Soft tissue tumors and lesions. Bone pathology.

Oral pathology

Soft tissue tumors and lesions



- mesenchymal tumor-like lesions
- true neoplasms
- majority are rare in oral cavity
- Diff. dg: hyperplastic lesions

Fibrous lesion of the oral mucosa



Hyperplastic lesions:

- epulides (fibrous, vascular, giant cell);
- pyogenic granuloma;
- fibroepithelial polyp;
- denture irritation and papillary hyperplasia

Neoplastic and neoplastic-like lesions:

- peripheral odontogenic fibroma
- fibrosarcoma
- fibrous histiocytoma
- nodular fasciitis
- fibromatosis

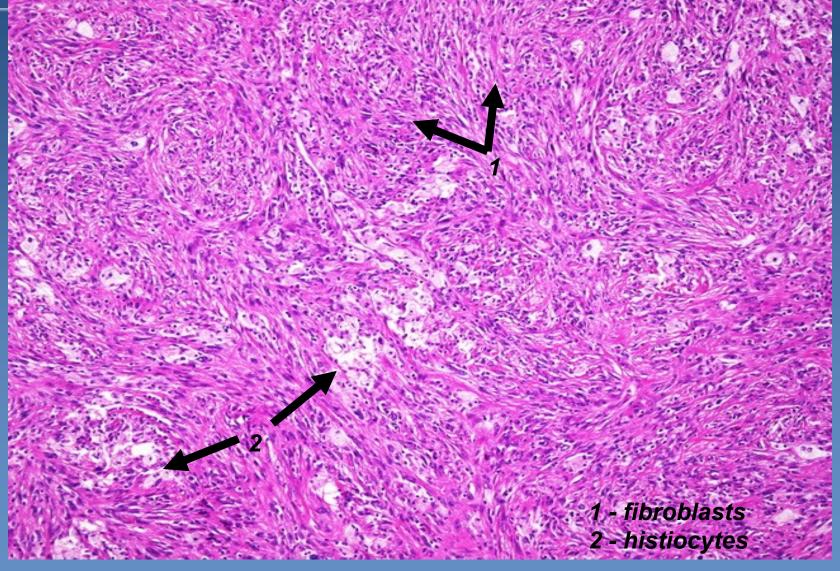
Fibrous histiocytoma



- show both fibroblastic + histiocytic differentiation
- middle-aged and older adult
- *buccal mucosa and vestibule
- nodular mass vary in size

Fibrous histiocytoma





Nodular fasciitis



- rare in oral cavity, superficial
- rapidly growing pseudosarcomatous lesion
- *self-limiting growth of a clone of neoplastic cells that contain a fusion gene (*USP6* rearrangement)
- painfull, tenderness
- histologically may resemble fibrosarcoma

Fibromatosis



- non-neoplastic but infiltrative fibrous lesion do not metastasize!!!
- children or young adults (juvenile fibromatosis)
- paramandibular soft tissue region
- vary in size
 → facial disfigurement
- firm mass with rapid growth + destruction adjacent bone

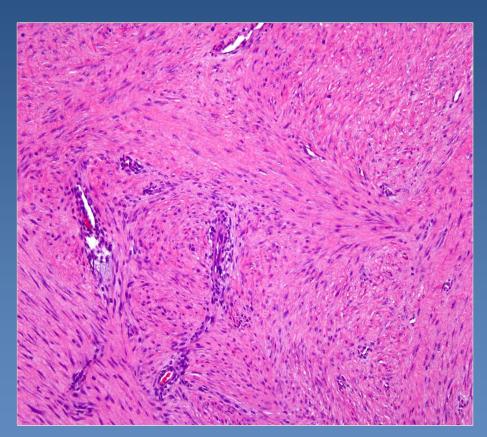
Micro: proliferation of spindle-shaped cells, no cytonuclear atypia

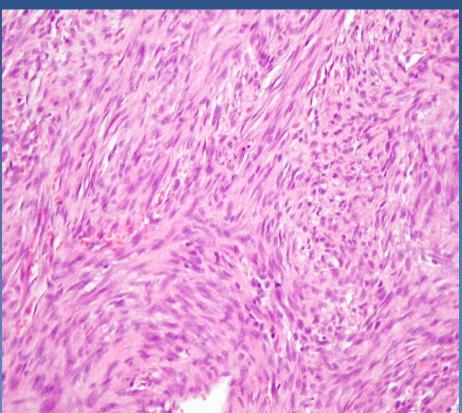
Treatment: wild excision

recurrence rate 23%

Fibromatosis







proliferation of spindle-shaped cells, without cytonuclear atypia





- malignant tumor of fibroblasts
- rare in oral cavity

have good prognosis

young adults and children

Micro: fascicles of fibroblasts that forms "herringbone" pattern

Fibrosarcoma









Lipoma – benign tumor of adipose tissue

- 40 yrs or older
- mucosa of cheeks and tongue

!!! ulcerated tumor-like masses of partly necrotic fat in very young children - the result of traumatic herniation of cheek's mucosa

