Fibro-osseous Lesions

A group of lesions affecting the *craniofacial skeleton* and characterized microscopically by *fibrous stroma* containing various combinations of bones and/or cementum-like material fall under the term ***benign fibro-osseous lesions***. They include a wide variety of lesions of *developmental*, *dysplastic*, and *neoplastic* origins with different clinical and radiographic presentation & behavior.

Fibro osseous lesions:

- **1. Fibrous dysplasia.**
- **2. Ossifying fibroma.**
- **3. Cemento Osseous Dysplasia (COD).**

FIBROUS DYSPLASIA

- Developmental tumor-like lesion characterized by replacement of normal bone by an excessive proliferation of cellular fibrous connective tissue mixed with irregular bone trabiculae.

- Results from mutation in GNAS 1 (Guanine nucleotide-binding protein, α-stimulating activity polypeptide 1). **Severity** of the condition depend on the **Time** during fetal or postnatal life that the mutation of GNAS1 occurs

CLINICAL TYPES:

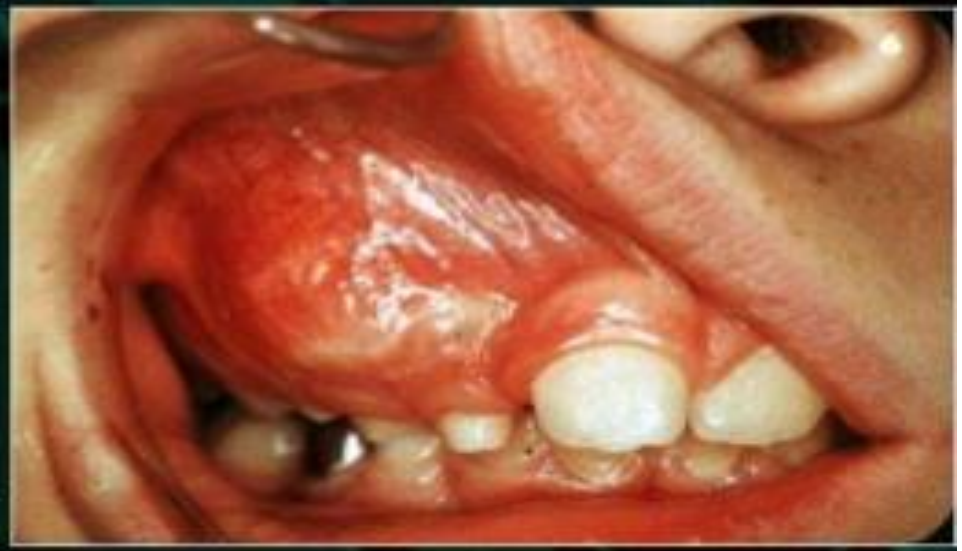
- **Monostotic fibrous dysplasia-** only one bone is involved.
- **Polyostotic fibrous dysplasia-** more than one bone is involved:

- **Jaffe type**; fibrous dysplasia involving variable number of bones, accompanied by pigmented lesions of the skin or ***café-au-lait*** spots.
- **Mc-Cune albright's syndrome**; this is severe form of fibrous dysplasia involving nearly all bones in the body, accompanied by pigmented lesions of the skin plus endocrinal disturbance of various types.

- **A craniofacial form-** In which the maxilla and adjacent bones are involved.

MONOSTOTIC FORM

CLINICAL FEATURES



- Limited to a single bone.
- It is common than the polyostotic form
- Accounts for 70% to 80% of all cases
- Occurs most commonly at the age of 20 to 30 years.
- M:F = 1:1
- Most common sites in order of affection are ribs, femur, tibia, maxilla and mandible.
- In Jaw bones **Maxilla > Mandible**.
- Unilateral painless facial swelling or enlarging deformity of alveolar process.
- Slow growth, become static with skeletal growth completion
- Teeth involved are either malaligned, tipped or displaced.

Polyostotic Fibrous dysplasia (Jaffe-Lichtenstein Syndrome; McCune – Albright Syndrome)

- Involvement of two or more bones.
- When seen with *café au lait* pigmentation → Jaffe-Lichtenstein syndrome.
- Polyostotic fibrous dysplasia + *café au lait* pigmentation + multiple endocrinopathies (precocious puberty, goiter, hyperthyroidism, acromegaly) → McCune Albright Syndrome.

CLINICAL FEATURES

- In children usually less than 10 years of age.
- male: female = 1:3
- May present with facial asymmetry
- Clinical features usually dominated by symptoms related to long bone lesions – Pathologic fractures with pain and deformity
- Leg length discrepancy due to involvement of upper portion of femur ***(hockey stick deformity)***

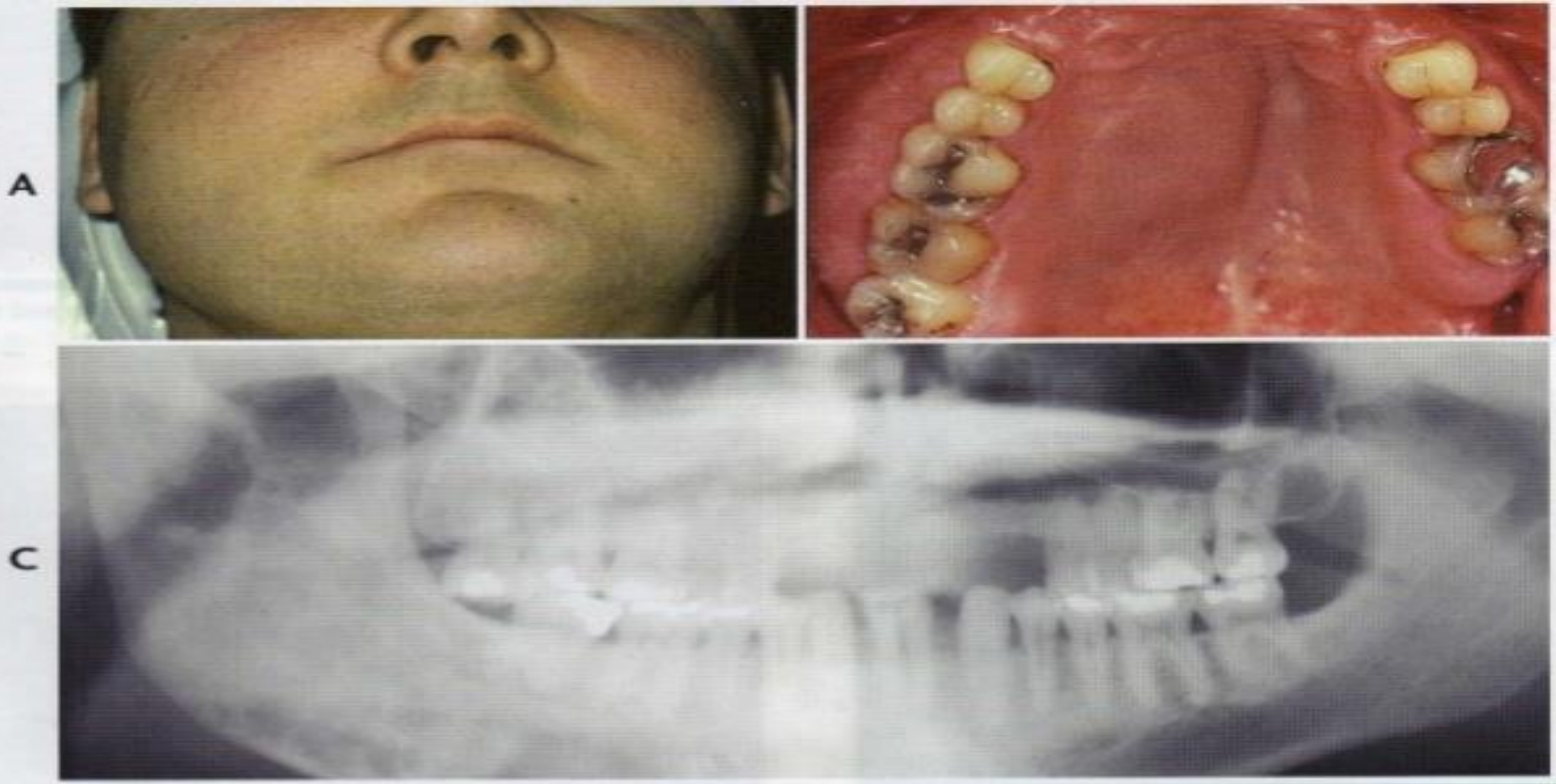


Figure 14-36 • **Polyostotic fibrous dysplasia.** Jaffe-Lichtenstein syndrome: **A,** young man exhibiting enlargement of the right maxilla and mandible; **B,** intraoral photograph showing unilateral maxillary expansion; **C,** panoramic radiograph showing ill-defined lesions of the right side of both jaws.

