BENIGN TUMOURS OF THE JAWS (PART 1 – ODONTOGENIC TUMOURS)

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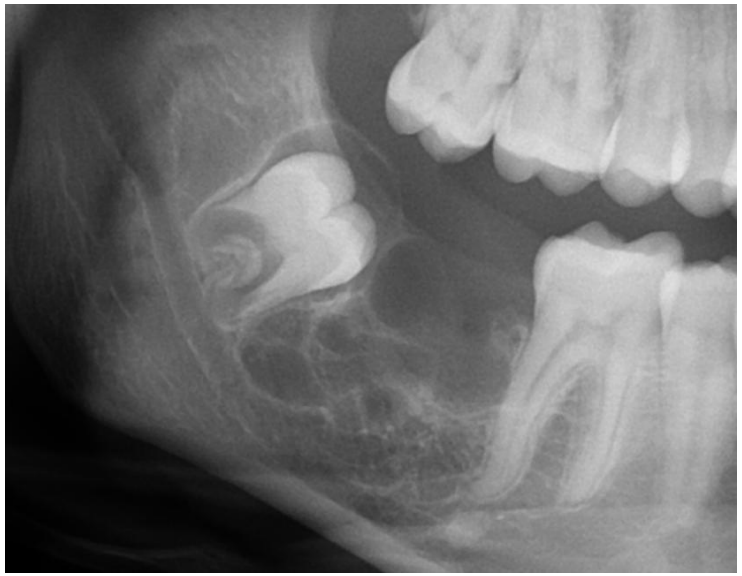
WHAT IS A BENIGN TUMOUR?

Definition: an abnormal mass of tissue that exhibits uncontrolled and uncoordinated growth, persisting even after the stimuli that initiated the growth have ceased. It tends to be slow-growing, well-differentiated and well-circumscribed, resemble the tissue of origin, and remain localised to the site of origin.

Clinical Features:

- Insidious onset with slow growth
- Painless
- Does not metastasise

GENERAL IMAGING FEATURES

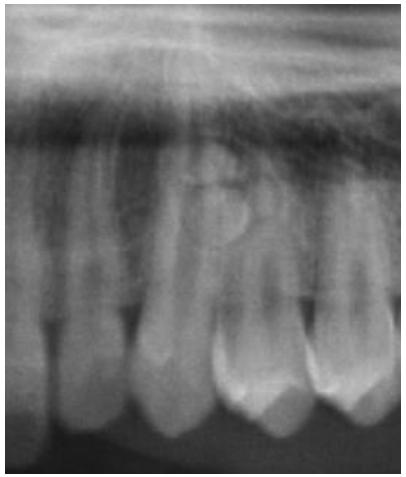


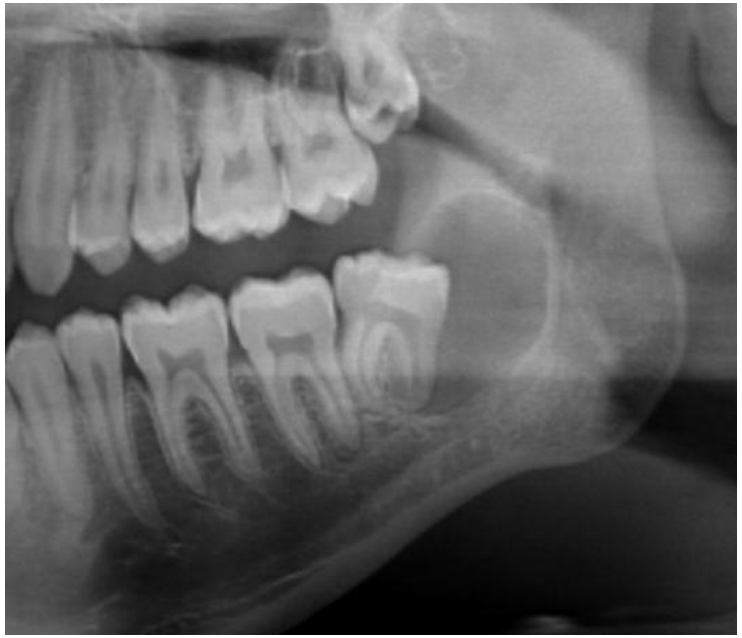
Location:

- Have specific anatomical predilection
- E.g. odontogenic tumours occur in the alveolar processes, above the IAC; cartilaginous tumours occur in jaw locations with residual cartilaginous cells

Periphery/Shape:

- Usually smooth, well-defined, sometimes corticated borders
  - Due to the slow enlargement by formation of additional internal tissue
- May be unilocular or multilocular
- A radiolucent band may be seen
  - This represents a soft tissue capsule





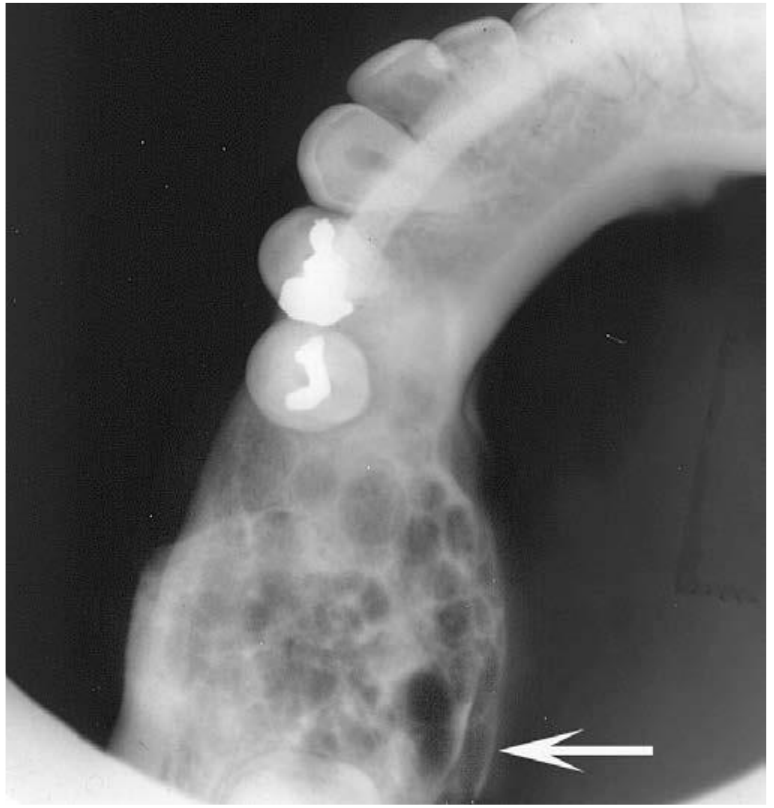
GENERAL IMAGING FEATURES

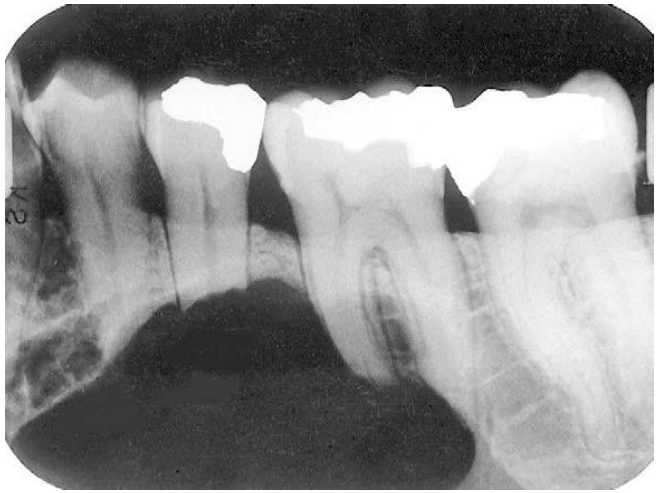
Internal Structure:

- Variable
  - May be totally RL, totally RO, or a mixture of RL/RO tissues
  - If the lesion contains RO elements, these usually represent residual bone, reactive bone formation, or a calcified material produced by the tumour
- The internal pattern may be characteristic for specific types of tumours
  - E.g. curved coarse septa are characteristic for ameloblastomas

Effects on Surrounding Structures:

- Displacement and resorption of teeth
- Expansion and thinning of the jaw cortices +/- perforation
- Displacement of the inferior alveolar canal (IAC)
- Elevation of the antral +/- nasal cortical floors





White and Pharoah, 2014

WHO CLASSIFICATION OF HEAD AND NECK TUMOURS 5TH ED (2022)

Odontogenic Benign Tumours

- Benign epithelial odontogenic tumours
  - Ameloblastoma
    - Ameloblastoma, conventional
    - Ameloblastoma, unicystic type
    - Ameloblastoma, extraosseous/peripheral type
    - Adenoid ameloblastoma
    - Metastasising ameloblastoma
  - Adenomatoid odontogenic tumour
  - Squamous odontogenic tumour
  - Calcifying epithelial odontogenic tumour
- Benign mixed epithelial & mesenchymal odontogenic tumours
  - Odontoma
  - Ameloblastic fibroma
  - Primordial odontogenic tumour
  - Dentinogenic ghost cell tumour
- Benign mesenchymal odontogenic tumours
  - Odontogenic myxoma/myxofibroma
  - Odontogenic fibroma
  - Cementoblastoma
  - Cemento-ossifying fibroma

Non-Odontogenic Benign Tumours

- Benign maxillofacial bone and cartilage tumours
  - Osteoma
  - Osteochondroma
  - Osteoblastoma
  - (Osteoid osteoma)
  - Chondroblastoma
  - Chondromyxoid fibroma
  - Desmoplastic fibroma of bone
- Soft tissue tumours
  - Vascular tumours
    - Haemangioma
  - Peripheral nerve sheath tumours
    - Neurofibroma
    - Schwannoma
    - Neuroma

PART 1: BENIGN EPITHELIAL ODONTOGENIC TUMOURS

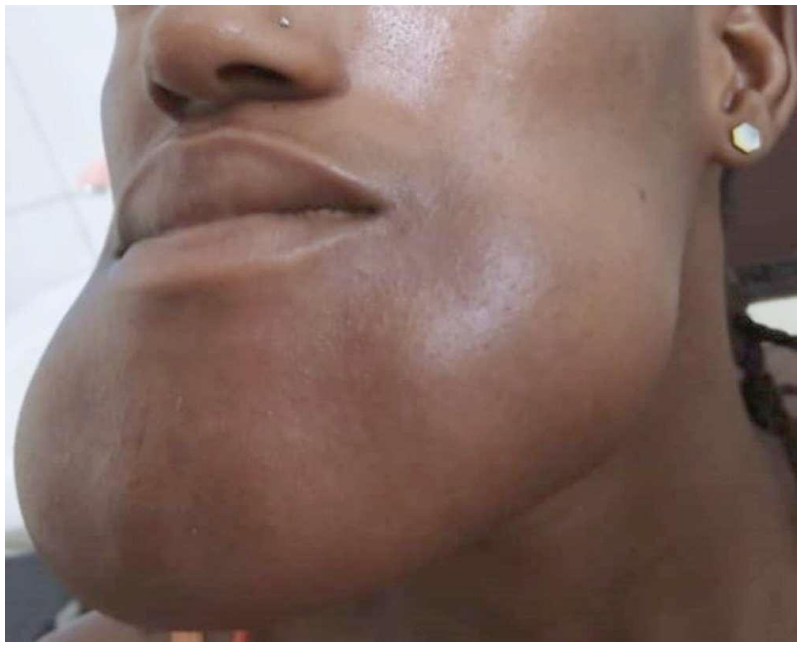
1. Ameloblastoma
    - Ameloblastoma, conventional
    - Ameloblastoma, unicystic type
    - Ameloblastoma, extraosseous/peripheral type
    - Adenoid ameloblastoma
    - Metastasizing ameloblastoma
2. Adenomatoid odontogenic tumour
3. Squamous odontogenic tumour
4. Calcifying epithelial odontogenic tumour

1.1 AMELOBLASTOMA

Definition	A benign but locally infiltrative epithelial odontogenic neoplasm of the jawbones characterised by ameloblast-like cells and stellate reticulum.
Types	1. Conventional (aka Solid-multicystic) (92%) • Follicular, plexiform, acanthomatous, granular cell, basal cell, desmoplastic 2. Unicystic (6%) 3. Extraosseous/Peripheral 4. Adenoid ameloblastoma 5. Metastasizing ameloblastoma
Prevalence	The most common odontogenic tumour in all ethnic groups (excluding odontomas) ~1% of all H+N neoplasms, with highest incidence in African and Afro-Caribbean populations
Age	Peak incidence = 4^{th} – 5^{th} decades Range = 8-92 years
Gender	M=F

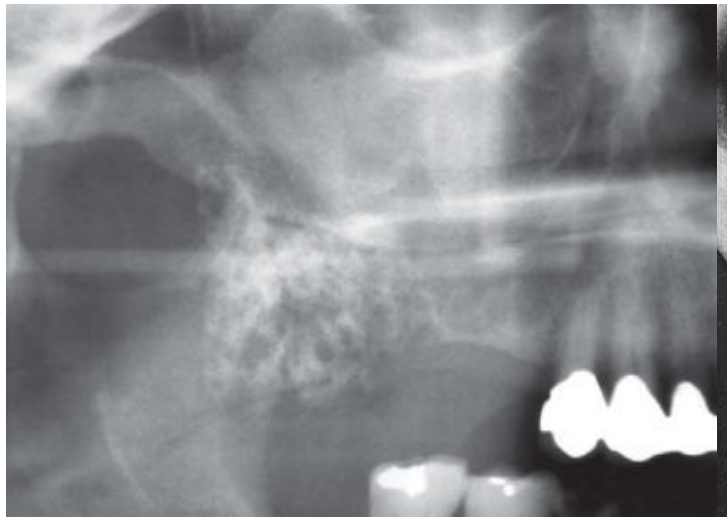
1.1.1 AMELOBLASTOMA

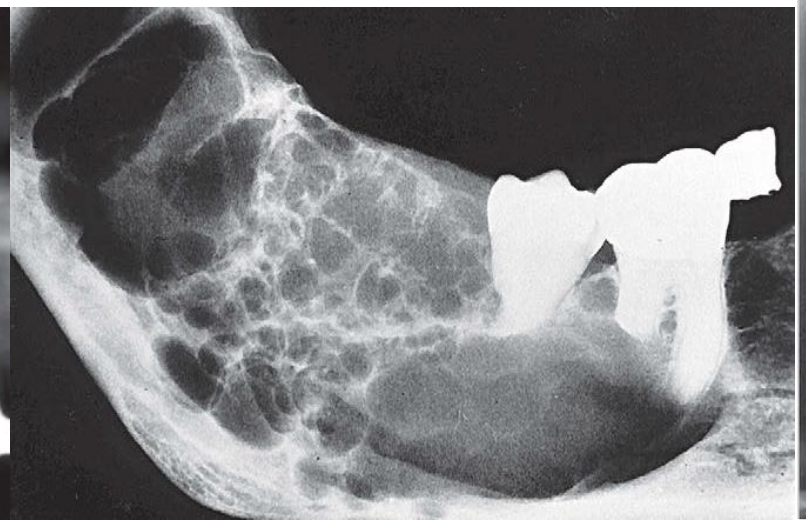
Aetiology	Arises from the dental lamina Mutations in the MAPK pathway present in almost 90% of ameloblastomas • BRAFV600E being the most common mutation
Clinical Features	A painless, slow-growing mass that, if untreated, reaches a large size, displaces and loosens teeth, expands and perforates the cortices, may cause paraesthesia and ultimately causes disfigurement and risks adjacent vital structures

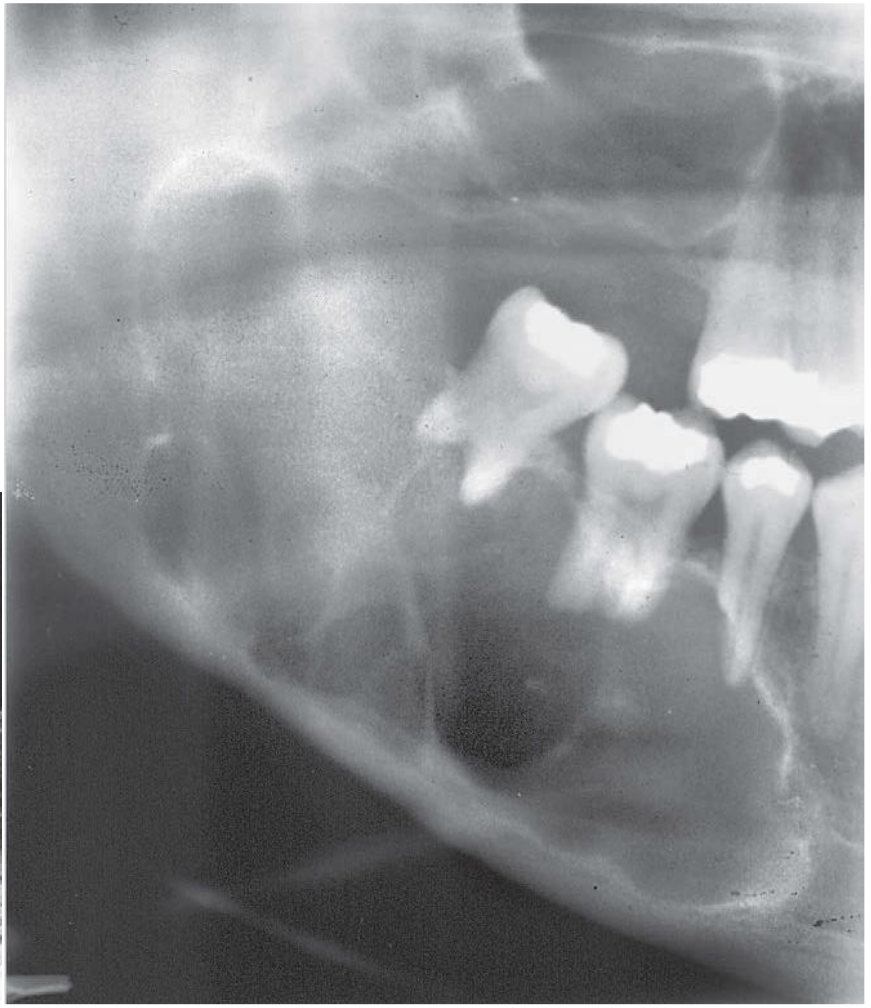


1.1.1 CONVENTIONAL AMELOBLASTOMA

Location	Mandible (87%) Post Md > Ant Md > Post Mx* > Ant Mx * Often extends into the Mx sinus & nasal floor NB: Desmoplastic ameloblastoma has predilection for anterior region of jaws, esp. Mx; Md = Mx NB: Ameloblastomas of the sinonasal tract are rare
Periphery/ Shape	Well-defined, corticated, curved NB: Mx lesions are often more ill-defined



