LAB 7
FIBRO OSSEOUS LESIONS

Fibrous dysplasia :-

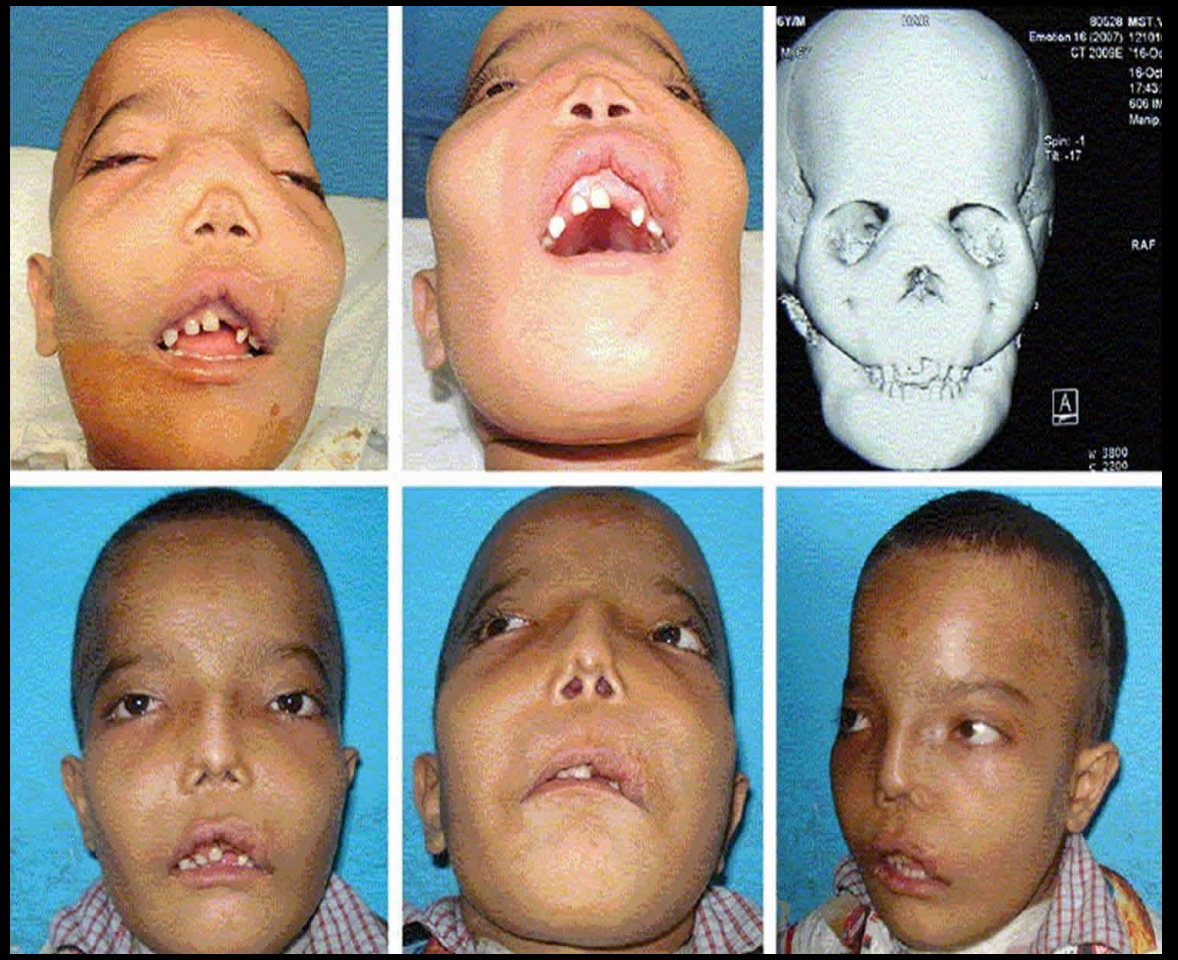
Fibrous dysplasia is a condition in which normal medullary bone is replaced by an abnormal fibrous connective tissue proliferation in which new, non-maturing bone is formed.

Clinical Features.

1- Most commonly presents as an asymptomatic, slow enlargement of the involved bone.

2- It may involve a single bone or several bones concomitantly.

3- *Monostotic fibrous dysplasia* is the designation used to describe the process in one bone.



Monostotic fibrous dysplasia more common than the polyostotic form

accounting for as many as 80% of cases.

Jaw involvement is common. Other bones that are commonly affected are the ribs and femur

more often in the maxilla than in the mandible.

Maxillary lesions may extend to involve the maxillary sinus, zygoma, sphenoid bone, and floor of the orbit. This form of the disease, with involvement of several adjacent bones, has been referred to as ***craniofacial fibrous dysplasia.***

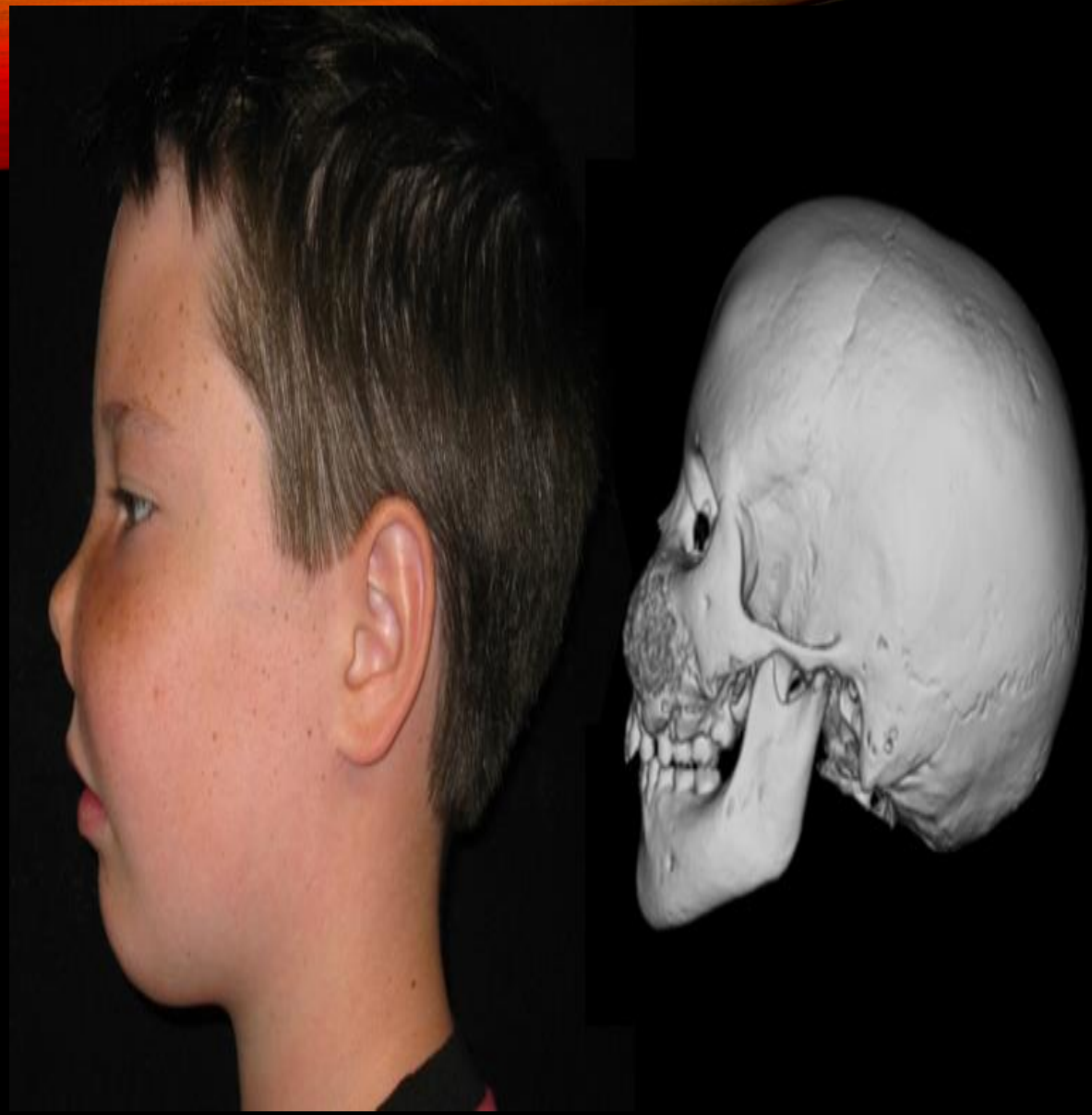
The most common site of occurrence with mandibular involvement is in the body portion.