SKIN PIGMENTATION

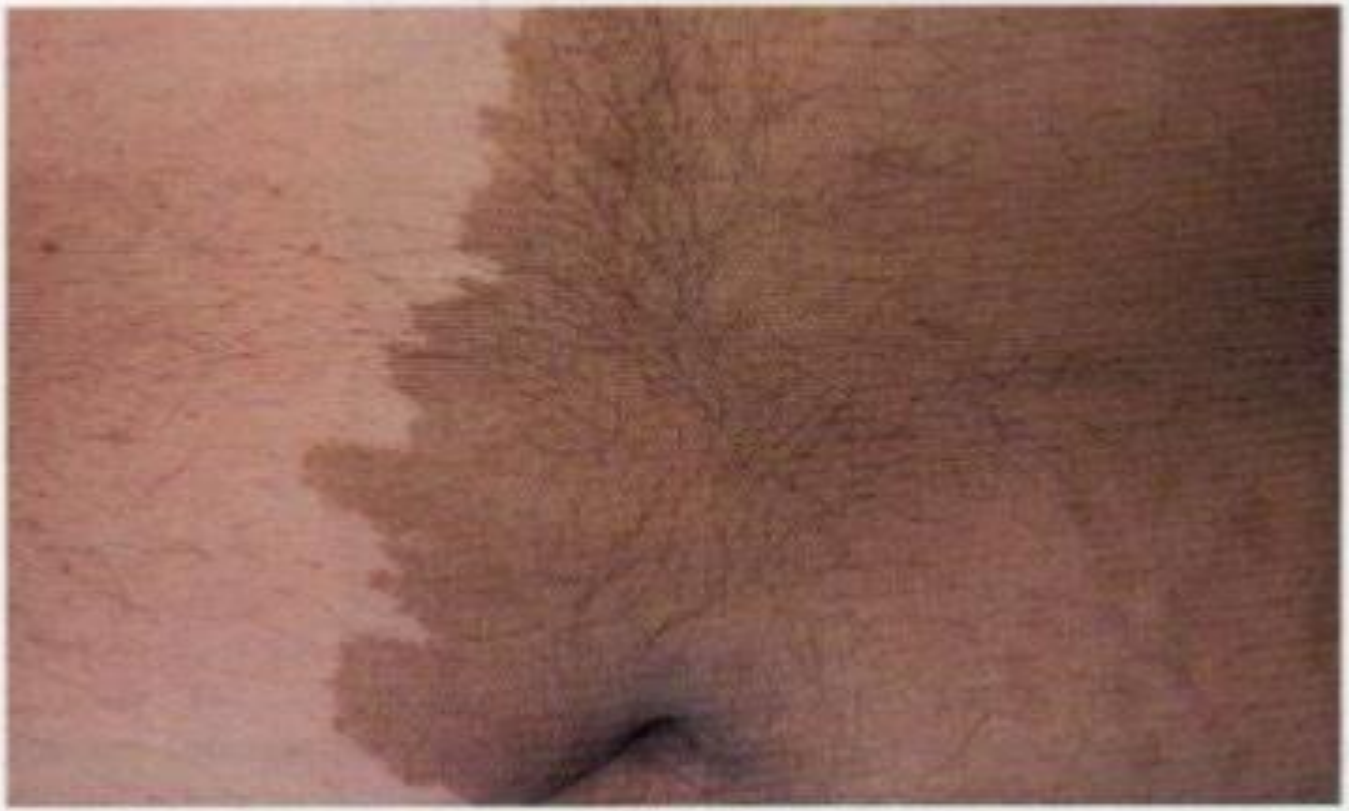


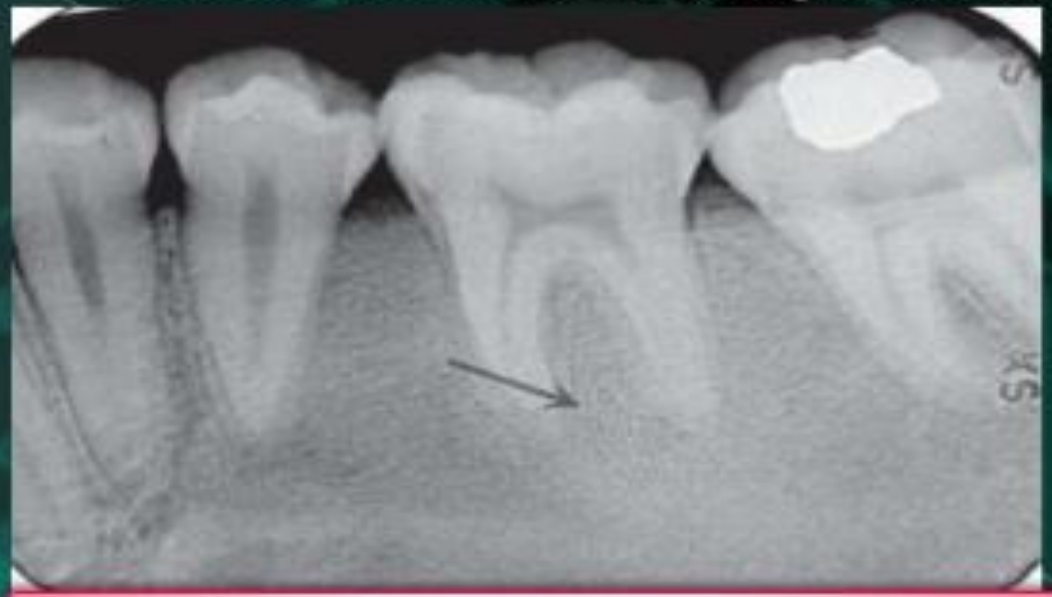
Figure 14-37 • **Polyostotic fibrous dysplasia.** Jaffe-Lichtenstein syndrome: *Café au lait* pigmentation of the abdomen. This is the same patient as shown in Figure 14-36.

Craniofacial fibrous dysplasia

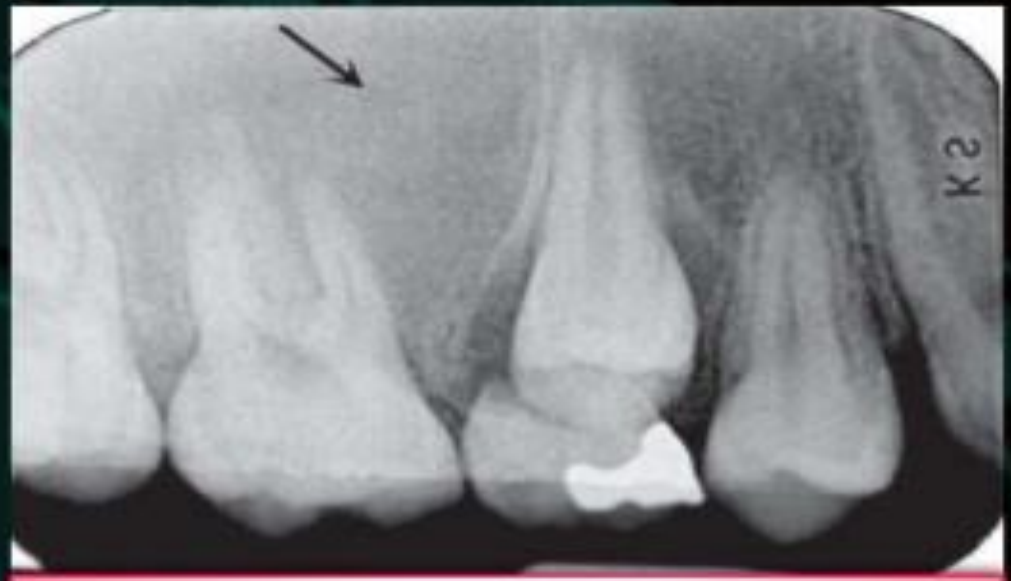
- Occurs in 10-25% pateint with monostotic form and in 50% with polyostotic form
- Peculiar form ***affecting skull bones***
- Not restricted to single bone, but confined to single anatomic site.
- Primarily affect maxilla, but may cross sutures into sphenoid, zygoma, frontonasal bones and base of skull.
- Hypertelorism, cranial asymmetry, facial deformity, visual impairment, exophthalmos, blindness, vestibular dysfunction,tinnitus and hearing loss may occur

Radiographic features:

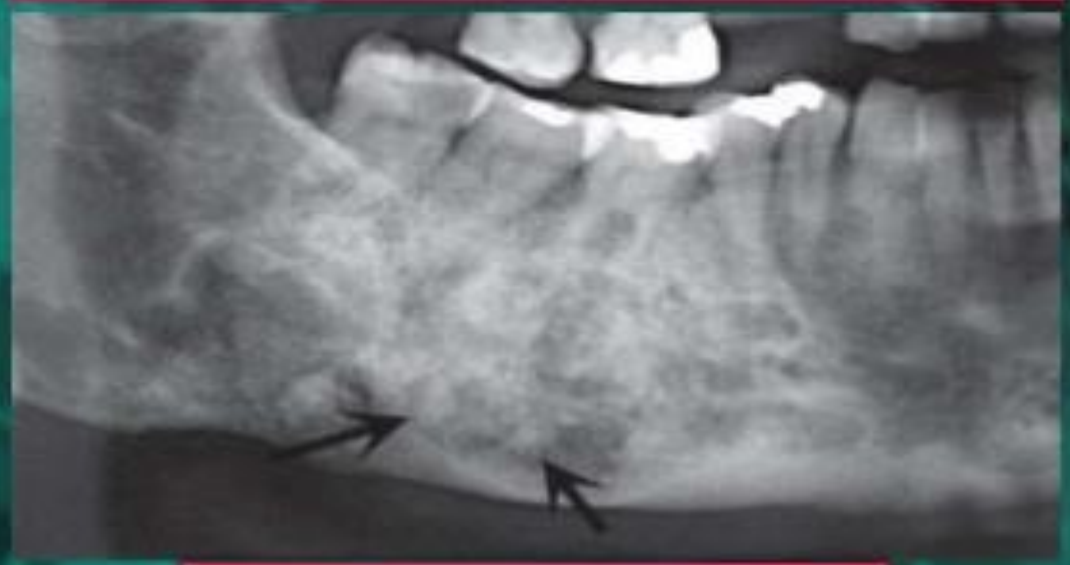
A series of films showing a variety of internal patterns of fibrous dysplasia.



