

# Soft tissue neoplasms



# Soft tissue neoplasms

## □ Arising from connective tissue

- n Fibroblasts
- n Blood vessels
- n Nerve
- n Muscle
  - p Skeletal
  - p Smooth



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# Soft tissue neoplasms

- ☐ Fibroma
  - ☐ Giant cell fibroma
  - ☐ Inflammatory papillary hyperplasia
  - ☐ Fibrous histiocytoma
  - ☐ Pyogenic granuloma
  - ☐ Peripheral giant cell granuloma
  - ☐ Peripheral ossifying fibroma
-



# Soft tissue neoplasms

## □ Neural:

- n Traumatic neuroma
- n Palisaded encapsulated neuroma
- n Neurilemmoma
- n Neurofibroma
- n Neurofibromatosis

## □ Neuro-endocrine:

- n Multiple endocrine neoplasia
  - n Melanotic neuroectodermal tumor of infancy
-



# Soft tissue neoplasms

## □ Vascular

- n Hemangioma
- n Vascular malformations
- n Sturge-weber syndrome
- n Hemangiopericytoma
- n Lymphangioma

## □ Muscle:

- n Leiomyoma
  - n Rhabdomyoma
-



# Malignant Soft tissue neoplasms

- ☐ Fibrosarcoma
- ☐ Malignant fibrous histiocyoma
- ☐ Liposarcoma
- ☐ Malignant schwannoma
- ☐ Malignant peripheral nerve sheath tumor
- ☐ Angiosarcoma
- ☐ Kaposi's sarcoma
- ☐ Leiomyosarcoma
- ☐ Rhabdomyosarcoma
- ☐ Synovial sarcoma
- ☐ Alveolar soft part sarcoma
- ☐ Metastasis to oral tissues.



# Fibroma

- Also called irritation fibroma, traumatic fibroma
- Most common tumor of the oral cavity
- It is a reactive hyperplasia of fibrous connective tissue in response to local irritation



# Clinical features

- Common in buccal mucosa along the occlusal line, labial mucosa, tongue and gingiva
  - Could represent fibrous maturation of a pre-existing pyogenic granuloma
  - Smooth surfaced, sessile, pink nodule about 0.5cm in diameter,
  - 4<sup>th</sup>-6<sup>th</sup> decades, in females
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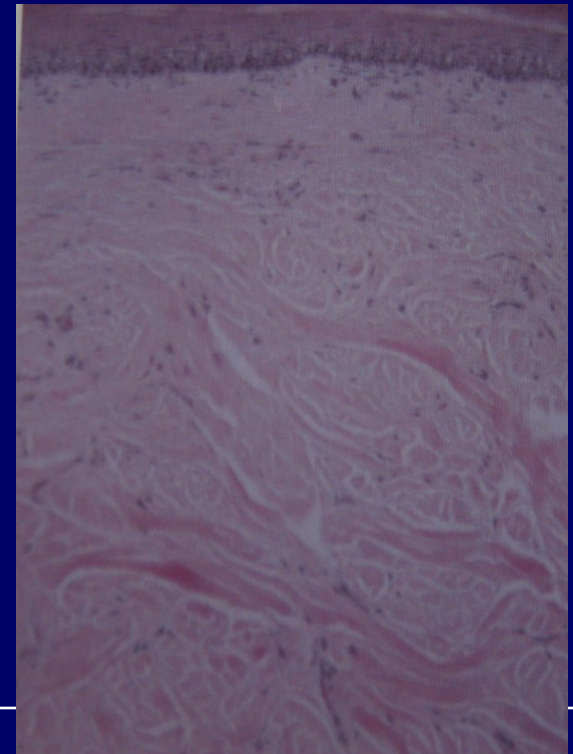
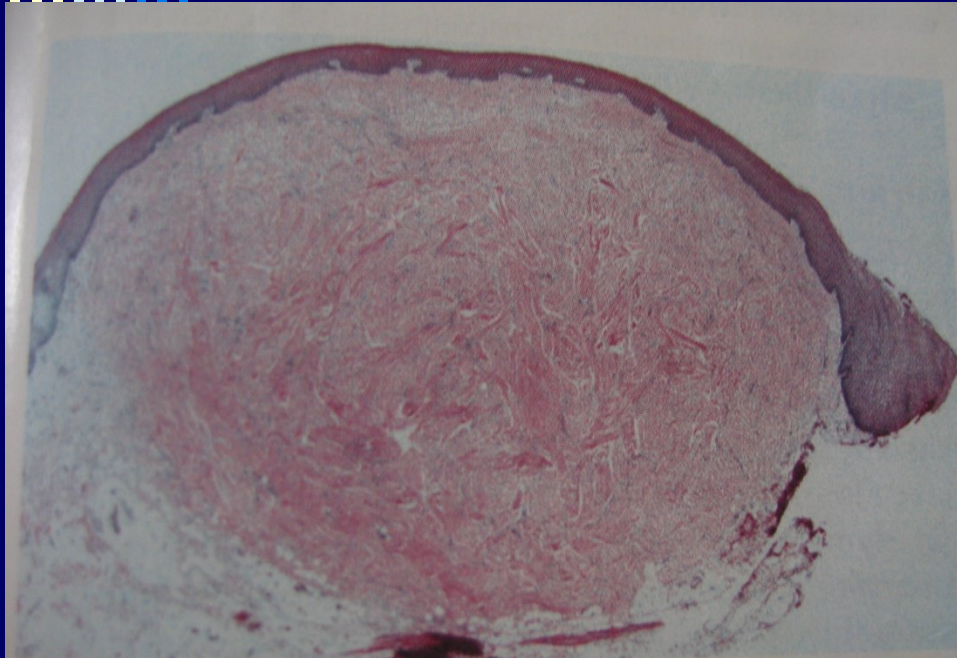
# Fibroma