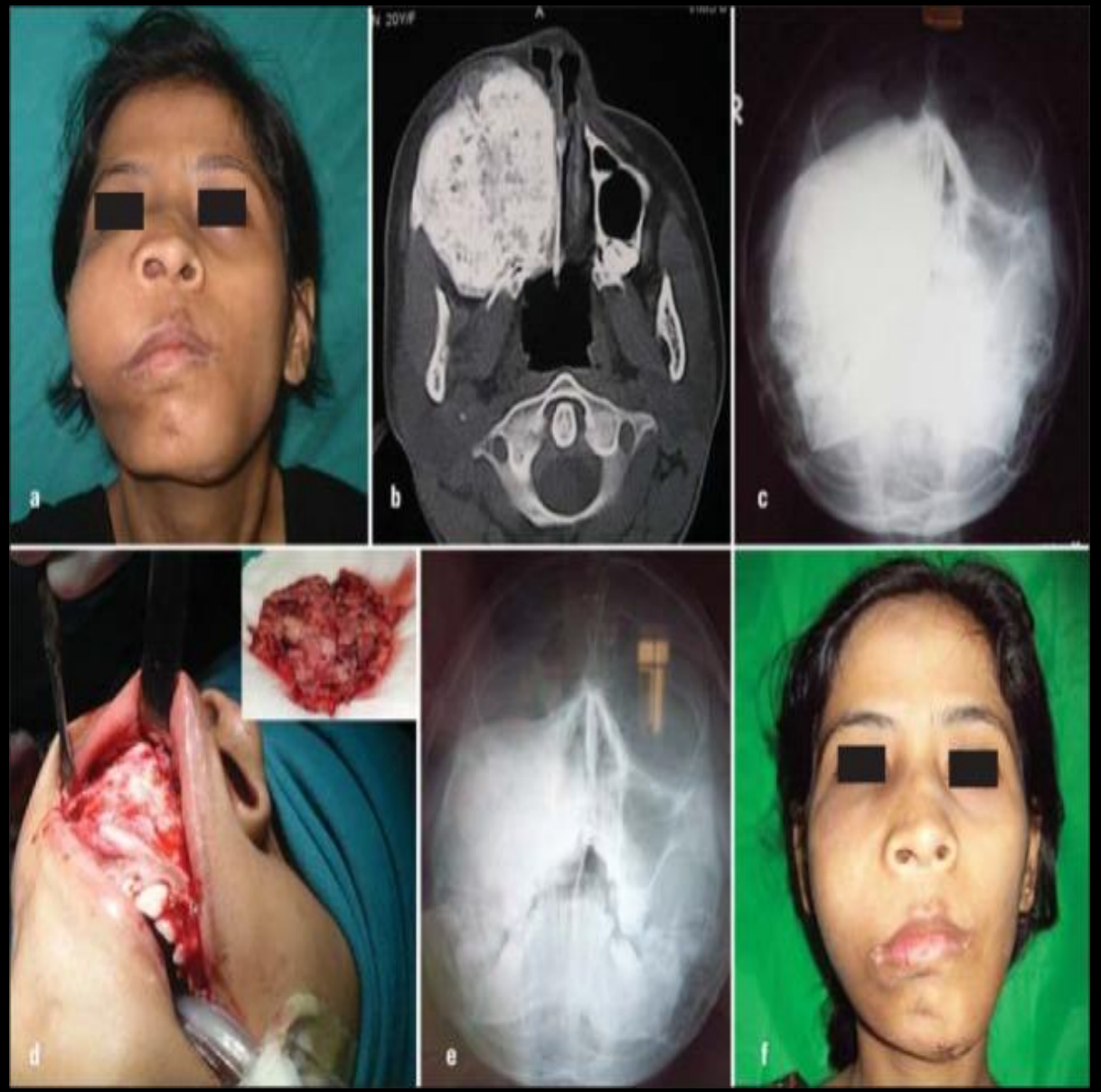
The slow, progressive enlargement of the affected jaw is usually painless and typically presents as a unilateral swelling. As the lesion grows, facial asymmetry becomes evident and may be the initial presenting complaint. The dental arch is generally maintained, although displacement of teeth, malocclusion, and interference with tooth eruption may occasionally occur. Tooth mobility is not seen.

8- Characteristically has its onset during the first or second decade of life.

9- Self-limiting and tend to stabilized during puberty .

10- ***Polyostotic fibrous dysplasia*** applies to cases in which more than one bone is involved.




a
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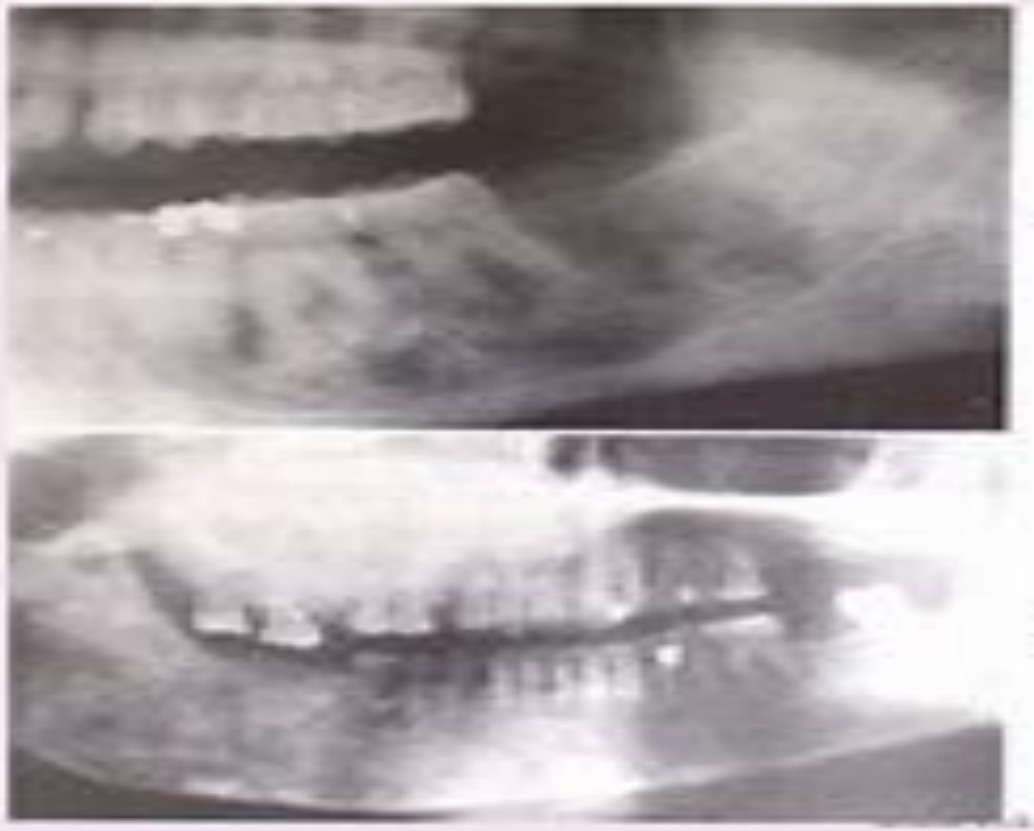
Radiographically :-