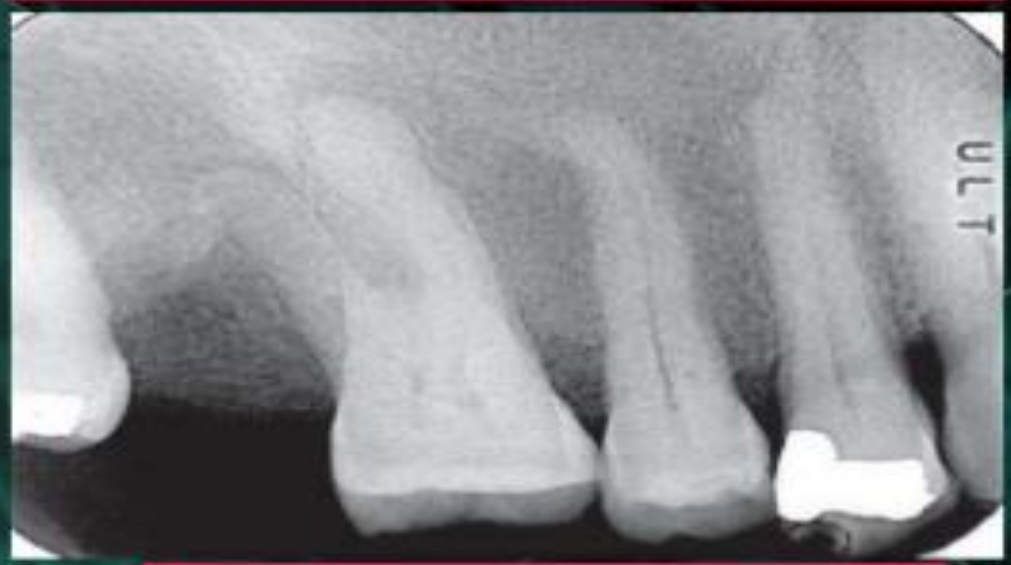
A fingerprint pattern around the roots of the first molar (arrow).



A granular, or ground-glass, pattern (arrow)



A cotton-wool pattern



An orange-peel pattern.

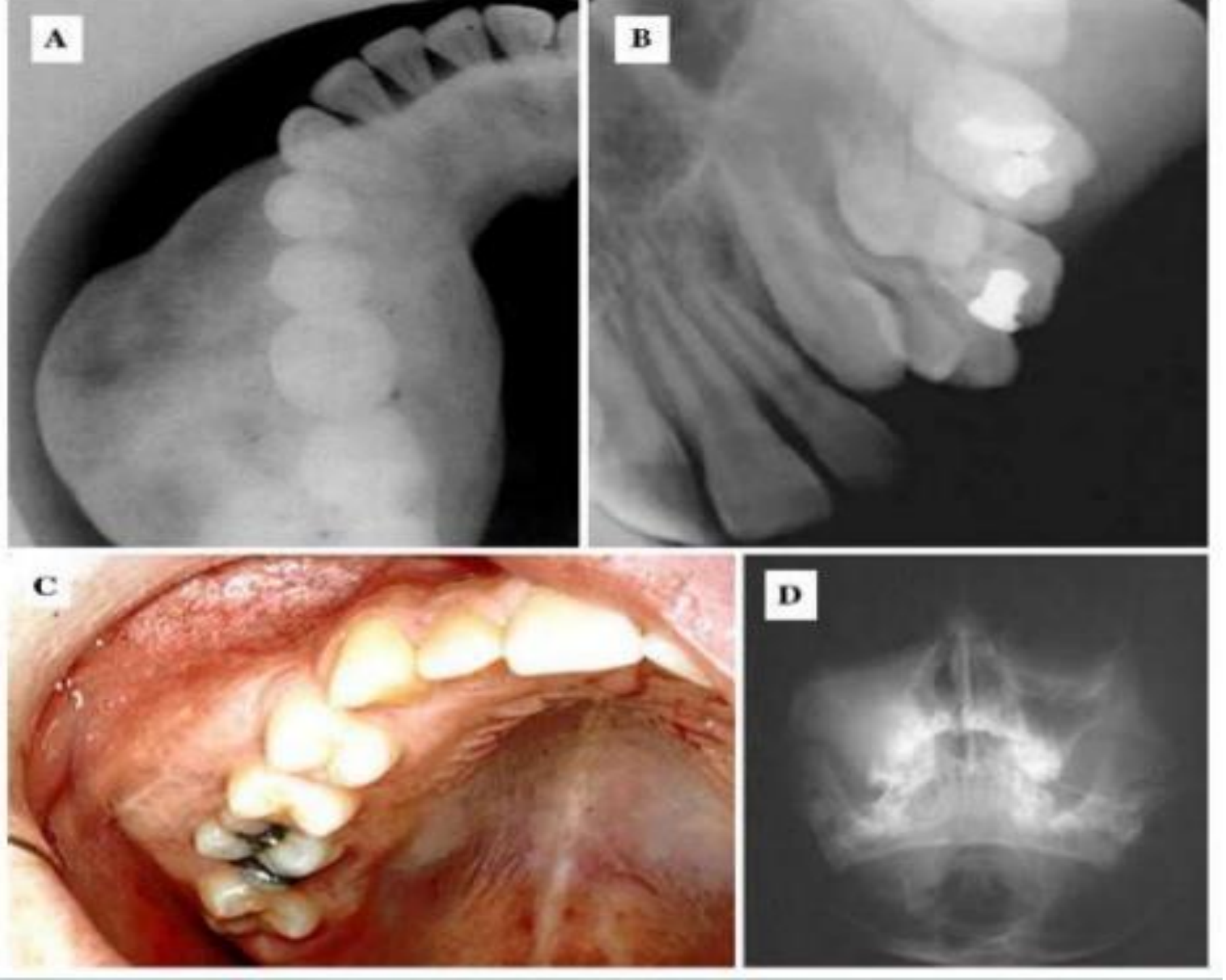


Fig. 2 Fibrous Dysplasia. (**a**) Expansile nonmarginated ground glass opacification in the mandible, (**b**). Ground glass pattern in the maxilla, (**c**). Clinical photograph demonstrating cortical expansion, (**d**). Diffuse unilateral opacification of the maxillary sinus

- **Histopathology:**

FD consists of a slight to moderate cellular fibrous connective tissue stroma that contains foci of irregularly shaped trabeculae of immature bone. The bone trabeculae assume irregular shapes linked to ***Chinese characters***and they do not display any functional orientation, without osteoblastic activity at the bone trabeculae margins.

Chinese characters

HISTOPATHOLOGY

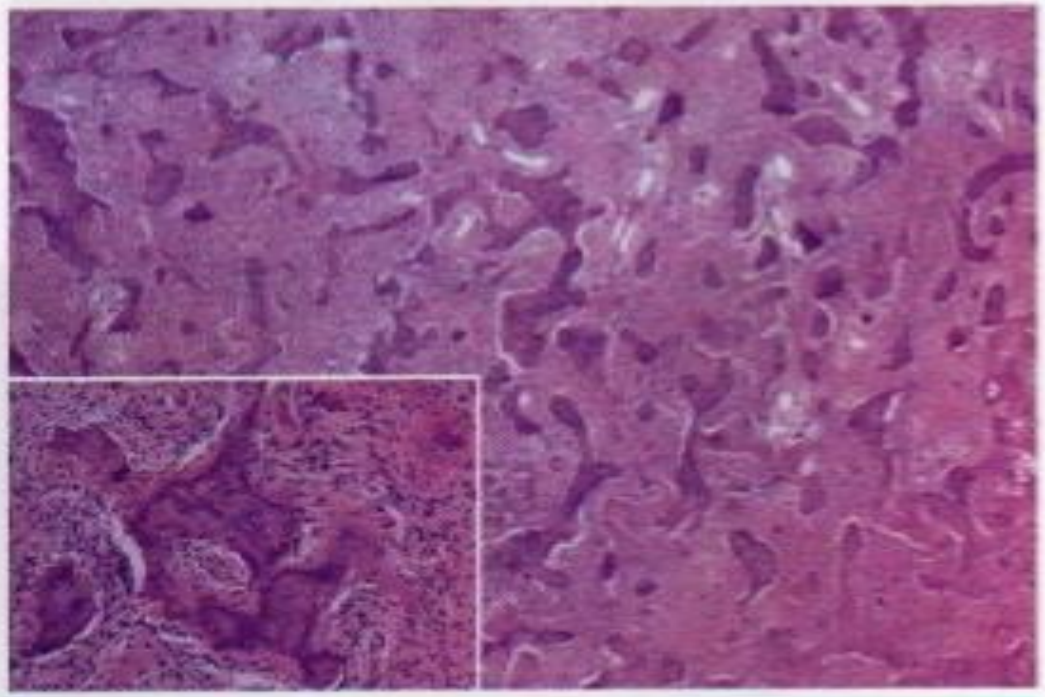


Figure 14-38 • **Fibrous dysplasia.** Irregularly shaped trabeculae of woven bone in a fibrous stroma.