# Histopathology

- Nodular mass of fibrous connective tissue covered by stratified squamous epithelium.
  - CT is dense and collagenized and it blends gradually into the surrounding connective tissue
  - Overlying epithelium could be atrophic and minimal inflammation.
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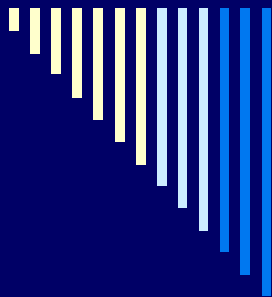
# Ttt and prognosis

- Surgical excision
  - Recurrence is extremely rare
-



# Giant cell fibroma

- Fibrous tumor not associated with chronic irritation
  - Asymptomatic, sessile or pedunculated nodule, <1cm in size, papillary surface
  - Usually in first three decades of life
  - Predominantly in gingiva – mandible
  - D/D Retrocuspid papilla – developmental lesion, bilateral, anatomical variation that disappears with age.
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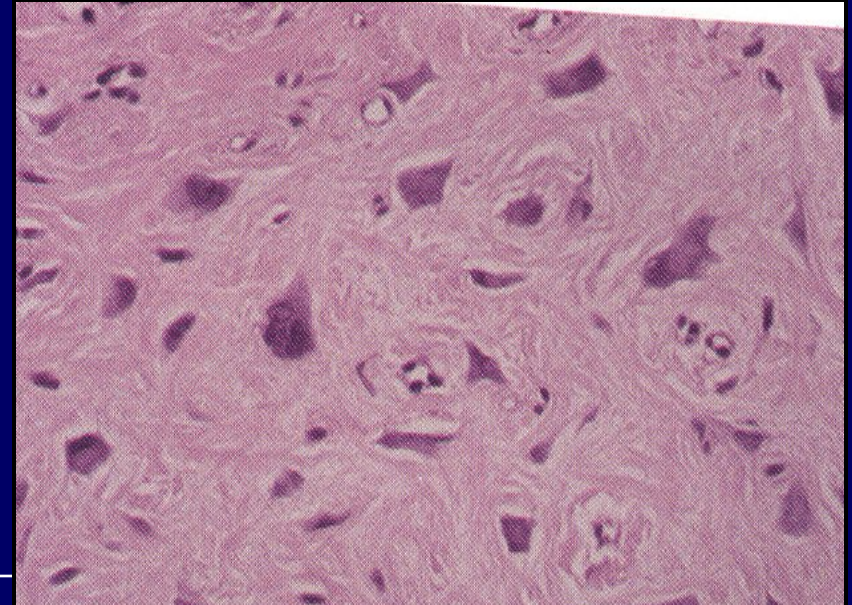


# Histopathology

- Mass of vascular, loosely arranged, fibrous connective tissue
  - Numerous large, stellate fibroblasts within the superficial connective tissue
  - Cells may have multiple nuclei
  - Overlying epithelium is thin and atrophic
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# GC fibroma