1- The classic lesion has been described as "ground glass" or "peaud' orange" effect. This characteristic image, which is most identifiable on intraoral radiographs, is not, however, pathognomonic.

2- Unilocular or multilocular radiolucencies, especially in long bones.

3- Additional radiographic features that have been described include a fingerprint bone pattern .

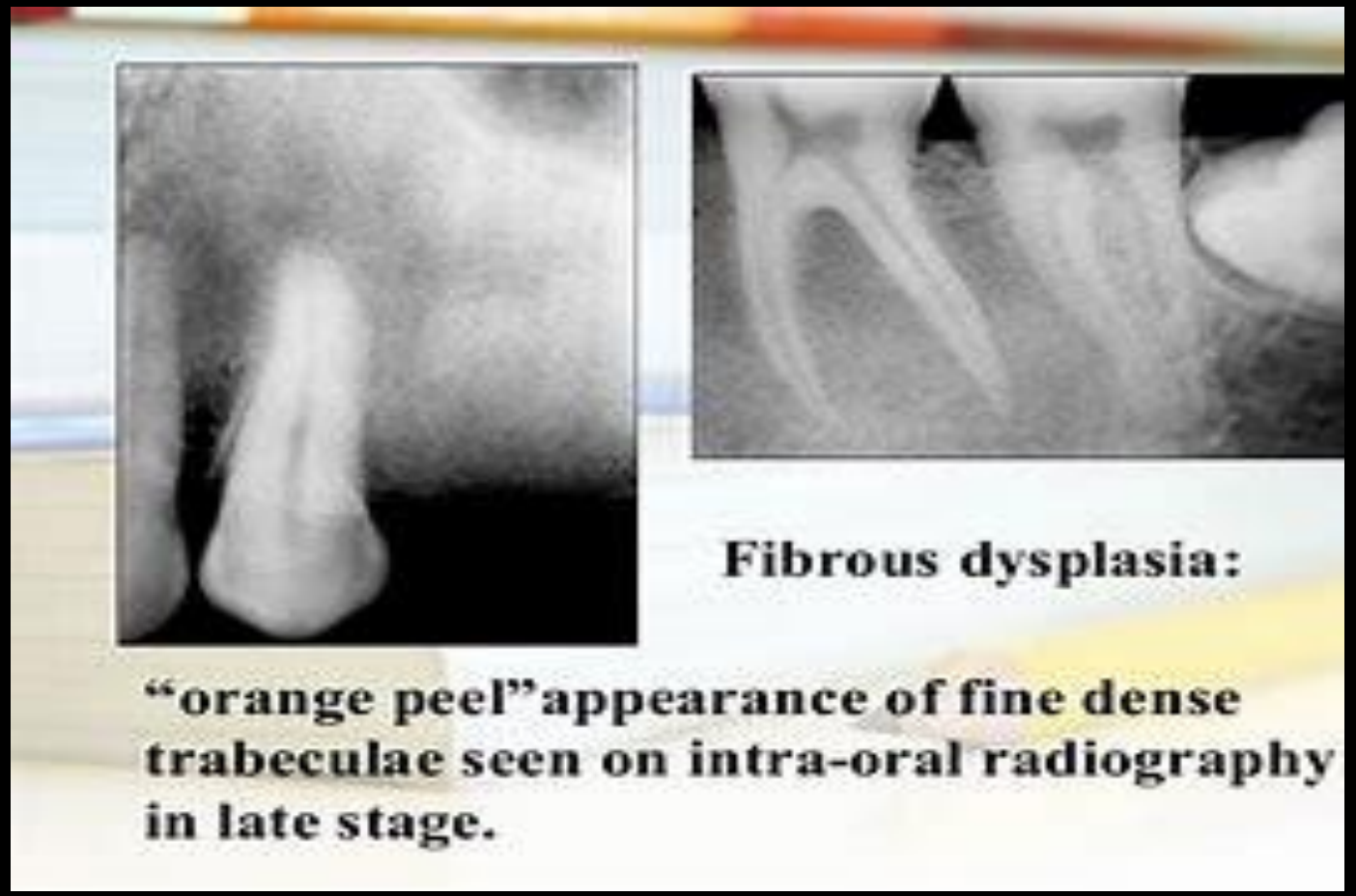
4- A uniformly radiopaque mass have been seen in long standing disease .

