BENIGN TUMOURS OF THE JAWS (PART 2 – NON-ODONTOGENIC TUMOURS)

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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 MAXILLOFACIAL BONE AND CARTILAGE TUMOURS

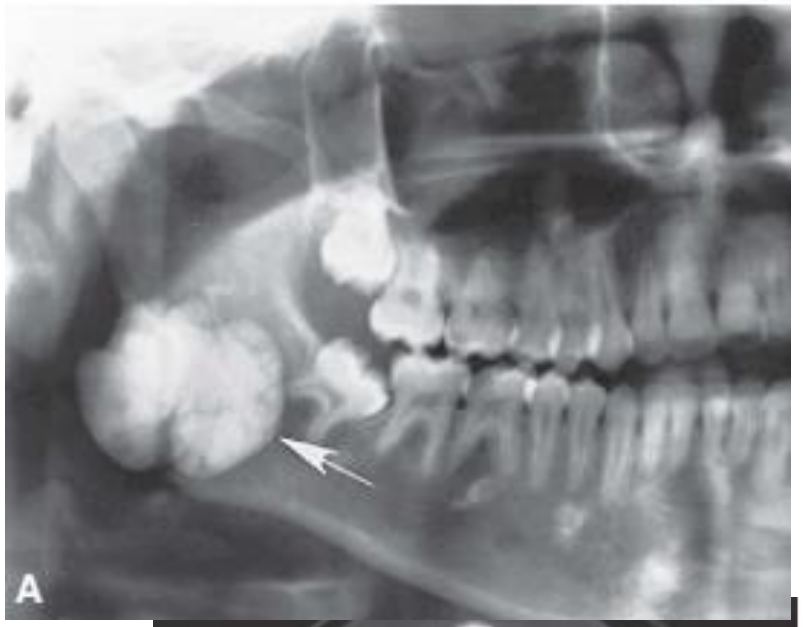
1. Osteoma
2. Osteochondroma
3. Osteoblastoma
(Osteoid osteoma)
4. Chondroblastoma
5. Chondromyxoid fibroma
6. Desmoplastic fibroma of bone

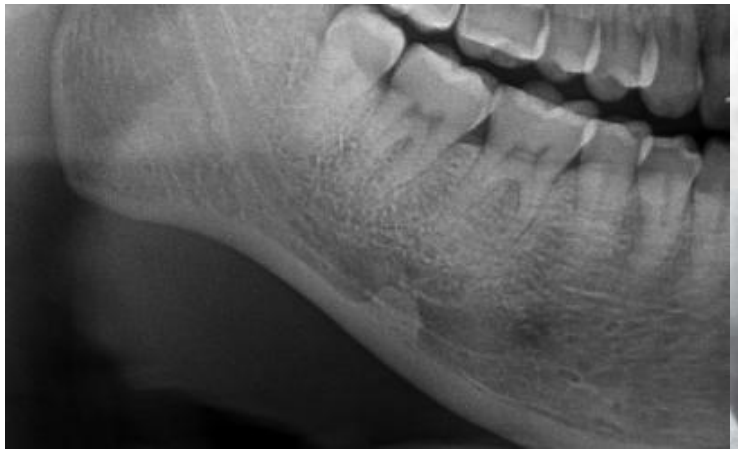
1.1 OSTEOMA

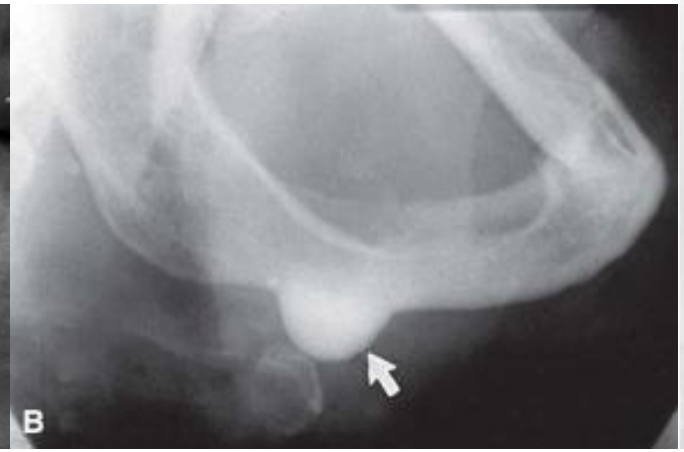
Definition	A benign bone forming neoplasm consisting of mature bone, restricted almost exclusively to the jaws and craniofacial bones.
Types	Surface (periosteal) Central (endosteal) – aka dense bone islands
Age	Wide age range, mostly 3rd to 5th decades
Gender	M=F
Aetiology	Unknown – debate whether they are benign neoplasms or hamartomas May be a manifestation of Gardners syndrome
Clinical Features	Central osteoma: often asymptomatic Surface osteoma: Slow growing swelling with facial distortion or altered dental occlusion Sinus/orbital osteomas: headache or pain

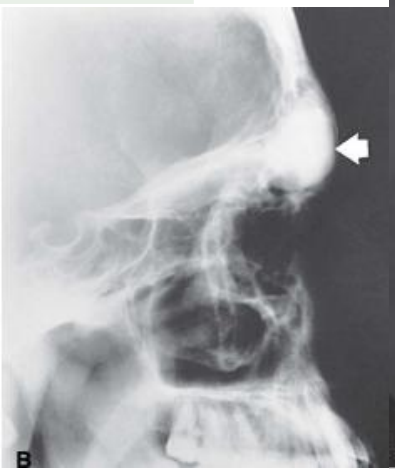
1.1 OSTEOMA

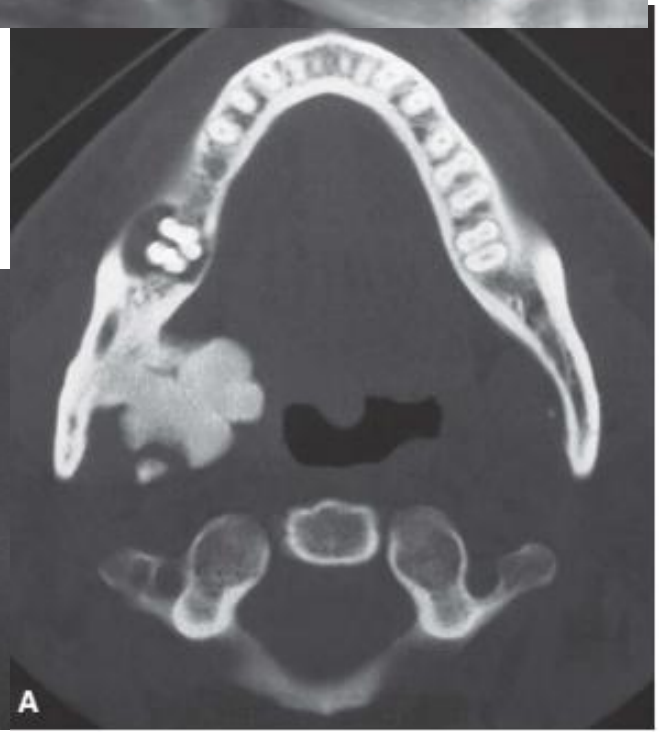
Location	Mandible > Maxilla (esp. lingual ramus & inferior border of Md) Paranasal sinuses (most commonly frontal sinus)
Periphery/ Shape	Well-defined Sessile or pedunculated Smooth or irregular
Internal Features	Compact bone (uniformly radiopaque) Cancellous bone (internal trabecular architecture)
Surrounding Features	When large, will displace adjacent soft tissues & cause dysfunction











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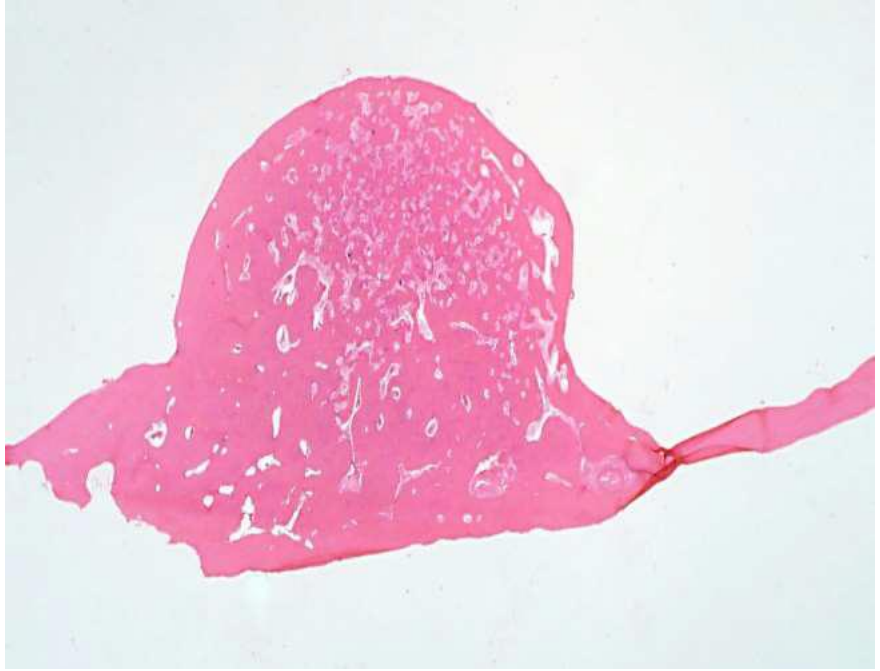
1.1 OSTEOMA

HISTOPATHOLOGY

- Composed mostly of lamellar bone (compact or trabecular) in which osteoblasts and osteoclasts are inconspicuous
  - +/- osteoblastoma-like areas (thought related to remodelling process rather than aggressive behaviour)
- Similar appearance to fibro-osseous lesions, sclerosing osteomyelitis, and ossification of a fibrous epulis – correlate with clinical and radiographic findings for definitive diagnosis



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MANAGEMENT/PROGNOSIS/RECURRENCE