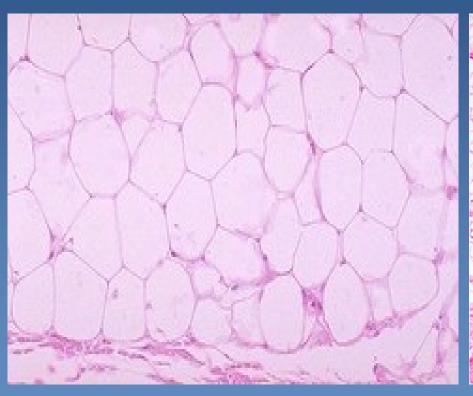
Gross: soft, yellowish-colored swelling

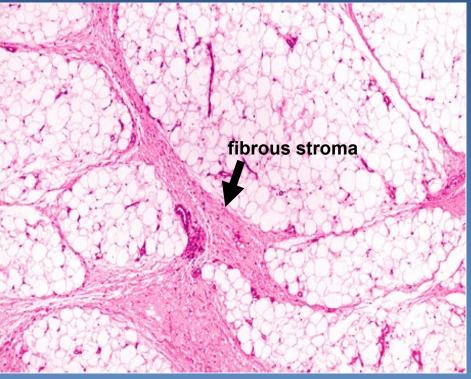
Micro: mature adipose tissue, thin fibrous capsule

Fibrolipoma – lipoma with increasing amount of fibrous stroma

Tumors of adipose tissue







lipoma

fibrolipoma



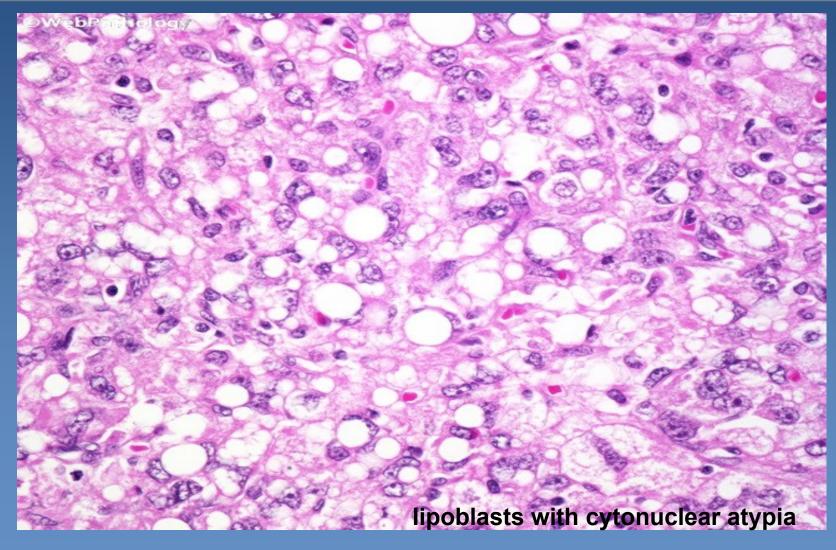


Liposarcoma – malignant tumor of adipose tissue

- peak 40-60 yrs
- cheeks, floor of the mouth, tongue
- resemble benign lipoma + atypical hyperchromatic nuclear
- myxoid, round-cell, well-differentiated, pleomorphic, dedifferentiated
- Have a good prognosis in oral cavity











Hemangioma — benign hamartomatous tumor

- 1-year-old children, F:M 3:1
- lips, tongue, cheeks or palate

Gross: solitary, flat or raised, dark reddish-purple in color typically blanch on pressure

Hereditary haemorrhagic telangiectasia – AD, multiple telangiectases in skin, mucous membrane, internal organs

Sturge-Weber syndrome – haemangiomatous lesions of the face (n. trigeminus) + heamangiomas and calcification of leptomeninges + limbs affecting





Histological types: capillary, cavernous, mixed

Complications: ulceration

thrombosis

organization

calcification

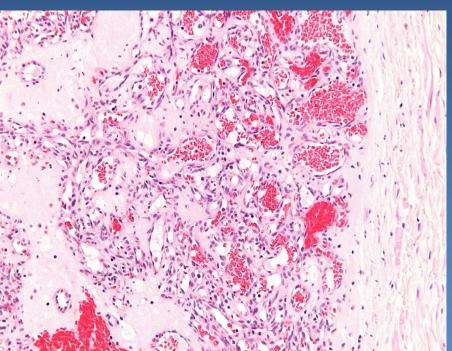
Treatment: "watchful neglect", occur regression

Sublingual varicosities - purplish venous ectasia on the ventral (undersurface) of the tongue after the age of fifty.