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# Ttt and prognosis

- Conservative surgical excision
  - Recurrence is rare
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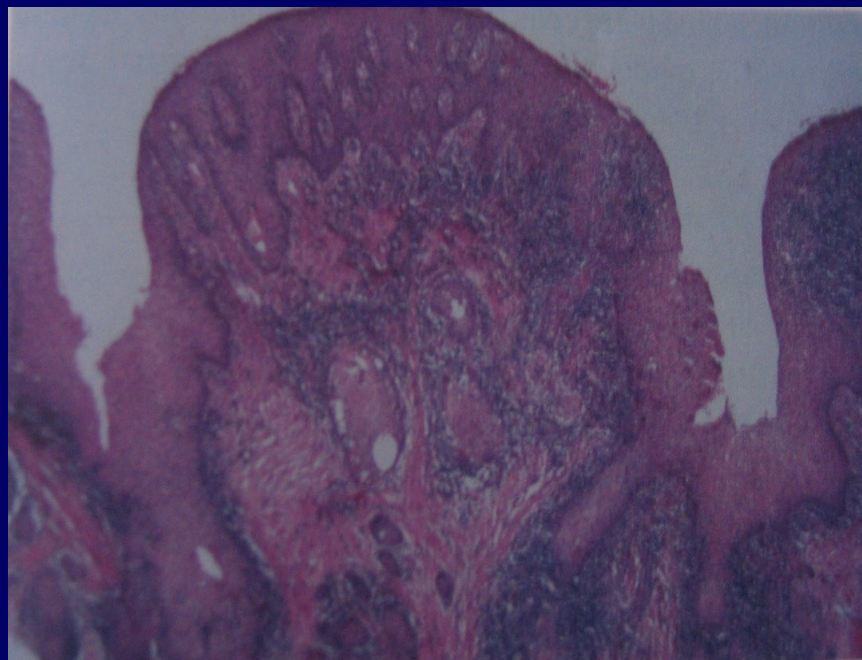
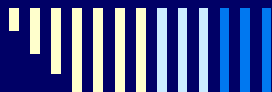
# Inflammatory papillary hyperplasia

- Called denture papillomatosis
  - Is a reactive tissue growth that usually develops beneath a denture
  - Related to
    - n Ill fitting denture
    - n Poor denture hygiene
    - n Wearing the denture 24hrs a day
  - Could be related to candida
-



# Clinical features

- In the hard palate beneath a denture base
  - Can also be seen in
    - n Mouth breathers
    - n With high palatal vault
    - n Candida associated with HIV
  - Asymptomatic, erythematous and has a pebbly or papillary surface
  - Early to advanced stages
-





# Histopathology

- Numerous papillary growths on the surface
  - Hyperplastic stratified squamous epithelium with pseudo-epitheliomatous hyperplasia – Mistaken for SCC
  - CT is loose and edematous to densely collagenized
  - Chronic inflammatory cell infiltrate with lymphocytes and plasma cells
-



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# Ttt and prognosis

- Removal of denture – lesion subsides
  - Excision in advanced stages
  - Use a tissue conditioner beneath the denture to facilitate healing.
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# Fibrous histioma

- Exhibits both fibroblastic and histiocytic differentiation
  - Cell of origin is not certain – arise from the tissue histiocyte which then assumes fibroblastic properties
  - It is a true neoplasm
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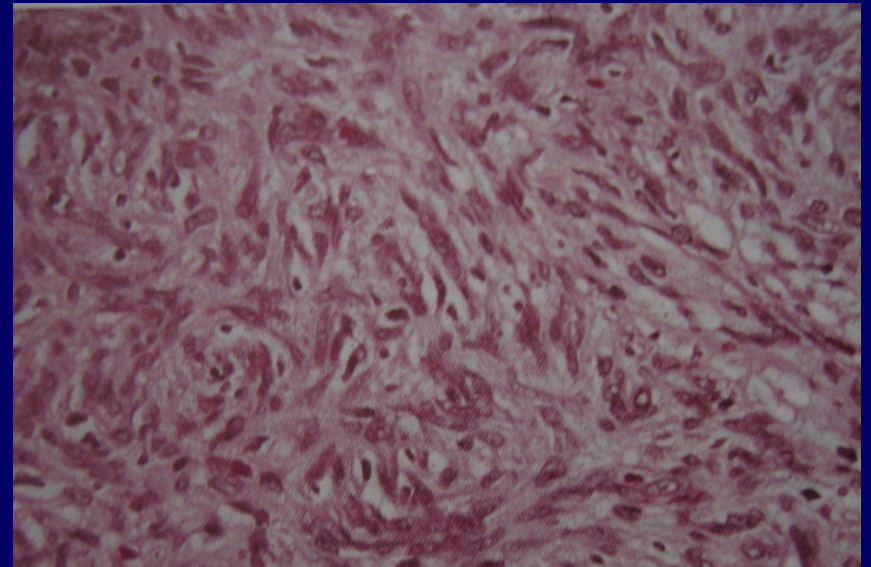


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# Clinical features

- In middle aged and older adults
  - Painless nodular mass
  - Uncommon in Oral and peri-oral region
  - Seen in buccal mucosa and vestibule
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# Histopathology

- Cellular proliferation of spindle shaped fibroblastic cells with vesicular nuclei.
  - Tumor cells are arranged in short, intersecting fascicles – storiform pattern – resembling a straw mat
  - Rounded histiocytic cells, xanthoma cells and multinucleated giant cells can be seen.
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# Ttt and prognosis