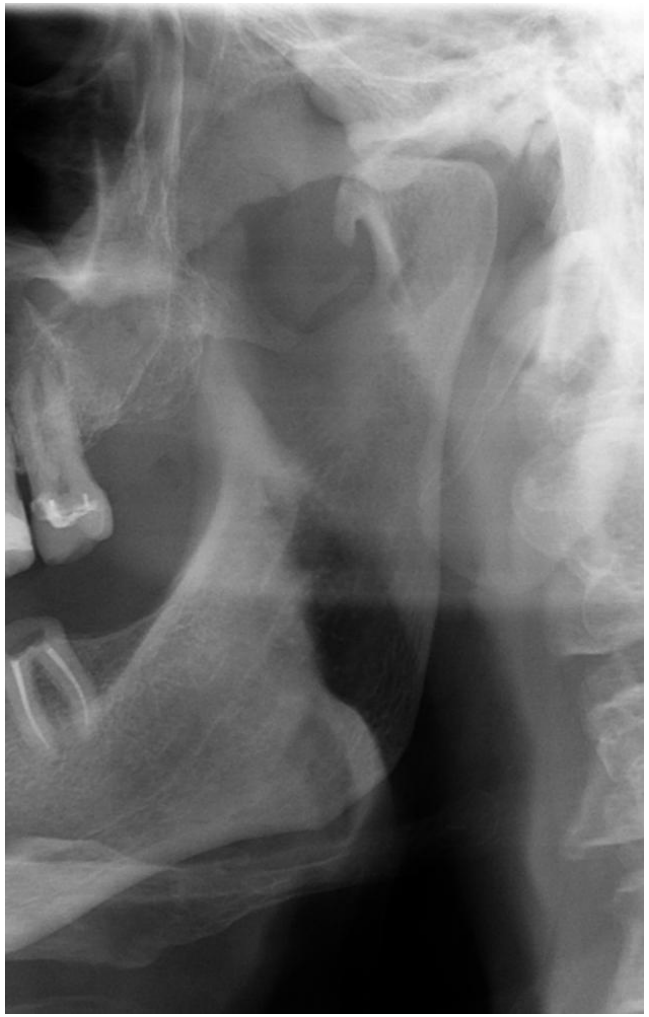
- Slow growth – most are monitored & resection only for symptomatic lesions
- Recurrence is unusual

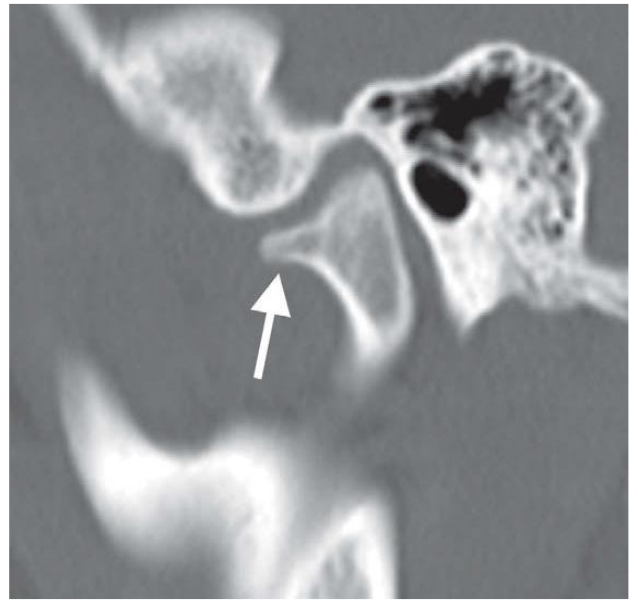
1.2 OSTEOCHONDROMA

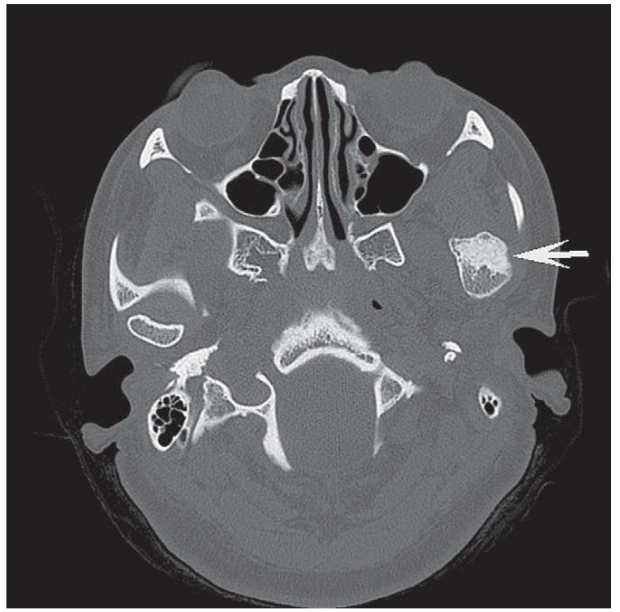
Definition	A benign neoplasm forming a bony projection with a cartilaginous cap, with continuity between the marrow cavity of the tumour and underlying bone
Prevalence	<1% of osteochondromas occur in the H+N (One of the most common lesions of the axial skeleton, but much less common in the facial bones)
Age	2nd to 4th decades
Gender	Slight F>M
Aetiology	Uncertain – trauma? Prior radiotherapy?
Clinical features	May be an incidental radiographic finding Swelling, asymmetry, trismus, malocclusion, and TMJ dysfunction May be multiple, but multiple lesions usually affect long bones

1.2 OSTEOCHONDROMA

Location	Skull base, maxillary sinus, zygoma, mandible (condyle & coronoid process)
Periphery/ Shape	Well-defined, lobulated Pedunculated or sessile Thin cartilaginous cap
Internal Features	Opacity with internal trabecular architecture
Surrounding Features	In continuity with the cortex and medulla of the bone of origin







1.2 OSTEOCHONDROMA

HISTOPATHOLOGY

- A cartilage capped bony projection from the external bone surface showing cortical and medullary continuity with the parent bone
- + surface layer of fibrous tissue
- + middle layer which resembles growth plate cartilage

MANAGEMENT/PROGNOSIS/RECURRENCE

- May not need intervention if asymptomatic
- Recurrence following excision is rare, unless incompletely removed
- Malignant transformation is very rare

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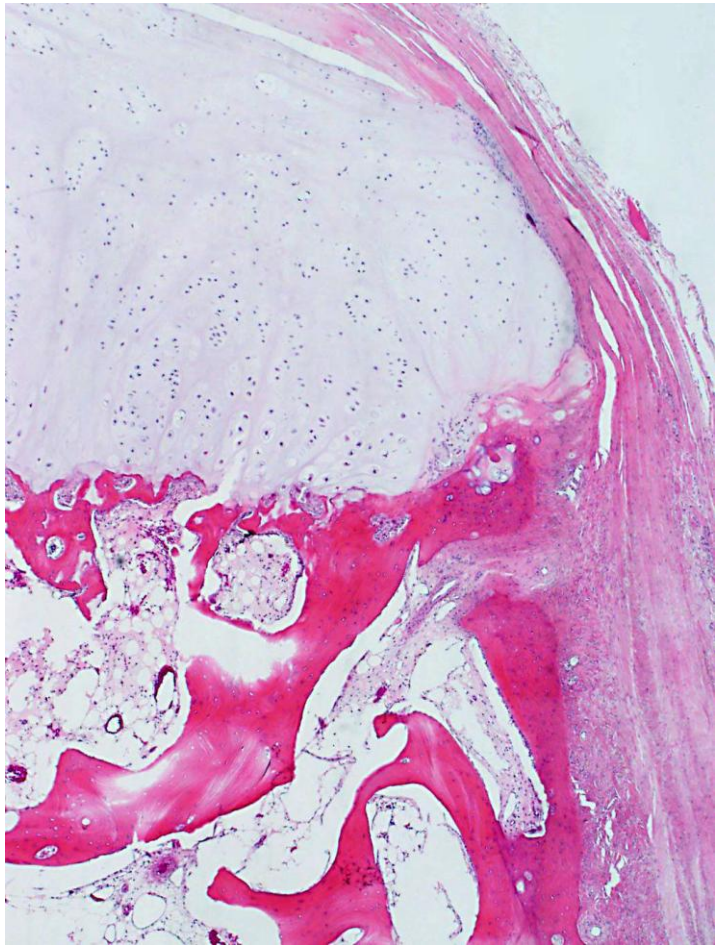
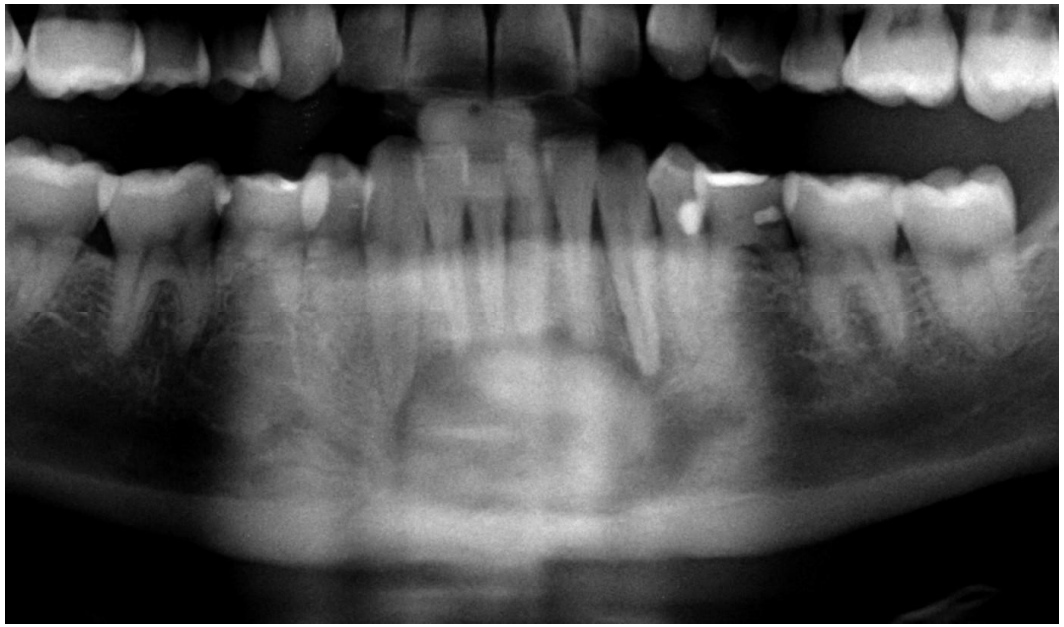