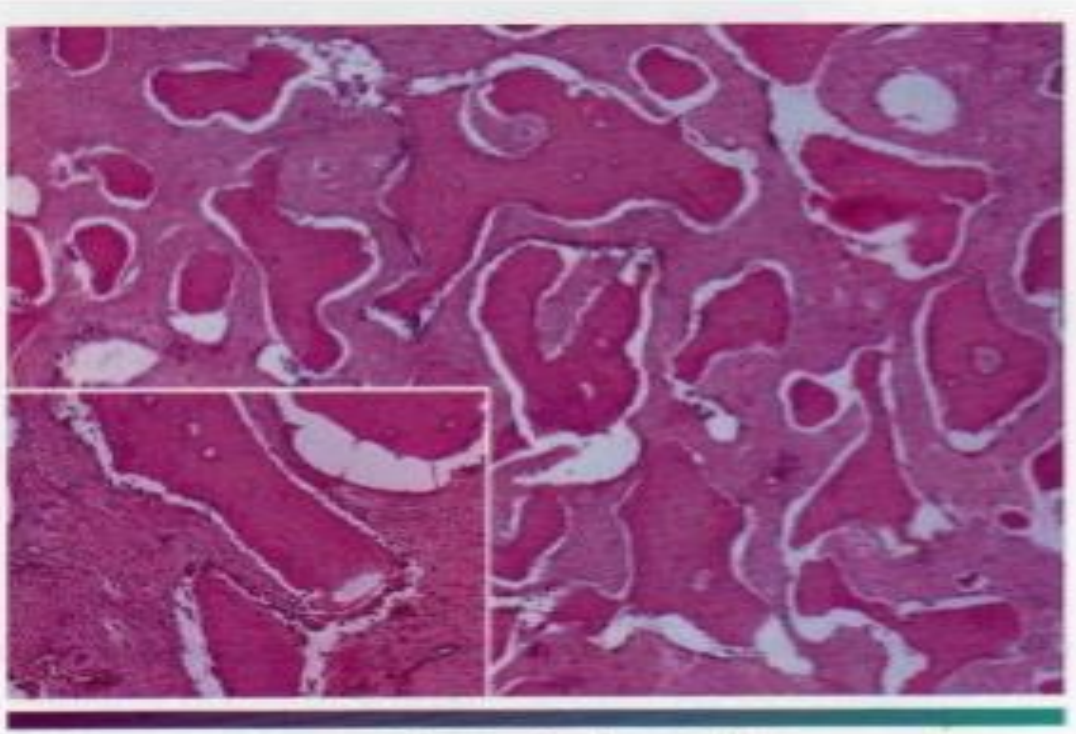


Figure 14-39 • **Mature fibrous dysplasia.** This long-standing lesion shows scattered trabeculae of bone within fibrous connective tissue. Note the lamellar maturation of the bone.

OSSIFYING FIBROMAS

(cementifying fibroma; cemento-ossifying fibroma)

- A true neoplasm with a significant growth potential.
- Composed of fibrous tissue that contains a variable mixture of bony trabeculae, cementum- like spherules, or both.
- Origin from odontogenic or periodontal ligament
- Cementum-like material present

Types

- Ossifying/ Cementifying Fibroma
- Juvenile Ossifying Fibroma
  - -Trabecular Juvenile Ossifying Fibroma
  - -Psammomatoid Juvenile Ossifying Fibroma

Ossifying/Cementifying Fibroma

- This bone tumour consists of highly cellular, fibrous tissue that contains varying amounts of abnormal bone or cementum-like tissue.
- Most common form of OF occurs in maxilla and mandible.

- Painless with expansion of both cortices.
- Larger lesions may expand the inferior aspect of mandible.
- Teeth are displaced superiorly (mandibular lesion) and inferiorly(maxillary lesion) and expand into the antrum.

Ossifying Fibroma

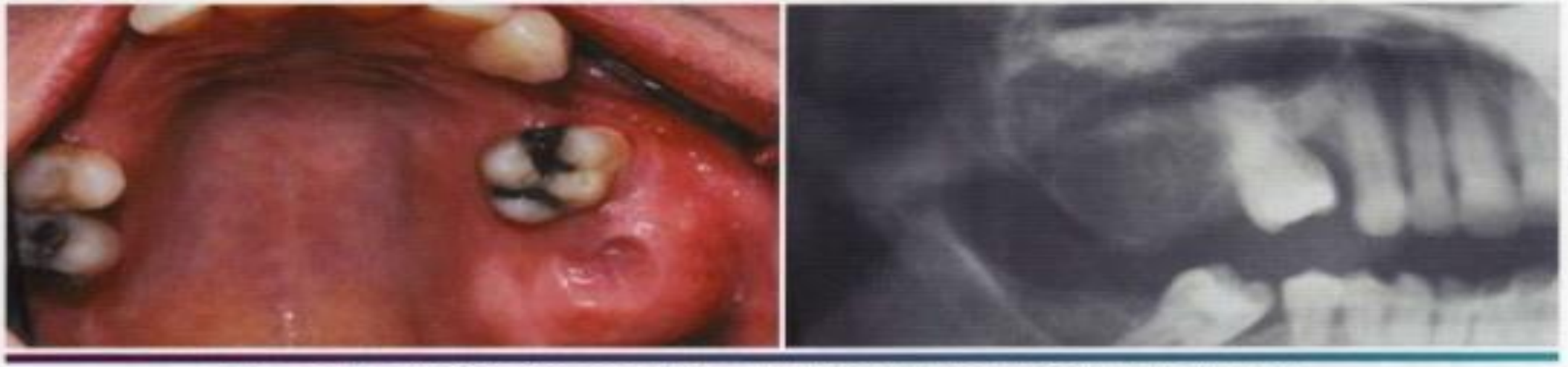


Figure 14-51 • **Ossifying fibroma. A,** Enlargement of the posterior maxilla caused by a large ossifying fibroma. **B,** Note the mixed radiolucent and radiopaque lesion expanding the posterior maxilla.

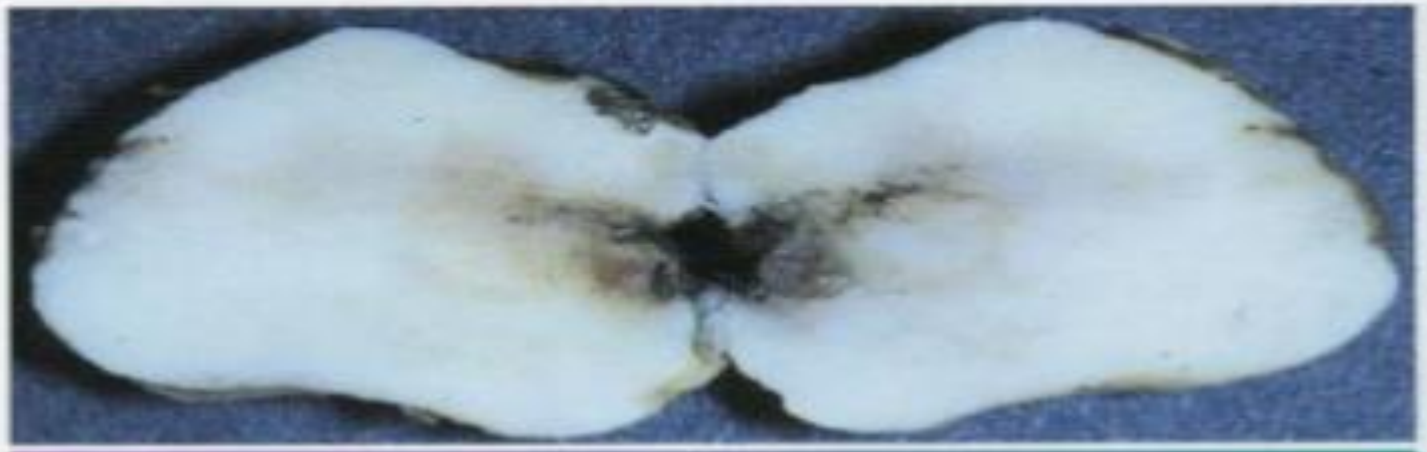
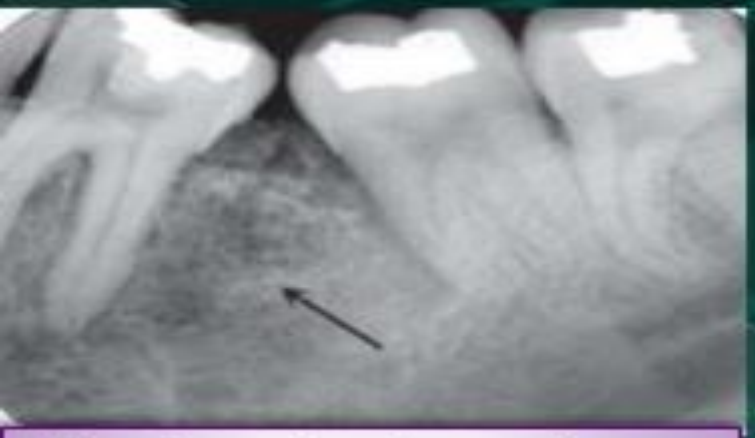


Figure 14-52 • **Ossifying fibroma.** Gross specimen showing a well-circumscribed tumor that shelled out in one piece.