- Local surgical excision
  - Recurrence is rare
-



# Fibromatosis

- ❑ True neoplasm
  - ❑ Intermediate between benign fibrous lesions and fibrosarcoma
  - ❑ In children and young adults
  - ❑ In paramandibular soft tissue region
  - ❑ Spindle shaped cells in streaming fascicles, poorly circumscribed and infiltrates
  - ❑ No pleomorphism or hyperchromatism
  - ❑ Locally aggressive, wide excision and could recur.
-



# Myofibromatosis

- ❑ Rare spindle cell neoplasm with myofibroblasts – both smooth muscle and fibroblastic features.
  - ❑ Predilection for head and neck – mandible
  - ❑ Biphasic with nodular fascicles with cellular areas.
  - ❑ Spindle cells with tapered or blunt ended nuclei.
  - ❑ Surgical excision.
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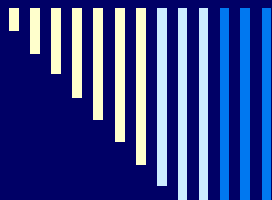
# Pyogenic granuloma

- ❑ Common tumor like growth – non-neoplastic
  - ❑ Thought to be caused by pyogenic organisms – now it is proven wrong
  - ❑ Represents an exuberant tissue response to local irritation or trauma
-



# Clinical features

- Most common in children and young adults.
  - Smooth or lobulated, pedunculated mass with an ulcerated surface
  - **Pink to bright red to purple** based on its duration
  - Mass is painless, it often bleeds easily because of its extreme vascularity.
  - **Maxillary anterior gingiva**, lips, tongue and buccal mucosa
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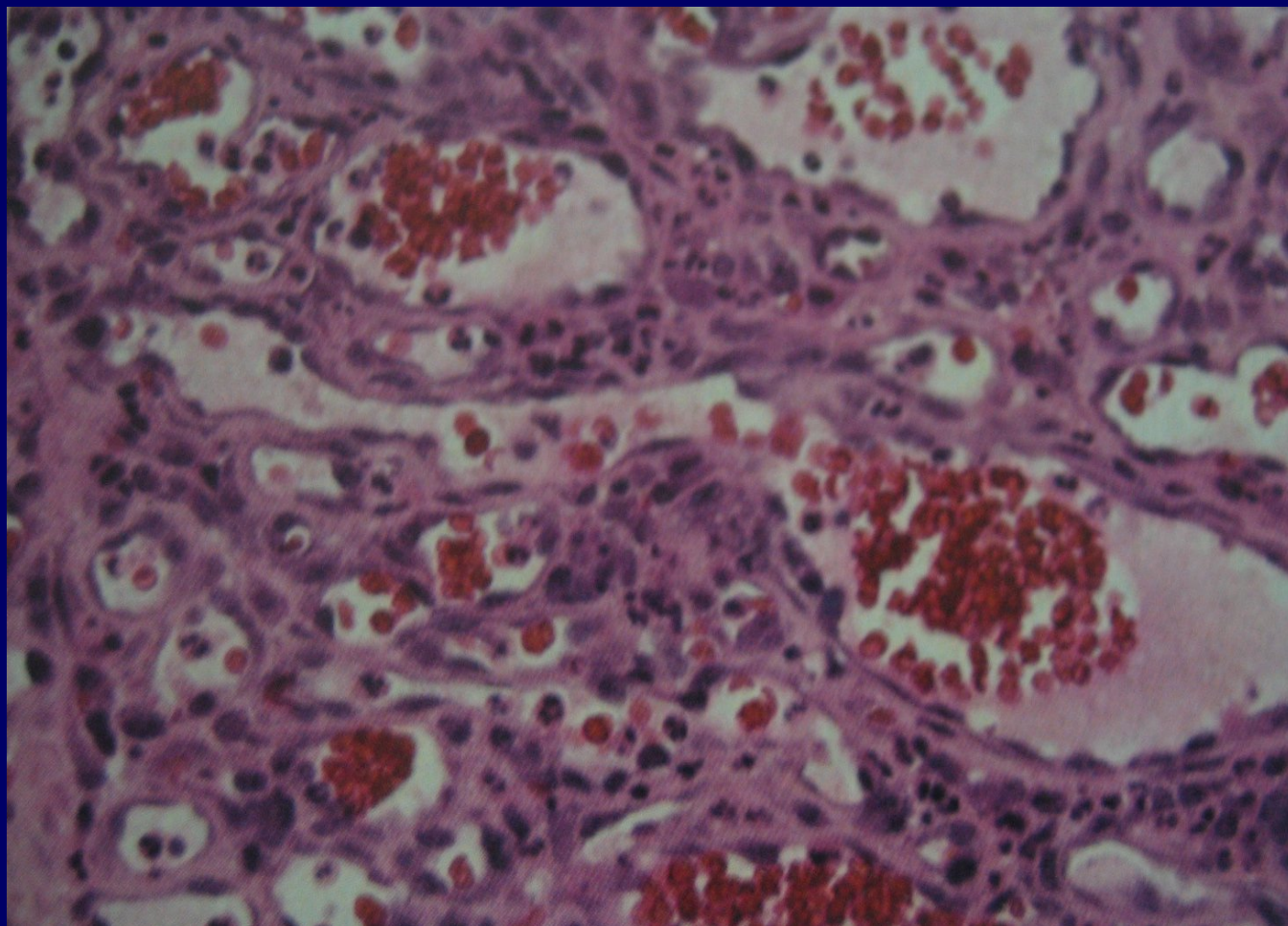
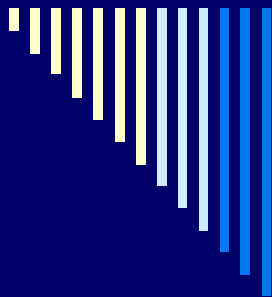
# Clinical features