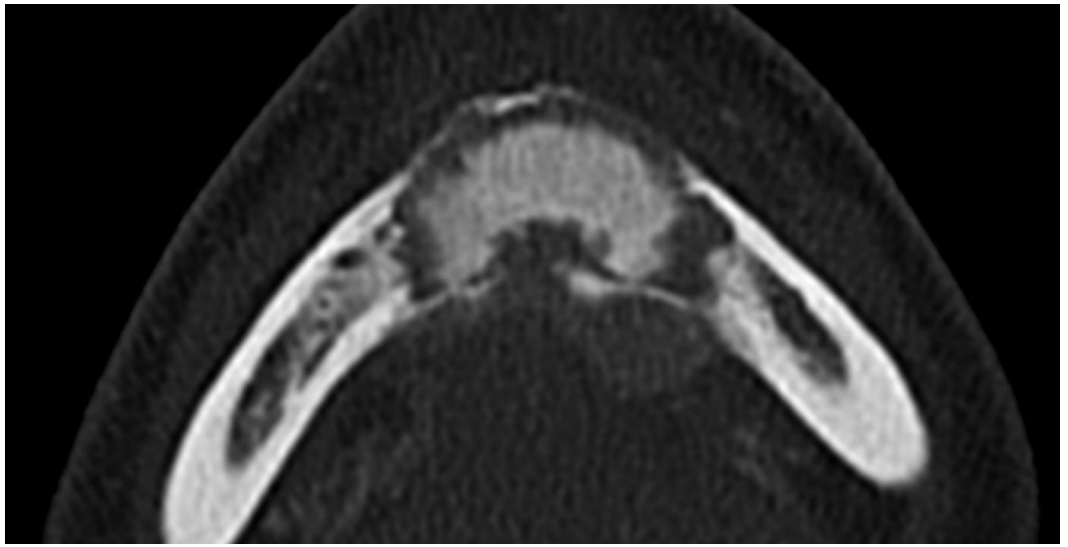
1.3 OSTEOBLASTOMA

Definition	A benign but aggressive bone forming tumour with large osteoblasts forming sheets and prominent osteoblastic rimming on woven bone, and greater than 20mm in diameter
Prevalence	Rare
Age	Occurs mostly in the 2nd to 3rd decades
Gender	Slight F>M
Aetiology	Unknown
Clinical Features	May be asymptomatic or present with localised swelling and pain

1.3 OSTEOBLASTOMA

Location	• 10% in the craniofacial bones, most often in the body of the mandible • May be intra-osseous or periosteal
Periphery/ Shape	• Well circumscribed, round/oval shaped • Usually no sclerotic border • >20mm in size





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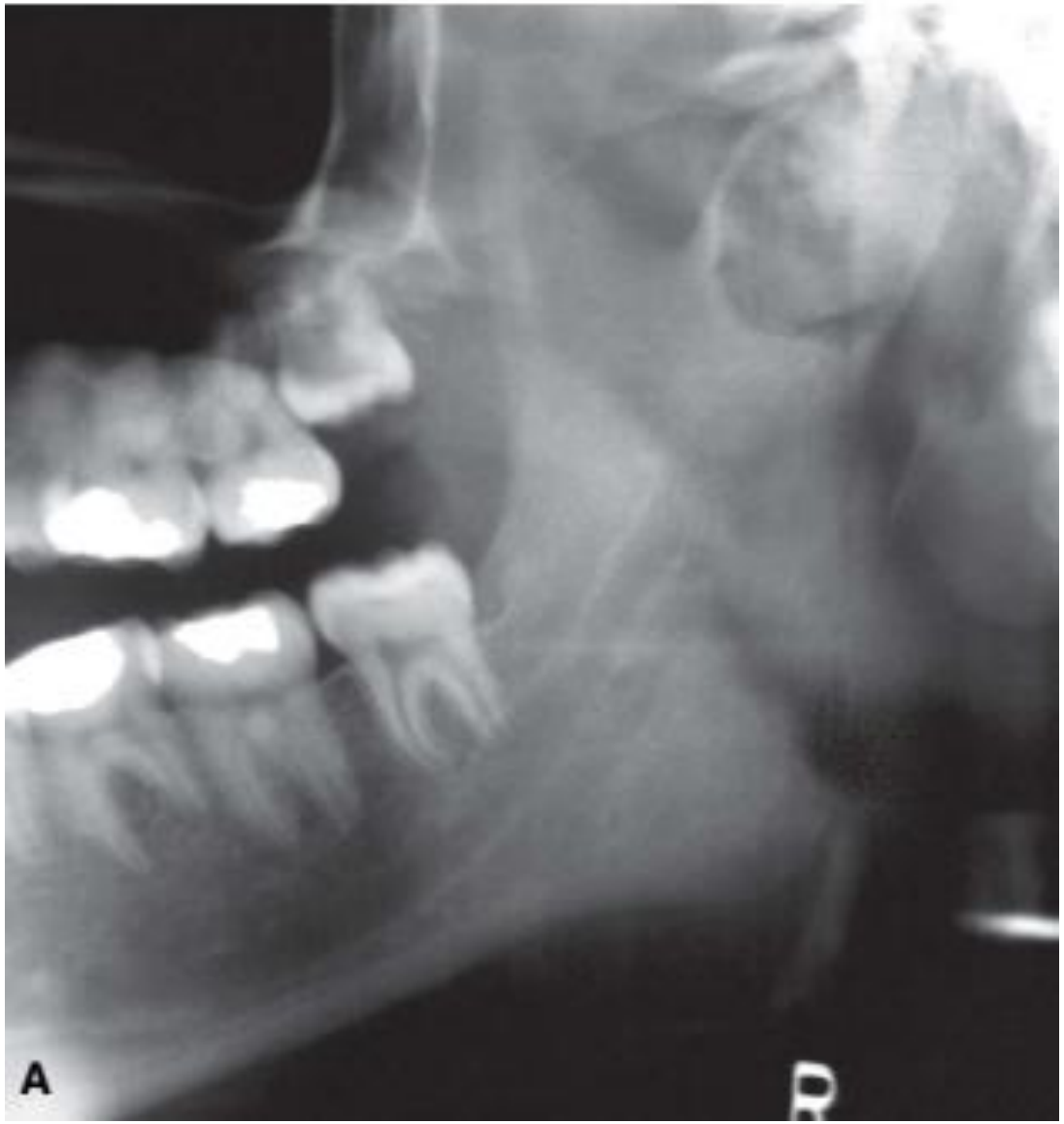
1.3 OSTEOBLASTOMA

Internal Features	• Early lesions are radiolucent • Varying degrees of calcific material as lesion progresses (may appear like fine granular bone trabeculae)
Surrounding Features	• Expansion (but cortex is maintained) • May invaginate into maxillary sinus • Root resorption is rare • Usually no periosteal reaction • May mimic malignancy radiologically



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1.3 OSTEOBLASTOMA

DIFFERENTIAL DIAGNOSIS

Osteoid osteoma	See next section
Cementoblastoma	Identical histologically, except for adherence to root surface with resorption "Blue-bone" appearance of osteoblastoma not common with cementoblastoma
Low-grade, well-differentiated osteosarcoma	Similar histopathological findings - look for benign vs malignant features (e.g. nuclear atypia, permeative growth into surrounding bone, atypical mitoses)
Large cemento-osseous dysplasia	Similar radiographic appearance with surrounding soft tissue capsule. Look for more aggressive features in osteoblastoma

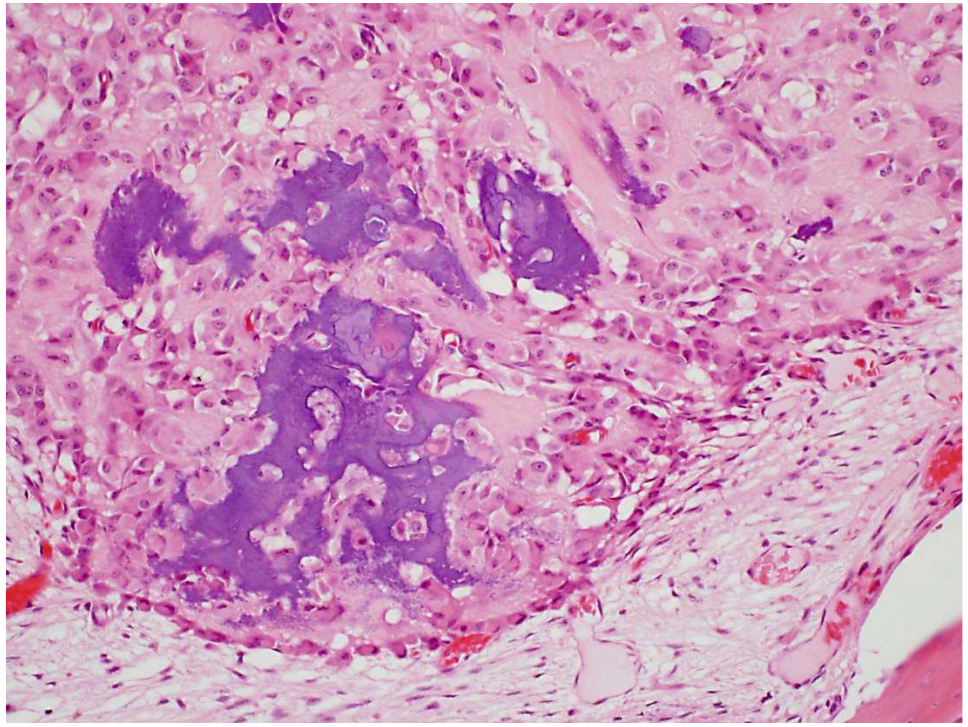
1.3 OSTEOBLASTOMA

HISTOPATHOLOGY

- Anastomosing trabeculae of bone and osteoid, rimmed by plump osteoblasts within a loose richly vascular fibrous stroma
- Characteristic “blue-bone” appearance
- Some osteoblasts may appear larger and epithelioid, but this does not indicate a clinically aggressive course