Hemangioma







sublingual hemangioma

capillary type – proliferation of capillary-sized vessels

Tumors of vascular tissue



Lymphangioma - benign hamartomatous tumor lymphatic vessels

- early childhood
- anterior 2/3 of the tongue (macroglossia)
 trauma ⇒ sudden increase in size

Gross: pebbly surface

Micro: endothelial-lined spaces containing lymph

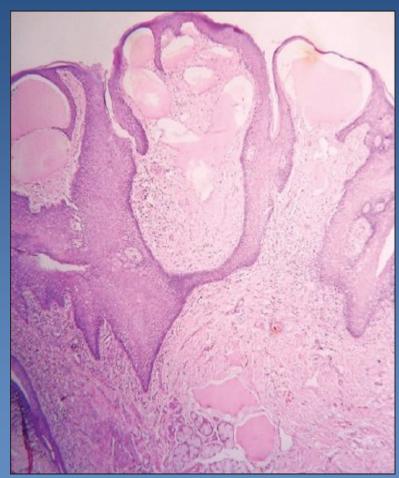
Cystic hygroma – large, fluctuant swelling (>10 cm) of the head and neck region, may extend to oral cavity.

Lymphangioma





pebbly surface of the tongue



dilated lymphatic vessels beneath the epithelium

Tumors of peripheral nerves



- 1. Neurofibroma
 - solitary
 - multiple
- 2. Neurinoma (Schwannoma)
- 3. Multiple mucosal neuromas (MEN sy)
- 4. Traumatic neuroma
- 5. Granular cell tumor

Tumors of peripheral nerves



Neurofibroma — mixture proliferation of Schwann cell and fibroblasts

- young adults, children

Solitary lesion – tongue, buccal mucosa, well-circumscribed nodules

Multiple lesions associated with neurofibromatosis (von Recklinhause's disease of nerves) – AD, mutations in NF1 gene, located 17q11.2

skin pigmentation (coffee and milk) + involving of cutaneous nerves + axillary freckling + oral lesions

!!! May be associated with tumors of CNS, leukemia, RMS, WT...

Tumors of peripheral nerves



Oral lesions:

- mucosal swelling of the tongue, gingiva
- enlargement of the fungiform papilla
- enlargement of mandibular foramen
- increased bone density

Risk of malignizatio⇒ MPNST (neurofibrosarcoma) 5-15%

Treatment: no specific therapy

Neurofibromatosis







Involving of cutaneous nerves + mucous swelling of the tongue, gingiva





Neurinoma (Schwannoma) - benign neoplasm of Schwann cell origin

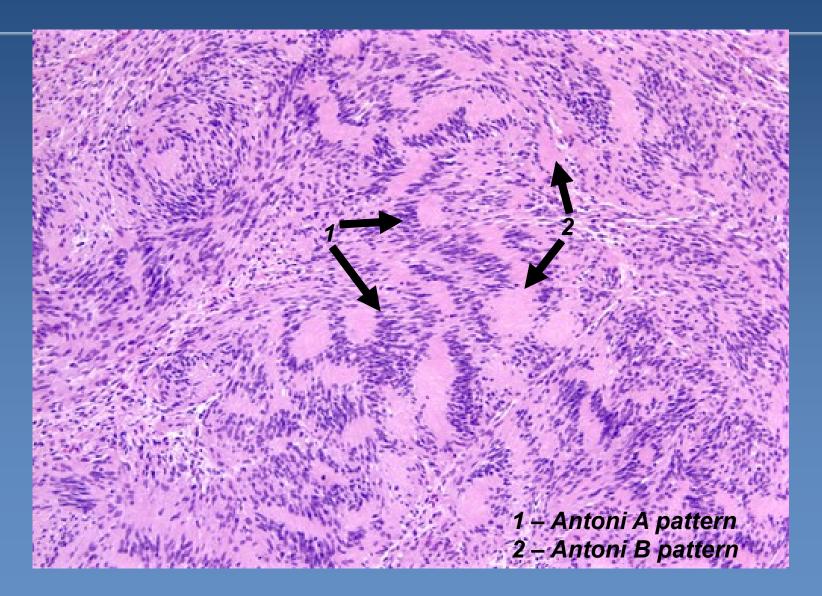
- young and middle-aged adults
- tongue, intraosseous in the posterior mandible

Gross: vary in size, encapsulated tumor

Micro: 2 patterns – Antoni A (palisaded nuclei, Verocay bodies)
Antoni B (less cellular fields)

Treatment: surgical excision

Neurinoma (Schwannoma)







Traumatic neuroma (amputation neuroma) – tumor-like reactive proliferation of Schwann cells

- cause: transection or other damage of a nerve bundle
- middle-age adults, F>M
- mental foramen area, tongue, lower lip
- painful lesion!!!