- In pregnant women – **Pregnancy tumor** or **Granuloma gravidarum**. Could be related to altered hormone levels.
- Hyperplastic growth of granulation tissue from the healing extraction sockets – **Epulis granulomatosa**



# Histopathology

- Highly vascular proliferation resembling granulation tissue
  - Numerous small and large endothelium lined channels with red blood cells
  - Surface is usually ulcerated and replaced by a thick, fibrinopurulent membrane
  - Mixed inflammatory infiltrate with
    - n Neutrophils near the surface
    - n plasma cells and lymphocytes in the deeper regions
  - Gingival fibromas could be matured pyogenic granulomas.
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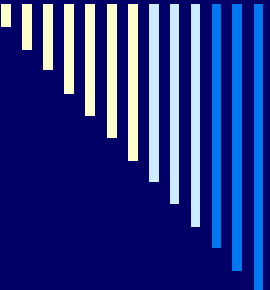




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# Ttt and prognosis

- Conservative surgical excision
  - Proper oral hygiene maintenance
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# Peripheral giant cell granuloma

- Reactive lesion to local trauma or irritation
  - The giant cells shows features of
    - n Osteoclasts
    - n From mononuclear phagocyte system
  - May represent the peripheral counterpart of central giant cell granuloma
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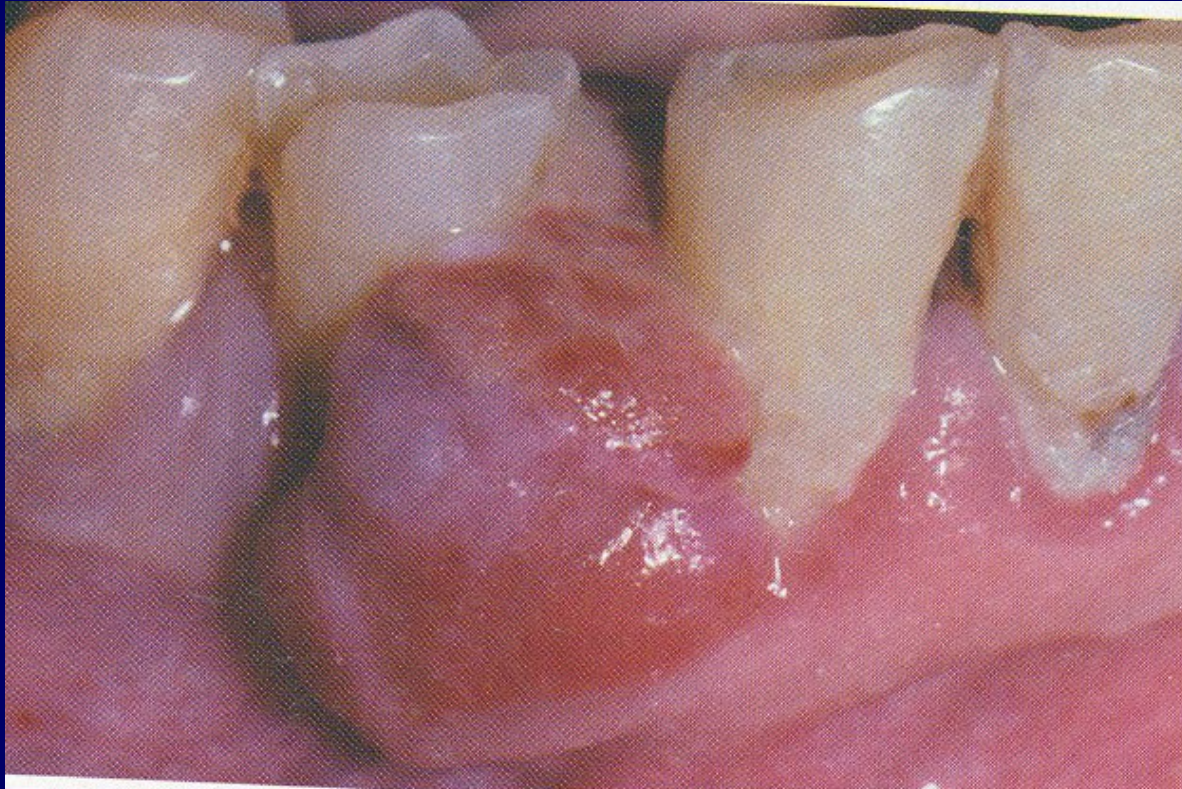


