jaws; disseminated juvenile fibrous dysplasia

CHERUBISM

Clinical features

Autosomal dominant

uncommon disease

2-5 years

clinical alteration typically progresses until puberty, then stabilizes and slowly regresses

CHERUBISM

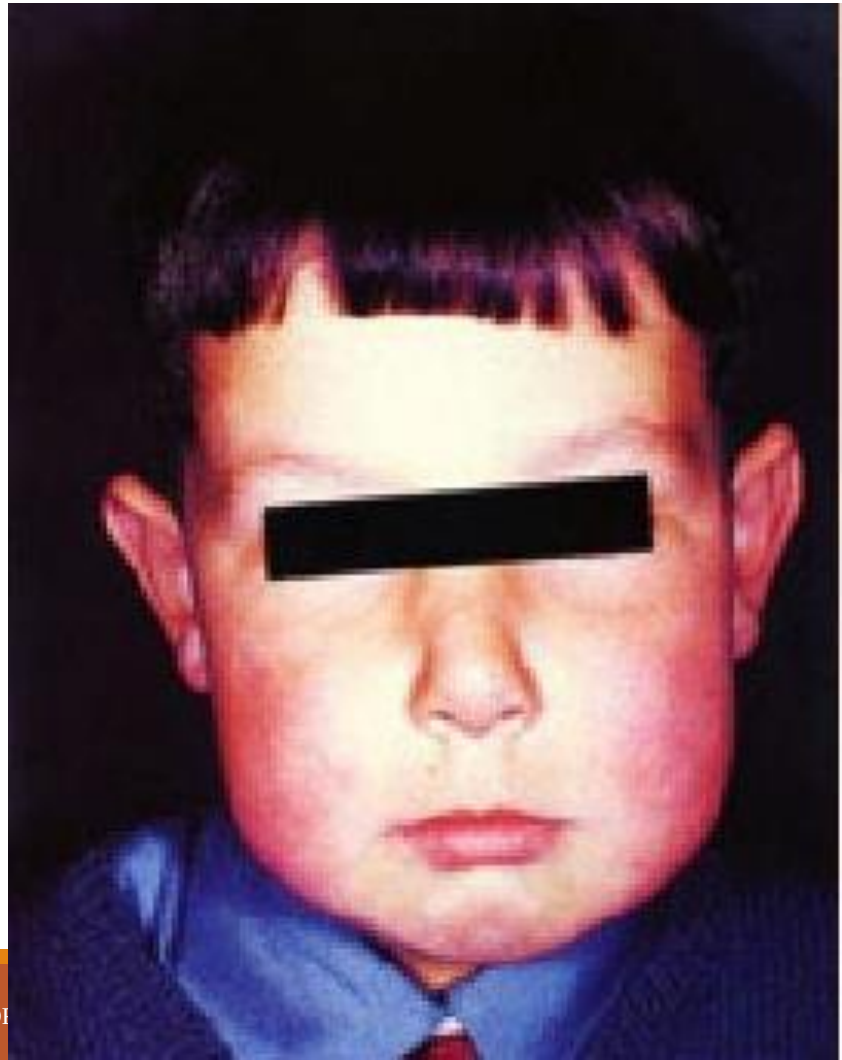
Bilateral involvement of posterior mandible