MANAGEMENT/PROGNOSIS/RECURRENCE

- Recurrence may follow curettage or incomplete removal



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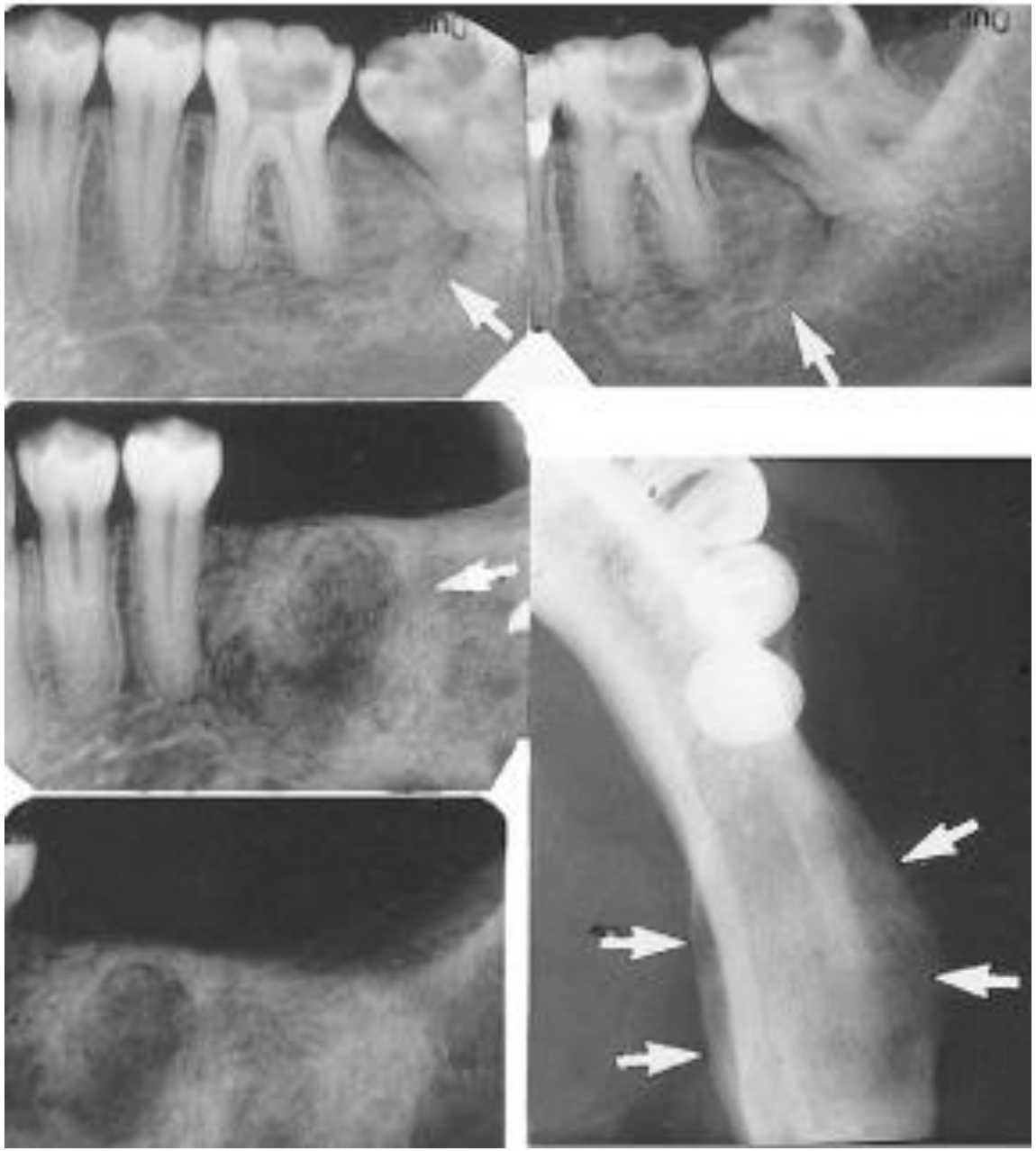
OSTEOID OSTEOMA

	Osteoblastoma	Osteoid Osteoma
Size	>20mm	≤20mm
Trabeculae	Larger, broader with wider trabecular spaces	
Clinical features	More aggressive Less pain than osteoid osteoma Does not occur at night Not relieved by salicylates	Pain disproportionate to lesion size Nocturnal pain Relieved by aspirin (extragnathic)
Imaging features	Usually within medullary bone Lucent rim (NO sclerotic border) surrounding central mixed RL/RO NO periosteal reaction	Usually develops in outer cortex Sclerotic rim surrounding central RL +/- periosteal reaction
Histopathology	More osteoclasts	Smaller size and surrounding zone of sclerotic bone

OSTEOID OSTEOMA



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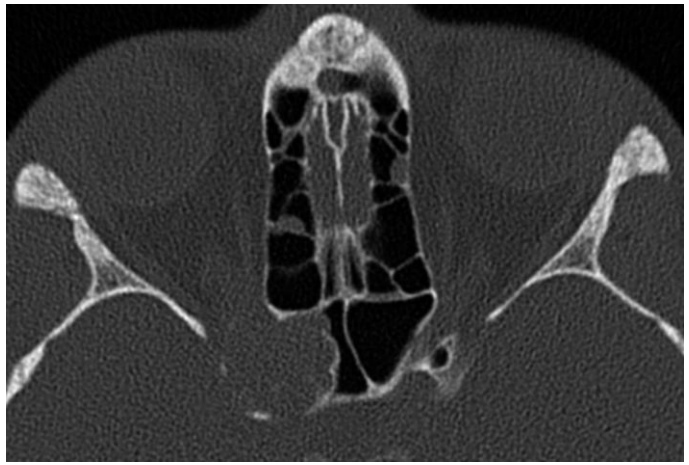
1.4 CHONDROBLASTOMA

Definition	A benign tumour of bone composed of chondroblasts forming sheets and islands of eosinophilic chondroid matrix, with or without chicken-wire calcifications
Prevalence	Rare in the H+N (about 100 cases reported in the literature)
Age	2nd decade of life Skull tumours present slightly older, in the 3rd or 4th decade
Gender	Slight M>F (1.3:1) for temporal lesions
Aetiology	H3-3A and H3-3B mutations
Clinical Features	Most commonly, pain +/- hearing loss, tinnitus, and vertigo (if tumour in temporal bone) +/- trismus (if TMJ)

1.4 CHONDROBLASTOMA

Location	Predominantly in the epiphyseal areas of long bones In H+N, mostly around TMJ and squamous part of temporal bone
Periphery/ Shape	Well-demarcated Lobulated
Internal Features	Foci of calcifications
Surrounding Features	Cortical expansion Displacement of adjacent soft tissues



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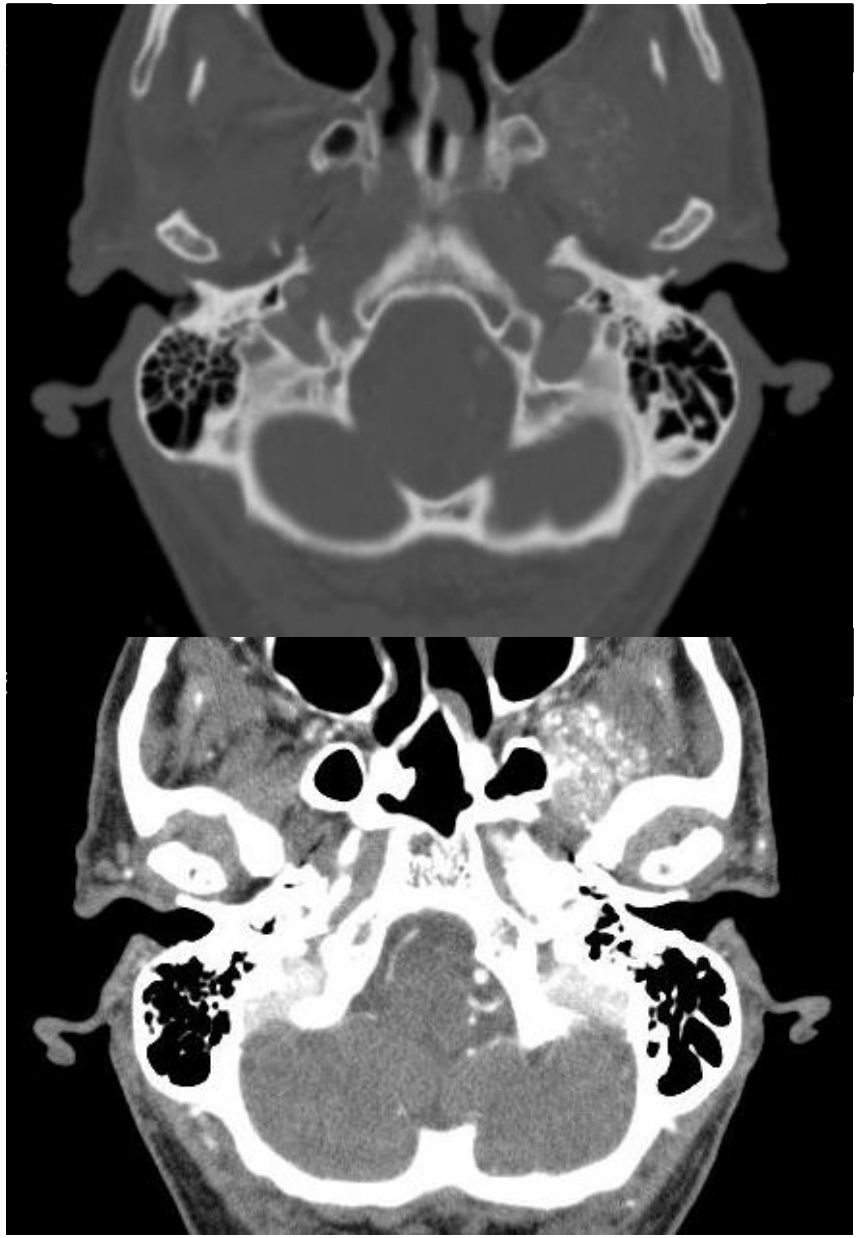


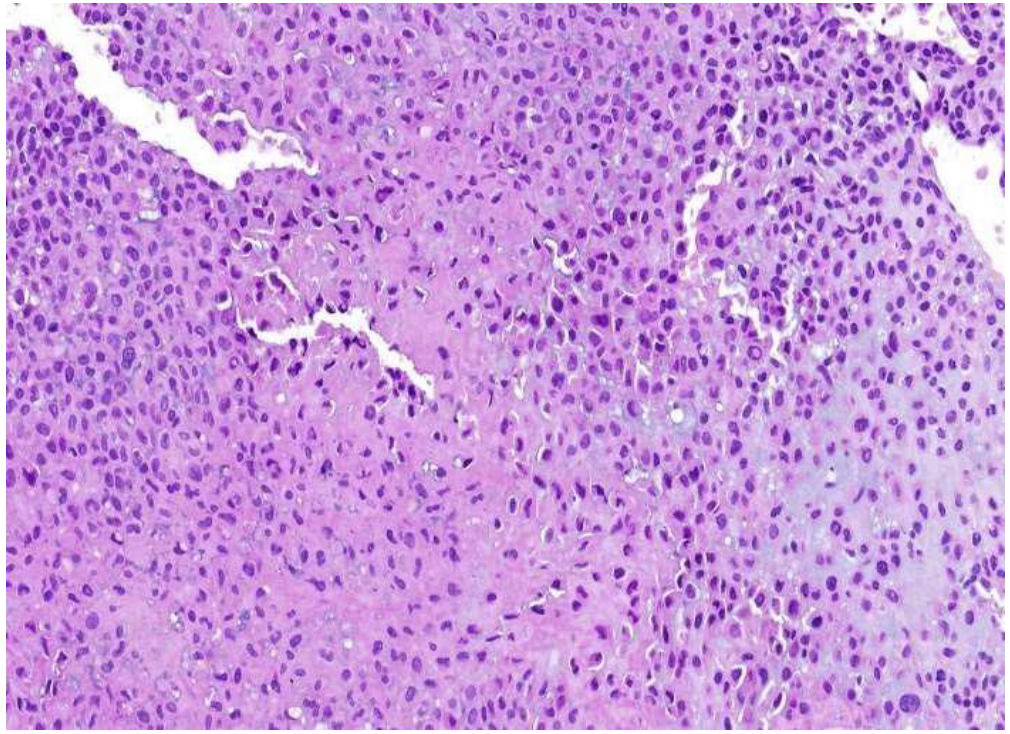
1.4 CHONDROBLASTOMA

HISTOPATHOLOGY

- Uniform, eosinophilic and polygonal cells intermingled with an amorphous eosinophilic matrix with osteoclast-like giant cells and varying amounts of 'chicken-wire' calcification
- +/- cystic haemorrhagic degeneration