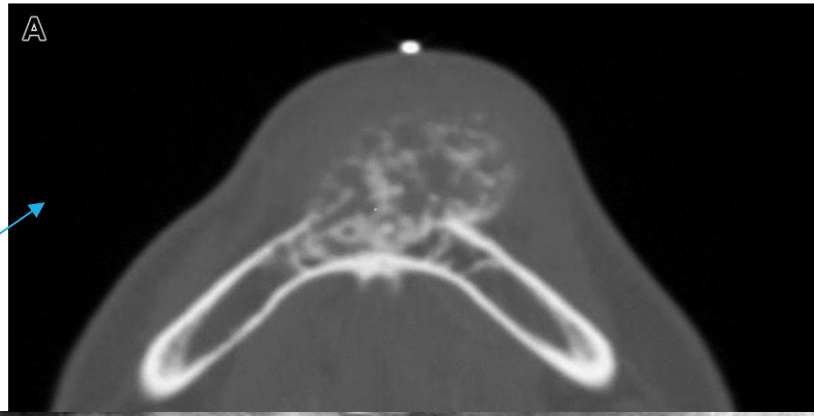


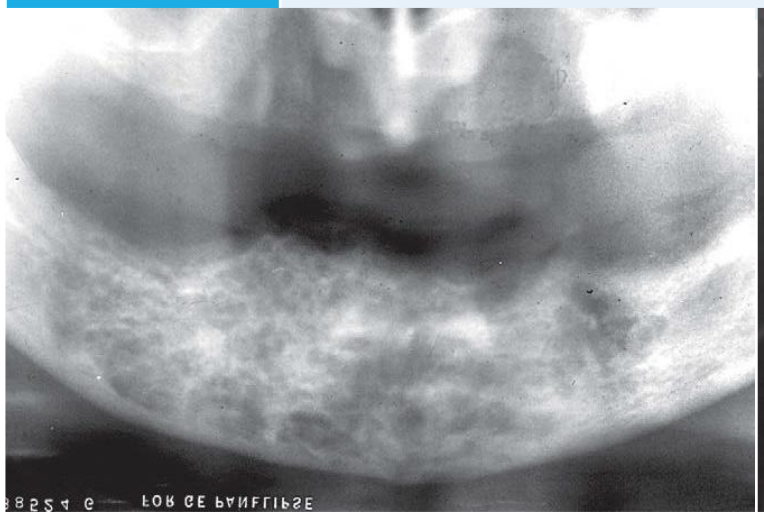


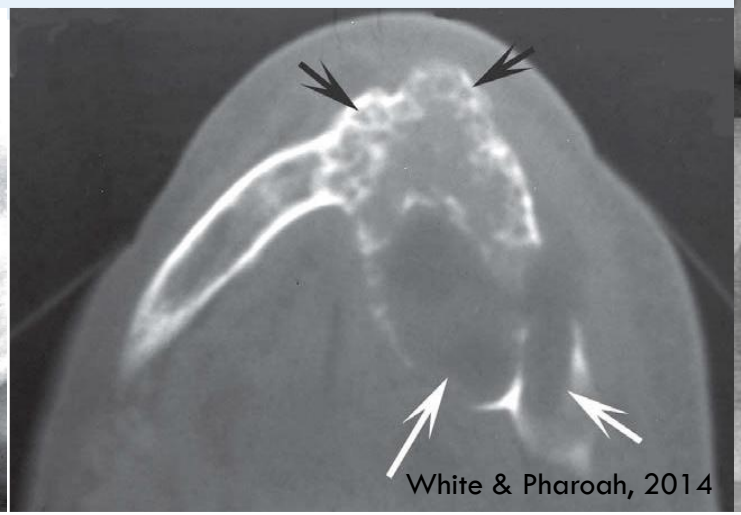
White & Pharoah, 2014

1.1.1 CONVENTIONAL AMELOBLASTOMA

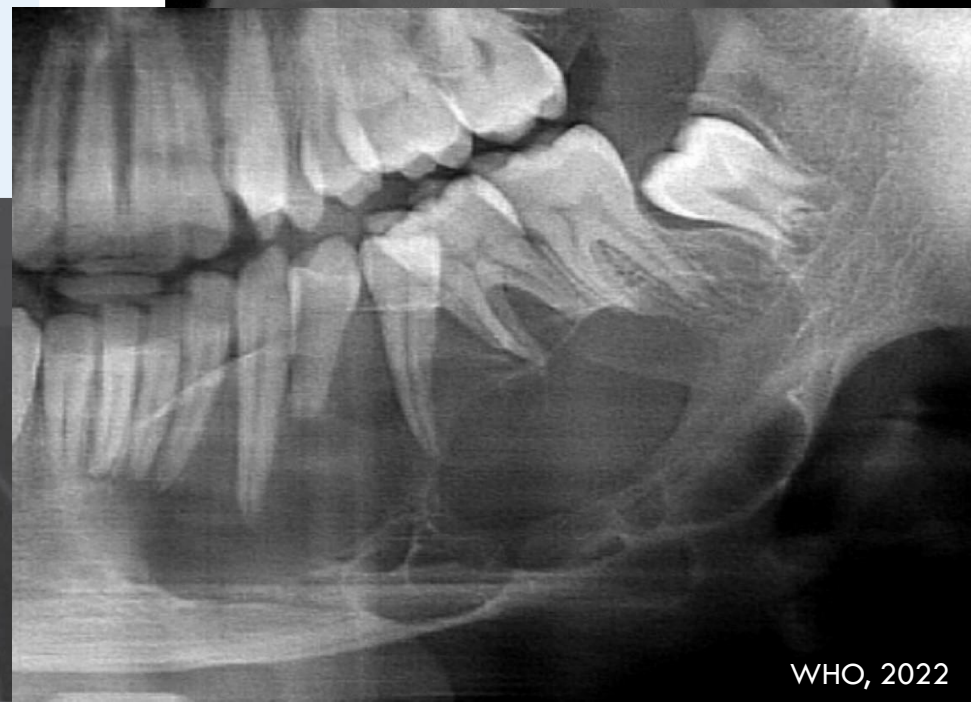
Internal Features	Multilocular soap-bubble or honeycomb radiolucency (most common) • Coarse, curved septae originating from normal bone trapped between cystic components • Locule size is smaller for anterior lesions and larger for posterior lesions Unilocular appearance is less common Desmoplastic AM may produce fine honeycomb mixed RL appearance resembling a fibro-osseous lesion
Surrounding Features	Buccal/lingual expansion ("eggshell") +/- perforation Straight edge blunting root resorption Tooth displacement (inc. apically) May be associated with UE/impacted tooth (18%)







White & Pharoah, 2014



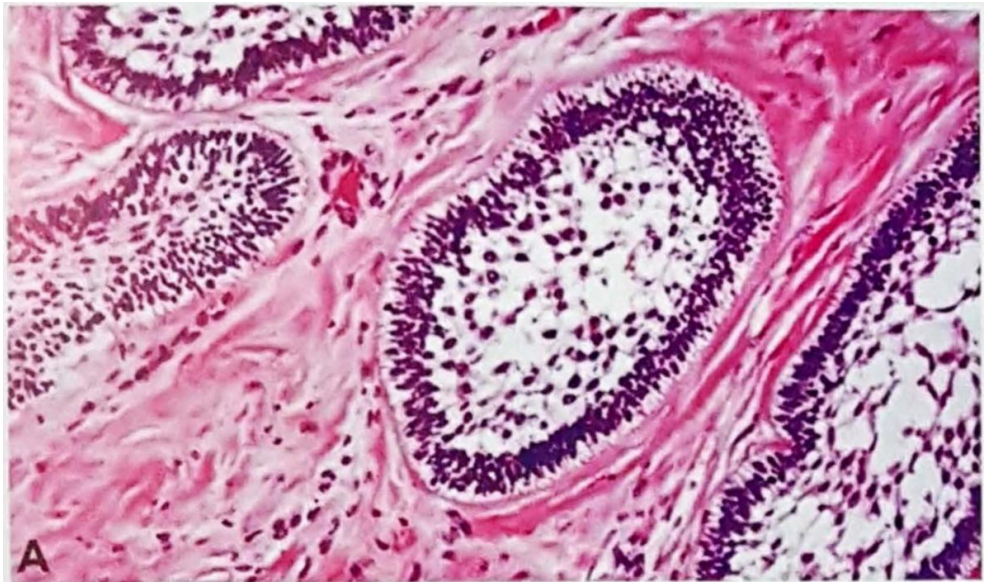
WHO, 2022

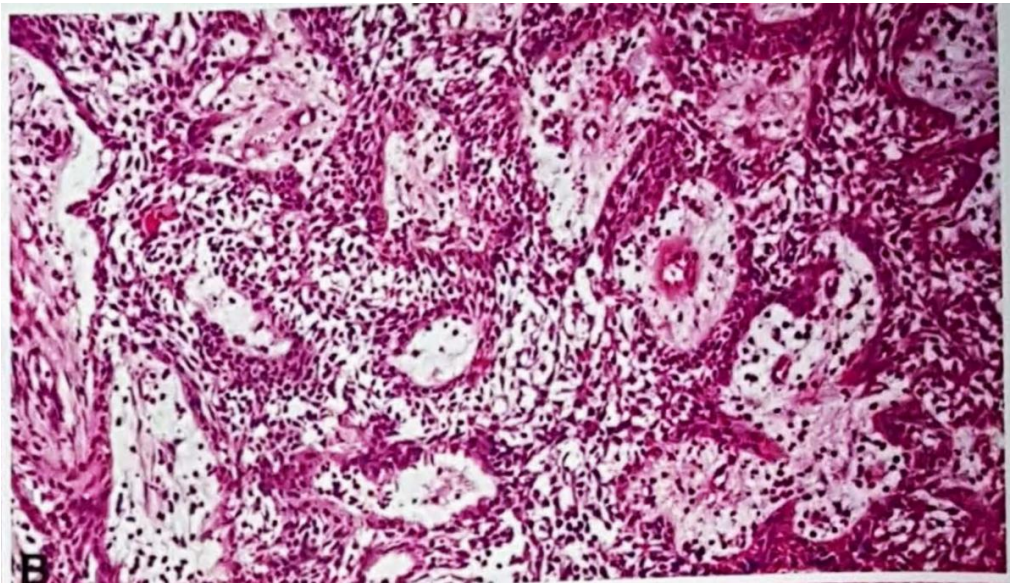
1.1.1 CONVENTIONAL AMELOBLASTOMA

DIFFERENTIAL DIAGNOSIS

Dentigerous cyst	Small unilocular AM located around crown of UE tooth may be indistinguishable
Odontogenic keratocyst	Tends to grow along bone without marked expansion May also have curved septa (uncommon)
Giant cell granuloma	**Younger** age group (unless brown tumour related to hyperparathyroidism) More granular or wispy, **ill-defined septa**
Odontogenic myxoma	Similar septa, but usually have 1-2 thin, sharp, **straight septa** Tend to grow along bone & **less expansile** than ameloblastoma
Ossifying fibroma	Septa are usually wide, granular, **ill-defined** Often there are small, irregular trabeculae
Aneurysmal bone cyst	**Fine internal septa** Typically **extremely expansive**

1.1.1 CONVENTIONAL AMELOBLASTOMA





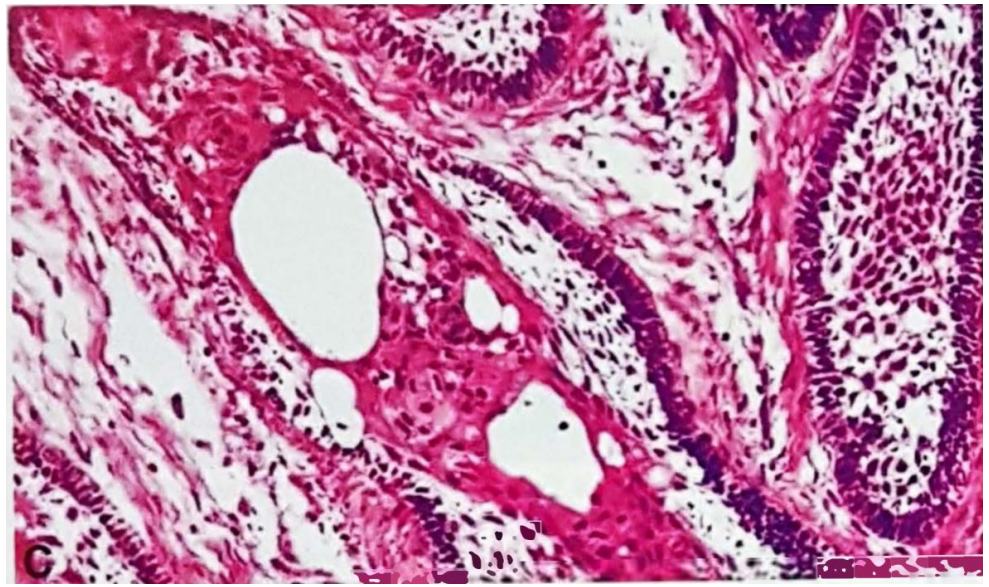
Follicular type (most common)

- Islands of odontogenic epithelium with **columnar peripheral cells**
- **Hyperchromatic nuclei** with **palisading pattern & reverse polarity**
- Reminiscent of the enamel organ (ameloblast-like peripheral cells with a central core of akin to stellate reticulum which often undergoes **cystic degeneration**)
- Connective tissue is moderate to highly collagenised

Plexiform type

- Strands & cords of ameloblastomatous epithelium that form anastomoses
- Peripheral cells: less pronounced
- Inconspicuous stellate reticulum
- Connective tissue: loose & often undergoes cystic changes

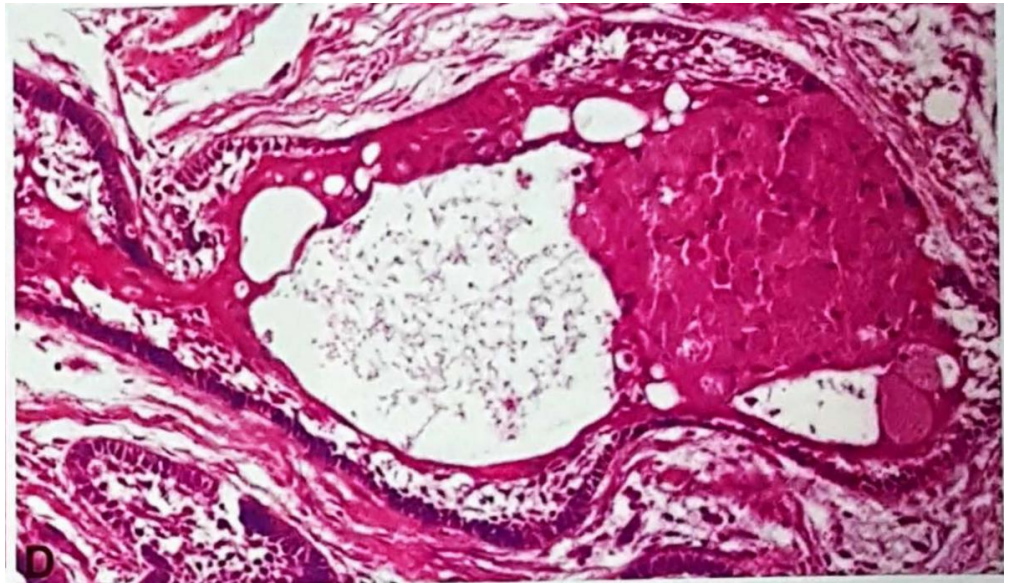
1.1.1 CONVENTIONAL AMELOBLASTOMA



Acanthomatous type

- Squamous metaplasia in stellate reticulum-like central areas
- May have keratin pearl formation

Other types: Basaloid, desmoplastic, clear cell
Mixed patterns are seen frequently
Infiltration into bony trabeculae is commonly seen



Granular type

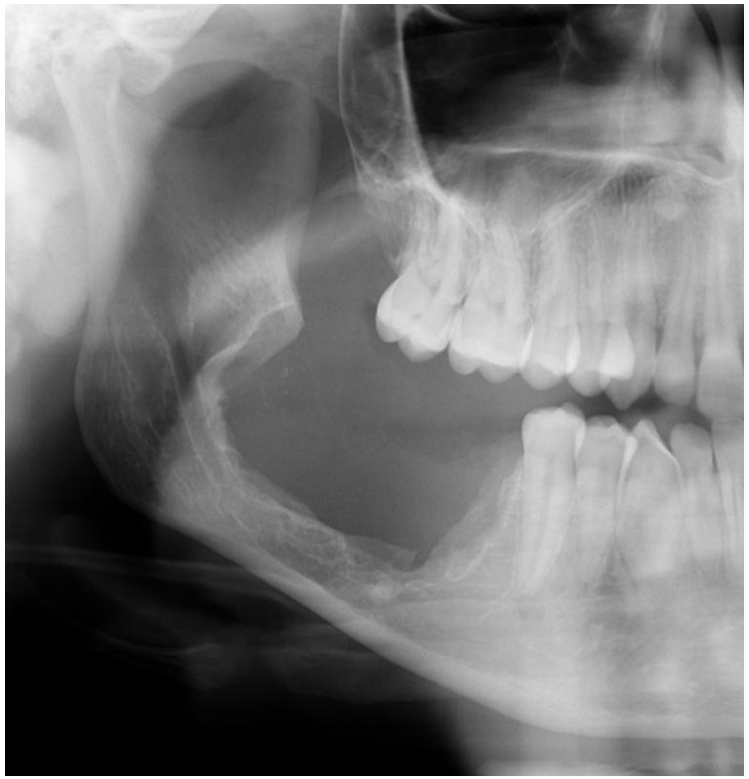
- Granular change in the stellate reticulum-like central areas

No clinical significance is associated with these histological patterns

1.1.1 CONVENTIONAL AMELOBLASTOMA

MANAGEMENT

- Complete excision with negative margins, irrespective of histopathological subtype
  - This may involve a segmental resection, mandibulectomy or maxillectomy, depending on lesion size
  - Requires removal of bone margin of at least 10mm beyond radiographic margin
  - Maxillary lesions are usually treated more aggressively
- BRAF inhibitor treatment has been proposed, alone or in combination with MAPK/ERK kinase (MEK) inhibitors



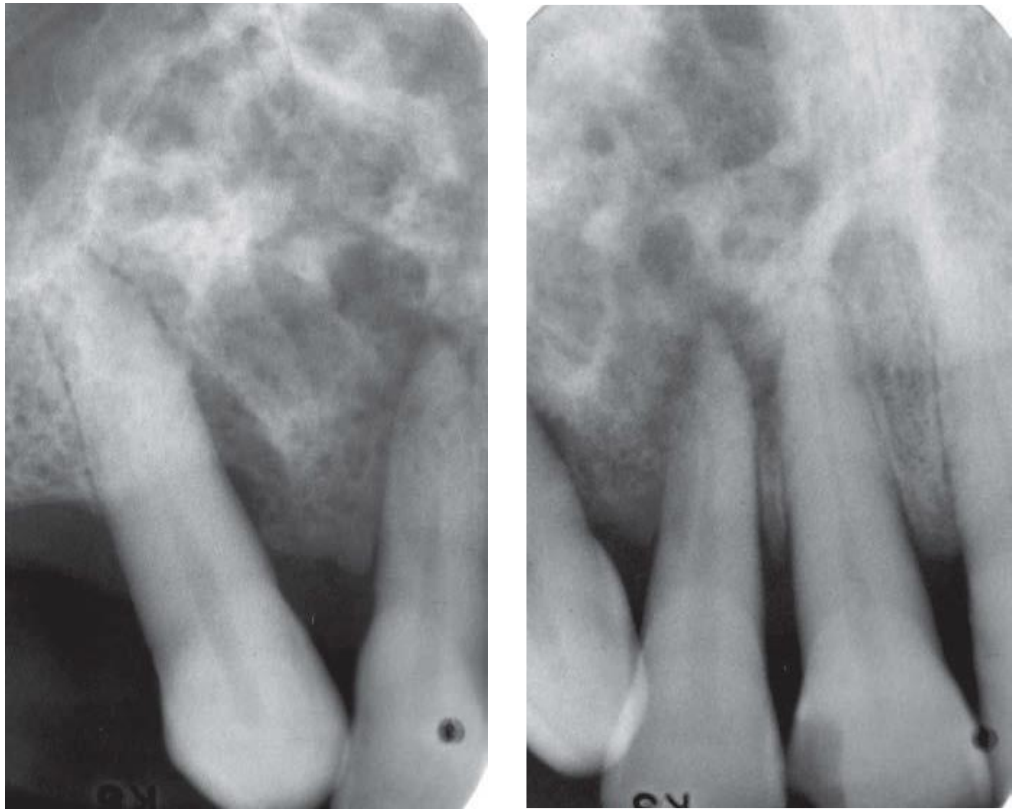
PROGNOSIS/RECURRENCE

- Recurrence rate = approx. 60-80% with conservative surgery
- Long follow up (1-2 decades) is mandatory – consider lifelong