Gross: small nodule

Micro: proliferation of mature nerve bundles, fibrotic stroma, mild chronic inflammation

Treatment: surgical excision (incl. involved nerve bundle)





Granular cell tumor — tumor of unknown origin (in the past was called the granular cell myoblastoma)

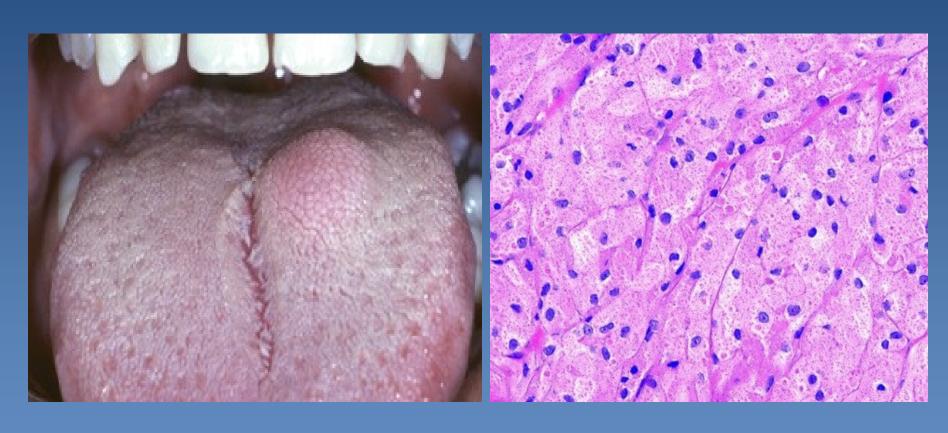
- wide age range
- dorsal surface of the tongue

Gross: non-encapsulated lesion

Micro: large cells with granular cytoplasm, pseudoepitheliomatous hyperplasia of the overlying epithelium

Granular cell tumor





non-encapsulated lesion of tongue's mucosa

large cells with granular cytoplasm



Tumors of muscle

Leiomyoma – benign smooth muscle tumors

probably, leiomyomas of the oral cavity have their origin from vascular smooth muscle

Leiomyosarcoma and rhabdomyosarcoma are both very rare in oral cavity



Bone pathology

Inherited and development disorders of bone

- 1. Osteogenesis imperfecta
- **∞**2. Osteopetrosis
- ×3. Cleidocranial dyspasia
- 4. Achondroplasia
- **★**5. Fibro-osseous lesions
- **★**6. Cherubism

Inherited disorders of bone

- *uncommon diseases
- jaw involvement variable
- orofacial manifestations include:
 - abnormalities in number, form, structure of teeth
 - malocclusion
 - abnormal facial appearances

Osteogenesis imperfecta



- *AD, mutations in the genes that code for type-1 collagen (80-90%)
- *generalized osteoporosis (slender bones)
 Clinically 4 main type:
- ➤ Type I (classic type) AD, blue sclera, deafness, +/dentinogenesis imperfecta
- ➤ Type II (perinatal lethal) AD
- **▼** Type III (progressively deforming) *AD/AR*, severe osteoporosis, progressive deformities, dentinogenesis imperfecta
- **▼** Type IV AD, similar to type I, but more severe

Micro: immature, woven bones of cortex

Osteogenesis imperfecta





Dentinogenesis imperfecta:

note delicate bone trabeculae + obliteration of pulp chambers

Osteopetrosis (marble bone disease)



- excessive density of all bones
- ★obliteration of marrow cavities → secondary anemia
- defect in osteoclastic activity, failure in the remodeling of the developing bone
- **★**bones mechanically week, common fractures!!!

Symptoms: delayed eruption of teeth, osteomyelitis (after tooth extraction)

Radiography: mandible>>maxilla, invisible roots of the teeth

Cleidocranial dysplasia (cleidocranial dysostosis)

- *AD, mutations to the RUNX2 gene
- disturbance of differentiation of osteoblasts from precursor cells
- *abnormalities of the skull, jaws, clavicle (partial/complete absence)
- maxilla with a high, narrow arched palate
- delayed or non-eruption of the permanent dentition, supernumerary teeth

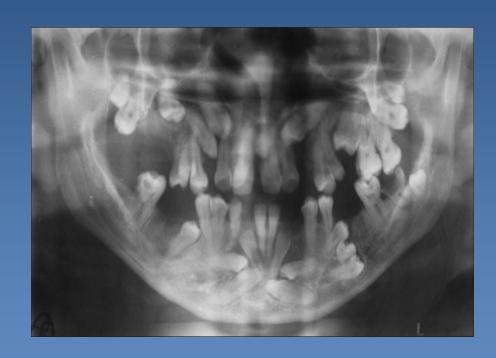
Radiography: thin teeth's roots

Cleidocranial dysplasia





Complete absence of clavicles



Retention of deciduous teeth + multiple impactions of permanent teeth

Fibro-osseous lesions



Divided into:

- I. Osseous dysplasia
 - 1. Fibrous dysplasia (monostotic/polyostotic)
 - 2. Cemento-osseous dysplasia
- II. Benign neoplasia (ossifying fibroma)
- replacement of normal bone by cellular fibrotic tissue
- contain woven bone + acellular islands of mineralized tissue develop

Fibrous dysplasia (FD)



development disorder, but not inherited

Monostotic FD: much more common!