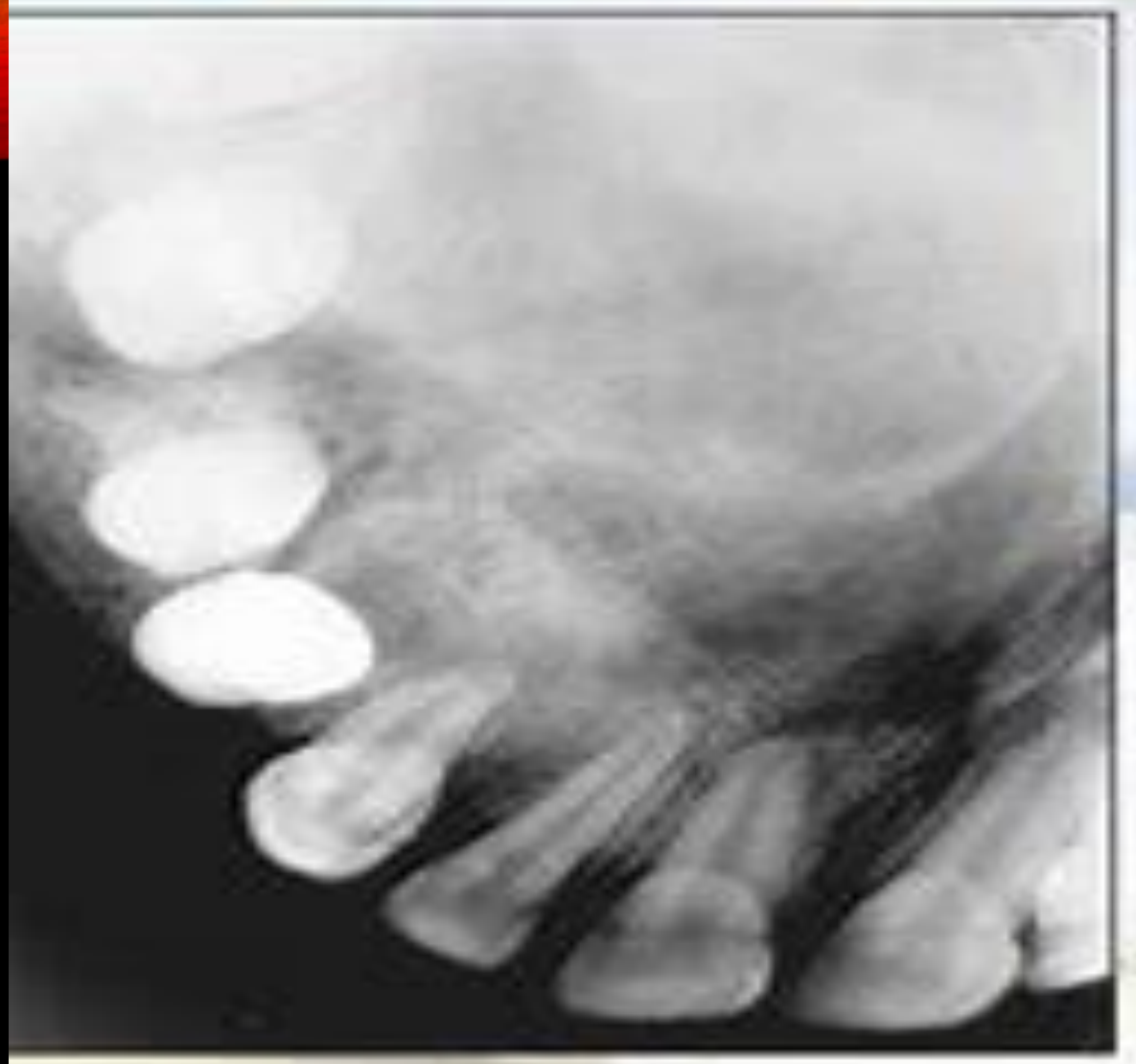


- Fibrous dysplasia (intermediate stage)
  A) ***Smoky or mottled*** appearance(also seen in B).

- B)Radiograph showing one lesion in maxilla & one in mandible.

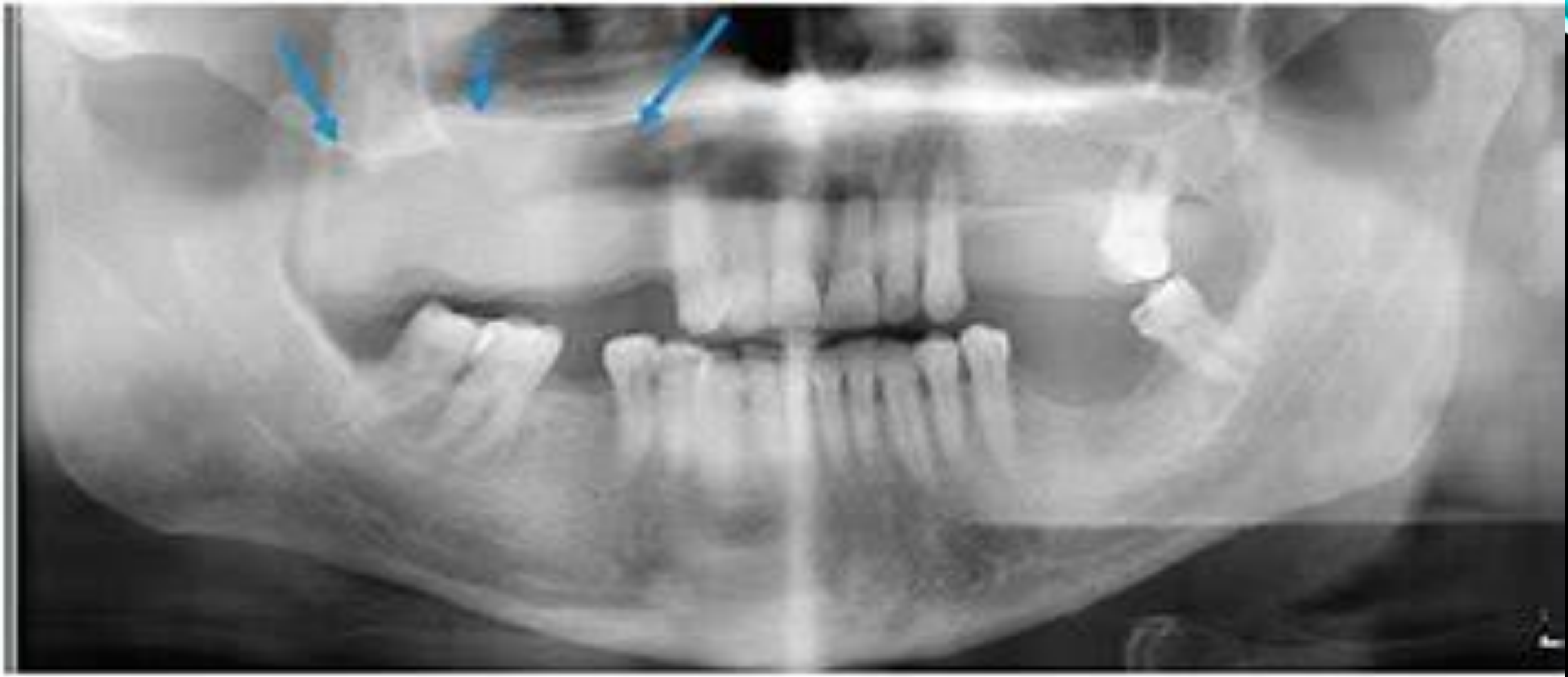
An important distinguishing feature of fibrous dysplasia is the poorly defined radiographic and clinical margins of the lesion. The process appears to blend into the surrounding normal bone without evidence of a circumscribed border.





Fibrous dysplasia:

note enlargement of affected area, buccal expansion of the maxilla and "orange peel" opacity (topographic occlusal view).