“ Eyes upturned to heaven’’ appearance

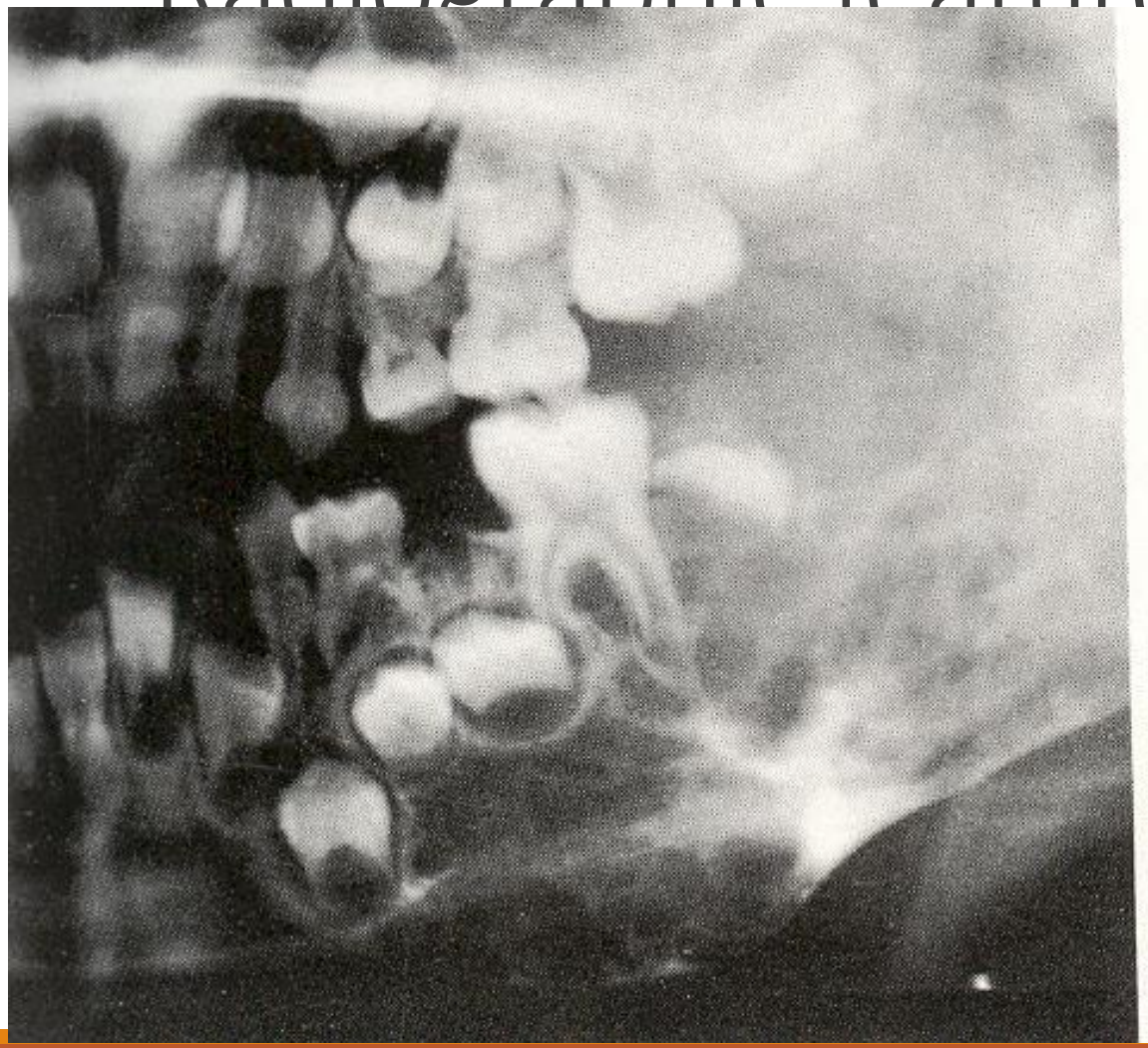
Cervical lymphadenopathy

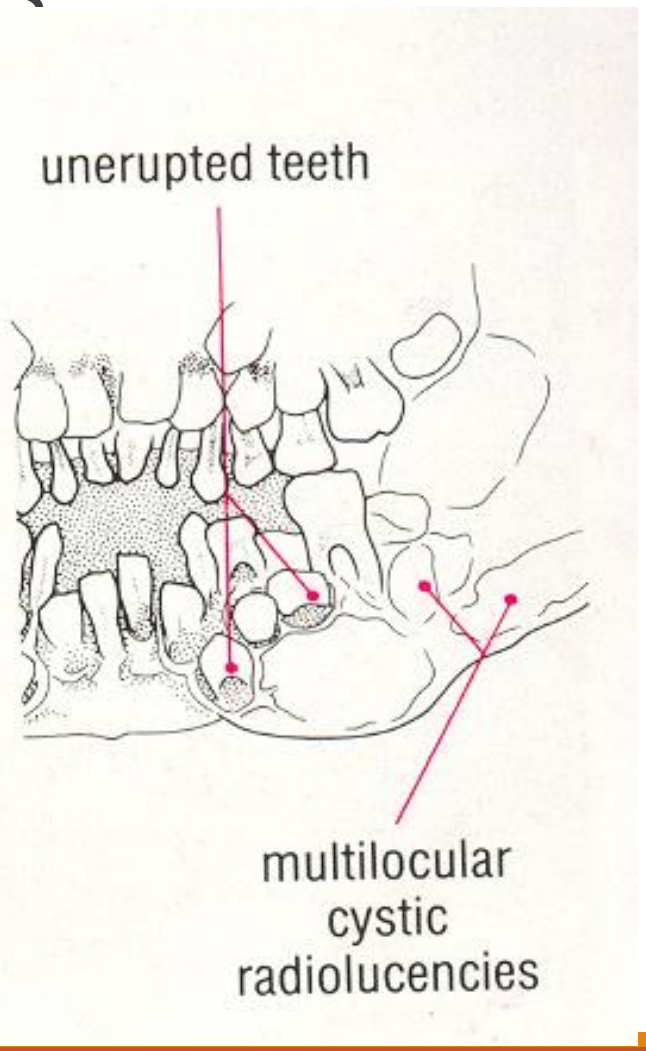
Painless and bilateral expansion

CHERUBISM



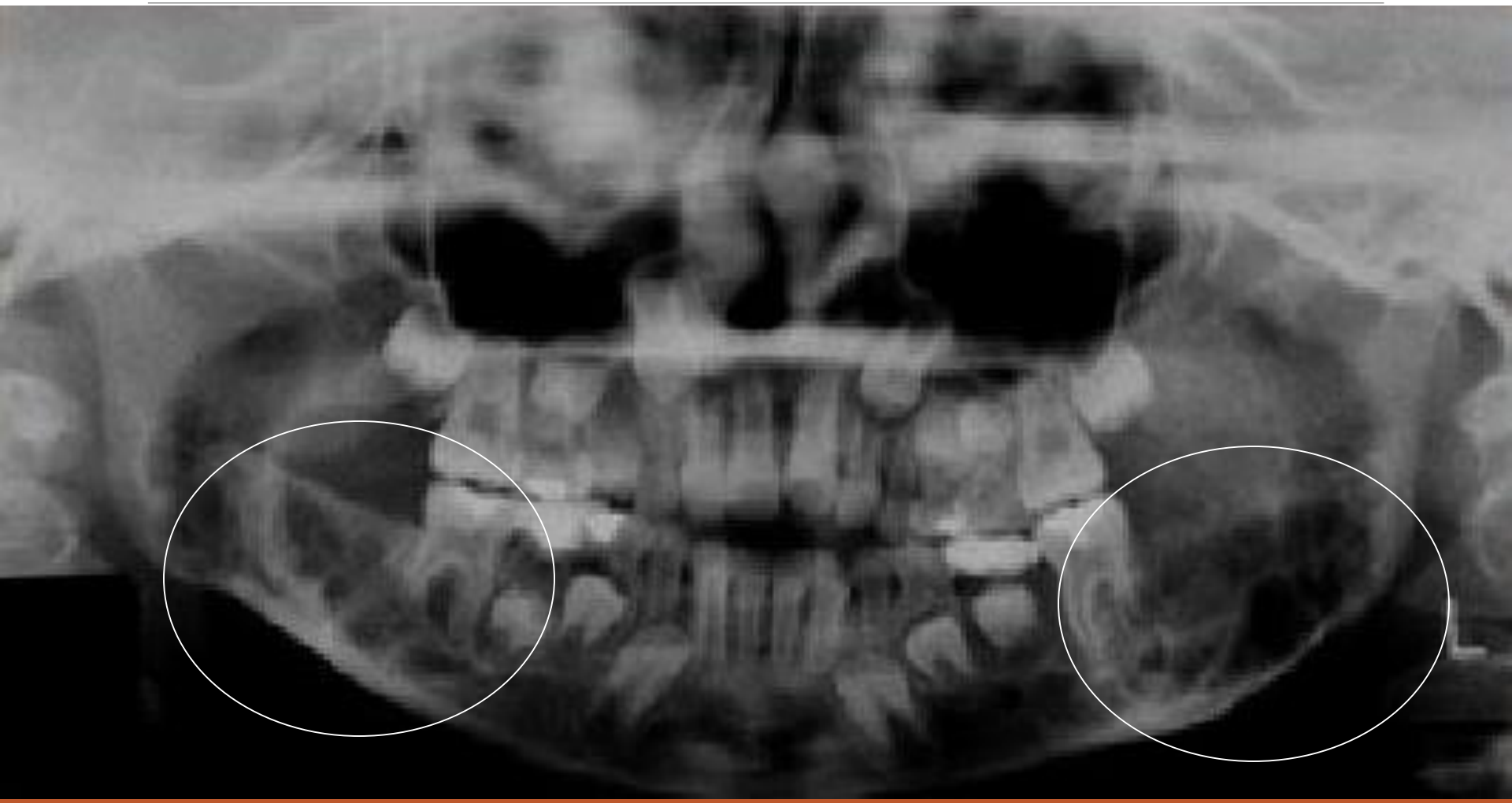
Radiographic features