1.1.1 RECURRENT AMELOBLASTOMA

Multiple small cyst-like structures with very coarse sclerotic cortical margins, sometimes separated by normal bone

More aggressive both radiographically & histologically than the original lesion



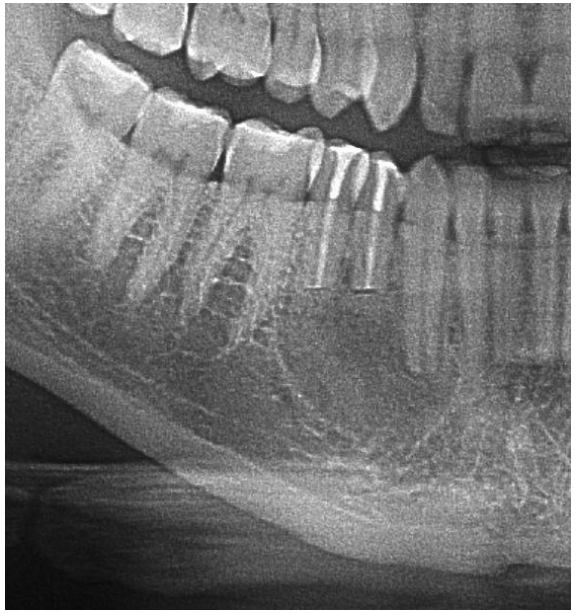
White & Pharoah, 2014

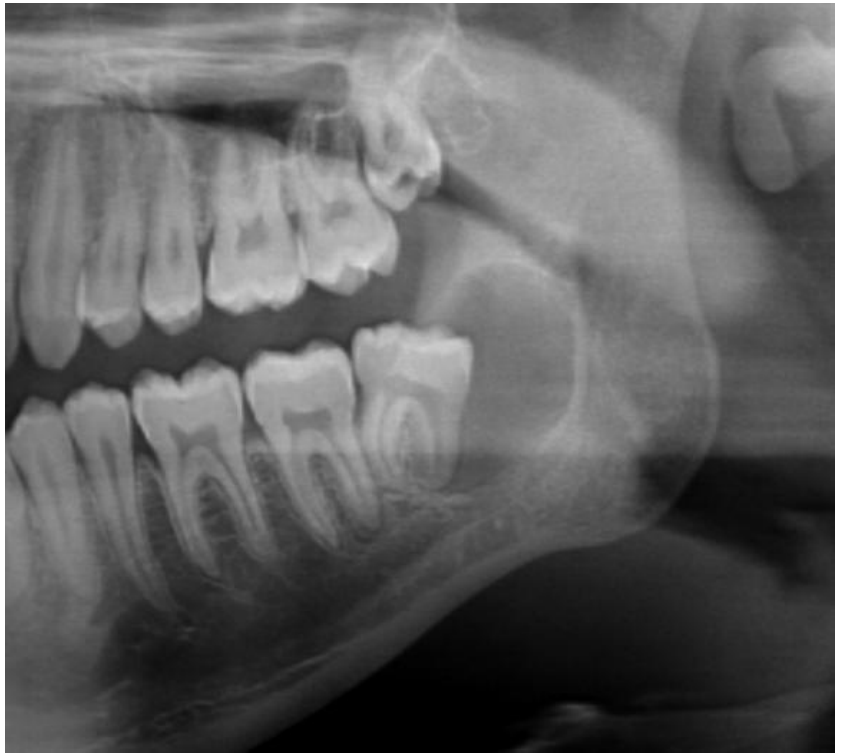
1.1.2 UNICYSTIC AMELOBLASTOMA

Definition	An intraosseous ameloblastoma with a single cyst cavity.
Subtypes	Luminal; Intra-luminal; Mural
Prevalence	5-22% of all ameloblastomas
Age	50% are diagnosed in 2nd decade of life Mean age = 16 years for cases associated with impacted tooth = 35 years in absence of impacted tooth Range = 1-79 years
Gender	Slight M>F But F more likely to have UAM not associated with impacted tooth
Aetiology	Dysregulated MAPK signaling pathways, with BRAF p.V600E the most common activating mutation in all subtypes
Clinical features	An asymptomatic jaw swelling

1.1.2 UNICYSTIC AMELOBLASTOMA

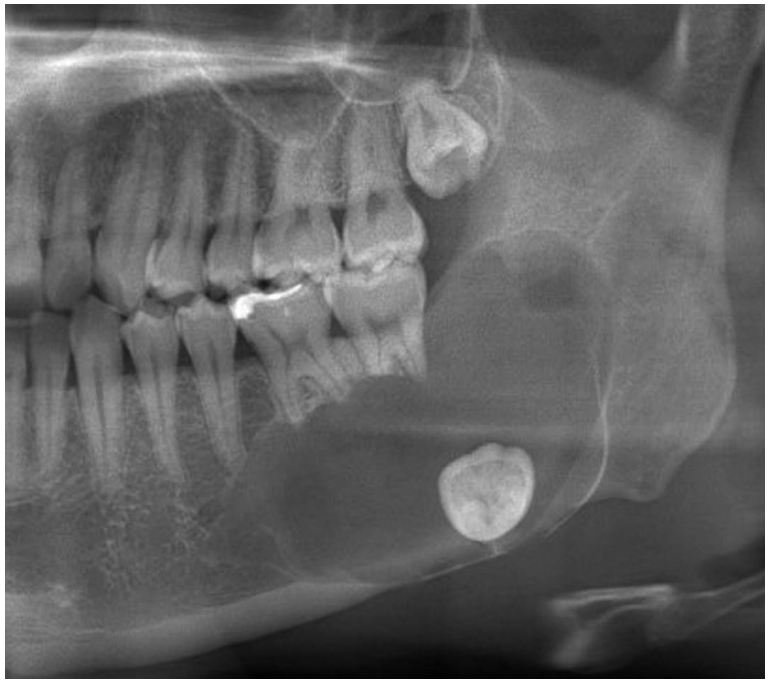
Location	Md: 3rd molar & ascending ramus > body > symphysis Most Mx cases occur in posterior areas May also be found in inter-radicular or periapical locations & edentulous areas
Periphery/ Shape	Well-defined, corticated Scalloped outline (for cases unrelated to an impacted tooth)





1.1.2 UNICYSTIC AMELOBLASTOMA

Internal Features	Unilocular radiolucency Often associated with UE tooth (most often Mand 8s)
Surrounding Features	Root resorption Cortical perforation in 1/3 of cases



WHO, 2022

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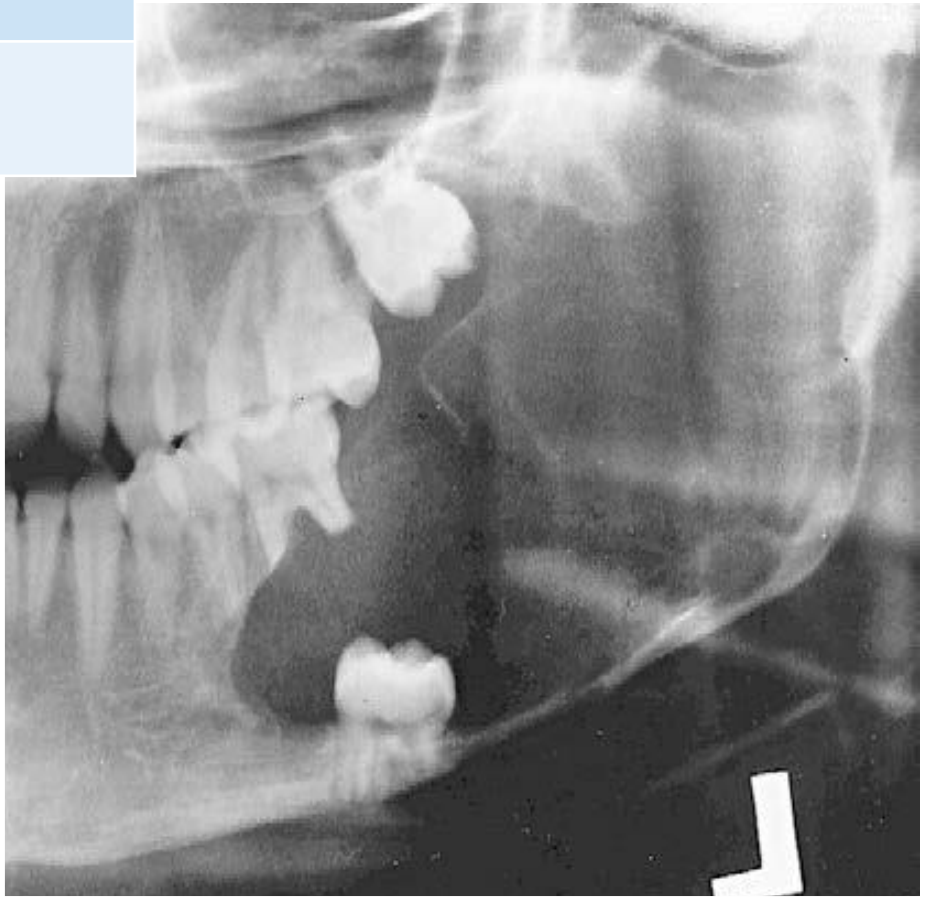


1.1.2 UNICYSTIC AMELOBLASTOMA

HISTOPATHOLOGY

- Single cyst lined by epithelium with palisaded columnar basal layer with reverse polarity and stellate reticulum-like upper layers in most of the lining constitutes the luminal subtype
- Additional plexiform epithelial masses may extend into the lumen only, constituting the intra-luminal type
- Additional islands of epithelium extending into the wall constitute the mural subtype
  - 50-66% of UAM may have a mural component → behave more aggressively like conventional ameloblastomas
- Extensive sampling is required for accurate definition of subtype
- Similar but focal changes may be seen in dentigerous and radicular cysts, especially in areas of inflammation

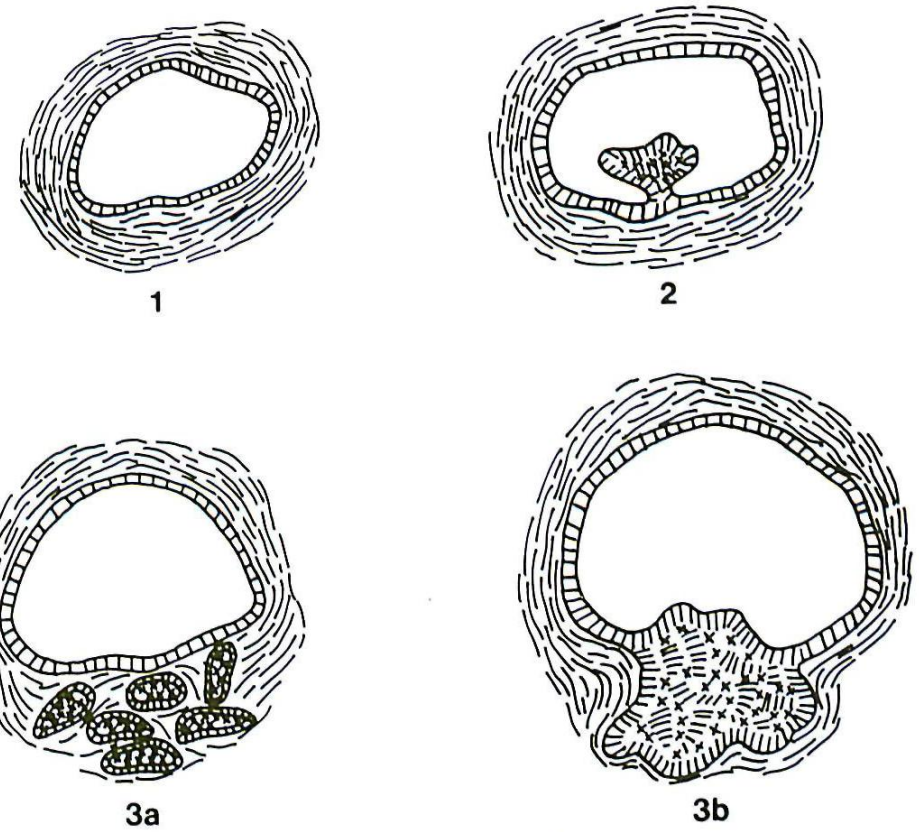


Fig. 1. Histologic types 1. Group 1 – cyst lined by variable, often nondescript, epithelium with no infiltration into fibrous cyst wall. 2. Group 2 – cyst showing intraluminal plexiform epithelial proliferation with no infiltration. 3. Group 3 – cyst with invasion of epithelium into the cyst wall in either (a) a follicular or (b) a plexiform pattern.

1.1.2 UNICYSTIC AMELOBLASTOMA

DIFFERENTIAL DIAGNOSIS

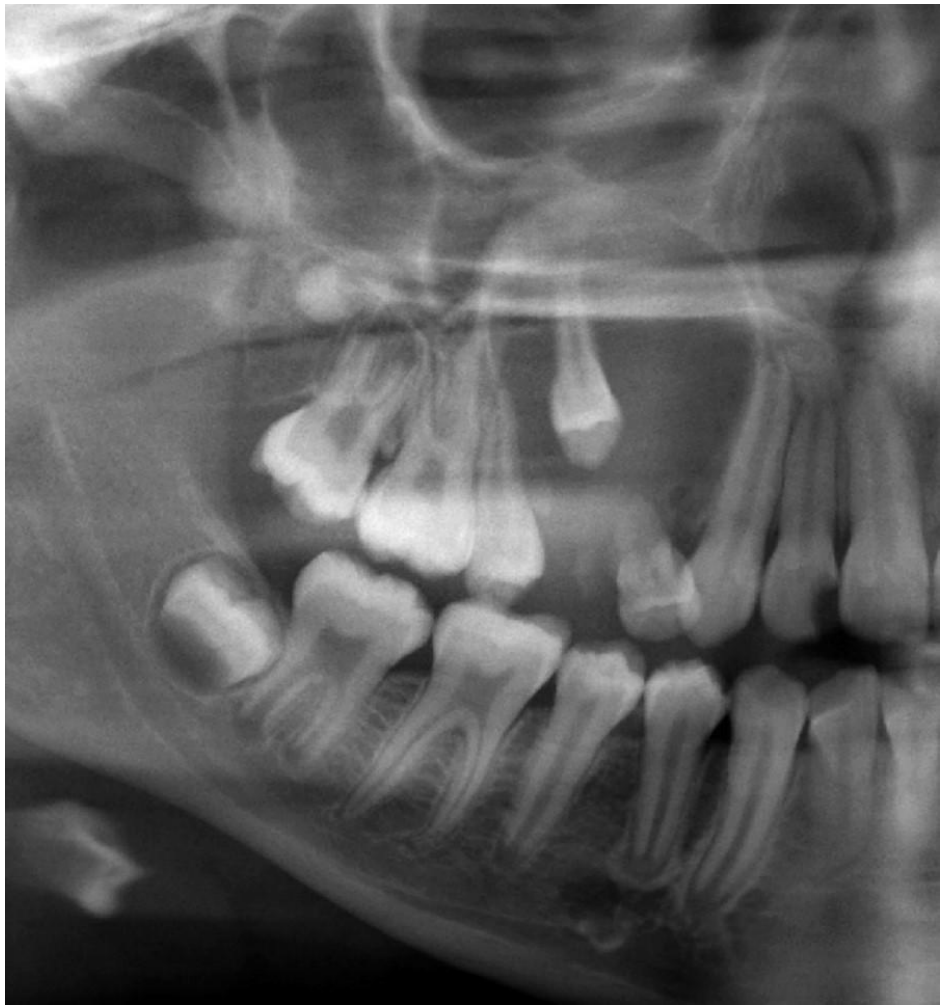
- May be indistinguishable from a dentigerous cyst or odontogenic keratocyst
- Histopathologic correlation required for definitive diagnosis

MANAGEMENT/PROGNOSIS/RECURRENCE

- Up to 30% may recur after enucleation, as definitive diagnosis prior to initial treatment is not possible
- Conservative marsupialisation followed by enucleation is proposed for luminal and intraluminal UAM
  - Risk of recurrence so long-term follow up required
- Mural UAM appears to be intermediate between UAM and conventional ameloblastoma
  - Might require consideration of more extensive surgery as for conventional ameloblastoma, depending on size, extent of intra-mural proliferation and radiological findings
  - However, accurate diagnosis often only follows definitive removal, allowing a period of follow up to confirm recurrence before more aggressive treatment.