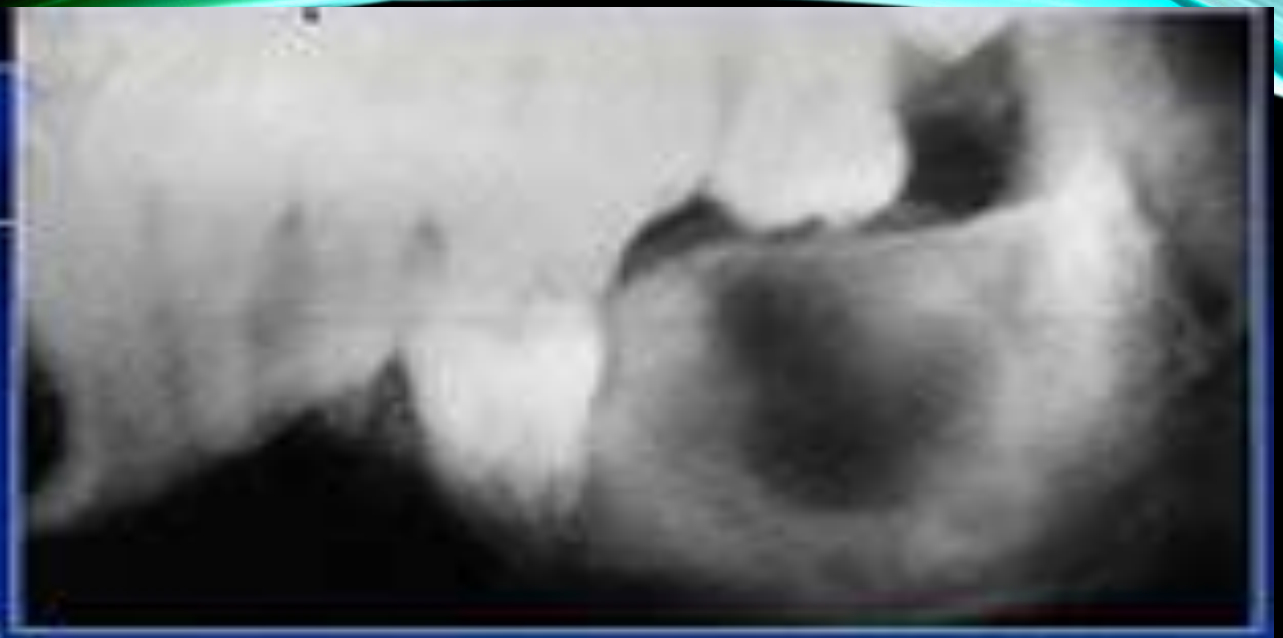


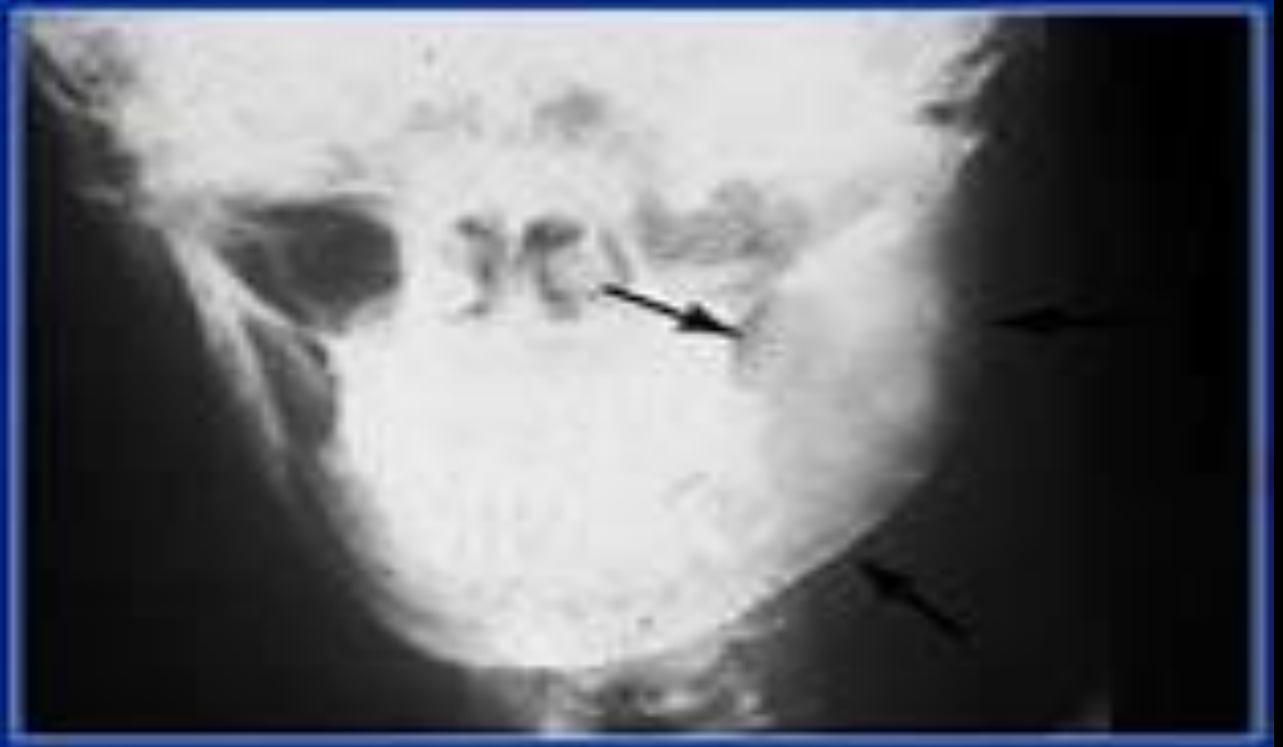
igure 2. Panoramic radiograph, diffuse granular appearar
nown as a "ground glass" appearance and expansion

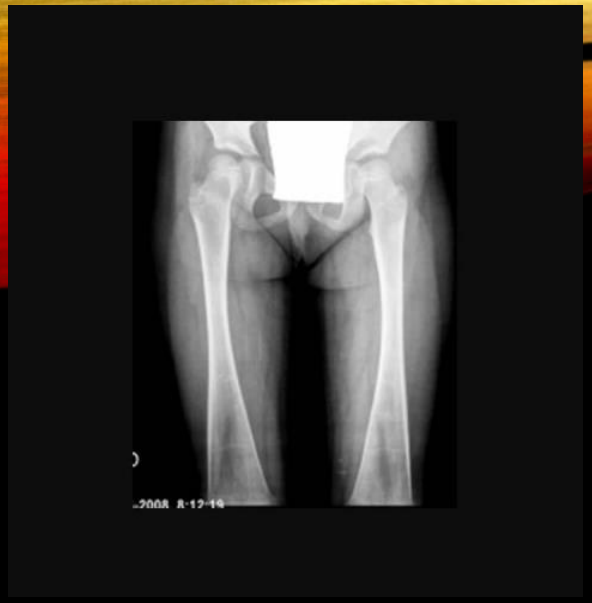
Fibrous Dysplasia
Clinical Variants

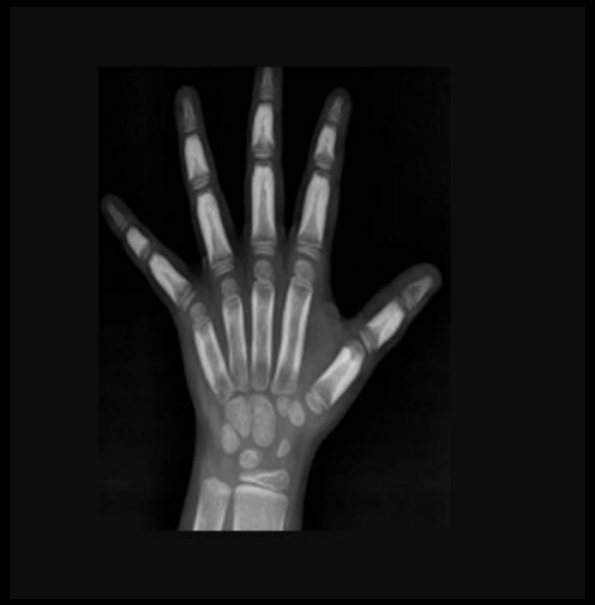
- Monostotic fibrous dysplasia
  - One bone involved
  - 85% of cases
  - Jaws: among most common sites