# Clinical features

- ❑ Exclusively in the gingiva or edentulous alveolar ridge
  - ❑ Red or reddish blue or **bluish purple** nodular mass
  - ❑ Peak prevalence in fifth and sixth decades – predominantly females
  - ❑ **Cupping resorption** of underlying bone could be seen.
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# Peripheral GCG



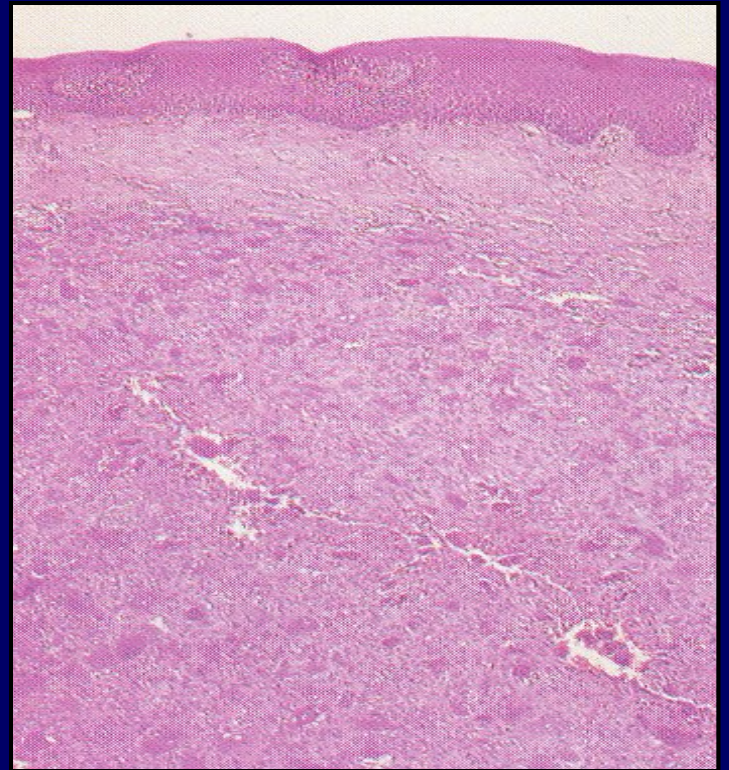
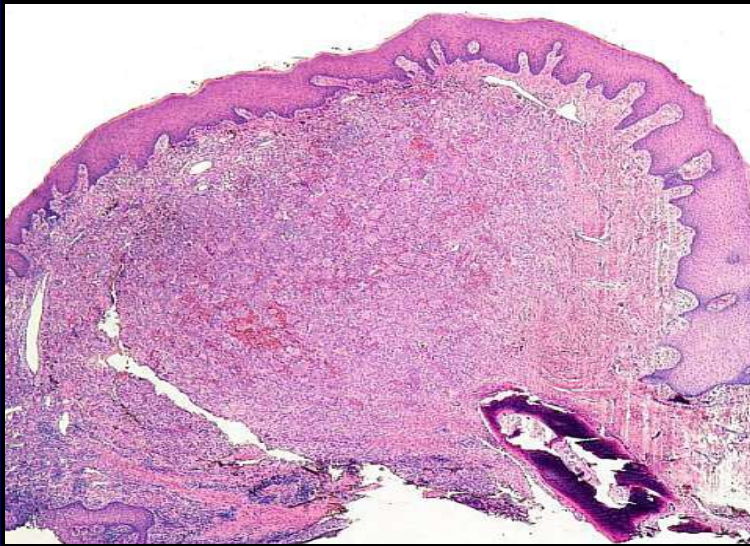