MANAGEMENT/PROGNOSIS/RECURRENCE

- Up to 50% of cases recur
- Metastasis has been reported only rare (<1%)



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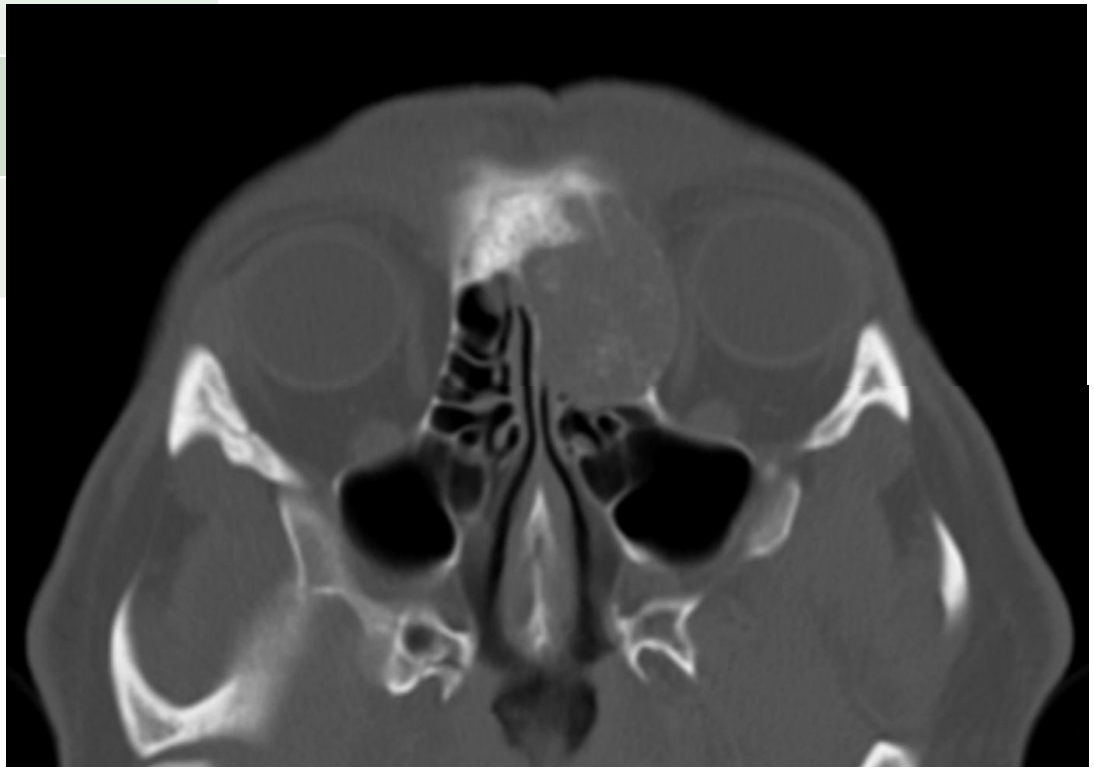
1.5 CHONDROMYXOID FIBROMA

Definition	A benign lobulated chondroid neoplasm with a zonal architecture composed of chondroid, myxoid, and myofibroblastic areas
Prevalence	Rare (5% of cases involve craniofacial bones)
Aetiology	Unknown, but majority involve a recombination of the glutamate receptor gene
Clinical Features	Depending on the site, there may be tinnitus, visual disturbances, headaches, hearing loss, and sinonasal congestion

1.5 CHONDROMYXOID FIBROMA

Location	Any craniofacial bone can be affected, but most commonly in the jaw and sinonasal bones
Periphery/ Shape	Well-demarcated Sclerotic rim
Internal Features	Primarily radiolucent 10% demonstrates focal mineralisations
Surrounding Features	Cortical thinning or erosion



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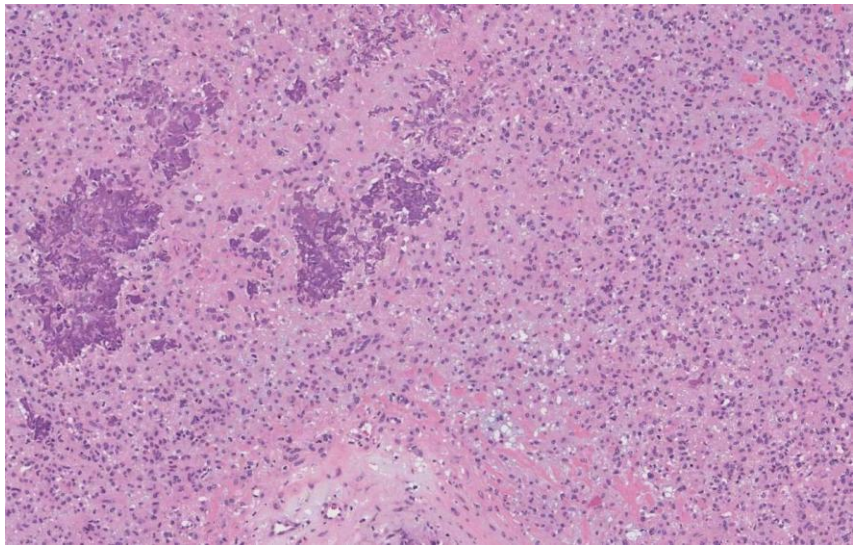
1.5 CHONDROMYXOID FIBROMA

HISTOPATHOLOGY

- A lobular proliferation of spindled and stellate cells with abundant eosinophilic cytoplasm and a chondromyxoid background
- 1/3 cases demonstrate scattered coarse calcifications

MANAGEMENT/PROGNOSIS/RECURRENCE

- Excellent prognosis, even for recurrent tumours
- Recurrence more common in craniofacial lesions, due to challenges obtatining clear surgical margins



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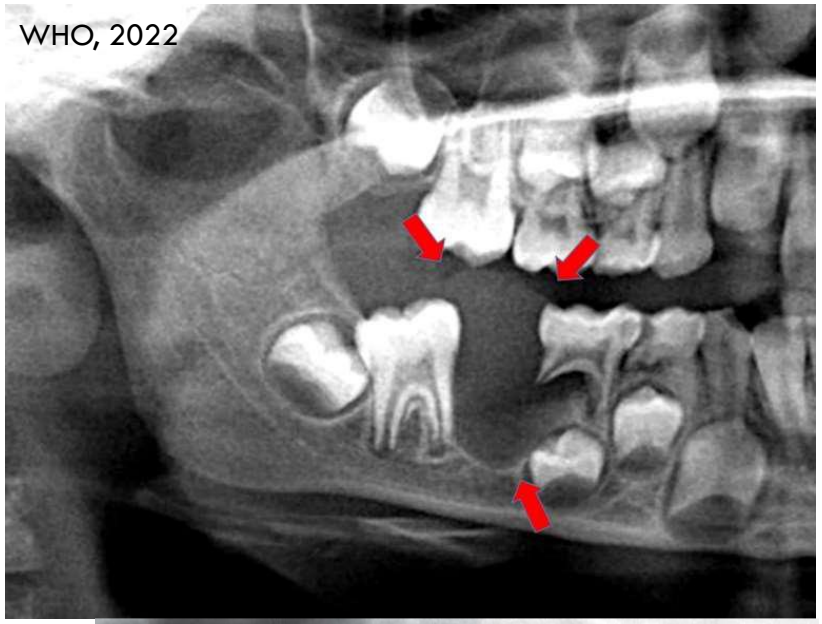
1.6 DESMOPLASTIC FIBROMA OF BONE

Definition	A locally aggressive fibroblastic/myofibroblastic tumour composed of benign spindle cells embedded in a collagenous background, mimicking desmoid-type fibromatosis.
Prevalence	Rare (5% of cases involve craniofacial bones)
Age	Wide age range (0.5 to 70 years) – mean age of diagnosis = 20 years 35% occur in 1st decade 70% occur before 30 years
Gender	Slight F>M (1.3:1)
Aetiology	Unknown
Clinical Features	Asymptomatic swelling or facial asymmetry (66%) Pain (15%), trismus (11%), mobile teeth (7%), infection (3%), and bleeding (3%)

1.6 DESMOPLASTIC FIBROMA OF BONE

Location	Mand (82%) > Max 70% posterior body and angle Maxillary lesions are usually anterior
Periphery/ Shape	Well-defined, some are ill-defined
Internal Features	Radiolucency without mineralisation Large lesions multilocular with very coarse, thick septa
Surrounding Features	Expansion, cortical erosion Tooth displacement and root resorption Often with soft tissue extension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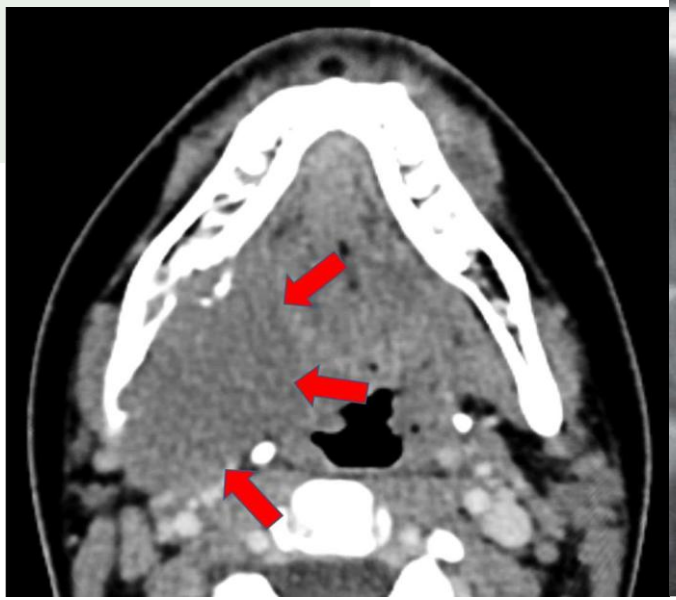