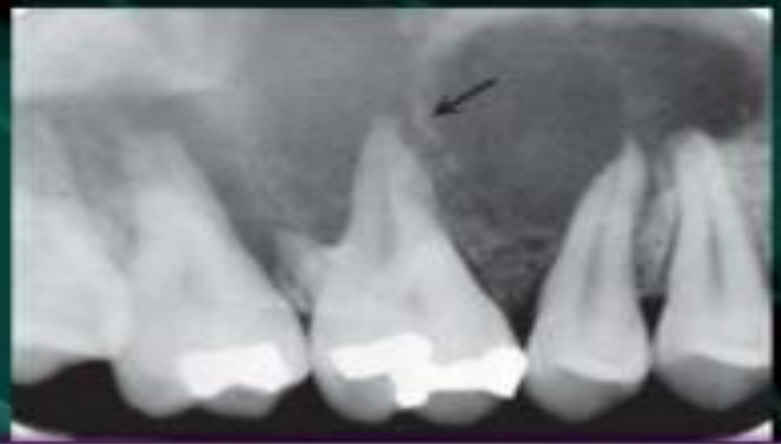
- **Radiographic Findings of COF:**

Well circumscribed, sharply demarcated border is the most common presenting radiographic feature, although OF may present as relatively lucent or opaque depending on the density of the calcification present. Also they may be unilocular or multilocular, mixed radiolucent-radiopaque image may be seen. The roots of the teeth present may be displaced & less commonly resorption is seen.

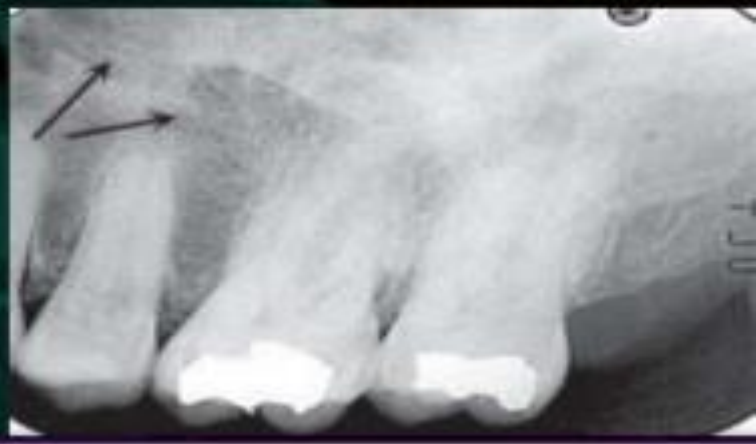
Various Bone Patterns Seen in Cemento-Ossifying Fibromas



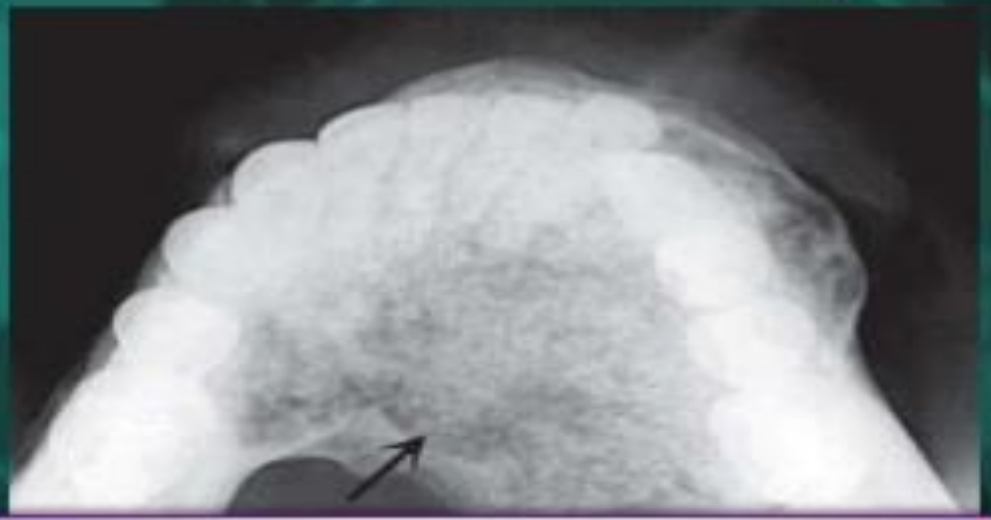
A wispy trabecular pattern *(arrow)*



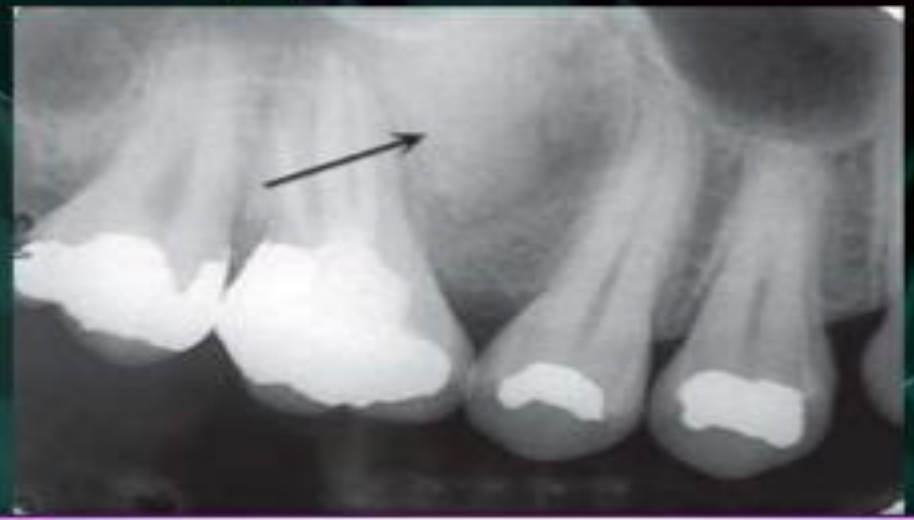
Radiolucent pattern with a few wispy trabeculae (arrow).



A fibrous dysplasia, granular-like pattern *(arrows).*



A flocculent pattern with larger tufts of bone formation *(arrow).*



A solid, radiopaque, cementum-like pattern (arrow).

- **Treatment & Prognosis:**

Surgical removal using ***curettage*** or ***enucleation***. The lesion can typically be separated easily from the surrounding bone. Recurrence is rare.

Juvenile Ossifying Fibroma:

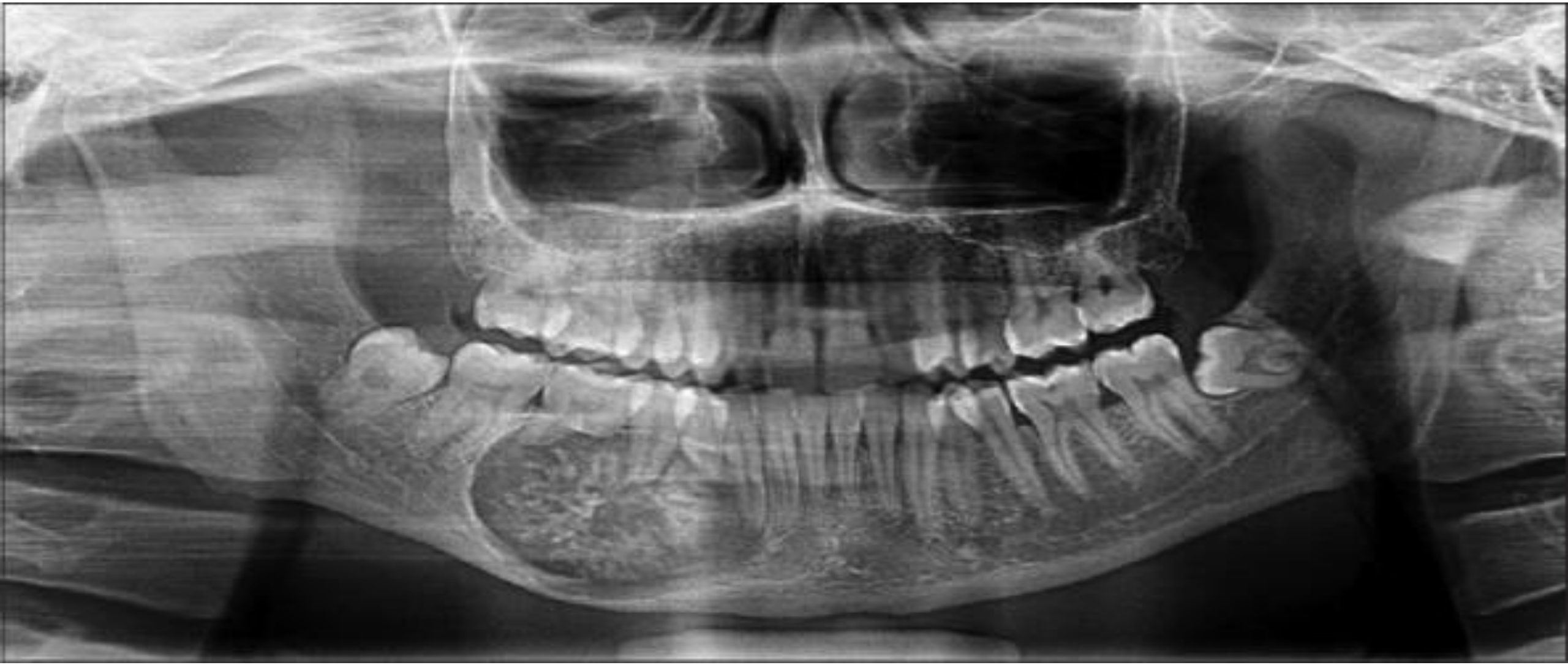
Is a well circumscribed rapidly growing neoplasm lack the continuity with adjacent normal bone. Lesions are circumscribed radiolucencies in some cases contain central radio-opacities (Ground glass) opacification may be observed. Those are present within a sinus may appear radiodense and create a clouding that could be confused with sinusitis. Two different neoplasm have been reported: (1) **Trabecular** and (2) **Psammomatoid**. The latter neoplasm occur more than the trabecular type in a ratio of approximately 4:1