1.2 ADENOMATOID ODONTOGENIC TUMOUR

Definition	A benign encapsulated epithelial odontogenic tumour that contains rosette or duct-like structures and has an indolent behaviour.
Prevalence	<10% of all odontogenic tumours
Age	Wide age range, but >80% are diagnosed in the 2nd and 3rd decades of life
Gender	F>M; 2:1
Aetiology	• Unknown, although KRAS p.G12V and p.G12R mutations are detected in approximately 70% of sporadic AOTs • Multiple AOTs can occur in Schimmelpenning syndrome
Clinical Features	• Most are asymptomatic • All have limited growth potential • Large AOTs present as bony hard swellings with cortical expansion but not perforation • Peripheral AOTs appear as small gingival nodules



WHO, 2022

1.2 ADENOMATOID ODONTOGENIC TUMOUR

Location	95% intraosseous (Mx>Md, Ant>Post) 75% cases are located around or alongside the crown of an UE permanent tooth, often extending apically past CEJ 5% extraosseous (anterior maxillary gingiva)
Periphery	Well defined, unilocular, symmetric with corticated/sclerotic border

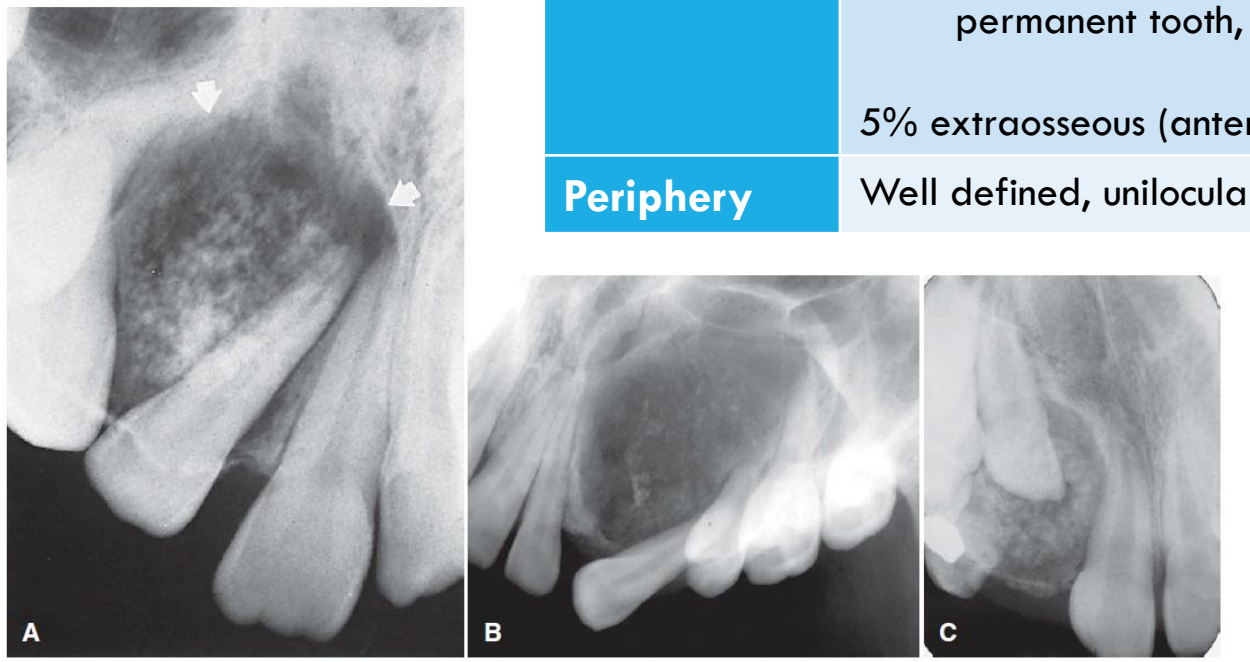


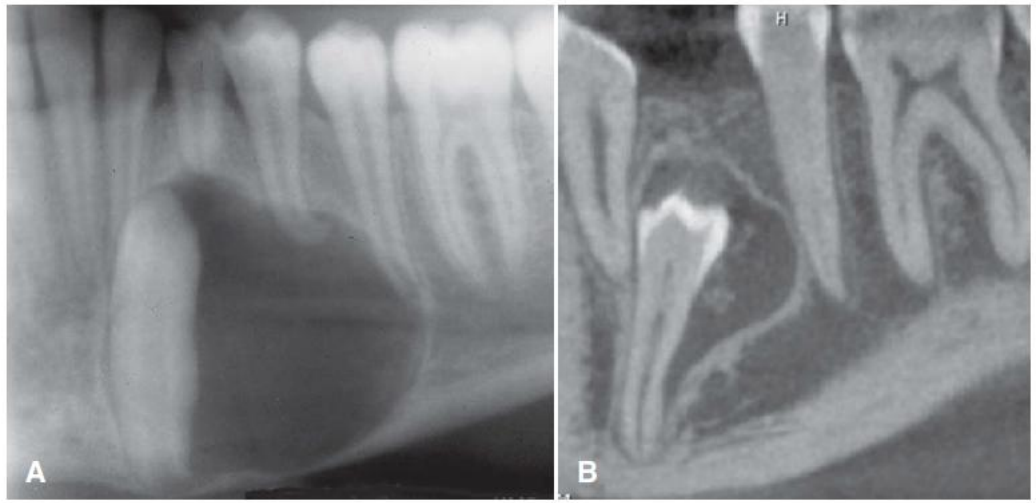
FIGURE 22-27 **A-C,** Intraoral images of adenomatoid odontogenic tumor *(arrows, Fig. A)* within the maxilla with various amounts of calcification, some of which have a pebble-like shape. **(A,** *Courtesy R. Howell, DDS, Morgantown, WV.)*

White & Pharoah, 2014

1.2 ADENOMATOID ODONTOGENIC TUMOUR

Internal Features	Radiolucent, 2/3rd of cases show small radiopaque foci – "snowstorm"
Surrounding Features	Tooth displacement frequent Root resorption rare Cortical expansion (perforation rare)

FIGURE 22-28 Adenomatoid odontogenic tumor within the mandible. **A,** Cropped panoramic image with no apparent internal calcifications. **B,** Cropped cone-beam CT image of a tumor related to the first premolar. Note the pebble-like calcifications distal to the premolar. *(Courtesy of Dr. Milan Madhavji, Toronto, Canada.)*



White & Pharoah, 2014

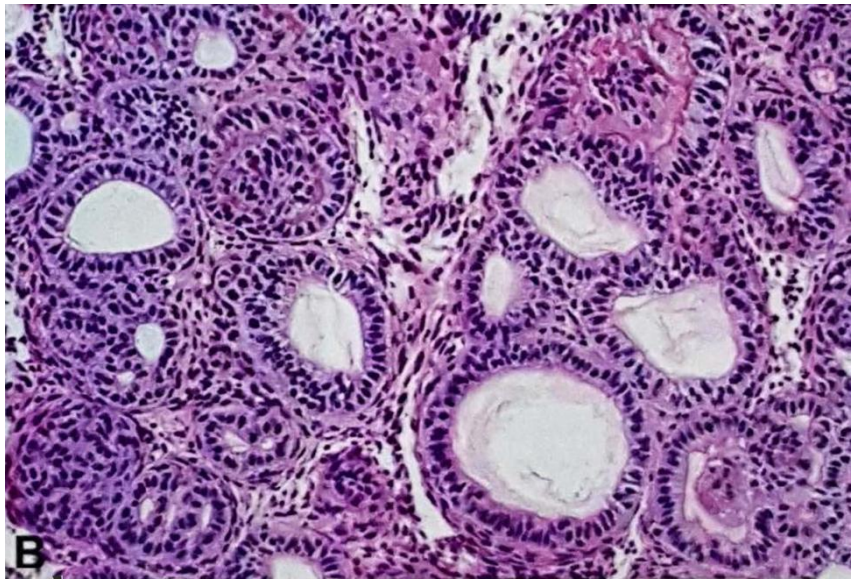
Extraosseous variant:

- Appears as a unilocular radiolucency located between, above, or superimposed upon the roots of erupted teeth
- May cause superficial erosion of the underlying alveolar bone

1.2 ADENOMATOID ODONTOGENIC TUMOUR

HISTOPATHOLOGY

- Encapsulated
- Contain variably sized nodules of spindle, cuboidal and columnar epithelial odontogenic cells with minimal stroma
- Within the nodules are rosette or duct-like structures, which produce the adenomatoid or gland-like appearance
- Eosinophilic amorphous secretory material similar to enamel matrix present between the epithelial cells and rosette-like structures
- +/- Small foci of calcification, dentinoid matrix, & haemorrhage
- +/- cysts lined by non-keratinising stratified epithelium



Characteristic duct-like spaces lined by cuboidal to columnar epithelium with nuclei away from the luminal surface. (WHO, 2017)

1.2 ADENOMATOID ODONTOGENIC TUMOUR

DIFFERENTIAL DIAGNOSIS

- AOT-like areas have been recognized within other odontogenic tumours (inc. odontomas)
- AOT may contain areas resembling calcifying epithelial odontogenic tumour with clear cells
- These histological overlaps makes radiological and clinical correlation essential for definitive diagnosis.

MANAGEMENT

- Enucleation (including capsule)

PROGNOSIS/RECURRENCE

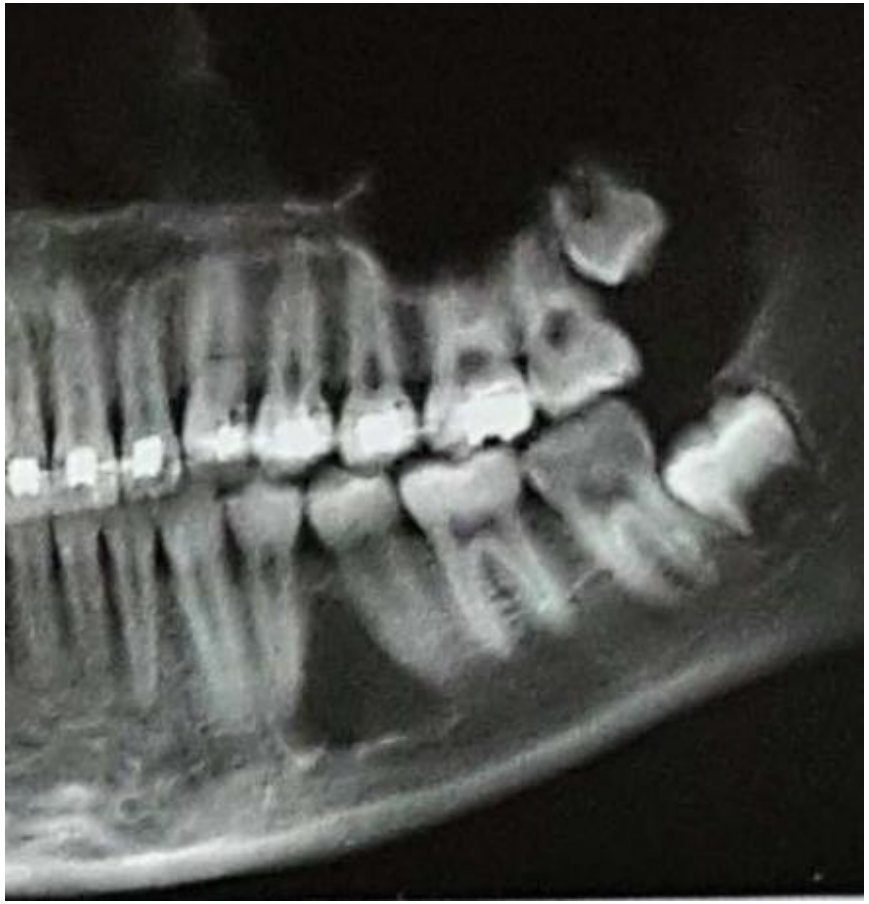
- Almost no risk of recurrence following conservative enucleation.

1.3 SQUAMOUS ODONTOGENIC TUMOUR

Definition	A benign, slow-growing epithelial odontogenic tumour with squamous differentiation.
Prevalence	Rare
Age	Wide age distribution (mean age = 34.8 years) Multiple SOTs: younger age group with marked predilection for African Americans
Gender	M=F
Aetiology	Unknown but a familial incidence has been reported Thought to originate from rests of Malassez, gingival surface epithelium or remnants of the dental lamina.
Clinical Features	Asymptomatic swelling Minority are associated with pain, tenderness, mobility of teeth or bone expansion.

1.3 SQUAMOUS ODONTOGENIC TUMOUR

Location	Tooth bearing parts of the jaw (Mx = Md) • Ant Mx & Post Md more common Multifocal or extraosseous lesions have been reported (rare)
Periphery/ Shape	Triangular or semi-circular Unilocular, well-defined +/- cortication Multilocularity reported for larger and extensive lesions
Internal Features	Radiolucent
Surrounding Features	Most lesions show continuity with 1+ tooth roots Root displacement common. Root resorption rare. Peripheral variant may cause saucerisation (due to pressure, not infiltration)

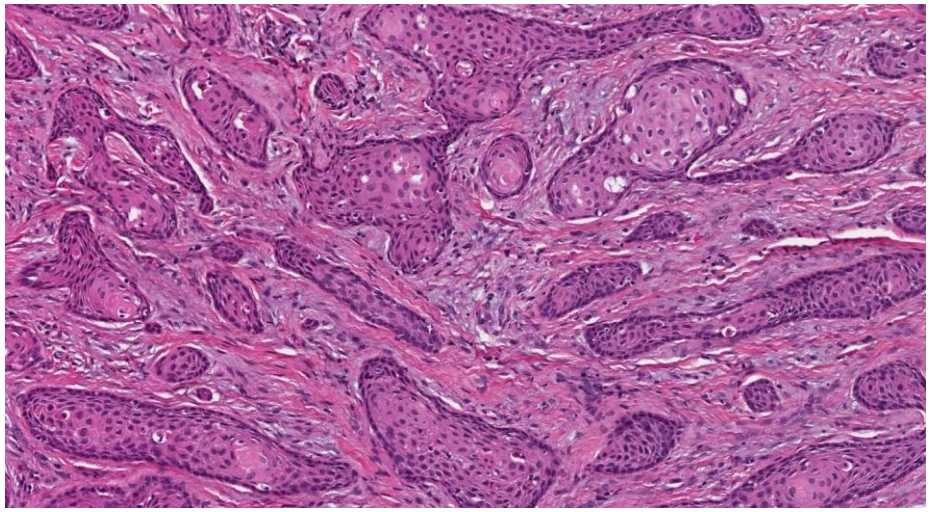


WHO, 2017

1.3 SQUAMOUS ODONTOGENIC TUMOUR

HISTOPATHOLOGY

- Islands of bland well-differentiated squamous epithelium in a fibrous or myxoid stroma
  - Peripheral cells of the islands are flat to cuboidal with very infrequent mitoses
  - Central cells have a tendency for cystic degeneration, individual cell keratinization, and calcification
- +/- Mucous metaplasia, sebaceous differentiation, and ghost cell-like areas



Islands of benign, squamous epithelium in a mature fibrous stroma. The peripheral cells are flattened, and central cells show areas of microcystic degeneration and individual cell keratinization. (WHO, 2022)

1.3 SQUAMOUS ODONTOGENIC TUMOUR

DIFFERENTIAL DIAGNOSIS

- Ameloblastoma – acanthomatous or desmoplastic variants
  - SOT lacks peripheral columnar cells & palisading nuclei
- Well-differentiated squamous cell carcinoma
  - SOT lacks peripheral palisading with reverse nuclear polarity, and the cytological features are bland (i.e. no dysplasia)
- SOT-like proliferation of epithelium may develop in the lining of odontogenic cysts

MANAGEMENT

- Enucleation

PROGNOSIS/RECURRENCE

- Unifocal SOT rarely recurs after conservative surgery.
- Multifocal or recurring SOT, especially of the maxilla, may require a more radical approach.

1.4 CALCIFYING EPITHELIAL ODONTOGENIC TUMOUR

Definition	A benign epithelial odontogenic tumour characterised by amyloid, which may calcify.
Synonyms	Pindborg tumour
Prevalence	Rare
Age	Wide age range (8-83 years) Maximum incidence in the 4th decade
Gender	M=F
Aetiology	Unknown (mutations in tumour suppressor genes and oncogenes have been reported)
Clinical Features	Asymptomatic Larger lesions cause slow growing, localised expansion of the jaw + tooth mobility

1.4 CALCIFYING EPITHELIAL ODONTOGENIC TUMOUR

Location	85% intraosseous, tooth-bearing areas of jaws (Md>Mx) Md body most common 50% are associated with UE teeth 15% extraosseous
Periphery/ Shape	Unilocular (70%) > Multilocular (30%) Variable definition and cortication

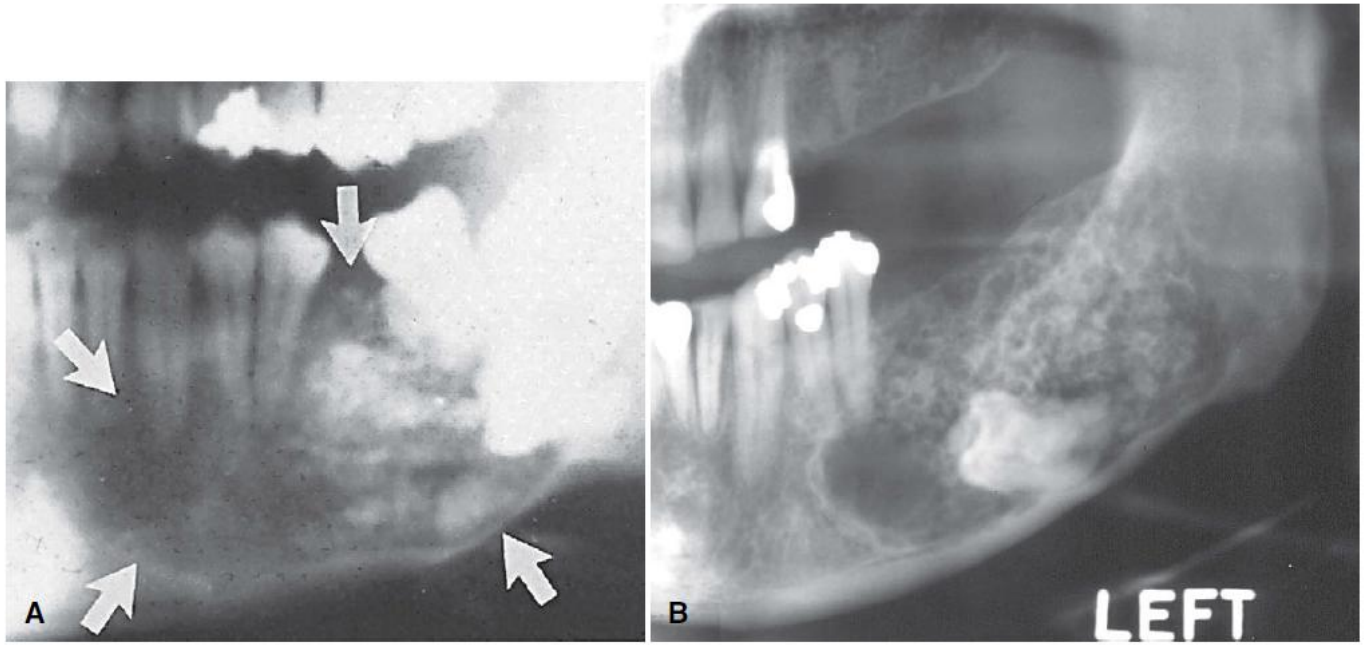
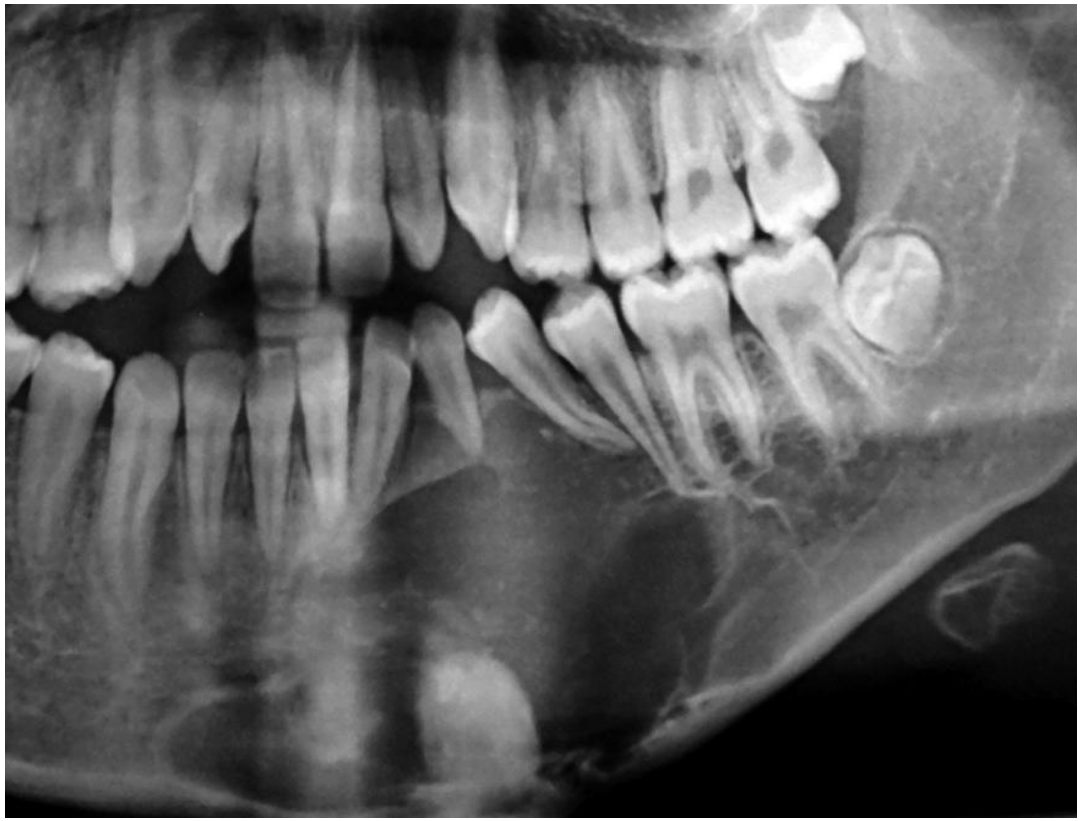