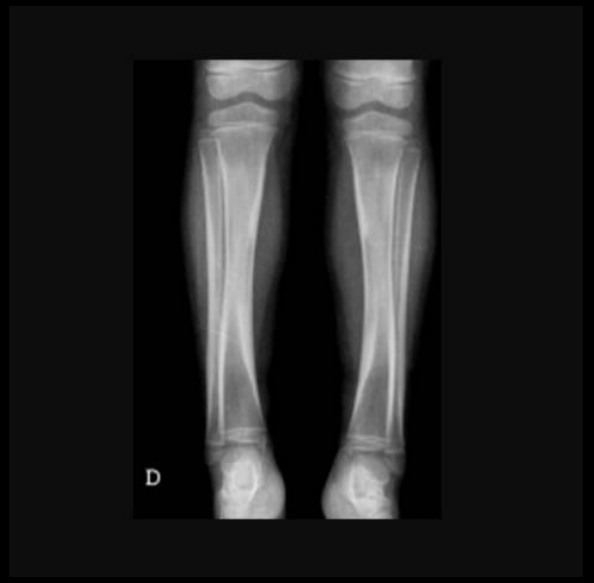
- Polyostotic fibrous dysplasia
  - Multiple bones involved
  - Jaffe-Lichtenstein syndrome
  - McCune-Albright syndrome

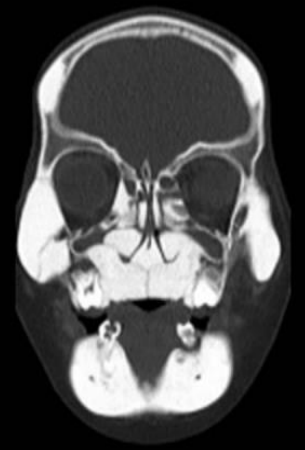












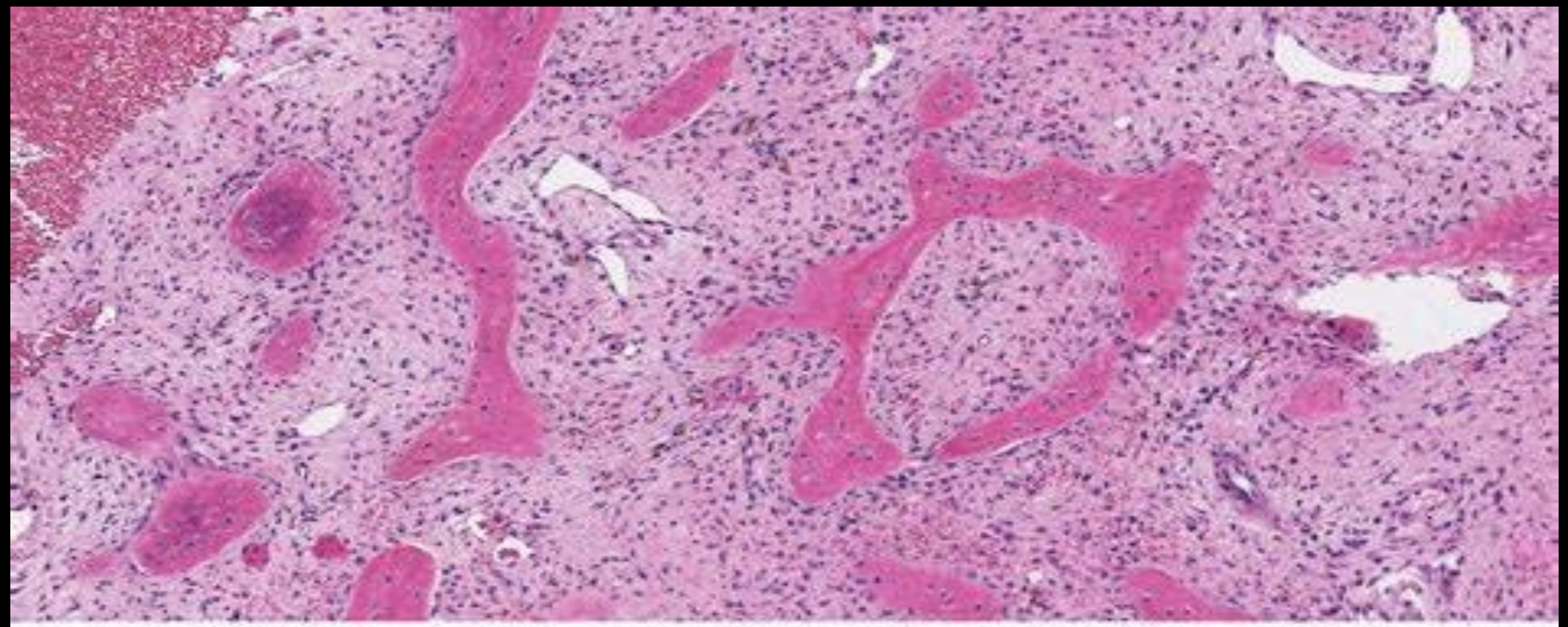
Polyostotic fibrous dysplasia with facial bones involvement

OIP.jpg

Histopathology:-

1- A slight to moderate cellular fibrous connective tissue stroma that contains foci of irregularly shaped trabeculae of immature woven bone.

2- The bony trabeculae assume irregular shapes likened to *Chinese characters*. Capillaries typically are prominent and uniformly distributed.



High power view of fibrous dysplasia. Notice the lack of osteoblastic rimming in the bone trabeculae. The spindle cells present in the stroma are cytologically bland.

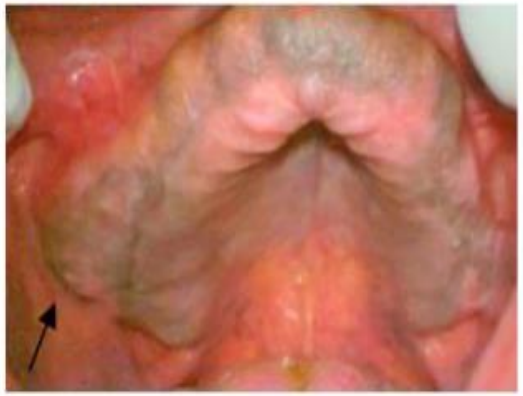


Figure 2. A single well-defined swelling evident on edentulous area on buccal aspect in relation to 15 to 17 region

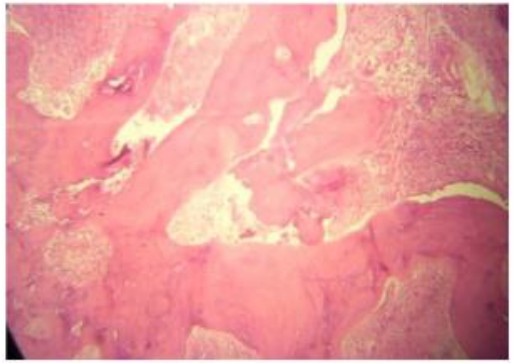


Figure 5. Histopathologic section revealed a fibrous connective tissue containing many slender irregular trabecular pattern of woven bone