- childhood, adolescencereactivation of quiescent lesion during pregnancy
- affected 1 bone: limb, skull bones, particularly the jaws maxilla>>mandible

Craniofacial fibrous dysplasia – 1 bone is affected (maxilla) + involvement of adjacent bones

Symptoms: painless swelling of the maxilla (buccall → facial asymmetry rapid and extensive grow → exophthalmos mandibular lesion → fusiform expansion + displacement of teeth

Gross: ill-defined smooth enlargement

Radiography: ground-glass/orange-peel-stippling effect displaced teeth, separated roots

Fibrous dysplasia (FD)





Fibrous dysplasia

Monostotic form more common in the craniofacial region Ground glass change with areas of sclerosis (arrows) More ill-defined border compared to ossifying fibroma





Polyostotic FD

- *affected several bones, segmentally lesions
- *affected sites: limb (lower), skull bones, vertebrae, ribs, pelvis
- childhood, F:M 2-3:1

expansion usually stops with skeletal maturation

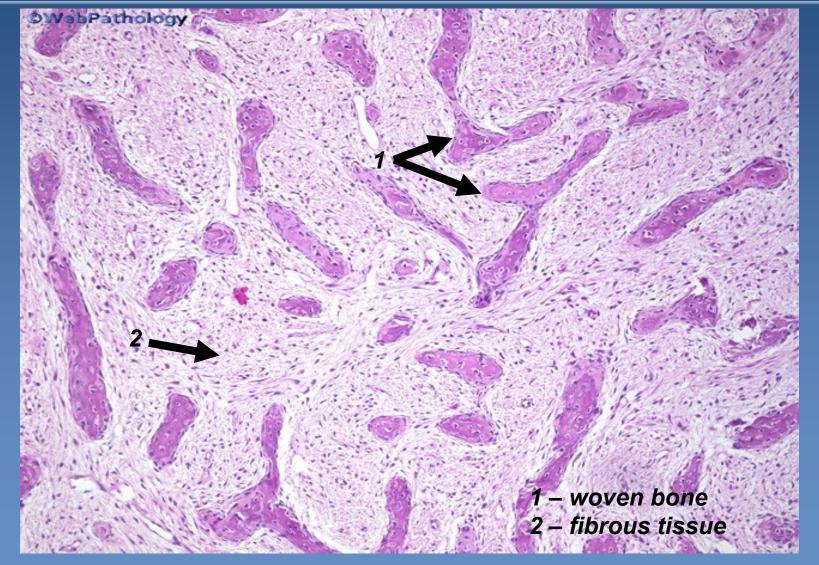
McCune-Albright syndrome – bone lesions are accompanied by skin pigmentation, sexual precocity, endocrine abnormalities

Micro: delicate trabeculae of woven bone + fibrous tissue
Remodelling of woven to lamellar bone may occur with increasing age!!!

Treatment: not radiosensitive !!! (risk of malignant transformation to fibrosarcoma)

Fibrous dysplasia (FD)





Cemento-osseous dysplasia



- osseous dysplasia of jaws, which involves the tooth-bearing areas
- *F>>M, over 30 yrs old, mandible>>maxilla
 Based on the clinical and radiographic features:

 periapical, focal and florid cemento-osseous dysplasia

Clinically:

multiple and small <1 cm	multiple and large >1 cm
associated with apical areas of the mandibular incisors	involve 1 or more quadrants in one or both jaws

Micro: fibrous tissue + bone/calcified accelular tisue develop

Radiography: radiolucent/mixed/radiopaque

Inflammatory diseases of bone

- 1. Alveolar osteitis (dry socket)
- *2. Focal sclerosing (condensing) osteitis
- 3. Osteomyelitis
- *****4. Chronic periostitis
- ★5. Radiation injury and osteoradionecrosis

Osteomyelitis



- now is a rare disease
- polymicrobial infection

Predisposing factors:

Local factors

- trauma
- radiation injury
- Paget's disease
- osteopetrosis
- major vessel disease

Systemic factors

- immune deficiency states
- *****immunosuppresson
- **≭**DM
- *malnutrition
- extremes of ages

Suppurative osteomyelitis

- clinically: acute, chronic (>1 month)
- mandible>maxilla
- **▼**source of the infection − dental abscess, fractures, penetraiting wounds, extractions

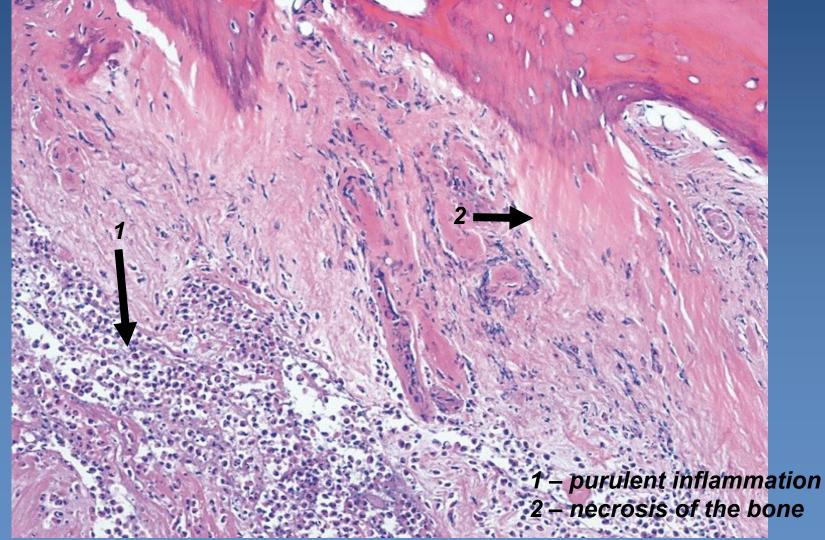
Symptoms:

acute lesion- pain, swelling, pyrexia, malaise, mobility of teeth chronic – discharge of pus through 1 or more sinuses

Micro: suppurative inflammation, necrosis of the bones, pus within marrow spaces, vascular thrombosis

Complication: sequestrum (exfoliated through a sinus) surgical removing

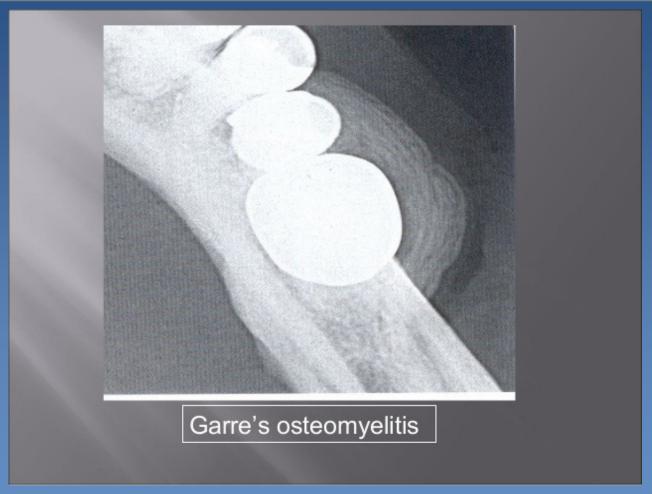




Chronic osteomyelitis with proliferative periostitis

- *syn. Garré's osteomyelitis, periostitis ossificans
- type of sclerosing osteomyelitis
- mandible, children and young adults
- Gross: swelling on the outer surface of the mandible
- Micro: subperiosteal mass of trabeculae of woven bone + chronic inflammation in fibrous marrow

Chronic osteomyelitis with proliferative periostitis



Subperiosteal mass in mandible

Metabolic and endocrine disorders of bone



- **≭**1. Osteoporosis
- 2. Primary hyperparathyroidism
- *3. Secondary hyperparathyroidism
- *4. Rickets and osteomalacia
- **★**5. Acromegaly

Osteoporosis



- excessive bone loss/when the apposition of bone is reduced
- **≭**F:M 2:1
- *postmenopausal women (rate of bone's loss 1-8% per year)
- edentulous patients (mandible)
- *accentuated in Cushing syndrome, thyrotoxicosis, primary hyperparathyroidism
- osteoporotic bone is reduced in quantity

Radiography: increased radiolucency, thin cortex





Primary

- secretion of parathormone (adenoma/Ca, hyperplasia PG)
- hypercalciemia, hypercalciuria + pathological metastatic calcification

Micro: brown tumor (haemosiderin + fibrotic tissue + multinucleated, osteoclast-like giant cells)

Secondary

- response to chronic hypocalciemia (CRI)
- may associated with rickets and osteomalacia

Micro: uncalcified osteoid + brown tumor

may affect jaws

Paget's disease of bone



- *form of osteodystrophy, disorganized formation and remodeling of bone
- **x**aetiology unclear genetic and environmental factors, paramyxovirus infection
- >40 yrs, more common in maxilla Phases:
- 1. Osteolytic
- 2. Mixed osteolytic and osteogenesis
- 3. Osteoblastic

Paget's disease of bone



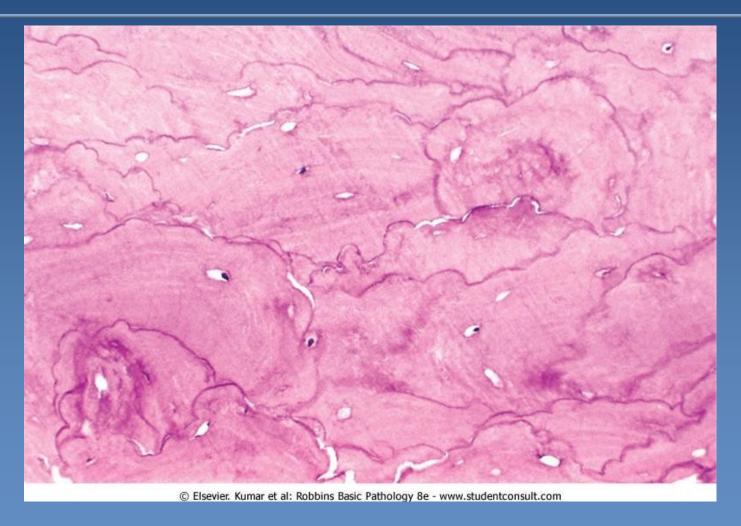
Symptoms: bone pain, cranial nerve compression, facial deformity, difficulties in wearing dentures hypercementosis, ankylosis ⇒ difficulty in extraction root resorption (1 phase) increased alkaline phosphatase