FIGURE 22-19 **A,** Calcifying odontogenic tumor or Pindborg tumor *(arrows)*. **B,** The tumor appears as a mixed radiolucent-radiopaque lesion associated with an unerupted tooth. *(**A,** Courtesy M. Gornitsky, DDS, Montreal, Canada. **B,** Courtesy Dr. D. Lanigan, University of Saskatchewan.)*

White & Pharoah, 2014

1.4 CALCIFYING EPITHELIAL ODONTOGENIC TUMOUR

Internal Features	75% mixed radiolucent/radiopaque • Classic "driven snow" appearance • May be crescent/donut shaped • ****Radiopacities close to crown of embedded tooth**** • Degree of calcification correlates with age of lesion Remaining 25% CEOTs are radiolucent (but totally radiopaque lesions are also seen)
Surrounding Features	May displace teeth or prevent its eruption Expansion of jaw (without perforation)

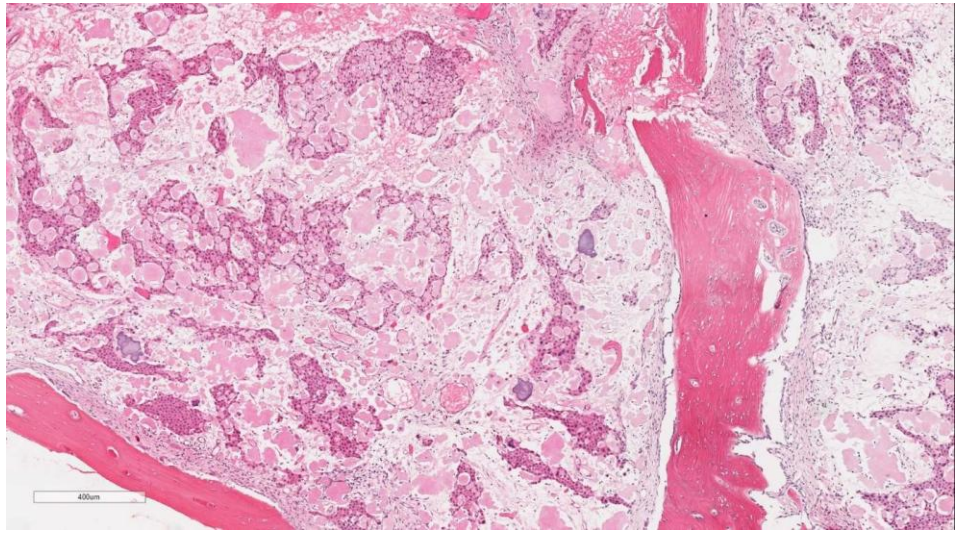


WHO, 2022

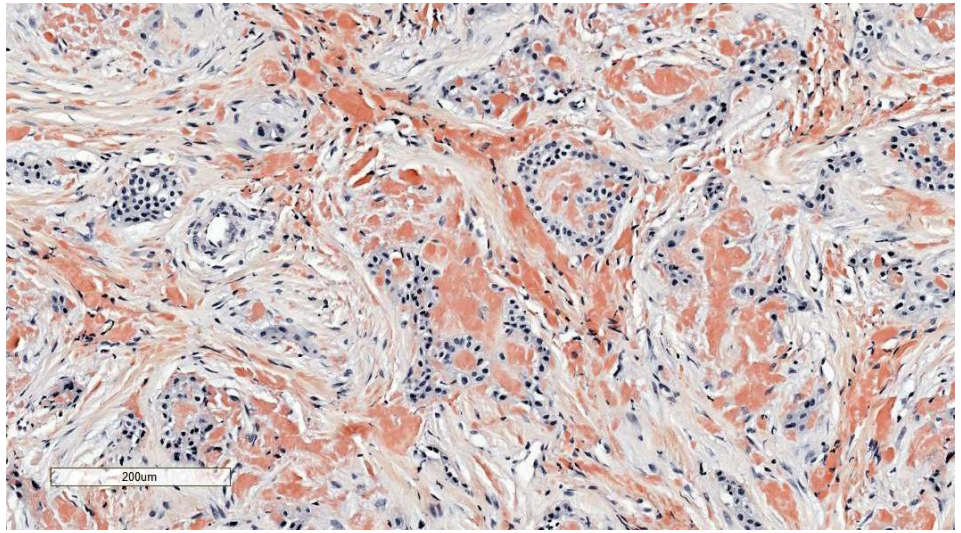
1.4 CALCIFYING EPITHELIAL ODONTOGENIC TUMOUR

HISTOPATHOLOGY

- Variable appearance with 4 subtypes:
  - Clear cell
  - Cystic/microcystic
  - Non-calcifying/Langerhans cell rich
  - AOT subtype
- Comprise sheets, cords or nests of polyhedral epithelial cells with distinct cell borders and prominent intercellular bridges
  - +/- nuclear pleomorphism (mitoses are rare)
- Deposits of amorphous lightly eosinophilic amyloid composed of ameloblast-associated proteins (stains with Congo red)
  - These may calcify forming large masses or small round concentric densely basophilic calcifications with Liesegang rings



Islands of odontogenic epithelium, with amyloid and focal calcification. (WHO, 2022)



Congo Red stain demonstrating the amyloid material. (WHO, 2022)

1.4 CALCIFYING EPITHELIAL ODONTOGENIC TUMOUR

DIFFERENTIAL DIAGNOSIS

- May extend into adjacent medullary spaces in an infiltrative pattern worrying for malignancy
- Foci resembling CEOT may be found in odontomas and the follicles of unerupted teeth

MANAGEMENT

- Local surgical removal

PROGNOSIS/RECURRENCE

- Recurrence rate = approx. 13%
  - Varies with treatment modality, being much higher in those treated by curettage
  - No histological parameters predict recurrence
- Malignant transformation rare

PART 2: BENIGN EPITHELIAL & MESENCHYMAL ODONTOGENIC TUMOURS

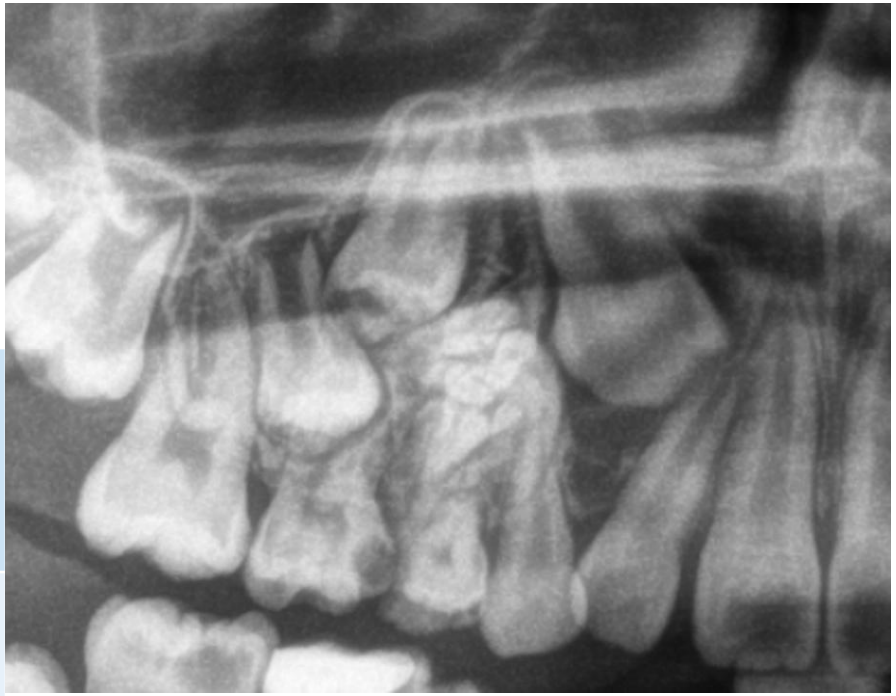
1. Odontoma
2. Ameloblastic fibroma
3. Primordial odontogenic tumour
4. Dentinogenic ghost cell tumour

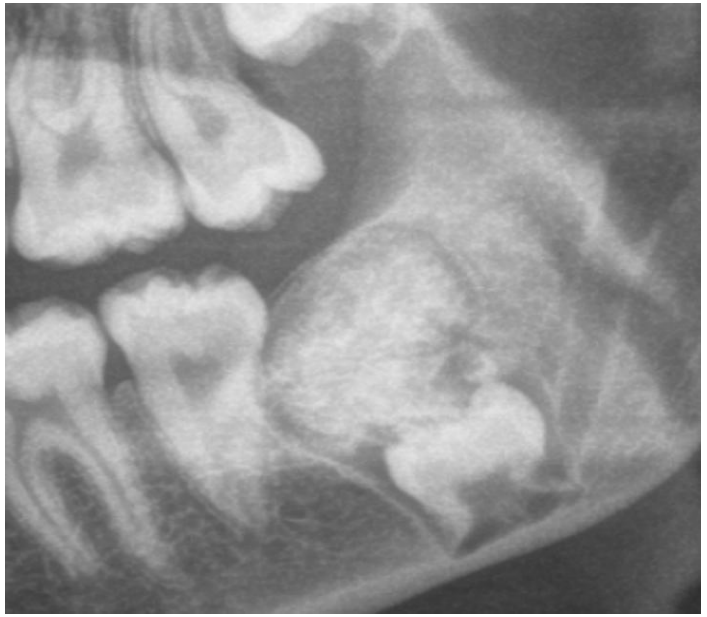
2.1 ODONTOMA

Definition	Mixed odontogenic hamartomas that mature from soft tissue to predominantly dental hard tissues with a small amount of residual odontogenic epithelium and ectomesenchyme
Subtypes	Compound; Complex
Prevalence	Most common odontogenic tumour (some consider it to be a hamartoma)
Age	2nd and 3rd decades
Gender	M=F
Aetiology	Unknown, genetic mutation of tooth germ is possible factor
Clinical Features	Frequently associated with UE tooth Asymptomatic (unless secondarily infected)

2.1 ODONTOMA

Location	Any tooth-bearing area; typically between roots or superior to crowns *Compound:* Mx ant *Complex:* Md post > Mx ant May be multiple
Periphery/ Shape	Well-defined with lucent band and adjacent corticated periphery Typically 10-30mm (up to 80mm has been reported)
Internal Features	*Early stage:* RL with focal areas of calcification *Compound:* numerous tooth-like structures (denticles) *Complex:* disorganised mass of calcified tissue
Surrounding Features	Associated with an unerupted tooth Expansion of jaw (larger lesions)





(WHO, 2022)

2.1 ODONTOMA

HISTOPATHOLOGY

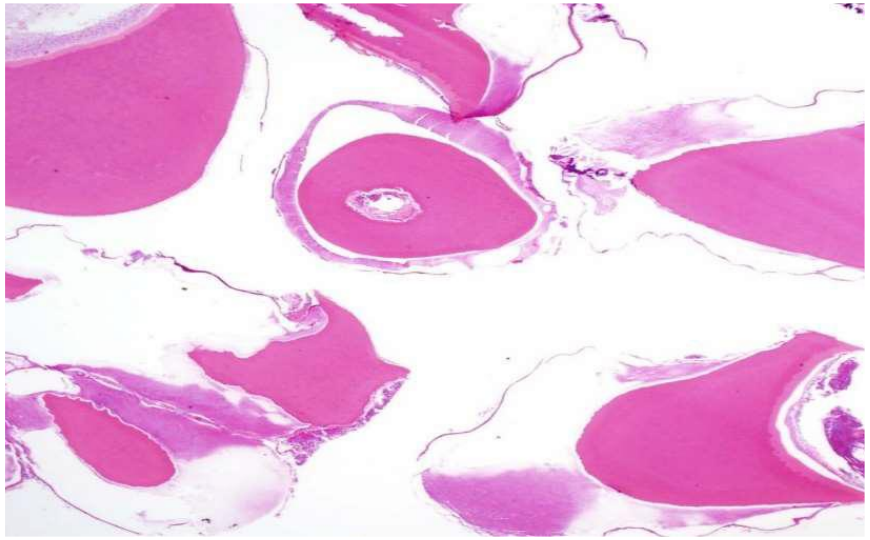
- *Compound odontoma:* contains multiple rudimentary teeth exhibiting dentin and enamel matrix. Dentine in odontomas usually shows some irregularity of tubule structure.
- *Complex odontoma:* consists of a disorganized mass of mature tubular dentine intermixed with rounded zones of enamel matrix where enamel has been lost on decalcification, with areas of dental pulp and cementum.

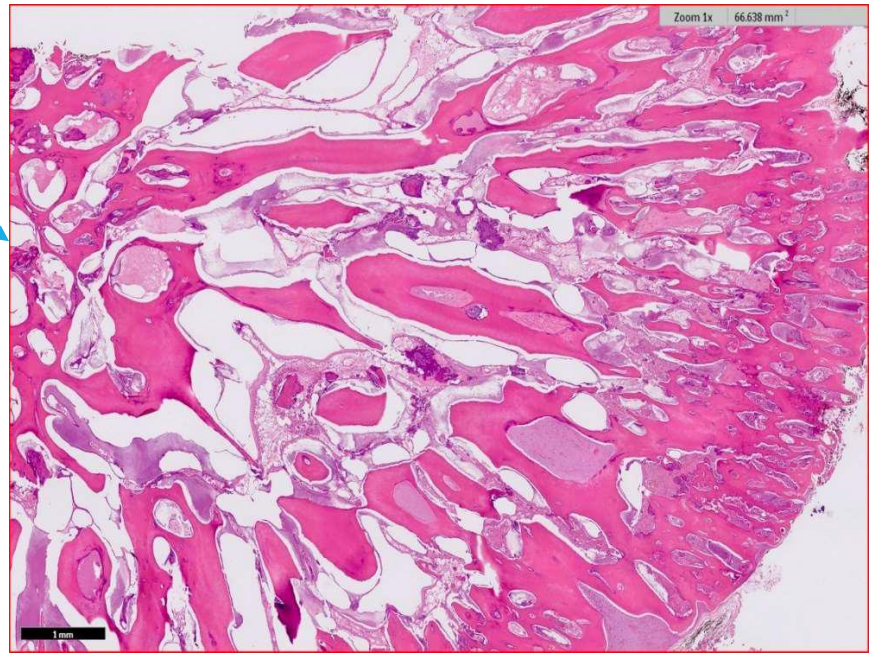
MANAGEMENT

- Local surgical removal

PROGNOSIS/RECURRENCE

- Do not recur (unless incomplete removal)





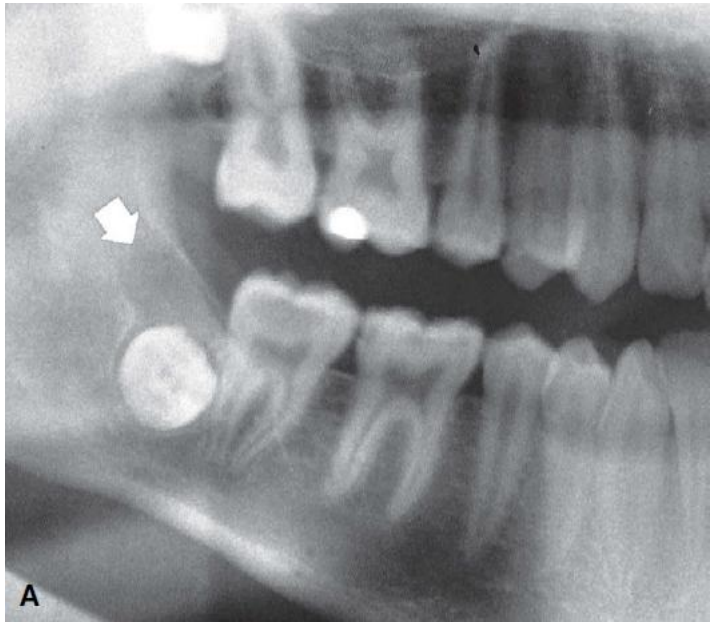
WHO, 2022

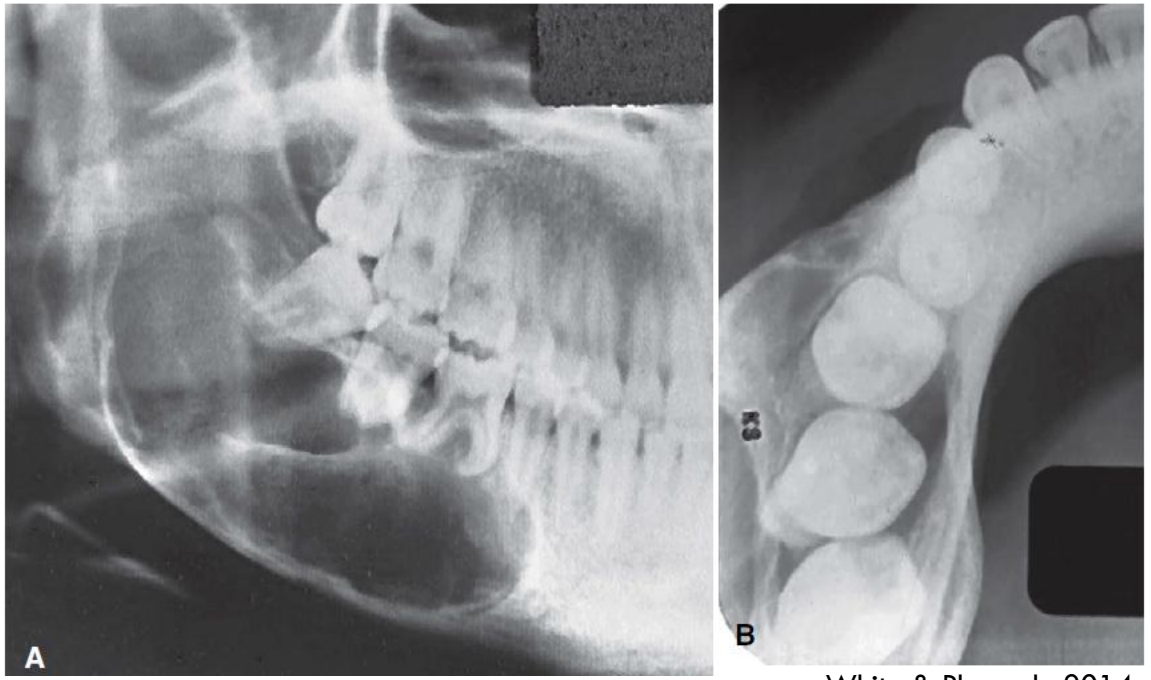
2.2 AMELOBLASTIC FIBROMA (AF)