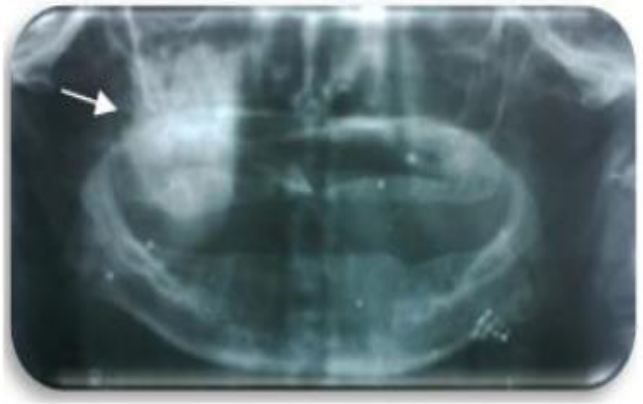


Figure 3. OPG revealed a mixed radiopaque-radiolucent area in right maxillary quadrant

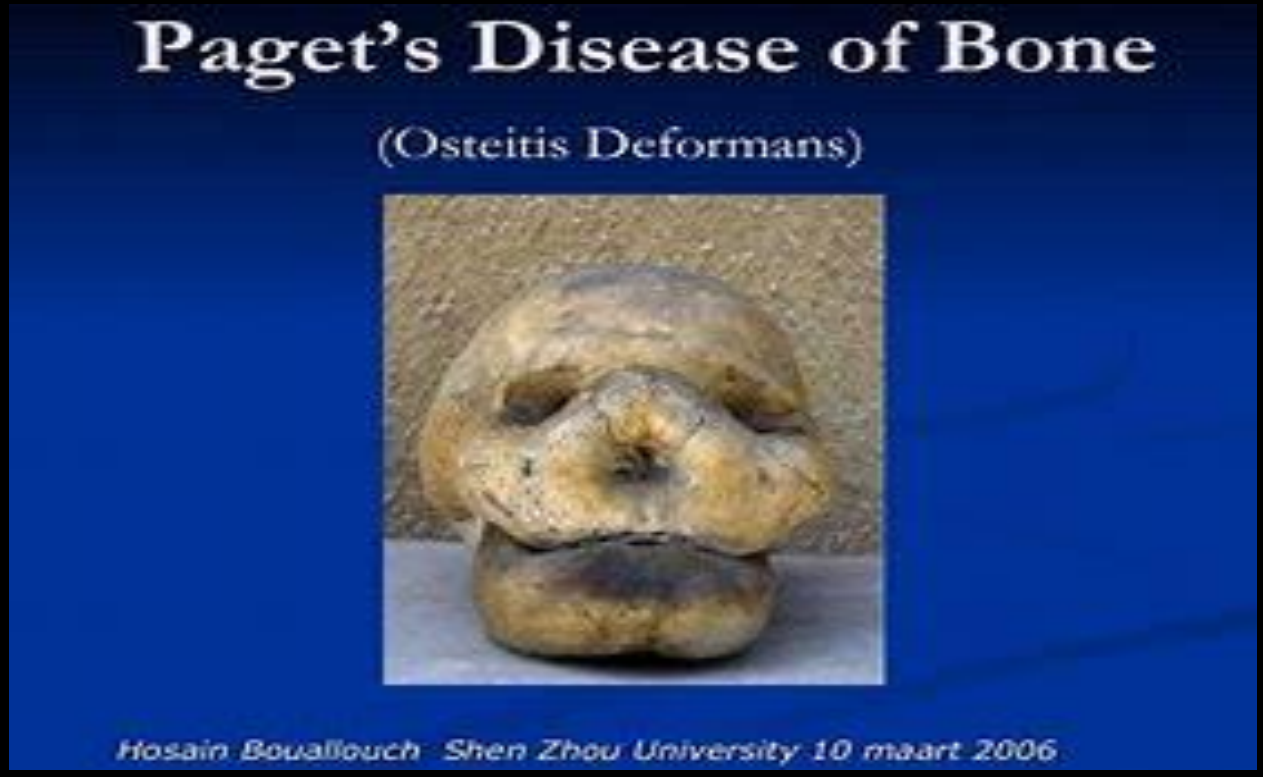
Treatment and Prognosis:-

- *After a variable period of growth, fibrous dysplasia characteristically stabilizes, although a slow advance may be noted into adulthood.*
- *Small lesions may therefore require no treatment other than biopsy confirmation and periodic follow-up.*
- *Large lesions that have caused cosmetic or functional deformity may be treated by surgical recontouring. This procedure is generally deferred until after stabilization of the disease process.*
- *Malignant transformation is a rare complication of fibrous dysplasia ,Many of the patients reported on were treated with radiation therapy, suggesting a role for radiation in the transformation process, although malignant change has been documented in the absence of radiation treatment.*

Paget's disease of bone

Is a condition characterized by abnormal resorption and deposition of bone, resulting in distortion and weakening of the affected bones. The cause of Paget's disease is unknown, but inflammatory, genetic, and endocrine factors may be contributing agents.

osteitis deformans



Clinical and radiographic features

- Paget's disease is relatively common.

- The disease principally affects older adults and is rarely encountered in patients younger than 40 years of age.

- Men are affected more often than women, and whites are affected more frequently than blacks.

- Asymptomatic disease often is discovered in radiographs taken for unrelated reasons or from an unexpected elevation in serum alkaline phosphatase.

-Although the disease may be ***monostotic*** (i.e., limited to one bone), most cases of Paget's disease are **polyostotic** (i.e., more than one bone is affected). Some patients may remain relatively asymptomatic.

Clinically called ***Lion face*** also

Symptoms vary. Bone pain, which may be quite severe, is a common complaint.

- Pagetic bone often forms near joints and promotes osteoarthritic changes, with associated joint pain and limited mobility.

- The lumbar vertebrae, pelvis, skull, and femur are the most commonly affected bones.

- Affected bones become thickened, enlarged, and weakened. Involvement of weight-bearing bones often leads to a bowing deformity, resulting in what is described as a simian (monkeylike) stance.

- Paget's disease affecting the skull generally leads to a progressive increase in the circumference of the head.

- Jaw involvement is present in approximately 17% of patients diagnosed with Paget's disease.

- Maxillary disease, which is far more common than mandibular involvement, results in enlargement of the middle third of the face.