Juvenile ossifying fibroma

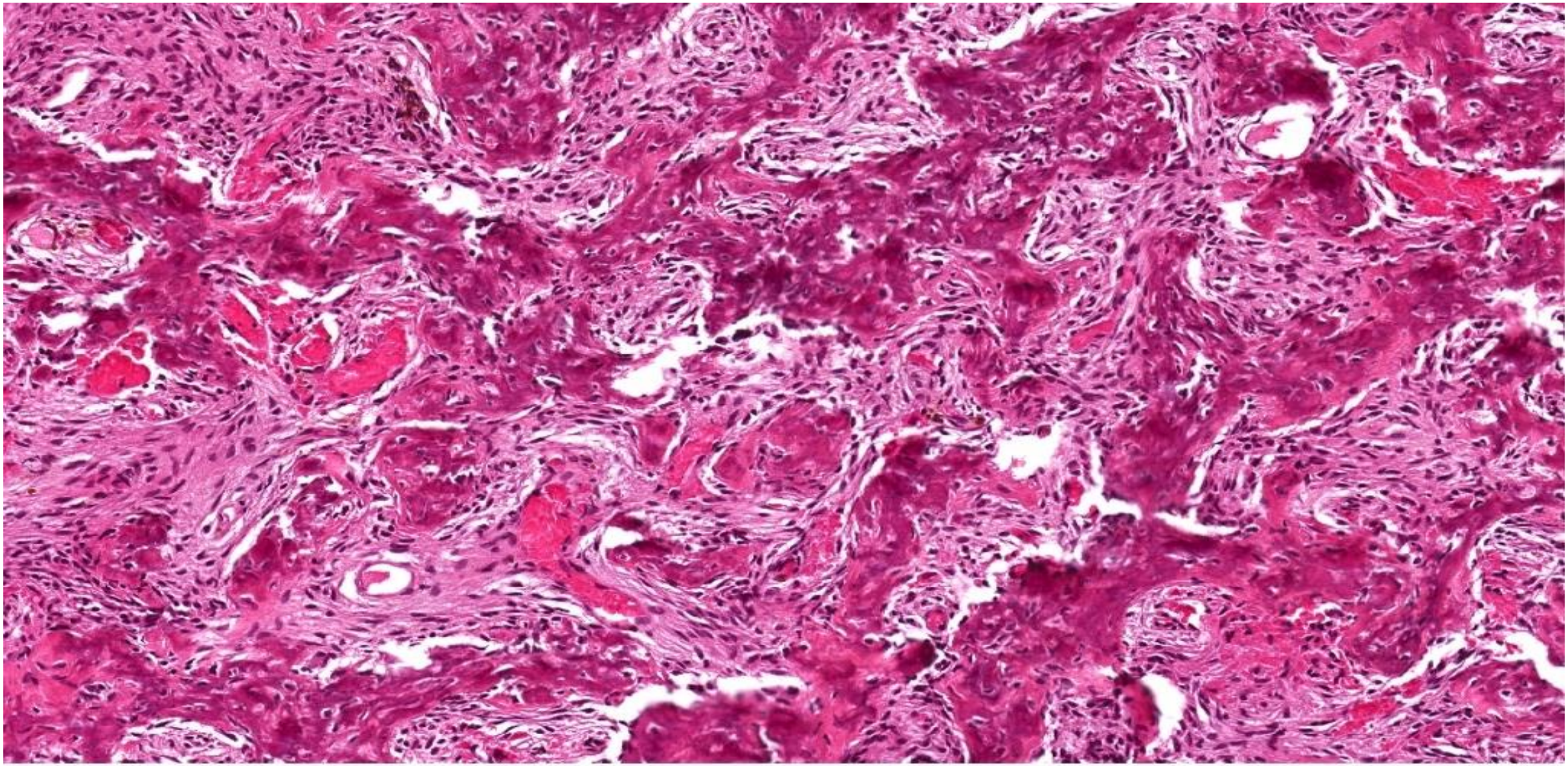
Rev Esp Cir Oral Maxilofac. 2017;39:234-8

Juvenile ossifying fibroma

Juvenile ossifying fibroma

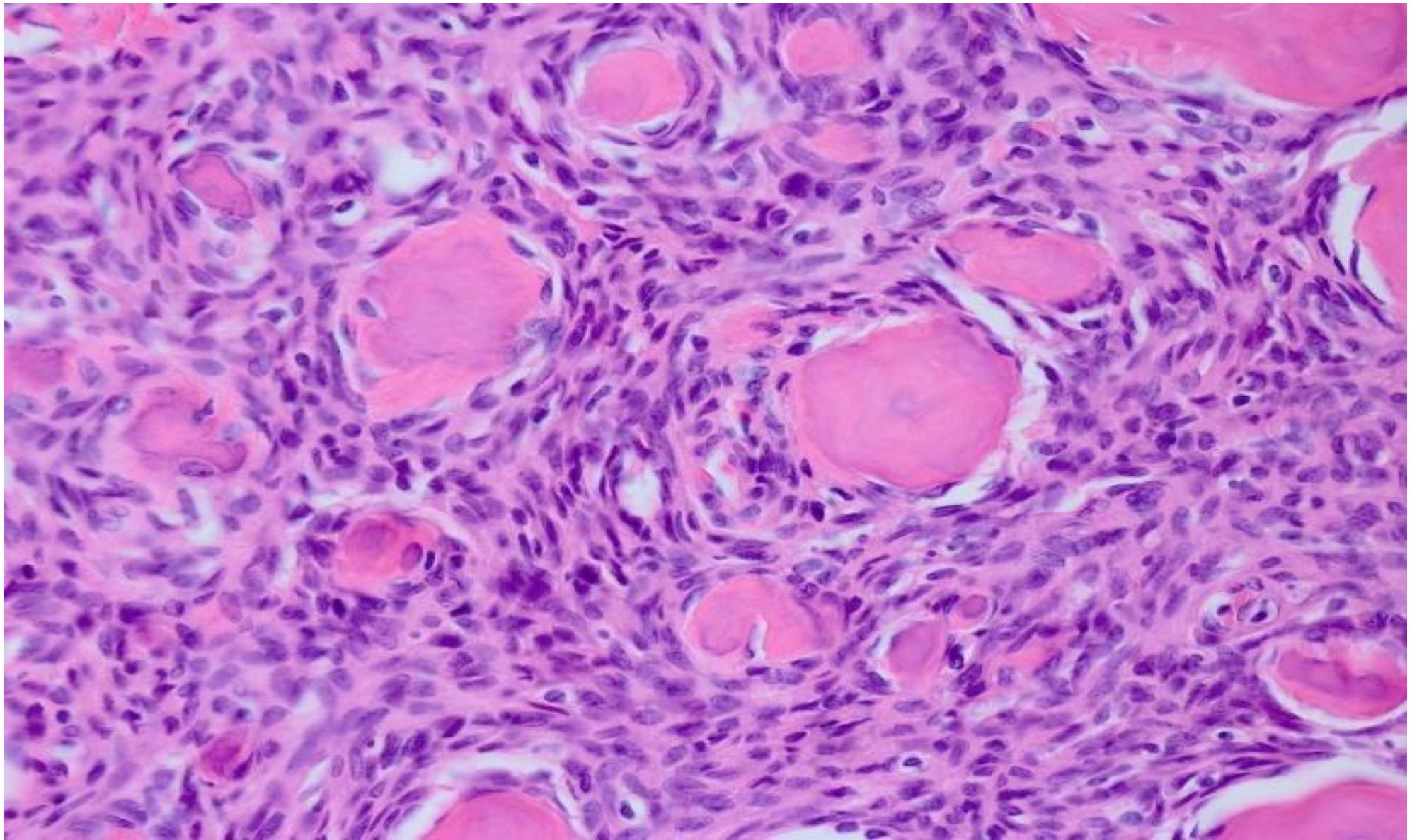


Trabecular juvenile ossifying fibroma



Juvenile trabecular ossifying fibroma, 10x

Psammomatoid juvenile ossifying fibroma



Cemento-osseous Dysplasia (COD):

The term COD refers to a disease process of the jaws for which the precise etiology is unknown.

COD includes:

–Periapical COD.

–Focal COD.

–Florid COD.

All the 3 disease processes have the same features, only distinguished on the basis of the extent of involvement of the affected portions of the jaw.

1. Periapical COD:

Represents a reactive or dysplastic process rather than a neoplastic one. It may represent an unusual response of periapical bone & cementum to some undetermined local factor.