Definition	A rare, benign, mixed odontogenic tumour comprising cellular mesenchymal tissue resembling dental papilla and an epithelial component resembling early developing enamel organ, without dental hard tissue or matrix.
Prevalence	2% of all odontogenic tumours
Age	Mean = 15 years Range = 7 weeks – 61 years 80% occur <22 years (before end of odontogenesis)
Gender	M>F; 1.4:1
Aetiology	Unclear, but BRAF p.V600E mutations in 46% of cases
Clinical Features	Asymptomatic, slow-growing jawbone expansion (78%)

2.2 AMELOBLASTIC FIBROMA

Location	Mand>Max; 3:1 Post (82%) > Ant (~10%) Common locations: • At crest of alveolar ridge • Follicular relationship with UE tooth (occlusal to tooth) • Where a tooth has failed to develop
Periphery/ Shape	Well-defined Corticated
Internal Features	Unilocular (60%) – totally radiolucent Multilocular (large) – indistinct curved septa 80% associated with an impacted tooth (usually 6s or 7s)
Surrounding Features	Tooth displacement (resorption uncommon) • Typically in apical direction • Tooth may be inhibited from eruption Jaw expansion (perforation uncommon)





White & Pharoah, 2014

2.2 AMELOBLASTIC FIBROMA

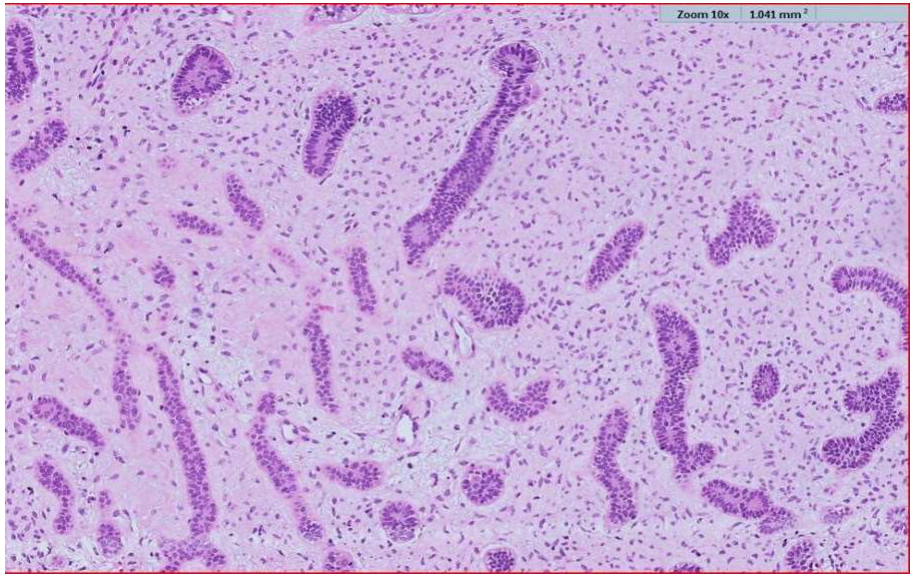
DIFFERENTIAL DIAGNOSIS

Hyperplastic follicle	Difficult to differentiate from small AF
Dentigerous cyst	Difficult to differentiate from small AF Less likely to be DC if margins not at CEJ or at root surface within 2-3mm of CEJ
OKC	Minimal expansion for size, internally lucent, only a few septa when large
Ameloblastoma	Coarser septa than AF (AF septa are infrequent & fine) Substantial root resorption is a feature Older age group
Central giant cell granuloma	Epicentre anterior to 6s Characteristically granular & ill-defined septa Tendency for substantial root resorption Typically lobulated expansion
Aneurysmal bone cyst	Extremely expansile, unless small; MRI shows fluid-fluid levels
Odontogenic myxoma	Straight septa Limited expansion for size Older age group

2.2 AMELOBLASTIC FIBROMA

HISTOPATHOLOGY

- *Mesenchymal component:* myxoid and evenly hypercellular; resembles the dental papilla of the tooth bud
- *Epithelial component:* forms long narrow cords and strands of bilaminar cuboidal to columnar palisaded cells with occasional thickenings
- No dental hard tissue is normally present but extensive sampling may reveal small foci



MANAGEMENT/PROGNOSIS/RECURRENCE

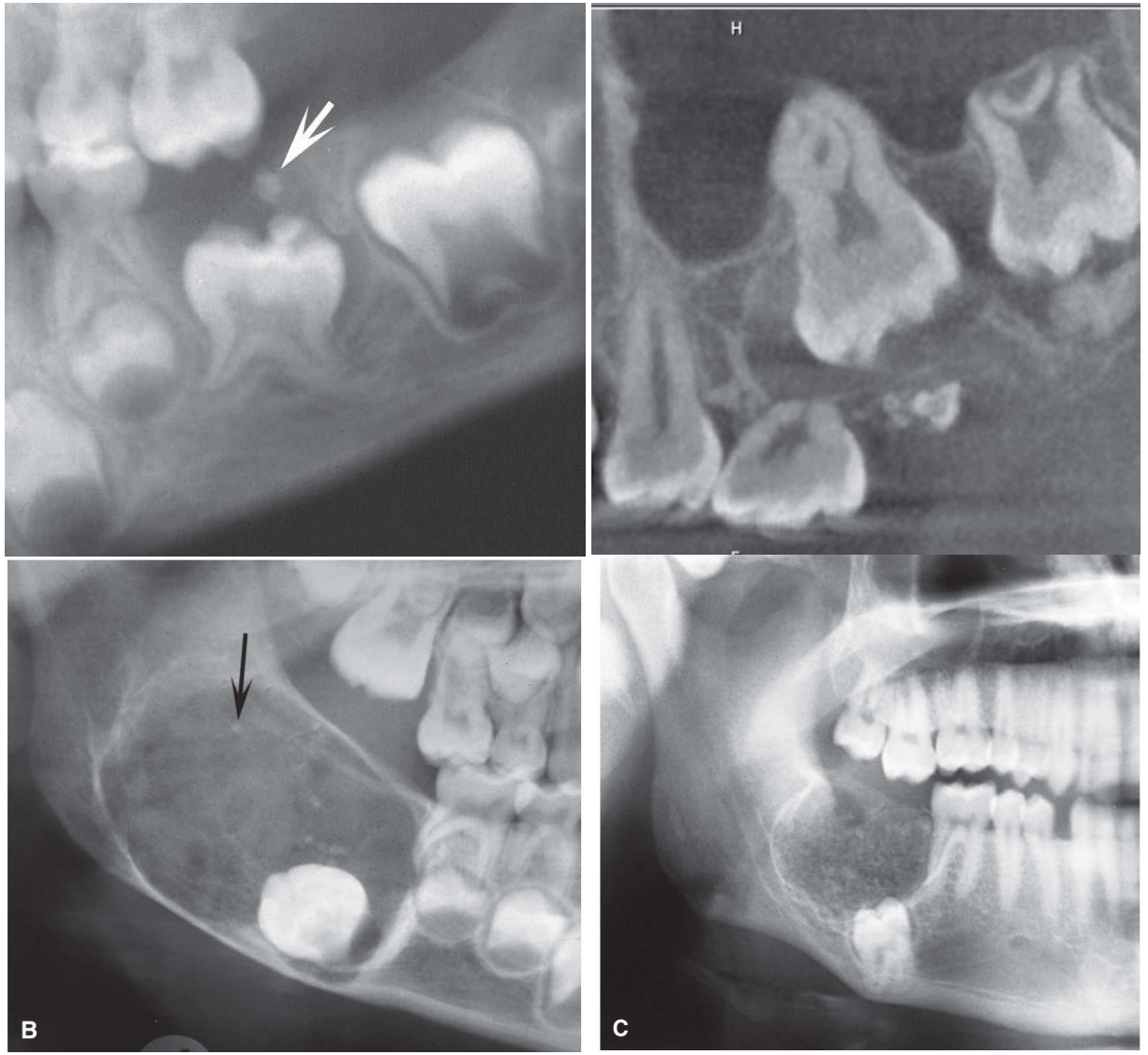
- 19% recurrence with conservative removal
- Extensive, destructive and recurrent tumours should be treated radically.
- Rare sarcomatous transformation

AMELOBLASTIC FIBRO-ODONTOMA (AFO)

Definition	A benign mixed odontogenic tumour involving ectomesenchymal tissues with the presence of enamel & dentine, demonstrating histological features of ameloblastic fibroma & complex odontoma. ~Atlas of Oral & Maxillofacial Radiology, 2017
Age	Identified 1st & 2nd decades of life
Gender	M=F
Clinical features	Slow growing, expansile Often interrupts tooth eruption Often associated with missing tooth

AMELOBLASTIC FIBRO-ODONTOMA

Location	Md > Mx Post > Ant Majority within alveolar process, often approximating alveolar crest Epicentre often occlusal to tooth or towards alveolar crest
Periphery/ Shape	Well defined, corticated
Internal Features	RL/RO Most often associated with an impacted tooth Majority of lesion is RL – with some calcifications May appear like donut (radiopaque enamel-like margin) or tooth-like structures (Small) enlarged follicles with 1-2 discrete RO (Large) More extensive calcified internal structure
Surrounding Features	Tooth displacement Impedes the eruption of involved tooth



AMELOBLASTIC FIBRO-ODONTOMA

DIFFERENTIAL DIAGNOSIS

AF	Difficult to differentiate unless internal calcifications present
Odontoma	AFO calcifications tend to be smaller & more diffuse; often large lucent regions within Compound odontoma: usually Mx ant; AFO not organised to resemble teeth Complex odontoma: usually one central mass of disorganised tissue
AOT/COC	More common in Mx anterior
CEOT	Rare

HISTOPATHOLOGY

- Resembles AF & complex odontoma

MANAGEMENT/PROGNOSIS/RECURRENCE

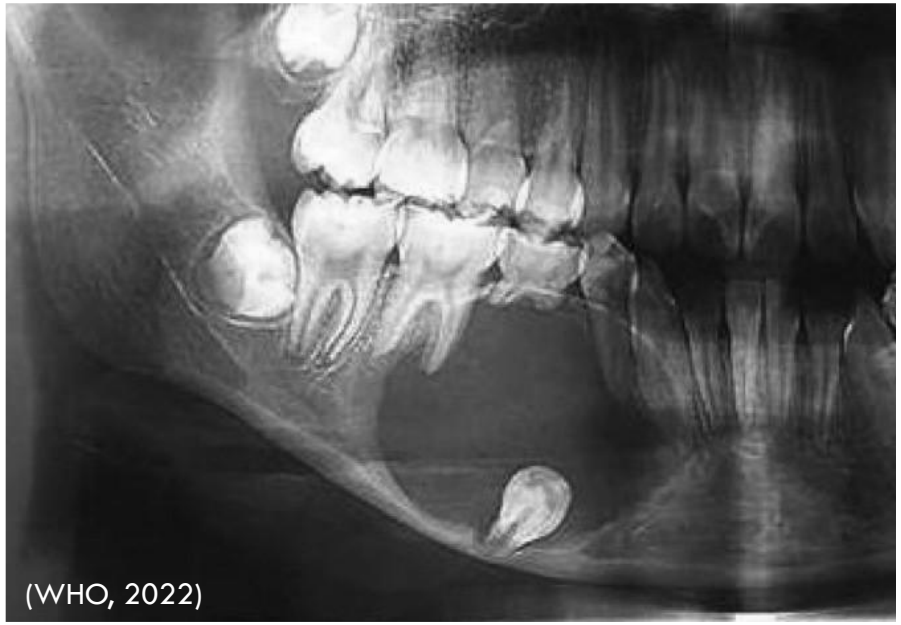
- Low recurrence with conservative excision & enucleation

2.3 PRIMORDIAL ODONTOGENIC TUMOUR

Definition	A benign tumour composed of variably cellular fibrous tissue with areas similar to dental papilla, surrounded by epithelium resembling the internal epithelium of the enamel organ.
Age	Mean = 11.4 years Range = 2-19 years
Gender	Slight M>F
Aetiology	Unknown
Clinical Features	Asymptomatic, although some produce marked cortical expansion

2.3 PRIMORDIAL ODONTOGENIC TUMOUR

Location	Tooth-bearing areas of jaws (Md>Mx) Intraosseous
Periphery/ Shape	Well-defined Unilocular
Internal Features	Radiolucent Associated with UE tooth (most commonly Md 8s resulting in pericoronal relationship)
Surrounding Features	Cortical expansion Tooth displacement and resorption



(WHO, 2022)

2.3 PRIMORDIAL ODONTOGENIC TUMOUR

HISTOPATHOLOGY

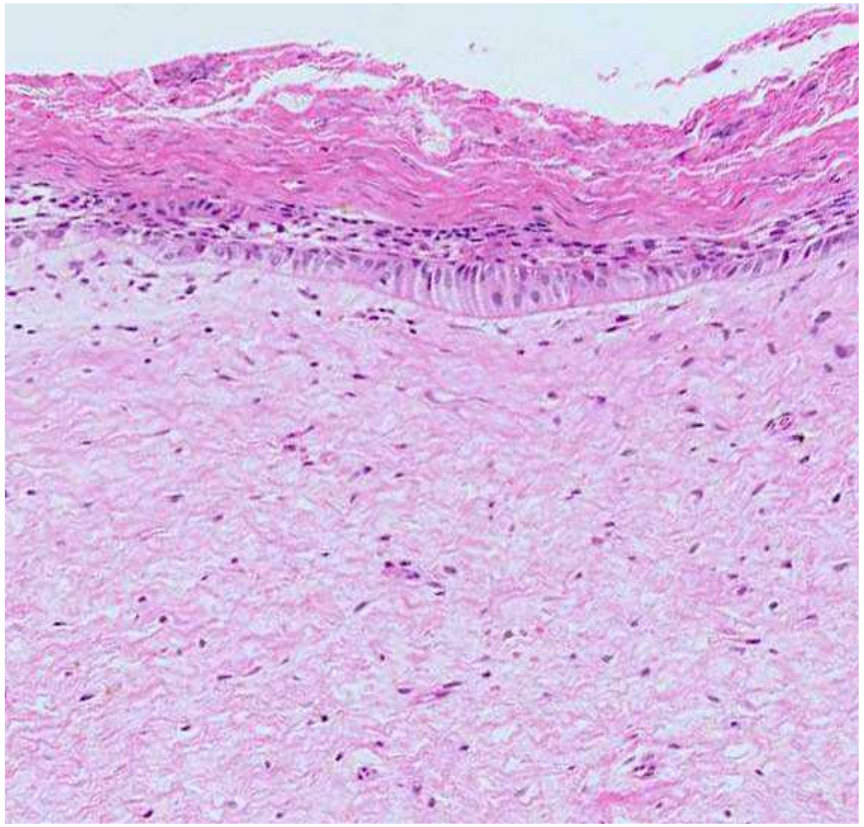
- Loose or myxoid fibrous tissue with variable numbers of fusiform and stellate fibroblasts, with areas of cellular odontogenic mesenchyme
- Periphery is covered by a single layer of columnar or cuboidal epithelium enclosed by a thin fibrous capsule

MANAGEMENT

- Conservative excision

PROGNOSIS/RECURRENCE

- Do not recur

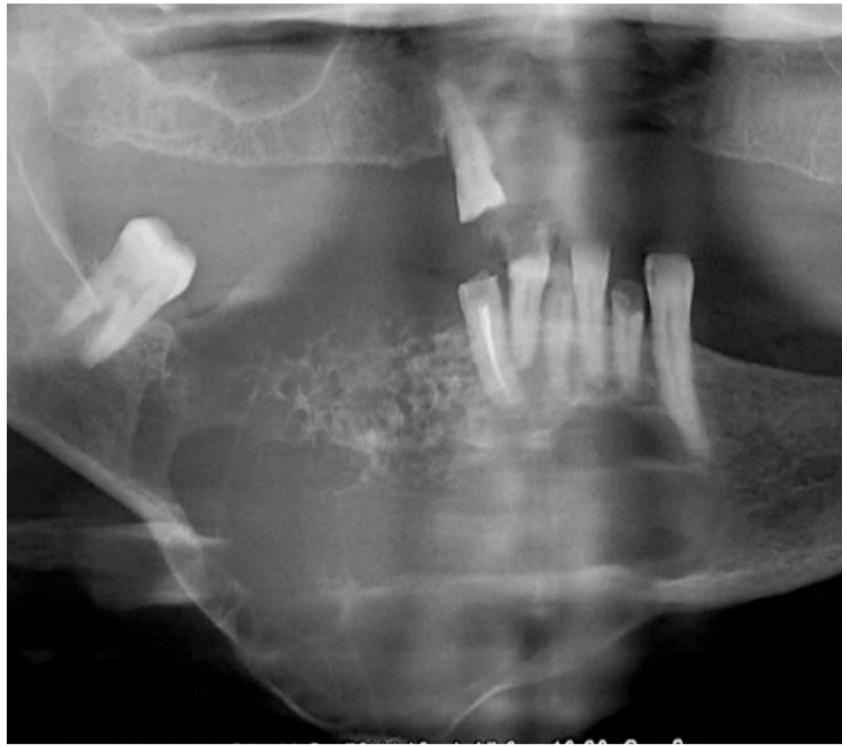


2.4 DENTINOGENIC GHOST CELL TUMOUR

Definition	A benign but locally infiltrative odontogenic tumour characterized by ameloblastoma-like sheets and islands of epithelium with prominent ghost cell keratinization and varying amounts of dentinoid in the stroma. Both the ghost cells and dentinoid may mineralise.
Prevalence	Rare (0.3-0.5% of all odontogenic tumours)
Age	Most diagnosed between 3rd and 5th decades Mean age = 40-47 years Range (1-84 years)
Gender	M>F (2:1), particularly Asian males
Aetiology	Unknown
Clinical features	Slow growing tumour with cortical expansion and occasional cortical perforation +/- mild pain and facial deformity

2.4 DENTINOGENIC GHOST CELL TUMOUR

Location	Mand=Max Post > Ant 25% are extraosseous (gingiva or alveolar ridge mucosa)
Periphery/ Shape	Well-defined (67%) > ill-defined (33%) Unilocular (75%) > Multilocular (25%)
Internal Features	Mixed RL/RO
Surrounding Features	Tooth displacement & resorption common in larger lesions Have been reported in association with odontomas