# Histopathologic features

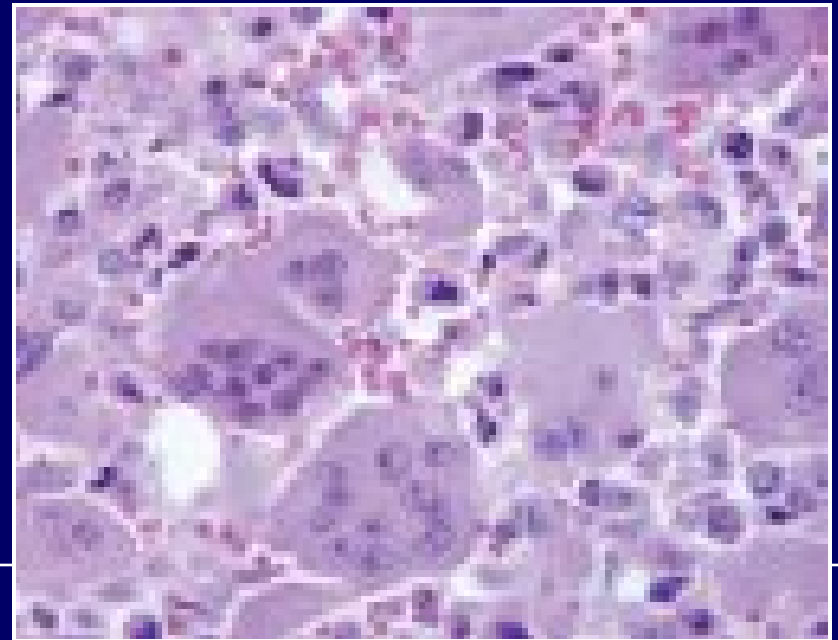
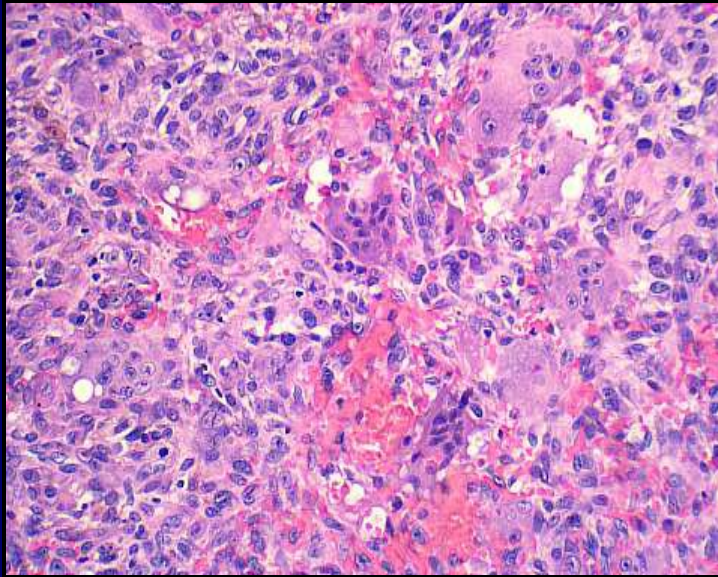
- ❑ Proliferation of multinucleated giant cells within a background of plump ovoid and spindle shaped mesenchymal cells
- ❑ Giant cells may have few nuclei upto several dozen nuclei
- ❑ May have large, vesicular to small, pyknotic nuclei
- ❑ Abundant hemorrhage throughout the mass, especially at the periphery
- ❑ Zone of dense connective tissue between the overlying epithelium and the giant cell proliferation.



# Peripheral GCG



# Peripheral GCG





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# Ttt and prognosis

- Local surgical excision
  - Rule out hyperparathyroidism – brown tumors
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# Peripheral ossifying fibroma

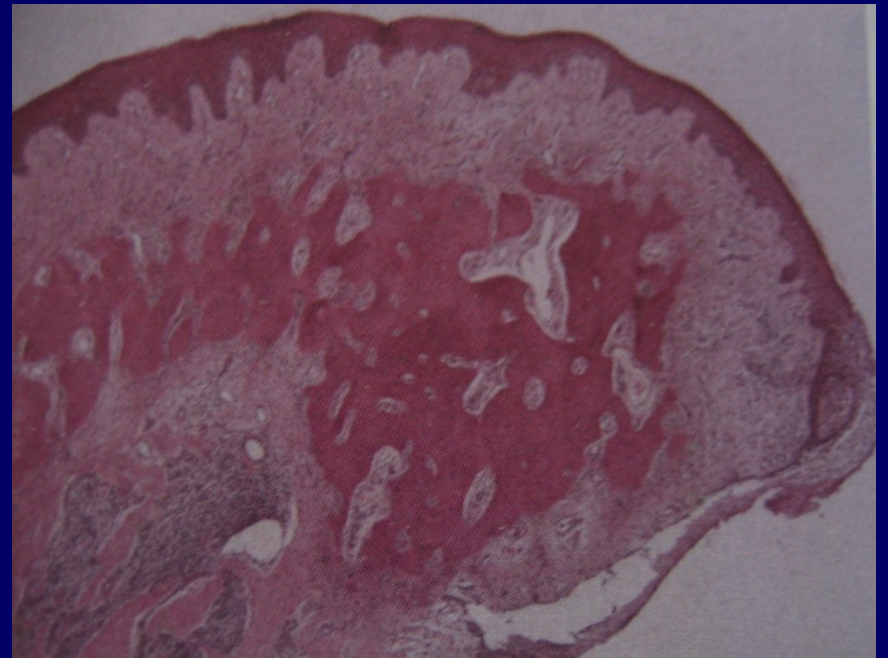
- Relatively common gingival growth
  - Mineralized product probably originates from cells of periosteum or periodontal ligament
  - Does not represent the soft tissue counterpart of the central ossifying fibroma
-