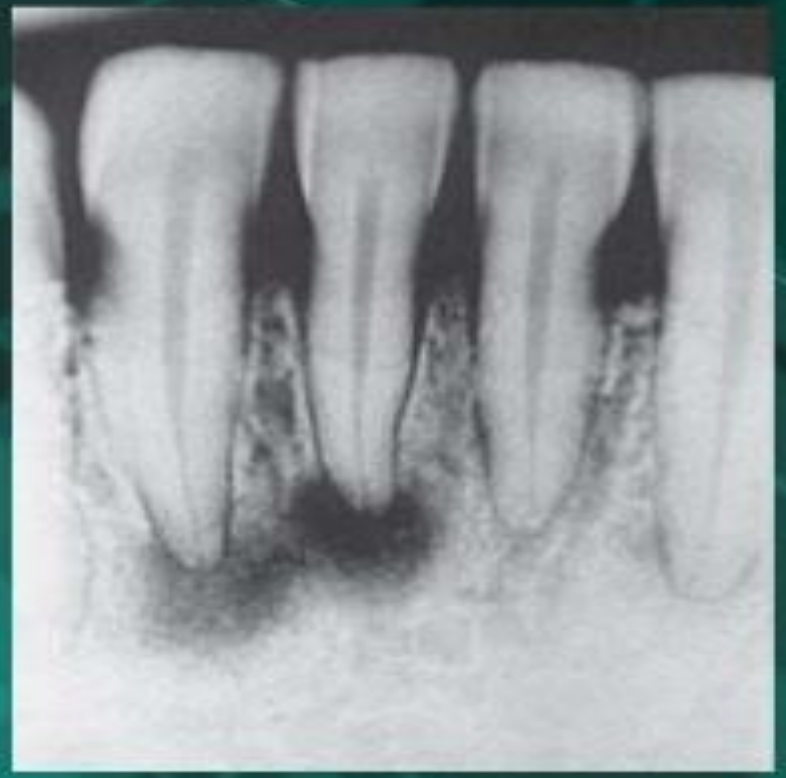
- When not associated with a tooth apex ⟶ ***Focal COD***.

- **Clinical Features:**

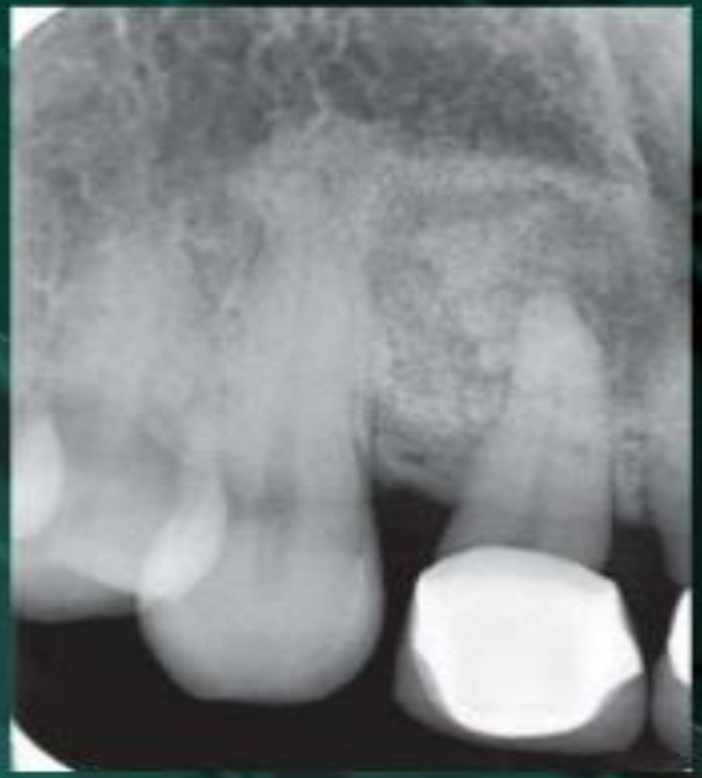
A common phenomenon, that occurs at the apex of vital teeth. A biopsy is unnecessary because the condition is usually diagnosed by clinical & radiographic features. Females are affected more than males. PACOD occurs in females at middle age (around 40 years) & rarely before the age 20. The mandible, especially the anterior periapical region, is far more commonly affected than other areas. More often, the apices of two or more teeth are affected.

- The condition appears 1st as a *periapical lucency* that is continuous with the periodontal ligament space. *To be differentiated from* ***Periapical granuloma*** → ***vitality test***.
- As the condition progresses, the lucent lesion develops into a mixed or mottled pattern because of bone repair.
- The final stage appears as a solid, opaque mass that is surrounded by a thin, lucent ring (after months – years).

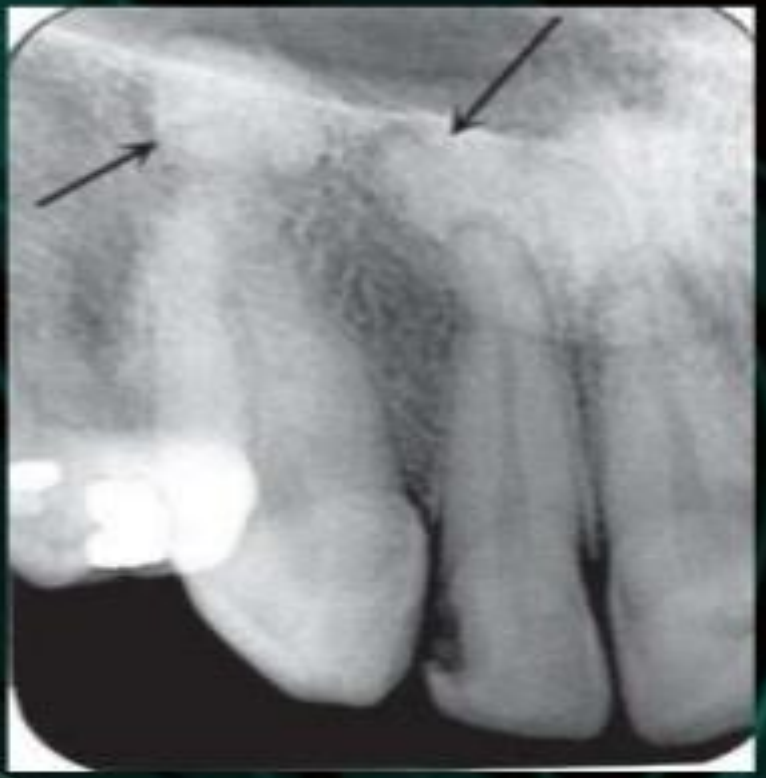
3 stages of Periapical Cemental Dysplasia



Radiolucent Stage



Mixed lesion.



Mature lesions *(arrows)*.

Focal COD:

RADIOGRAPHIC FEATURES

- Most lesions appear as radiolucent – radio-opaque areas with thin peripheral radiolucent rim.
- Lesion is well defined, borders are slightly irregular.
- In edentulous areas development of idiopathic bone cavities, result in bony expansion of affected area.

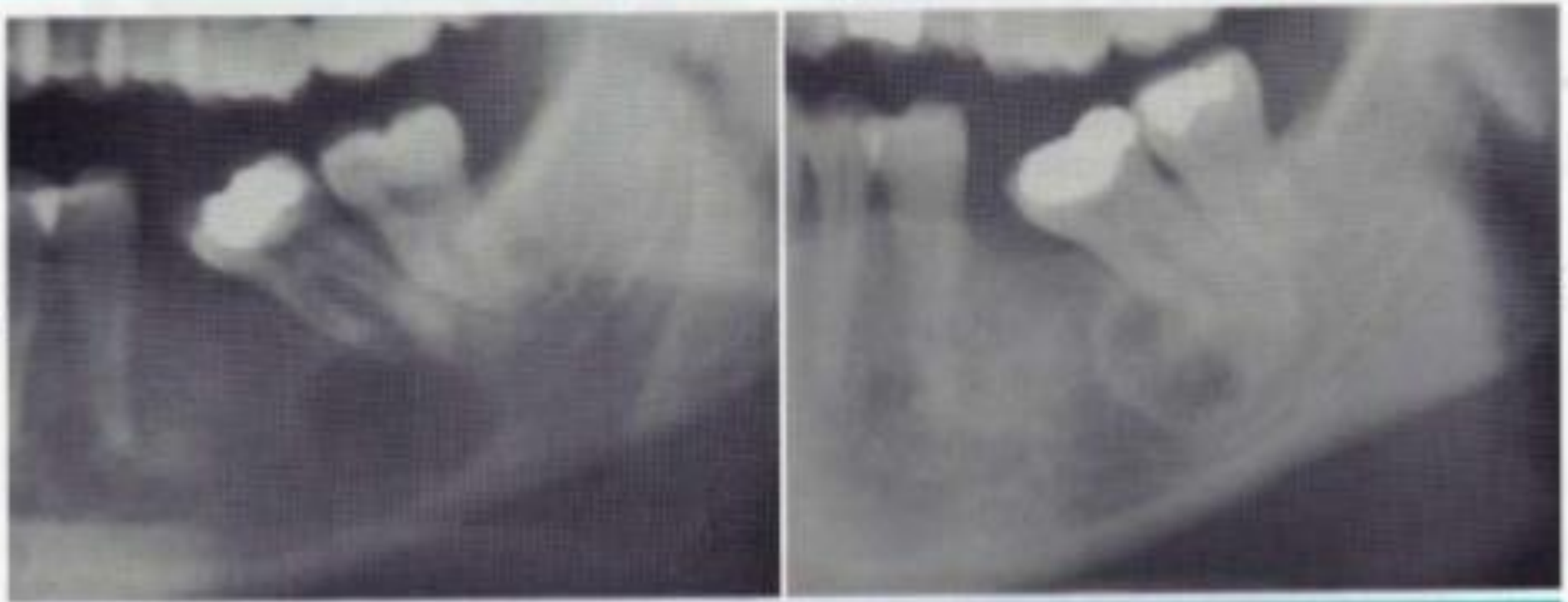


Figure 14-40 • **Focal cemento-osseous dysplasia. A,** A radiolucent area involves the edentulous first molar area and the apical area of the second molar. **B,** Radiograph of the same patient taken 9 years later showing a mixed radiolucent and radiopaque pattern.