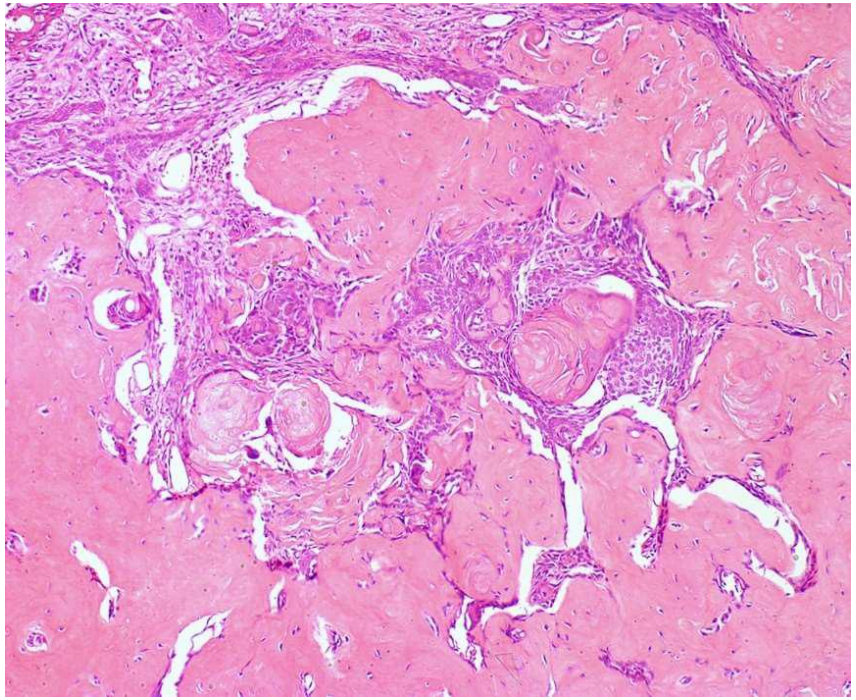


WHO, 2022

2.4 DENTINOGENIC GHOST CELL TUMOUR

HISTOPATHOLOGY

- Nests, islands, and sheets of odontogenic epithelium resembling conventional ameloblastoma, with cuboidal to columnar hyperchromatic basal cells that have reverse nuclear polarity
- +/- cystic degeneration
- Characteristic ghost cells which may undergo mineralisation



MANAGEMENT/PROGNOSIS/RECURRENCE

- 73% recurrence with conservative surgery
- 33% recurrence with radical surgery
- Long term follow-up recommended
  - Recurrence has been reported 1-20 years after treatment

PART 3: BENIGN MESENCHYMAL ODONTOGENIC TUMOURS

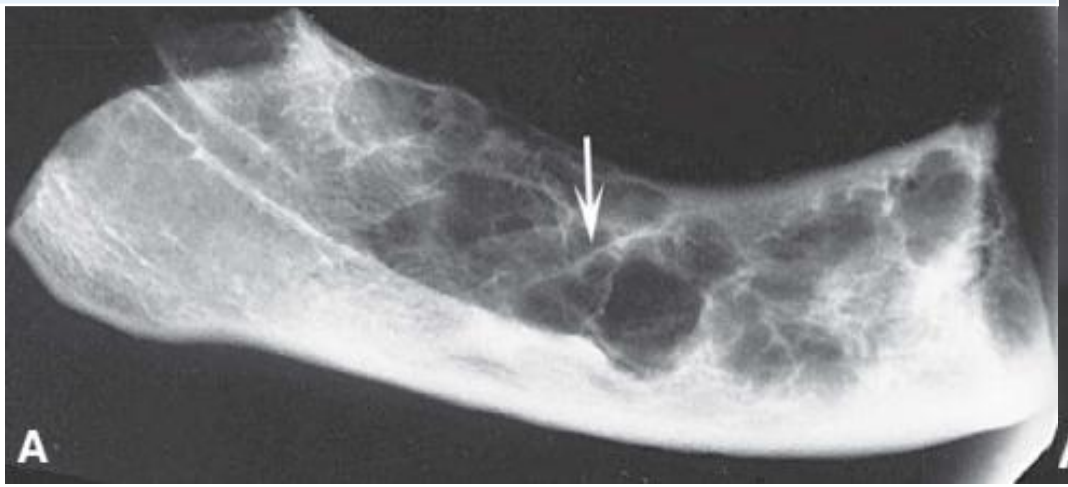
1. Odontogenic myxoma/myxofibroma
2. Odontogenic fibroma
3. Cementoblastoma
4. Cemento-ossifying fibroma

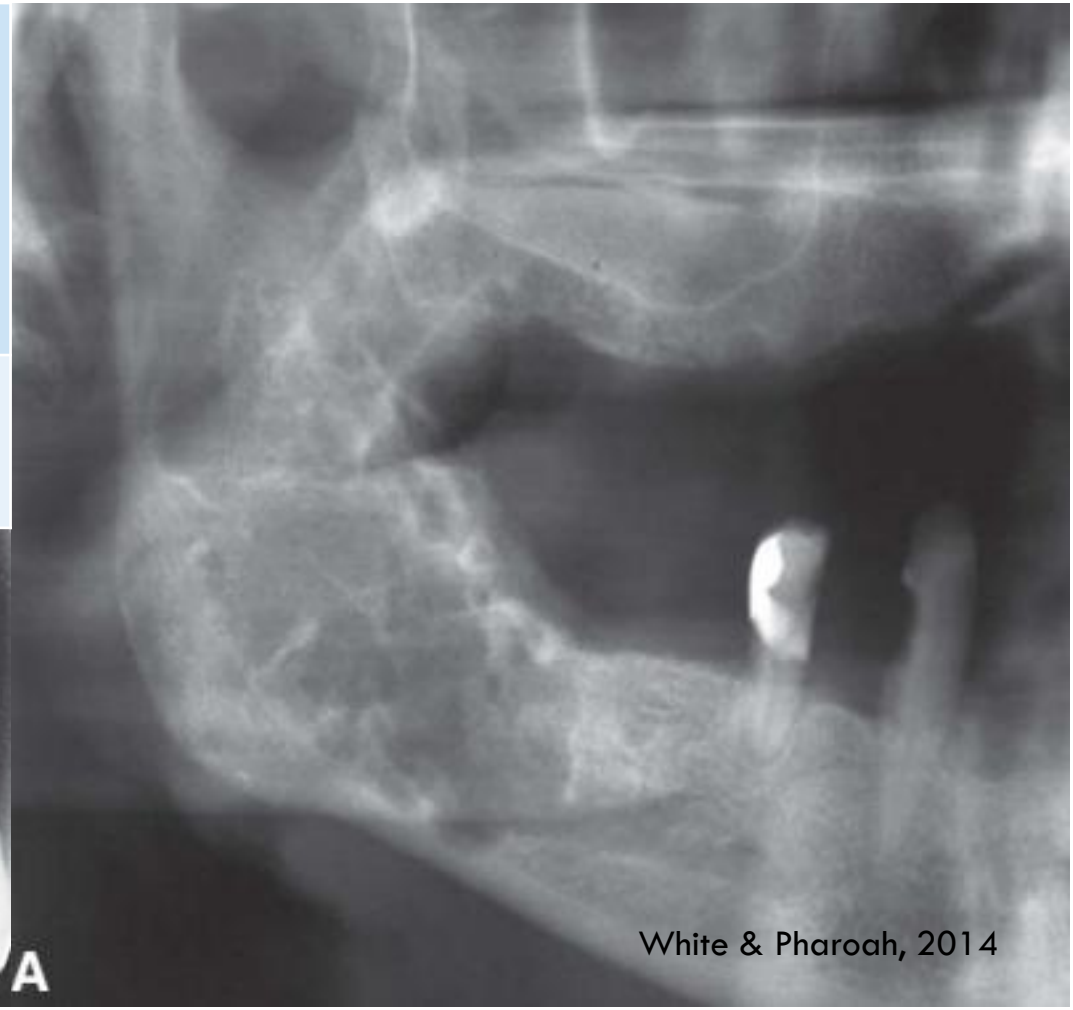
3.1 ODONTOGENIC MYXOMA

Definition	A benign neoplasm histologically resembling odontogenic ectomesenchyme and characterized by sparse spindle or stellate cells in a myxoid stroma.
Prevalence	<10% of odontogenic tumours 3rd most frequent after ameloblastoma and odontomas
Age	Wide age range Most commonly young adults (2nd and 3rd decades of life)
Gender	F>M (2:1)
Aetiology	Unknown
Clinical features	Slow permeative growth, causing bone destruction, expansion, mobility or absence of tooth, eventual cortical perforation and soft tissue infiltration Firm and not tender to palpation

3.1 ODONTOGENIC MYXOMA

Location	Md (2/3) >Mx (1/3) Most common in premolar/molar region Rarely in non-tooth bearing areas (ramus & condyle) Mx lesions usually involve premolar/molar alveolar process or zygomatic process – may extend to, expand, & obliterate sinus Extraosseous gingival lesions very rare
Periphery/ Shape	Unilocular or multilocular Well defined & corticated May also be poorly defined (esp. in Mx)

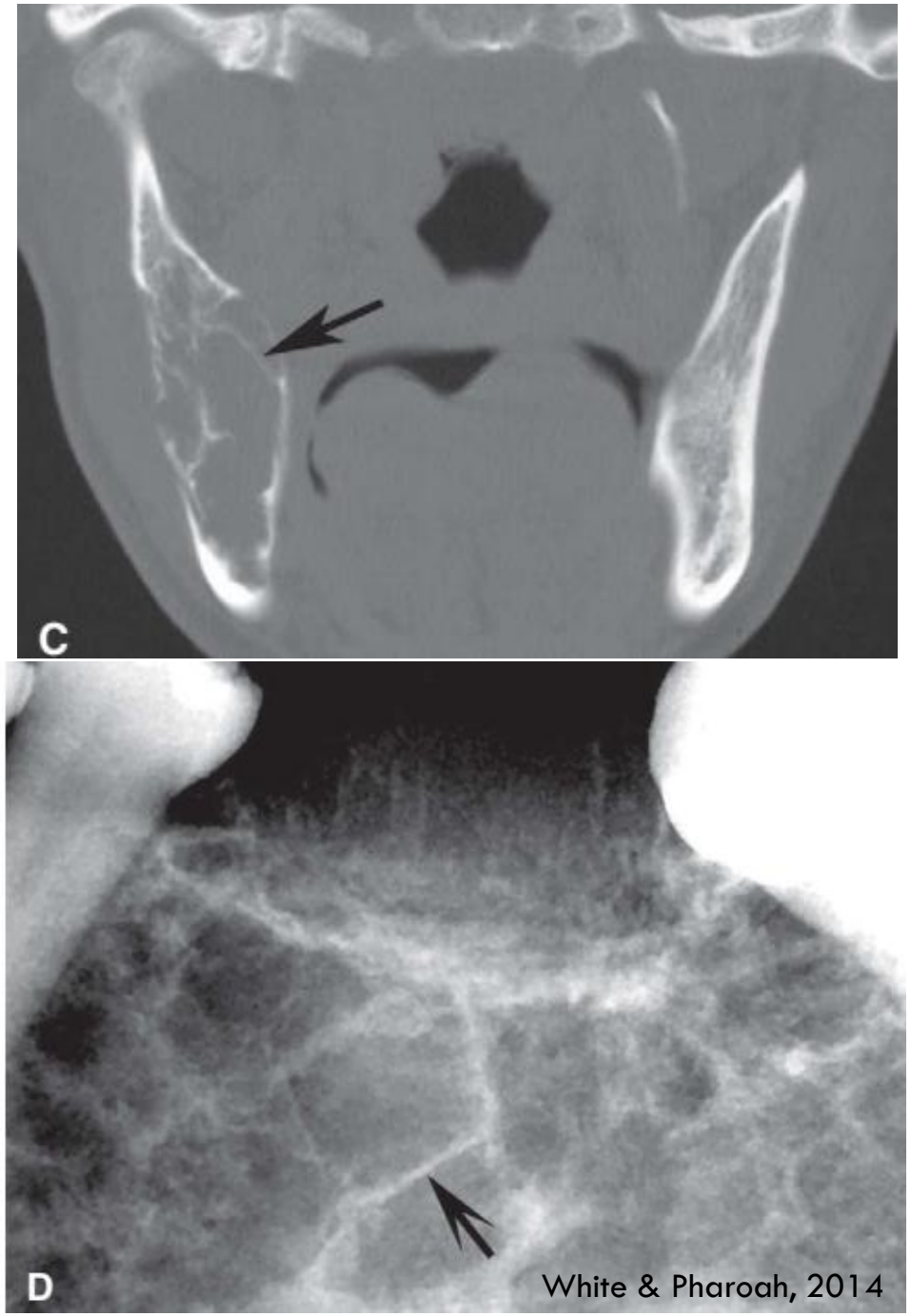




White & Pharoah, 2014

3.1 ODONTOGENIC MYXOMA

Internal Features	• RL (unilocular cyst-like when pericoronal) • Most have a mixed RL/RO internal pattern • Variable septa appearance - may be curved/straight & fine/coarse – **look for straight septa!** • May show fine soap-bubble or honeycomb appearance, occasionally with **fine straight tennis-racket trabeculations**
Surrounding Features	• Displaces & loosens teeth (root resorption rare) • Often scallops between roots of adjacent teeth • Tendency to grow along bone without the same amount of expansion as other odontogenic tumours • Considerable expansion (& perforation) when large • Mx lesions tend to obliterate the Mx sinuses as an early feature • May present with periosteal reactive bone layer (larger)



White & Pharoah, 2014

3.1 ODONTOGENIC MYXOMA

DIFFERENTIAL DIAGNOSIS

Other multilocular lesions	e.g. ameloblastoma, CGCG, central haemangioma, ABCs, OKC May need further imaging with MDCT or MRI Look for **thin straight septa with less than expected bone expansion**
Osteogenic sarcoma	May appear similar due to spiculated periosteal reaction Look for an intact outer cortex for OM
Odontogenic fibroma	Occasionally has same radiographic features & cannot be reliably differentiated

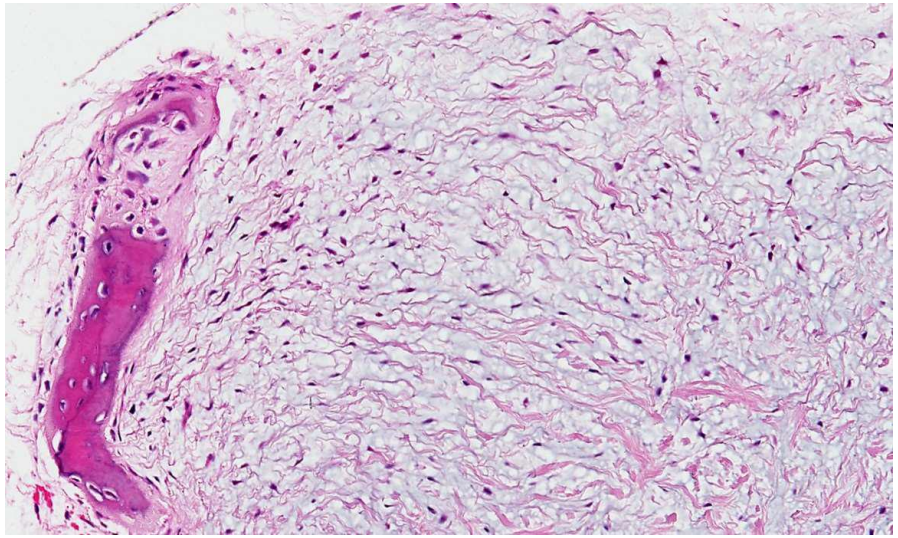
3.1 ODONTOGENIC MYXOMA

HISTOPATHOLOGY

- Predominantly loose myxoid stroma with a sparse population of stellate to spindle cells
- Small inactive rests of odontogenic epithelium are present in a small minority and are few in number
- Unencapsulated with permeative spread in medullary bone
- Appears histologically similar to the dental follicle or developing dental papilla (but clinical and radiologic features are different)

MANAGEMENT/PROGNOSIS/RECURRENCE

- Lack of capsule and permeative spread account for recurrence rates = 10 to 43%
- It is unclear whether recurrence increases with more conservative treatment methods
- Maxillary lesions are more likely to recur than mandibular lesions



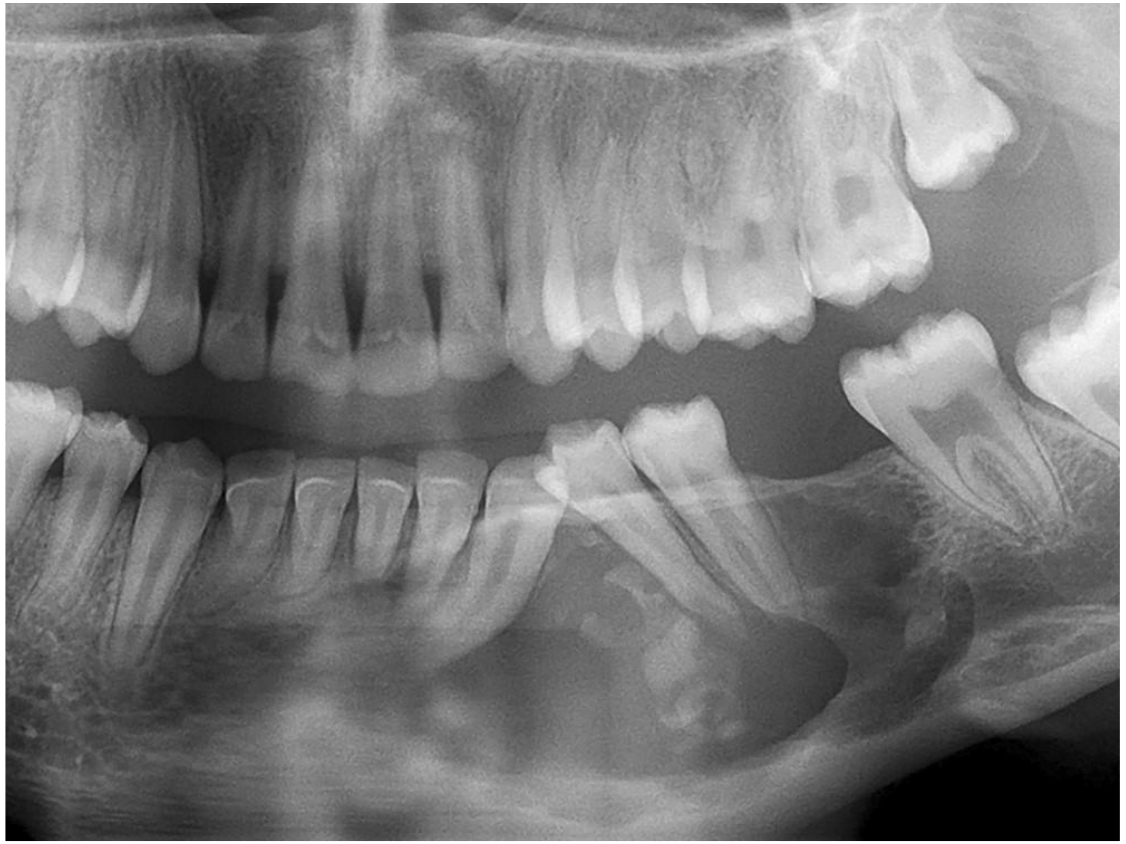
WHO, 2022

3.2 ODONTOGENIC FIBROMA

Definition	A neoplasm of mature fibrous or fibromyxoid connective tissue with variable amounts of inactive-appearing odontogenic epithelium, with or without associated mineralisations
Age	Wide age range; mean = 34 years
Gender	F>M (2.2:1)
Aetiology	Unknown
Clinical features	(Small) asymptomatic (Large) localised swelling, loosening of teeth, pain

3.2 ODONTOGENIC FIBROMA

Location	Max>Mand *Maxilla:* Anterior to first molar *Mandible:* Posterior to first molar
Periphery/ Shape	Well-defined, corticated
Internal Features	Radiolucent (90%) > RL/RO (10%) Unilocular (small); Multilocular (large)
Surrounding Features	Divergence or resorption of associated tooth roots