Multiple, expansile radiolucency

Radiographic features



Oral manifestations

Agenesis of second and third molars

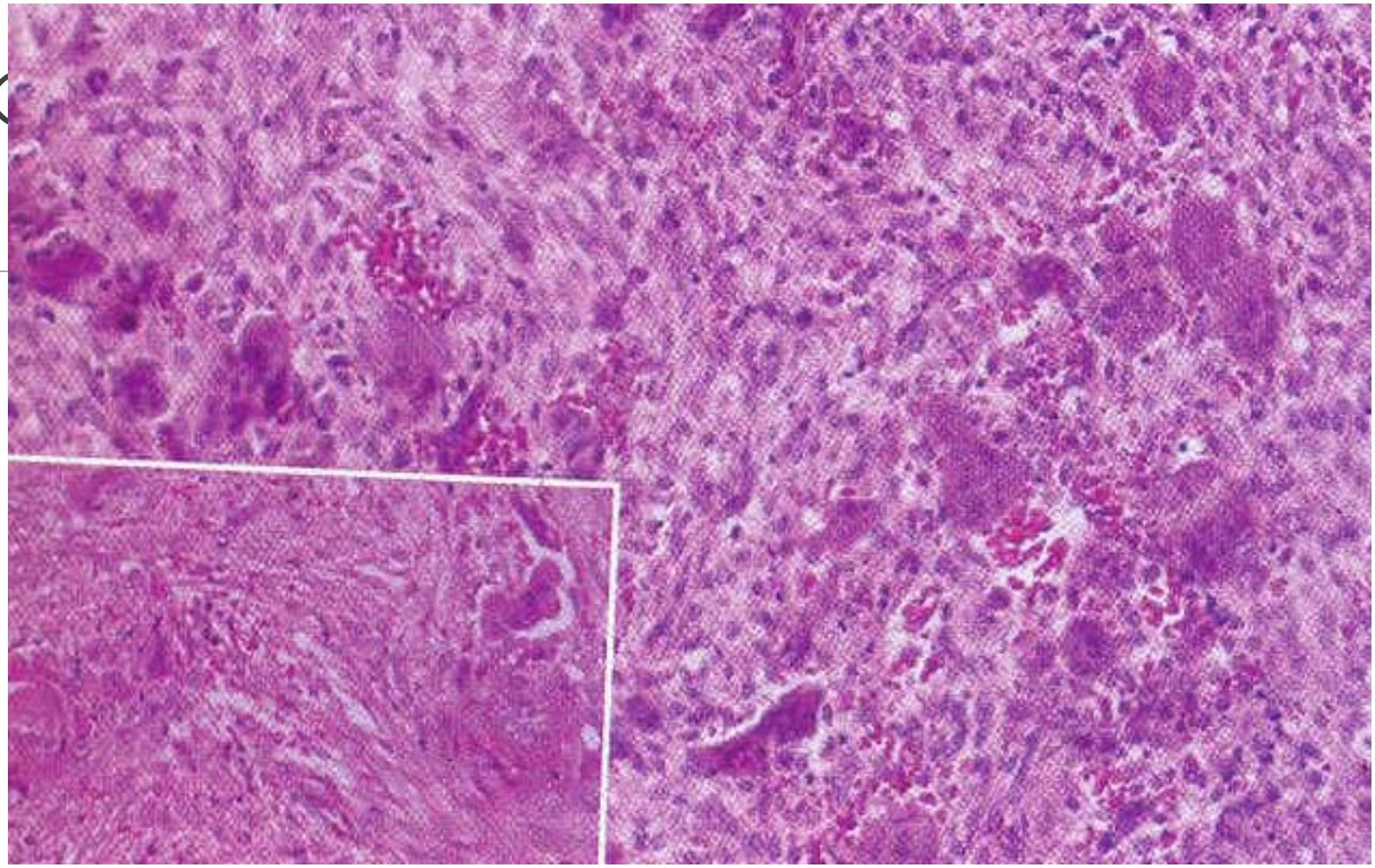
Displacement of teeth

Premature exfoliation of primary teeth

Delayed eruption of permanent teeth

Transposition and rotation of teeth

Histop



Scattered giant cells within background of cellular, hemorrhagic mesenchymal tissue.

The inset demonstrates perivascular Eosinophilic cuffing

Treatment & prognosis

No treatment required

Radiotherapy- contra indicated

Osteoradionecrosis