2.Florid COD:

The FCOD is an exuberant[1] form of PACOD. FCOD represents the severe end of the spectrum of this unusual process. The patient is asymptomatic except when complication of osteomyelitis occurs. Females are more commonly affected (**black women**); between 25-60 years of age. The condition is typically bilateral & may affect all four quadrants.

Radiographically, FCOD appears as diffuse radiopaque masses throughout the alveolar segment of the jaw. A ***ground-glass*** or cyst-like appearance may also be seen.

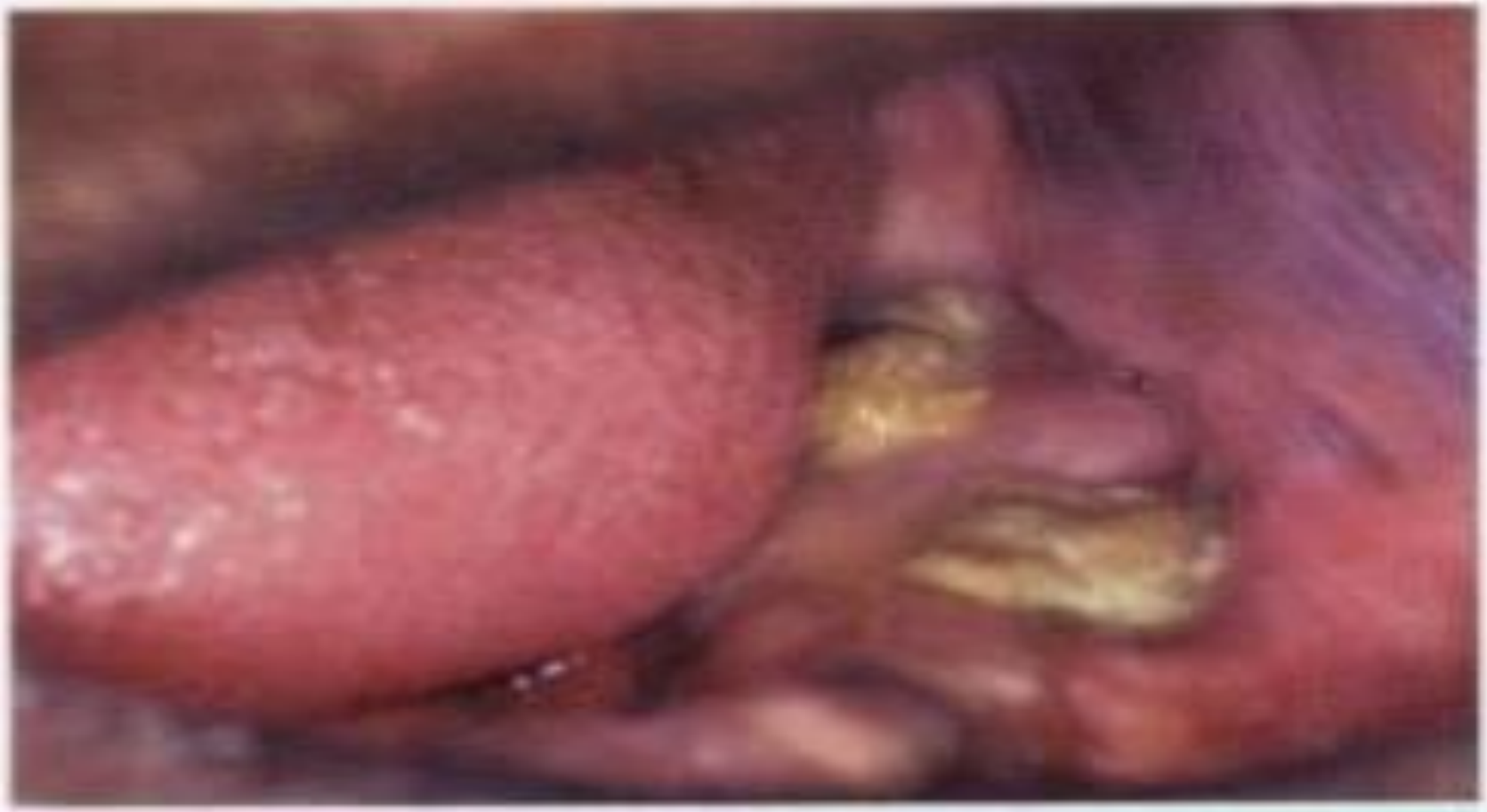
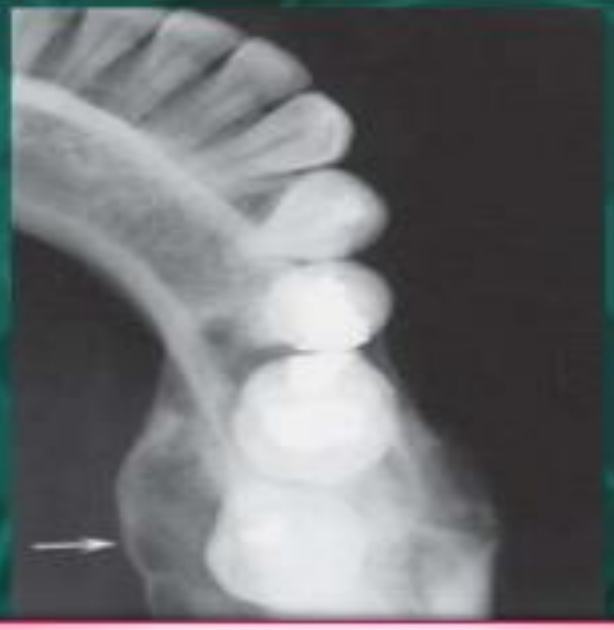
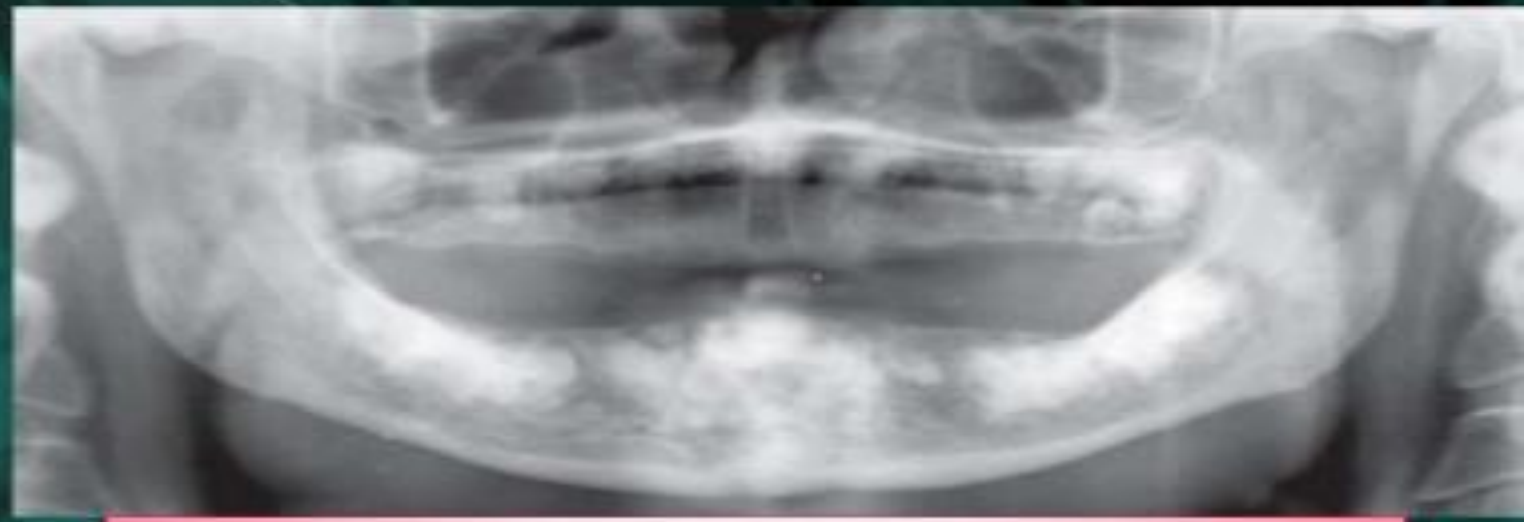


Figure 14-45 • **Florid cemento-osseous dysplasia.** Yellowish, avascular cementum-like material is beginning to exfoliate through the oral mucosa.

Florid cemento osseous dysplasia: multiple mixed radiopaque-radiolucent lesions in the periapical regions throughout the jaws *(arrows)*



Undulating expansion of the medial cortical plate *(arrow)*.



multiple, very mature, almost totally radiopaque lesions in edentulous jaws.