WHO, 2022

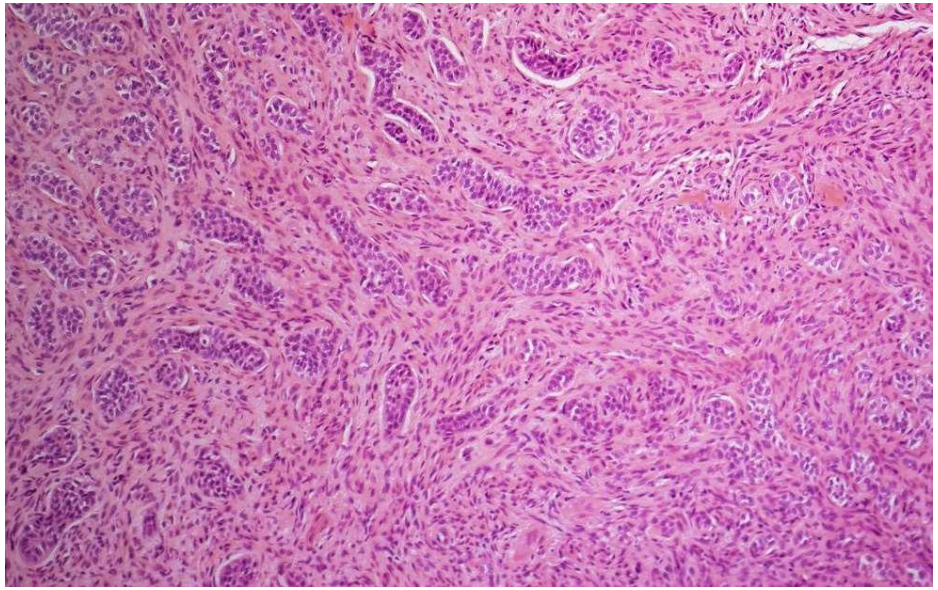
3.2 ODONTOGENIC FIBROMA

HISTOPATHOLOGY

- Moderately cellular bland fibrous tissue with moderate to dense collagen content, accompanied by varying amounts of dispersed inactive-appearing odontogenic epithelial nests and cords
- +/- minor hard tissue formation

MANAGEMENT/PROGNOSIS/RECURRENCE

- Almost never recur after enucleation and curettage



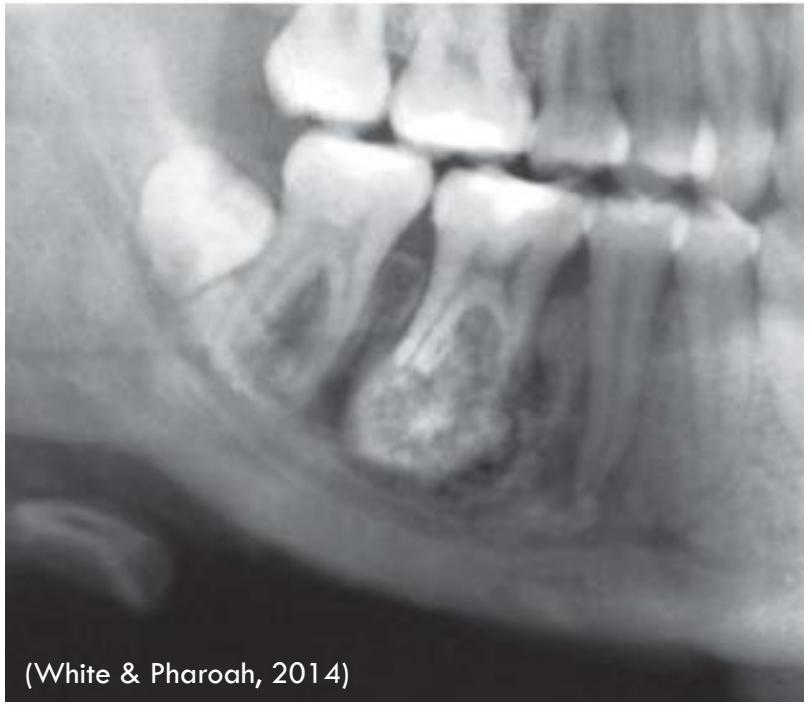
WHO, 2022

3.3 CEMENTOBLASTOMA

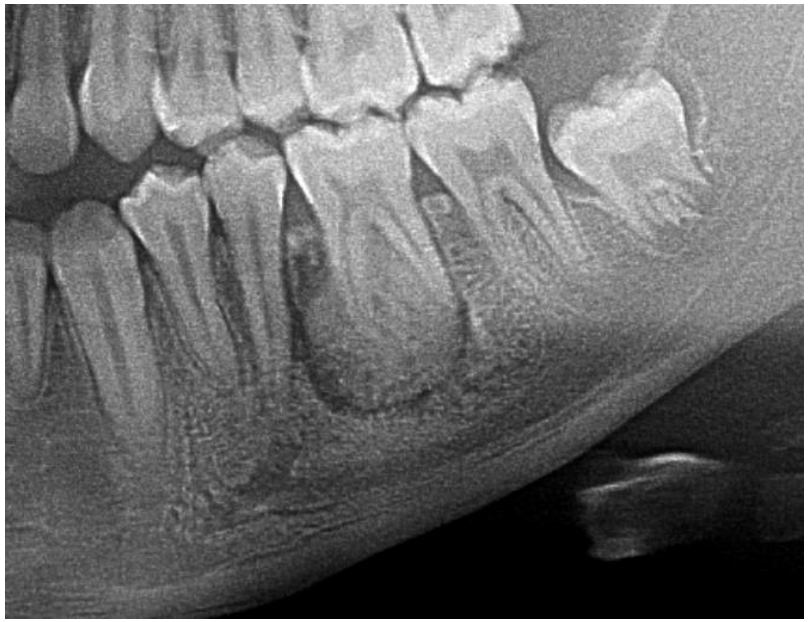
Definition	A benign odontogenic neoplasm that forms a rounded mass of cementum on the root of a tooth.
Terminology	Acceptable: benign cementoblastoma Not recommended: true cementoma; cementoma
Prevalence	Relatively rare, 3% of all odontogenic tumours
Age	Wide age range; highest frequency in 2nd – 3rd decades
Gender	M=F
Aetiology	Unknown
Clinical features	Slow-growing characteristically painful expansion of the jaw Associated tooth is vital in about 80% of cases

3.3 CEMENTOBLASTOMA

Location	Develops on the apical third of a tooth root Most common in the posterior mandible Permanent first molar > mandibular premolar and maxillary molar regions > other teeth Involvement of deciduous teeth is rare
Periphery/ Shape	Well-defined Characteristic radiolucent rim which is continuous with the PDL
Internal Features	• Mixed RO/RL; majority RO • Amorphous or wheel spoke pattern • Density of the cemental mass usually obscures outline of enveloped root • Maturation of lesion from centre to periphery (as evidenced by lucent band)
Surrounding Features	External resorption of involved root (2/3rd of cases) Cortical bone perforation & tooth displacement are rare



(White & Pharoah, 2014)



WHO, 2022

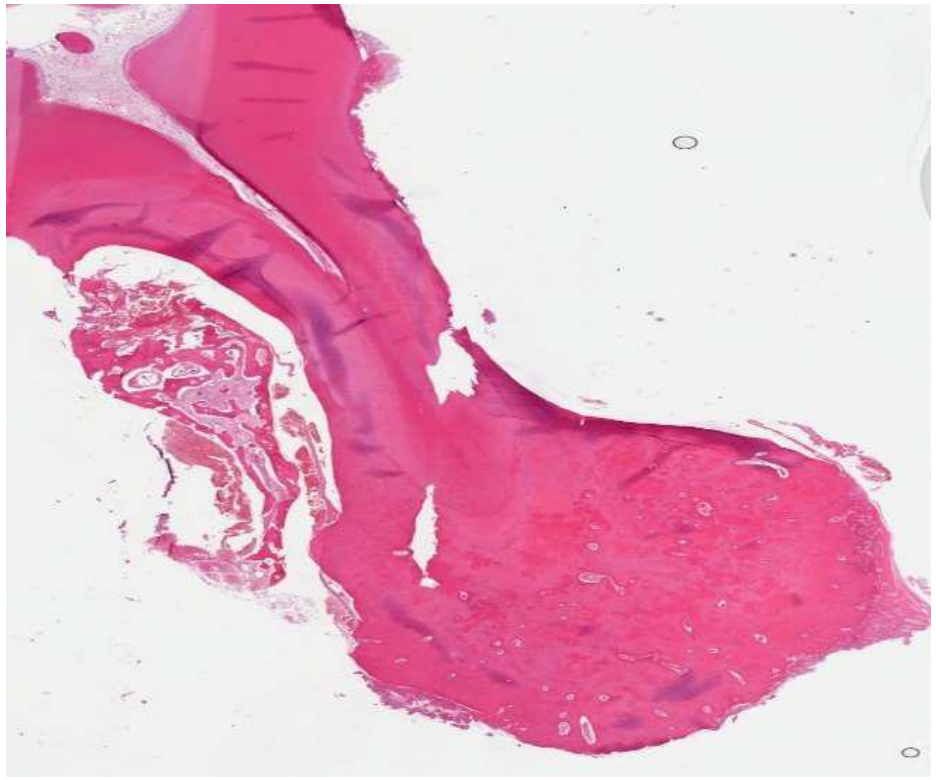
3.3 CEMENTOBLASTOMA

HISTOPATHOLOGY

- A mass of dense cellular cementum resembling bone, often with prominent reversal lines, fused with the resorbed surface of the tooth root.
- At the periphery there are radiating finger-like trabeculae of newly formed matrix, often associated with plump cementoblasts and cementoclasts and vascular immature fibrous tissue.

MANAGEMENT/PROGNOSIS/RECURRENCE

- Does not usually recur after extraction of the associated tooth and curettage



WHO, 2022

3.4 CEMENTO-OSSIFYING FIBROMA

Definition	A benign odontogenic fibro-osseous neoplasm arising in the jaws and characterised by production of bone and cementum-like calcifications in a fibrous stroma
Terminology	Acceptable: Ossifying fibroma, conventional type Not recommended: Cementifying fibroma; ossifying fibroma; ossifying-odontogenic fibroma; periodontoma
Age	Wide age range; peak 3rd to 4th decade
Gender	F>M (5:1); primarily Caucasians, followed by African descent
Aetiology	Odontogenic in origin, related to inactivating mutations in the tumour suppressor gene
Clinical features	Painless jaw expansion Slow-growing, but can reach considerable size if left untreated Usually solitary, but rare cases of multiple lesions (sporadic or as a component of hyperparathyroidism-jaw tumour syndrome)

3.4 CEMENTO-OSSIFYING FIBROMA

Location	Mand>Max (particularly premolar-molar region)
Periphery/ Shape	Well-defined, corticated Characteristic radiolucent rim
Internal Features	Radiolucent (early stages) Variable internal patterns
Surrounding Features	Cortical thinning and expansion Tooth displacement & root resorption Displacement of antral floor

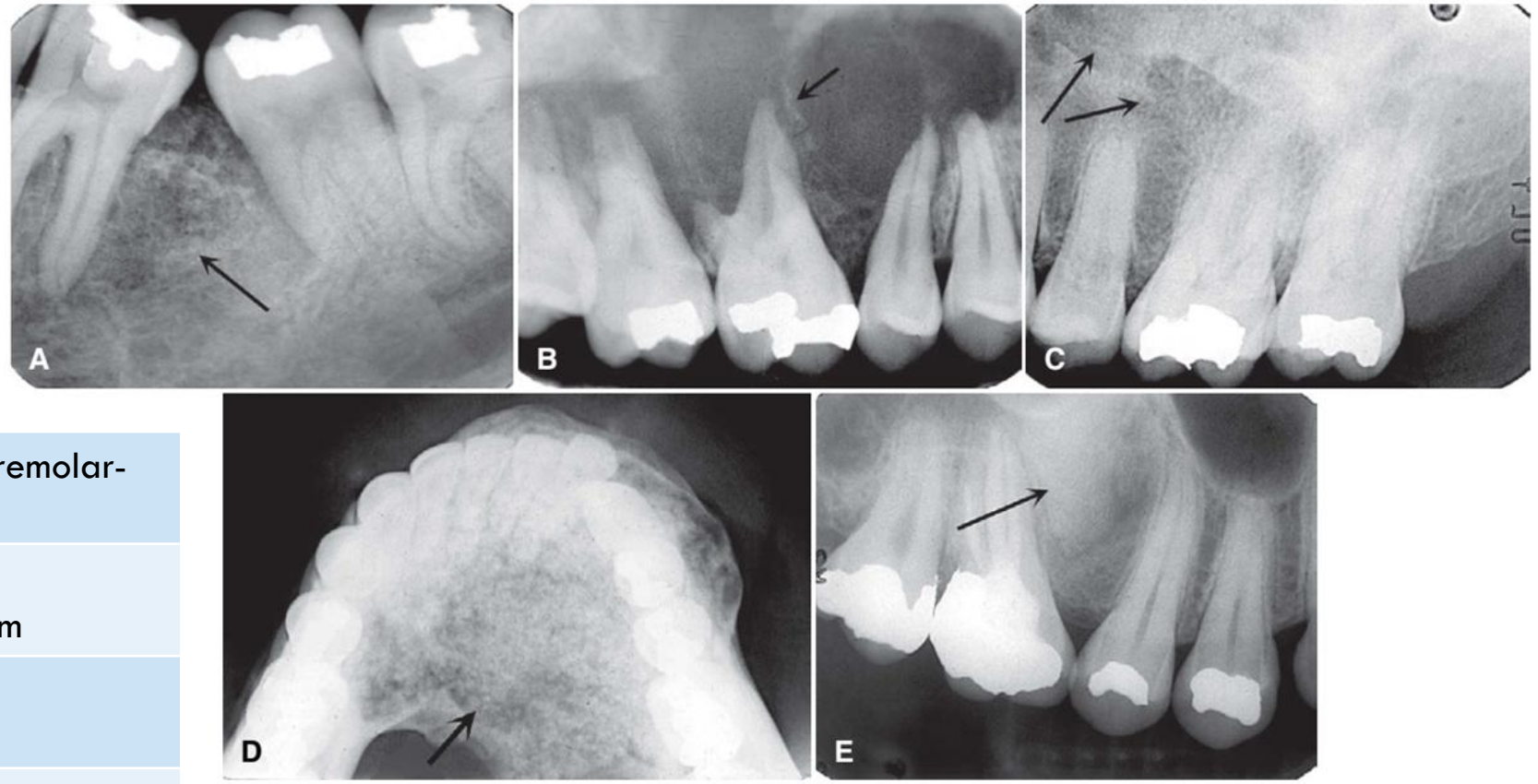


FIGURE 22-52 Various bone patterns seen in ossifying fibroma. **A,** Wispy trabecular pattern *(arrow)*. **B,** Most of this pattern is radiolucent with a few wispy trabeculae *(arrow)*. **C,** Fibrous dysplasia granular-like pattern *(arrows)*. **D,** Flocculent pattern with larger tufts of bone formation *(arrow)*. **E,** Solid, radiopaque, cementum-like pattern *(arrow)*.

White & Pharoah, 2014

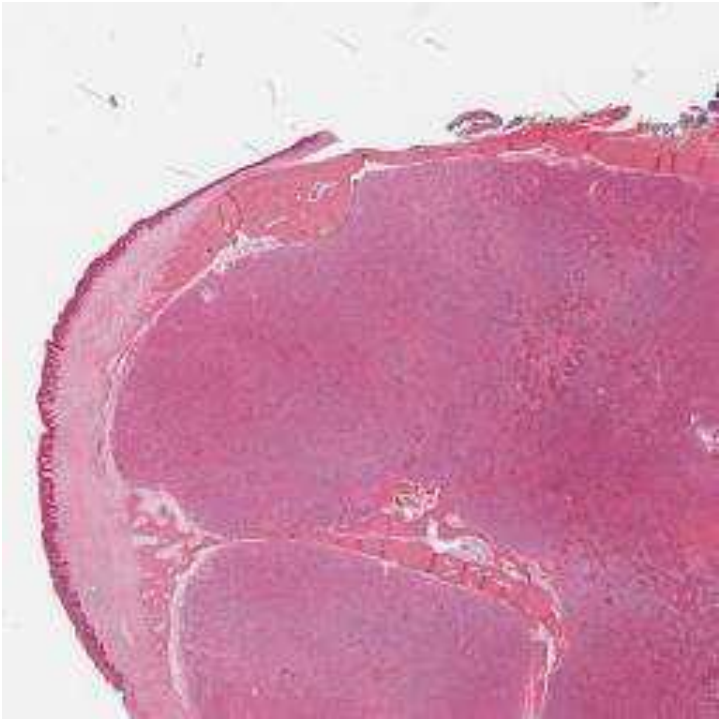
3.4 CEMENTO-OSSIFYING FIBROMA

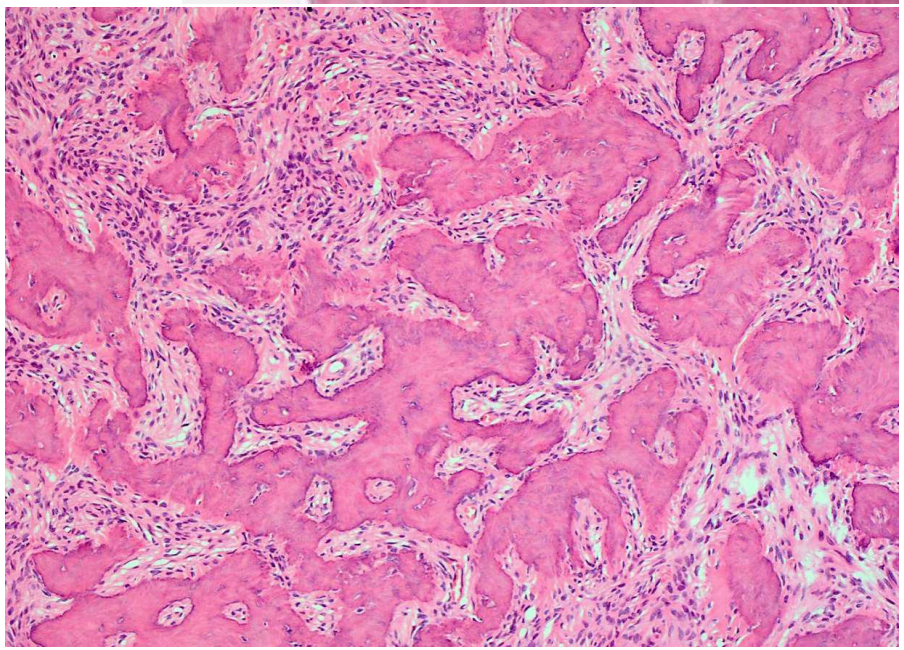
HISTOPATHOLOGY

- Variable proportions of fibrous and mineralised tissue, more heavily mineralised centrally and with a thin fibrous capsule or well demarcated margin from surrounding normal bone
- A fibro-osseous appearance of condensation of woven bone from stroma must be present at least focally
- Osteoblastic rimming is frequent and there is bone remodelling in the lesion with infrequent osteoclasts

MANAGEMENT/PROGNOSIS/RECURRENCE

- Rarely recurs with enucleation and curettage





WHO, 2022

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