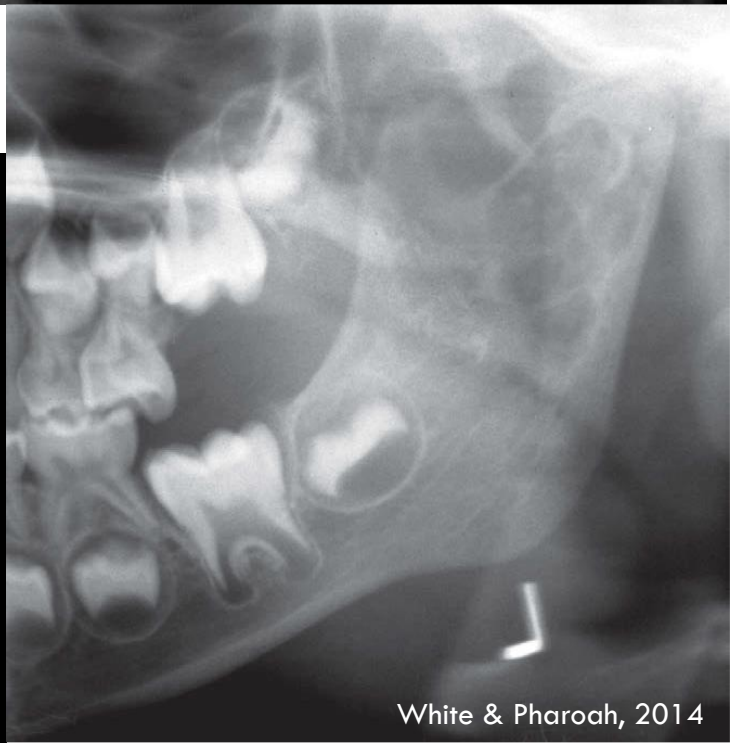
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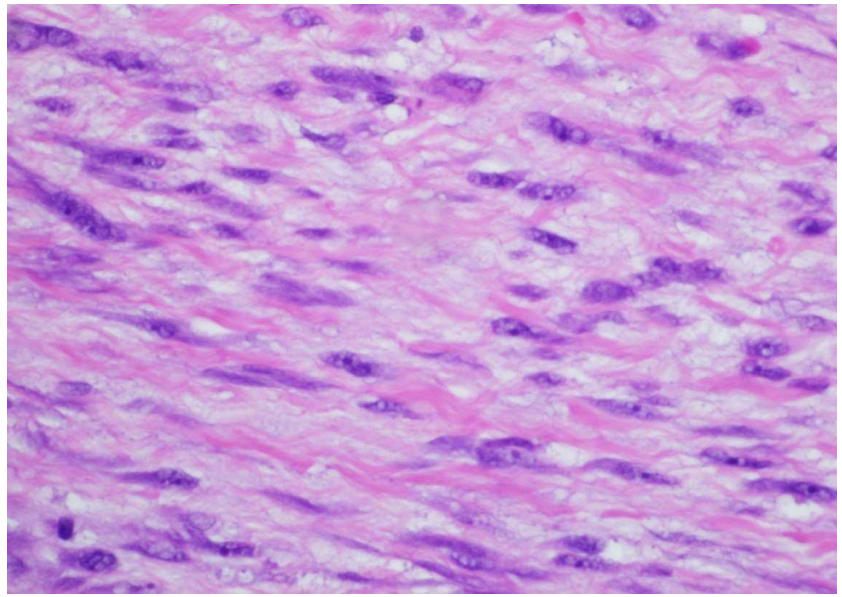
1.6 DESMOPLASTIC FIBROMA OF BONE

HISTOPATHOLOGY

- Uniform benign spindle cells with slender tapering nuclei, arranged in intertwining fascicles, without atypia or pleomorphism and only rare mitoses
- Due to the infiltrative margin, entrapped residual bone trabeculae may be present



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MANAGEMENT/PROGNOSIS/RECURRENCE

- Frequently recurs after curettage (31%) or enucleation (25%)
- Approximately 10% recurrence after resection
- Chemotherapy may be considered if excision is not feasible
- Radiotherapy is not recommended

PART 2:
SOFT TISSUE TUMOURS

1. Vascular tumours
    - Haemangioma
2. Peripheral nerve sheath tumours
    - Neurofibroma
    - Schwannoma
    - Neuroma

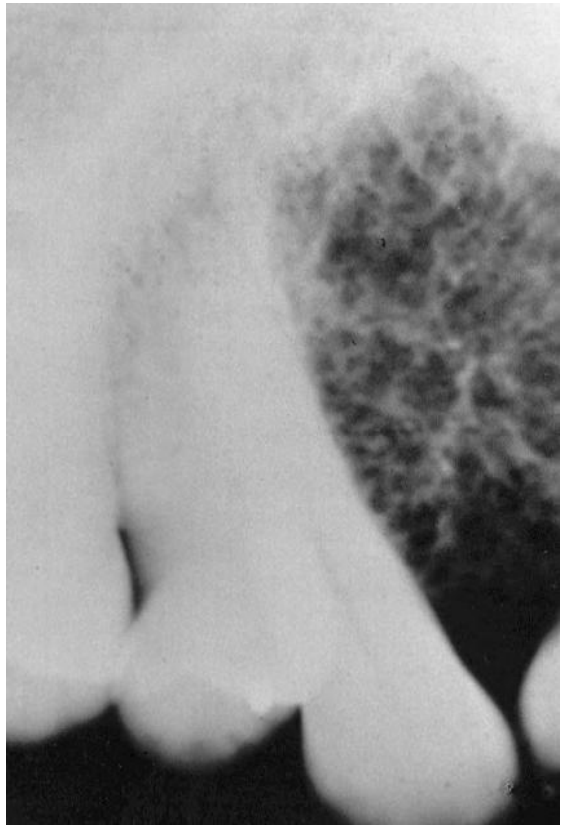
2.1 CENTRAL HAEMANGIOMA

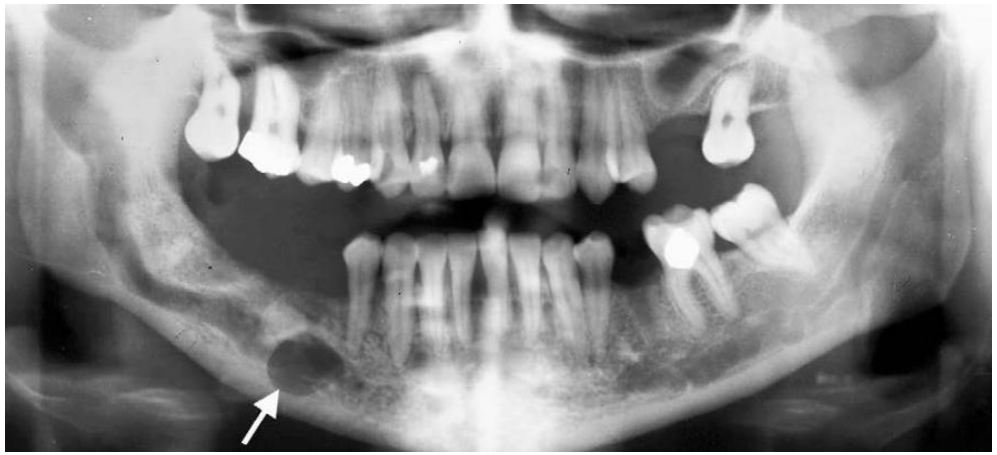
Definition	A benign vascular neoplasm
Age	First decade, although may occur later in life
Gender	F>M (2:1)
Aetiology	Trauma or developmental in origin
Clinical Features	Slow, non-tender, bony hard expansion of the jaw over several months/years +/- pain, compressible and pulsatile (bruit on auscultation) +/- anaesthesia of skin supplied by the mental nerve +/- loosening & migration of teeth (Grade III mobility) +/- bleeding around neck of affected teeth
Management	Should be treated without delay due to the risk of lethal exsanguination. May involve one or a combination of embolisation, surgery, and/or sclerosing techniques.

White & Pharoah, 2014

2.1 CENTRAL HAEMANGIOMA

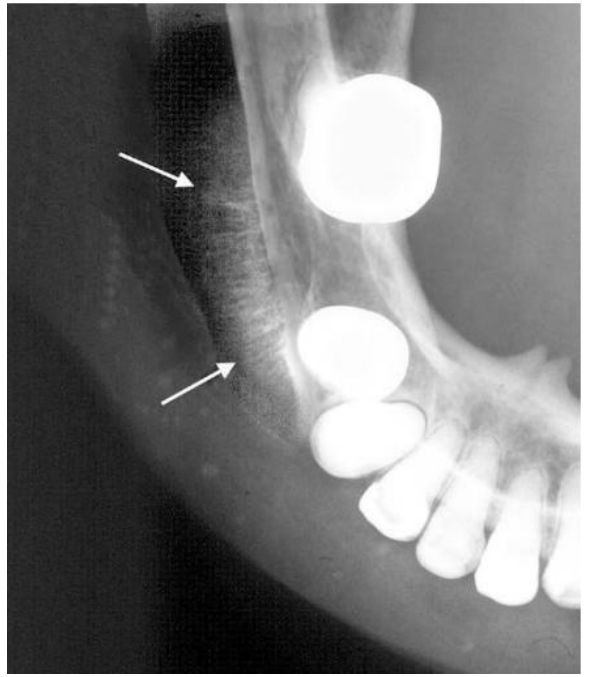
Location	More common in vertebrae or skull. Rarely in jaws. Posterior Mandible - body, ramus, or within the IAC
Periphery / Shape	Well-defined and corticated or ill-defined (simulating malignancy)
Internal Features	When small, unilocular and lucent. Variably multilocular with honeycomb pattern with coarse, dense, well-defined trabeculae If involving the IAC, the whole canal is enlarged with serpiginous shape

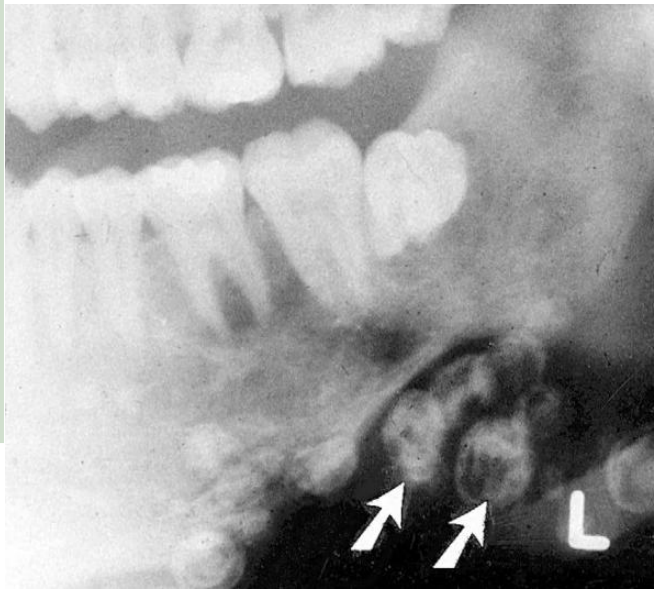




2.1 CENTRAL HAEMANGIOMA

Surrounding Features	• Tooth displacement and root resorption
	• IAC and associated foramina are often enlarged with serpiginous shape
	