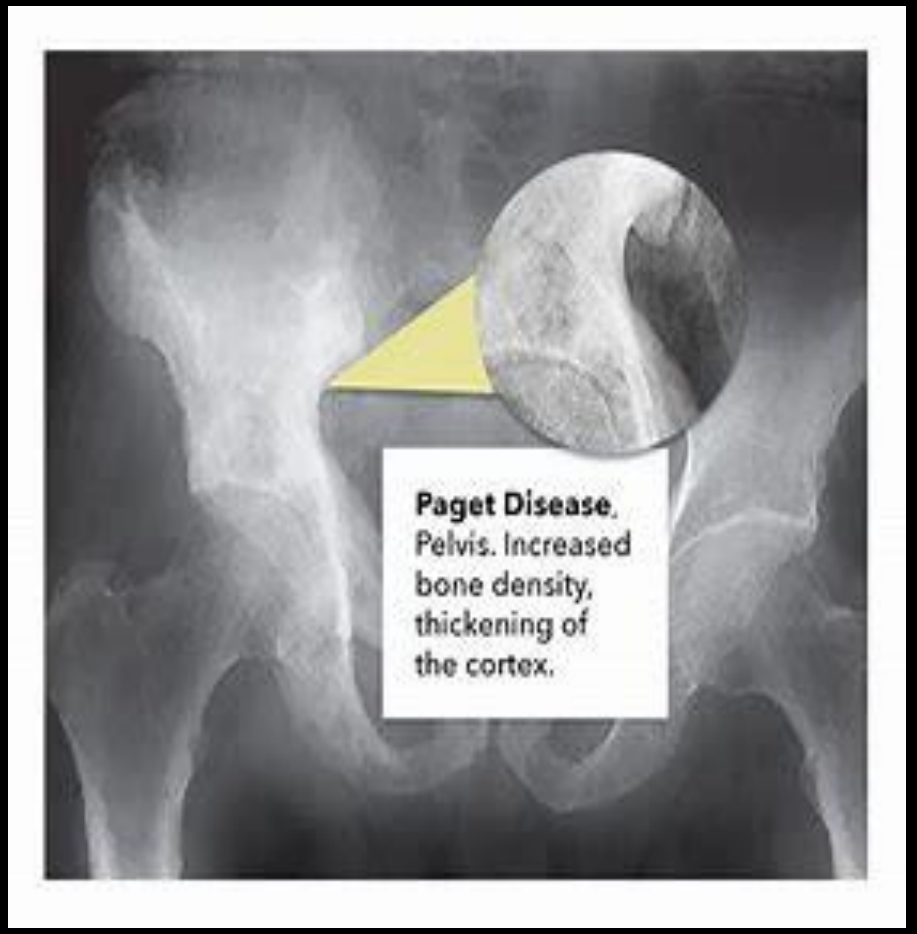
- The alveolar ridges tend to remain symmetrical but become grossly enlarged. If the patient is dentulous, then the enlargement causes spacing of the teeth. Edentulous patients may complain that their dentures no longer fit because of the increased alveolar size.

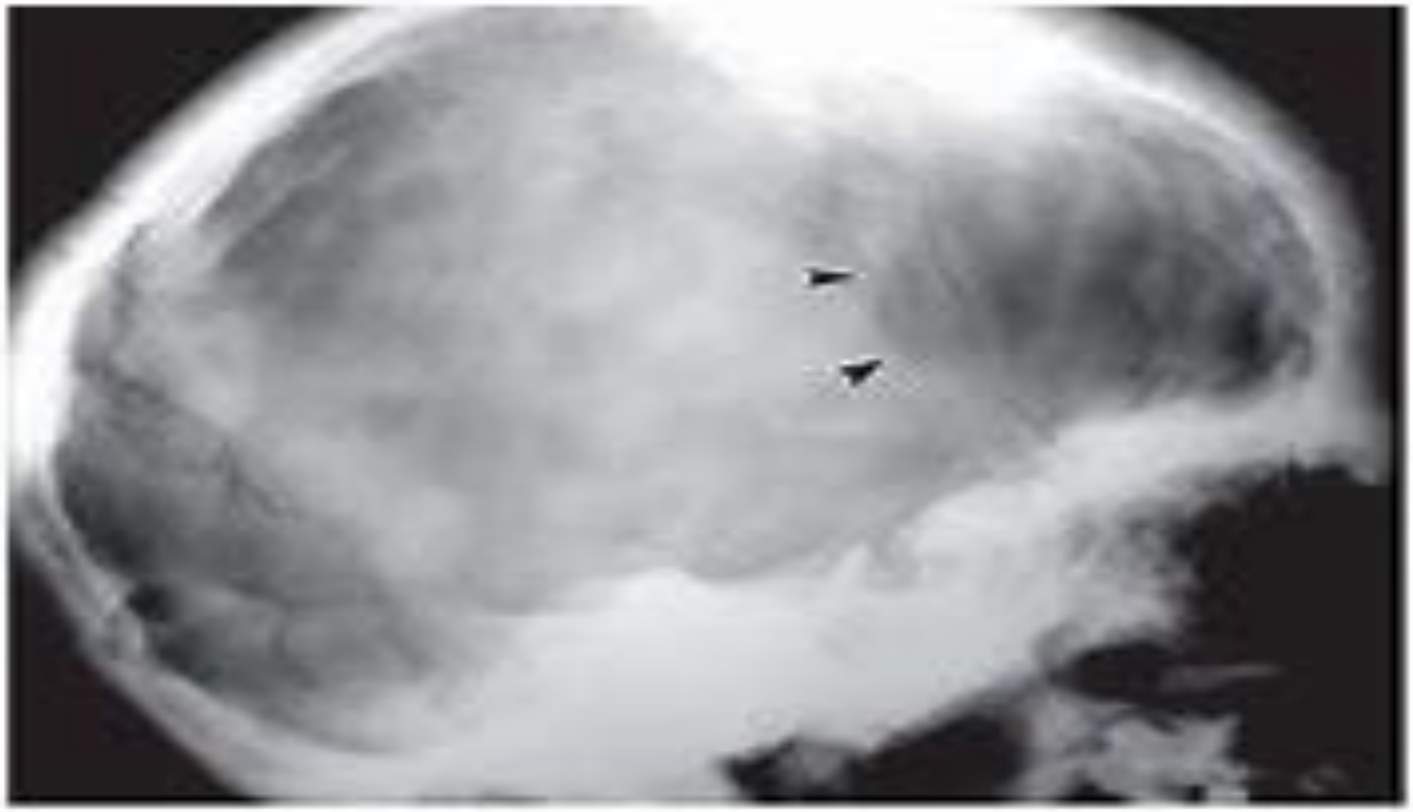
Radiographically:

- The early stages of Paget's disease reveal a decreased radiodensity of the bone and alteration of the trabecular pattern. Particularly in the skull, large circumscribed areas of radiolucency may be present ***(osteoporosis circumscripta).***

- During the osteoblastic phase of the disease, patchy areas of sclerotic bone are formed, which tend to become confluent. The patchy sclerotic areas often are described as having a ***"cotton wool"*** appearance.

- Teeth often demonstrate extensive hypercementosis





Lateral skull radiograph of 67-year-old woman with Paget disease shows area of osteolysis (*arrowheads*) *in frontal region that is designated*

"osteoporosis circumscripta" in lytic phase of Page disease .

Histopathologic features

uncontrolled alternating resorption and formation of bone.

A characteristic microscopic feature is the presence of ***basophilic reversal lines*** in the bone. These lines indicate the junction between alternating resorptive and formative phases of the bone and result in a "***jigsaw puzzle***," or "***mosaic***," appearance of the bone.

Diagnosis

- Elevations in serum alkaline phosphatase levels" which is sensitive marker "with normal blood calcium and phosphorus levels

- Urinary hydroxyproline levels often are markedly elevated.

The clinical and radiographic features, combined with supportive laboratory findings, are typically sufficient for diagnosis.

Paget Disease of Bone
Histopathology

- Chinese character bone
  - Mosaic; jig-saw bone
- Immature trabeculae
- Abundant osteoblastic activity
- Lesser osteoclastic activity
- Fibrous background stroma
- Many reversal/cement lines in the bone