Micro: criss-crossing reversal lines, mosaic bone

Complication: risk of malignant transformation (osteosarcoma)

Paget's disease of bone





criss-crossing reversal lines, mosaic bone

Tumors of bone



1. Bone-forming tumors

Benign: Osteoma

Osteoblastoma

Malignant: Osteosarcoma

2. Cartilage-forming tumors

Benign: Chondroma

Malignant: Chondrosarcoma

3. **Marrow tumors**: Myeloma

4. Histiocytic and dendritic cell neoplasms Langerhans cell histiocytosis

5. **Vascular tumors**: Haemangioma of bone

6. Fibrous tumors: Ossifying (cemento-ossifying) fibroma

7. Metastatic tumors





Osteoma – benign, slow-growing tumor

*adults, mandible>maxilla

Gross: solitary, well-circumscribe lesion

multiple osteomas of the jaws occur as a feature of Gardner sy

Micro: compact type: dense lamellar bone cancellous type: interconnecting trabeculae + fibrous marrow

Osteoblastoma – rare tumor in the jaws

Micro: cementoblastoma (!!!not related to the roots of the teeth)





Osteosarcoma – primary malignant Tu of bone

- 30 yrs and older
- relatively rare in jaws

Intramedullary type - arise centrally within the jaws

Juxtacortical type – peripherally in the relation to the periosteum, better prognosis

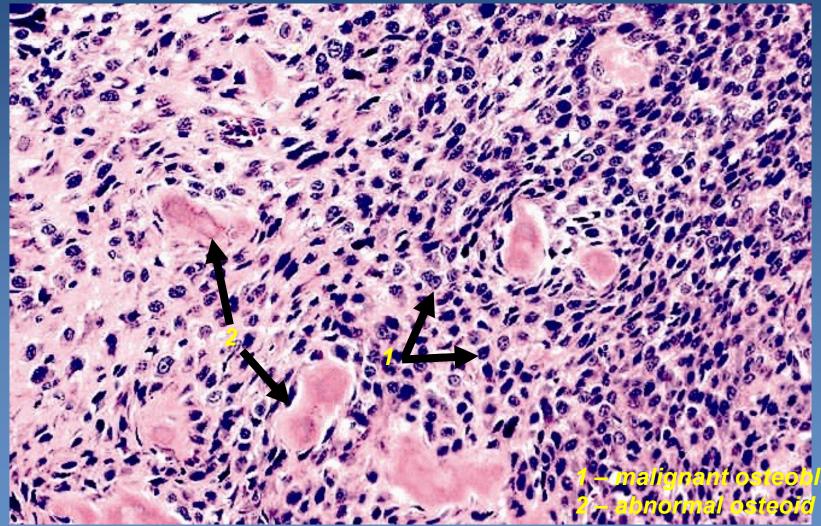
Micro: malignant osteoblasts + abnormal osteoid

Metastasis: RLN, lungs, brain

Treatment: neoadjuvant CT+ surgical removal + adjuvant CT

Osteosarcoma





asts

Cartilage-forming tumors



Chondroma – rare benign Tu in the jaws

- **×**3-4 decades
- Mandible (condylar process, posterior part)
 Maxilla (anterior part)

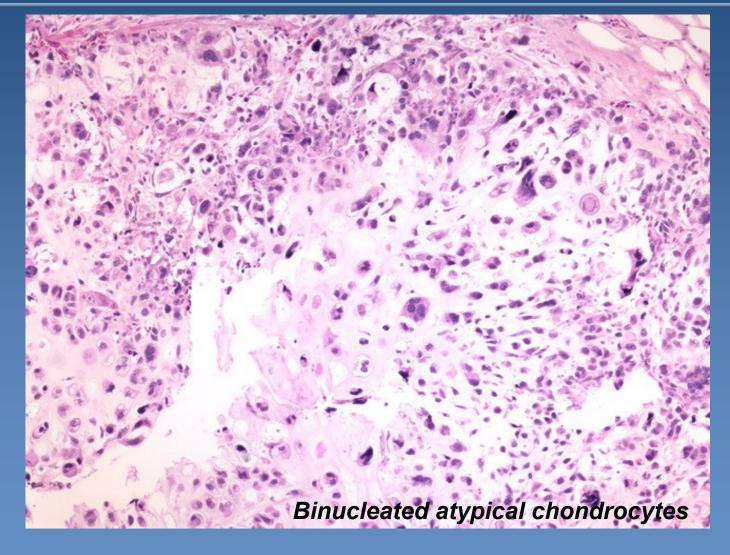
Micro: circumscribed mass of mature hyaline cartilage

↑ cellularity, binucleated cells ⇒susp well-differentiate
 chondrosarcoma !!!