	• Involved bone may be enlarged, with coarse trabeculae
	• Enlarged teeth with earlier eruption
	
	• Small, channel-like perforations through the cortices
	• May show spiculated, sun-ray-like periosteal response when it perforates the cortex and involves the periosteum
	
	• Intralesional phleboliths may be present
	  • Presence of phleboliths raise the suspicion of a vascular malformation, typically venous type
	  • Well-defined, round calcifications with targetoid appearance





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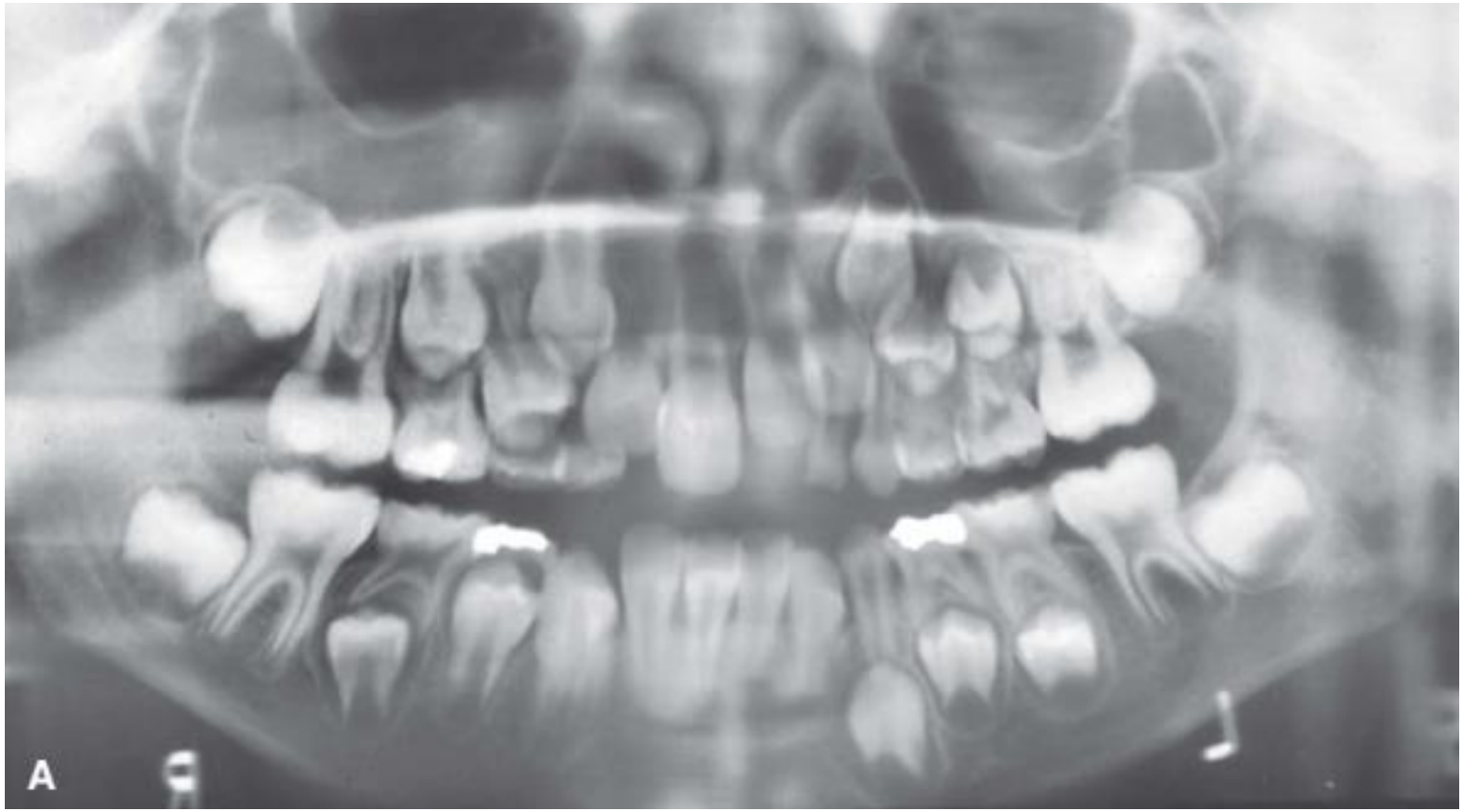
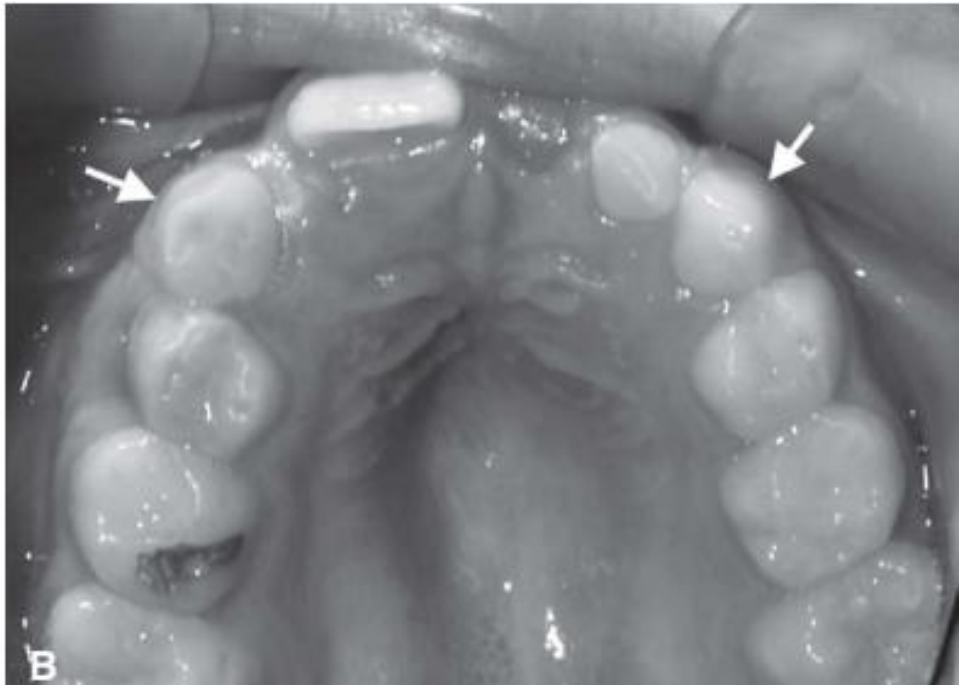
2.1 CENTRAL HAEMANGIOMA





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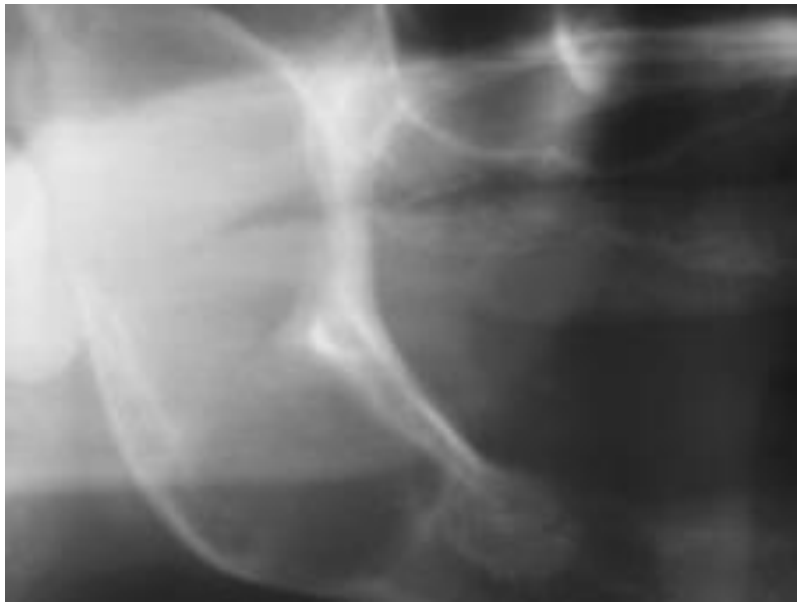
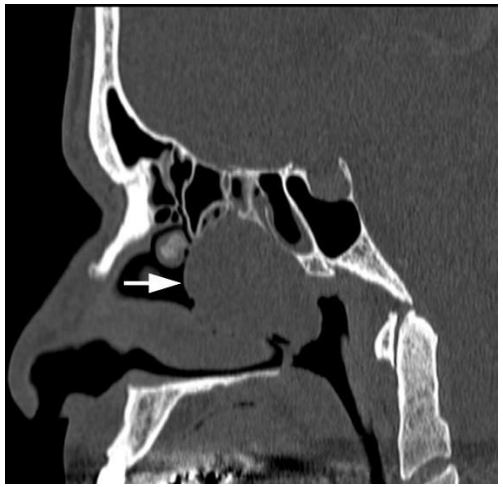
2.2 NEUROFIBROMA

Definition	A benign peripheral nerve sheath tumour consisting of differentiated Schwann cells, fibroblasts, perineurial-like cells, and residual interspersed axons set in a fibromyxoid matrix
Associations	Strongly associated with Neurofibromatosis type I
Prevalence	Most common benign peripheral nerve sheath tumour About 15% of neurofibromas occur in the H+N region
Age	Any age, more commonly 2nd to 4th decade
Gender	M=F
Clinical Features	Slowly growing, often circumscribed and sometimes painful mass
Management/ Prognosis/ Recurrence	Solitary central lesions seldom recur But lesion is non-encapsulated, so periodic review is recommended Potential exists for malignant transformation

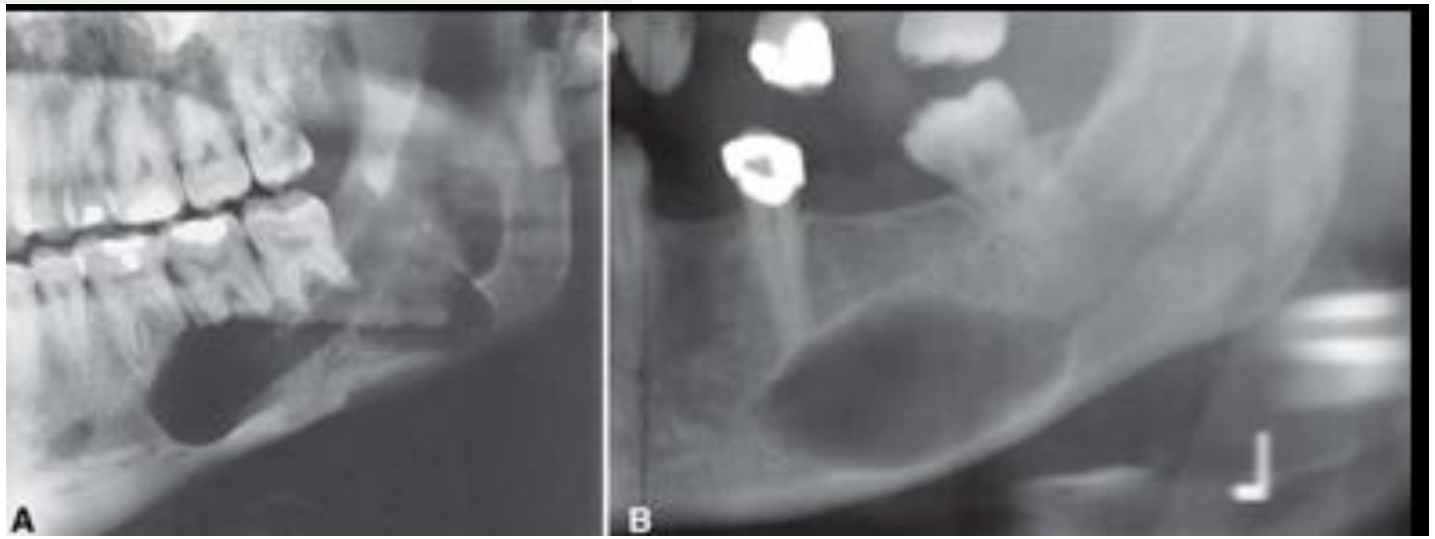
2.2 NEUROFIBROMA

Location	Skin or central lesions Central lesions: IAC, cancellous bone, or below the periosteum
Periphery/ Shape	Well-defined, corticated Some may have indistinct margins
Internal Features	Unilocular (occasionally multilocular)
Surrounding Features	Fusiform enlargement of MC Cortical expansion +/- perforation





WHO, 2022



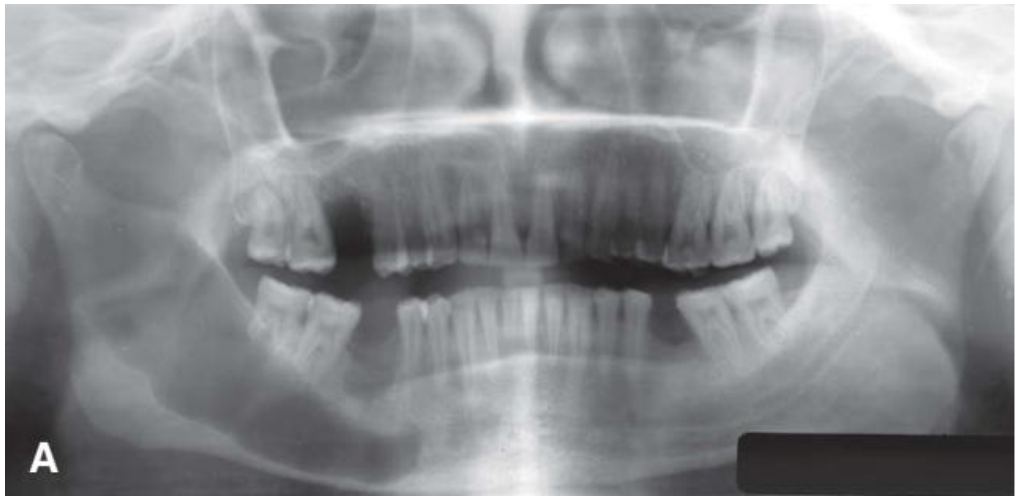
White & Pharaoh, 2014

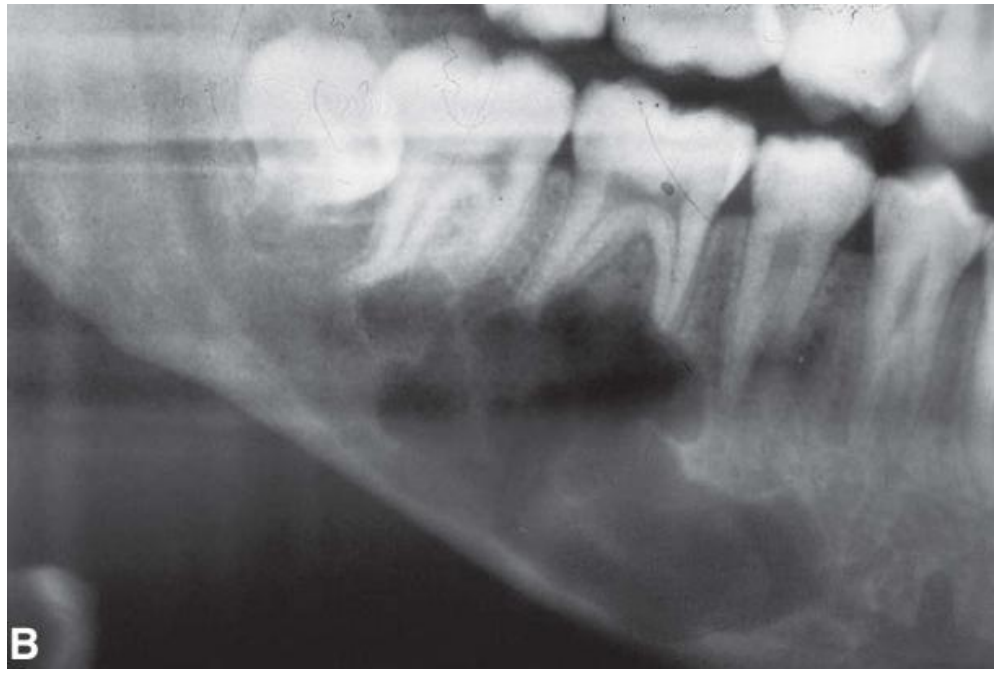
2.3 SCHWANNOMA

Definition	A nerve sheath tumour composed entirely or nearly entirely of differentiated neoplastic Schwann cells.
Prevalence	Up to 40% of schwannomas occur in the head and neck
Age	2nd to 5th decades
Gender	M=F
Aetiology	Most lesions are sporadic. Possible association with neurofibromatosis type 2 and schwannomatosis
Clinical Features	Slowly growing sometimes painful mass Other signs depend largely on size, nerve of origin, and localization
Management/ Prognosis/ Recurrence	Schwannomas are benign Multifocality may mimic recurrence or malignancy

2.3 SCHWANNOMA

Location	Intracranial (almost all involve cranial nerves) Mandible – most often within the IAC
Periphery/ Shape	Well-circumscribed, corticated. Small lesions are cyst-like, but end to be fusiform in shape Large lesions demonstrate scalloping outline.
Internal Features	Typically lucent. Calcifications rare. Larger lesion demonstrate more heterogenicity
Surrounding Features	• Enlargement of the mandibular/mental foramen • Outer cortex of canal is maintained • Expansion of canal is localised with a definite epicentre unless lesion is large • +/- root resorption





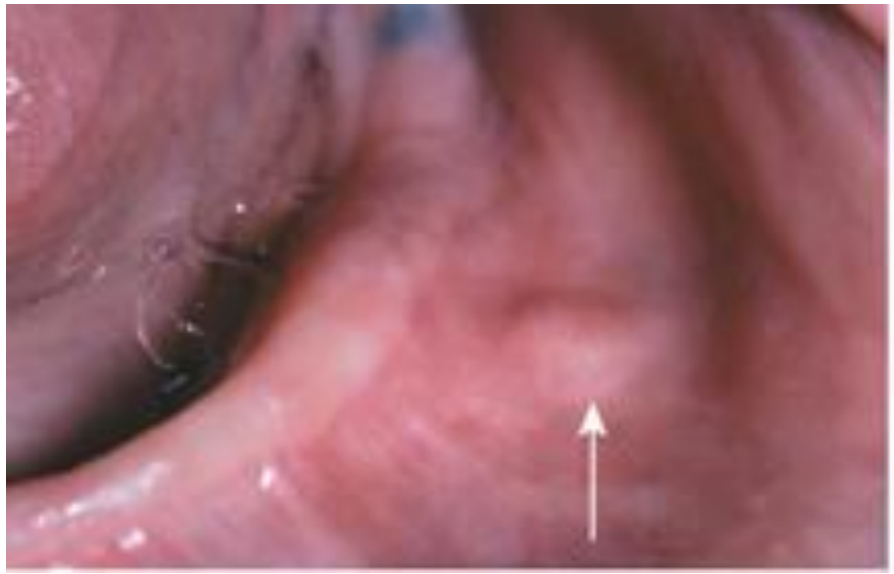
White & Pharoah, 2014

2.4 NEUROMA

Definition	Comprise a diverse group of peripheral nerve sheath tumors, some reactive and hyperplastic, including the Traumatic neuroma (TN).
Aetiology	Accidental or iatrogenic nerve injury or amputation.
Clinical Features	Slow-growing, reactive hyperplasias, seldom >1cm in diameter Severe pain (as the mass applies pressure within its bony cavity or a result of external trauma), reflex neuralgia, referred pain to eyes, face, and head.
Management/ Prognosis/ Recurrence	Simple excision when symptomatic. Recurrence is uncommon.

2.4 NEUROMA

Location	TN occurs in areas of nerve injury, including bones Mental foramen > anterior maxilla > posterior mandible (often within the IAC)
Periphery/ Shape	Well-circumscribed, corticated. Various shapes, depending on amount of resistance to expansion
Internal Features	Radiolucent
Surrounding Features	Expansion of IAC



WHO, 2017

REFERENCES

WHO Classification of Tumours Editorial Board. Head and neck tumours. Lyon (France): International Agency for Research on Cancer; 2022 (WHO classification of tumours series, 5th ed.; vol. 9).

Koong, B. (2017). *Atlas of Oral and Maxillofacial Radiology*. Chichester, UK: John Wiley and Sons.

White, and Pharoah. (2014). *Oral radiology : Principles and interpretation* (7th ed.). St. Louis: Elsevier/Mosby.