Prognosis: better for mandibular lesions

Chondrosarcoma







Marrow tumors

Myeloma – plasma cells neoplasm

Multiple myeloma – disseminated disease involving many bones Solitary myeloma (plasmocytoma) – solitary lesion

- **≈** 50-70 yrs
- ***skull**, vertebrae, sternum...(sites with red marrow)
- *abnormally high levels of single homogenous type *Ig* in serum (paraprotein)

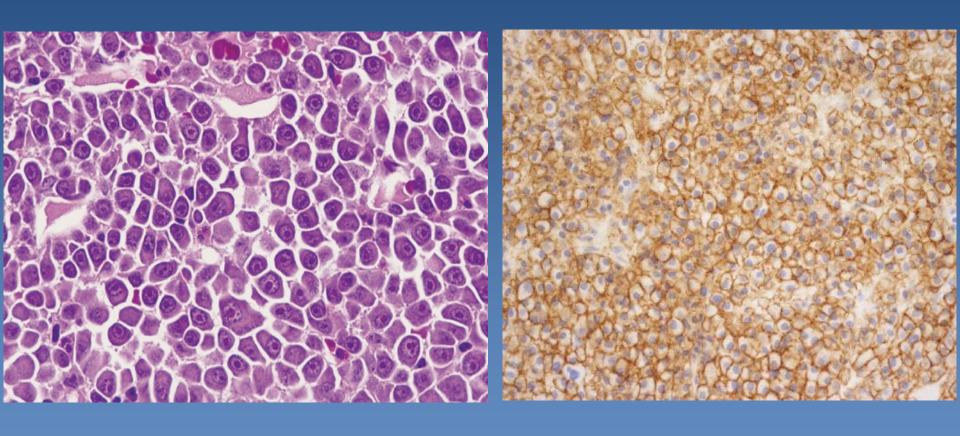
Radiography: osteolytic lesions (punched-out radiolucencies)

Micro: cellular sheets of Tu cells resemblance to plasma cells

IHC: positivity of CD20, CD138, kappa, lambda

Myeloma





Tumour's cells resemblance to plasma cells

IHC: positivity of CD138





Langerhans cell histiocytosis – clonal proliferation of Langerhanstype cells

3 forms: monoostotic (with solitary osteolytic lesion, "eosinophilic granuloma") polyostotic (with multiple osteolytic lesion, soft tissue involvement) disseminated and multisystem disease

Previous classification: eosinophilic granuloma, Hand-Schüller-Christian disease, Abt-Letterer-Siwe disease

Unifocal/multifocal "eosinophilic granulomas":

- <20 yrs, M:F 2:1

- cranuim and jaws (mandible)

Radiography: solitary/multiple osteolytic lesions

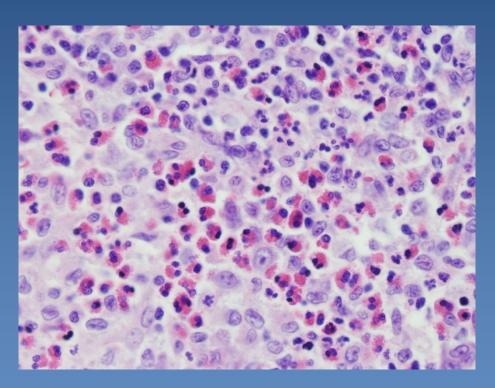
Micro: histiocytes + variable numbers of eosinophils

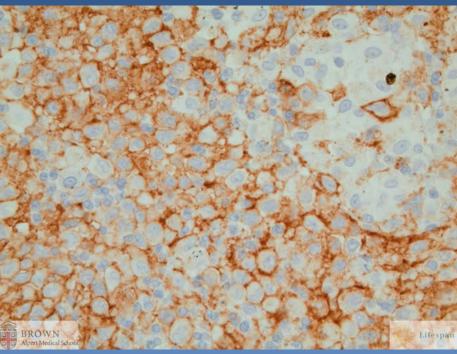
EM: Birbeck granules

IHC: positivity of CD1α, S100

Langerhans cell histiocytosis







Histiocytes + variable numbers of eosinophils

IHC: positivity of CD1a





Ossifying (cemento-ossifying) fibroma — benign well-demarcated !!! neoplasm

wide age range, F>M

rapid growth in children/adolescence - juvenile ossifying fibroma

Micro: well circumscribed cellular fibrous tissue + trabeculae of bone

Diff.dg: fibrous dysplasia

Juvenile ossifying fibroma – richly cellular + high mitotic activity + immature-looking woven bone (recc. rate 30-60%)

Diff. dg: osteosarcoma

Metastatic tumors



- ★1% of malignant Tu of oral cavity
- Mandible>>maxilla gingiva, alveolar mucosa, tongue
- Ca of breast, bronchus, kidney...
- Mts may cause: osteolytic changes osteoblastic changes