# Clinical features

- ❑ In teenagers and young adults – 2/3<sup>rd</sup> in females
  - ❑ Exclusively on the maxillary anterior gingiva
  - ❑ Pedunculated or sessile mass and it emanates from interdental papilla
  - ❑ Red to pink in color, frequently ulcerated surface
  - ❑ Most are <2cm in size
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# Histopathology

- Fibrous proliferation associated with formation of mineralized product
- If ulcerated, the epithelium is replaced by fibrinopurulent membrane
- Increased cellularity in areas of mineralization

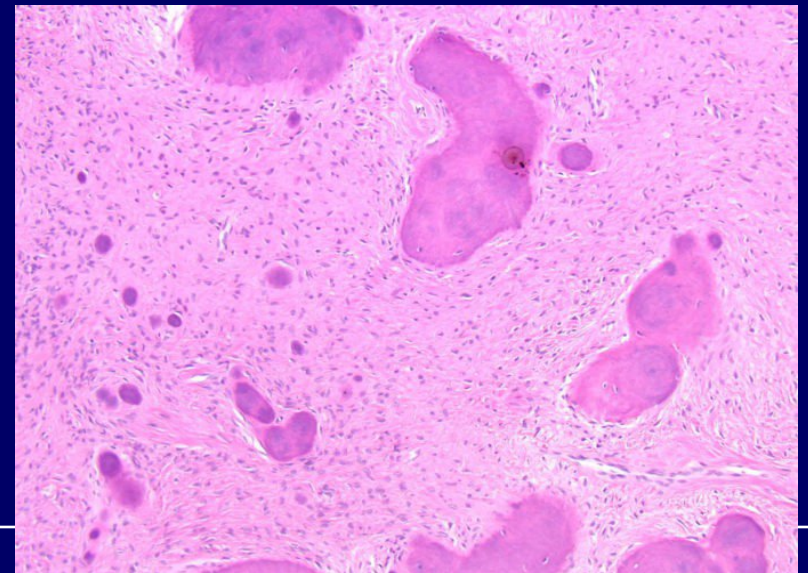
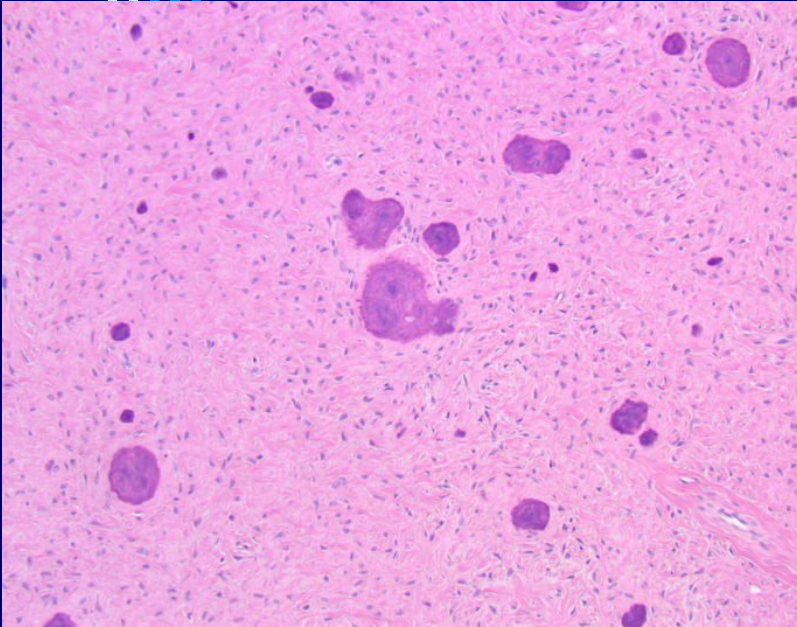


# Histopathology

- Mineralized component consists of bone, cementum like material or dystrophic calcifications
  - Usually occurs in combinations
    - n Bone is woven and trabecular in type
    - n Ovoid droplets of basophilic cementum like material
    - n Dystrophic calcifications – multiple granules, tiny globules or large, irregular masses of basophilic mineralized material.
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# Histopathology





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# Ttt and prognosis

- Local surgical excision till the periosteum



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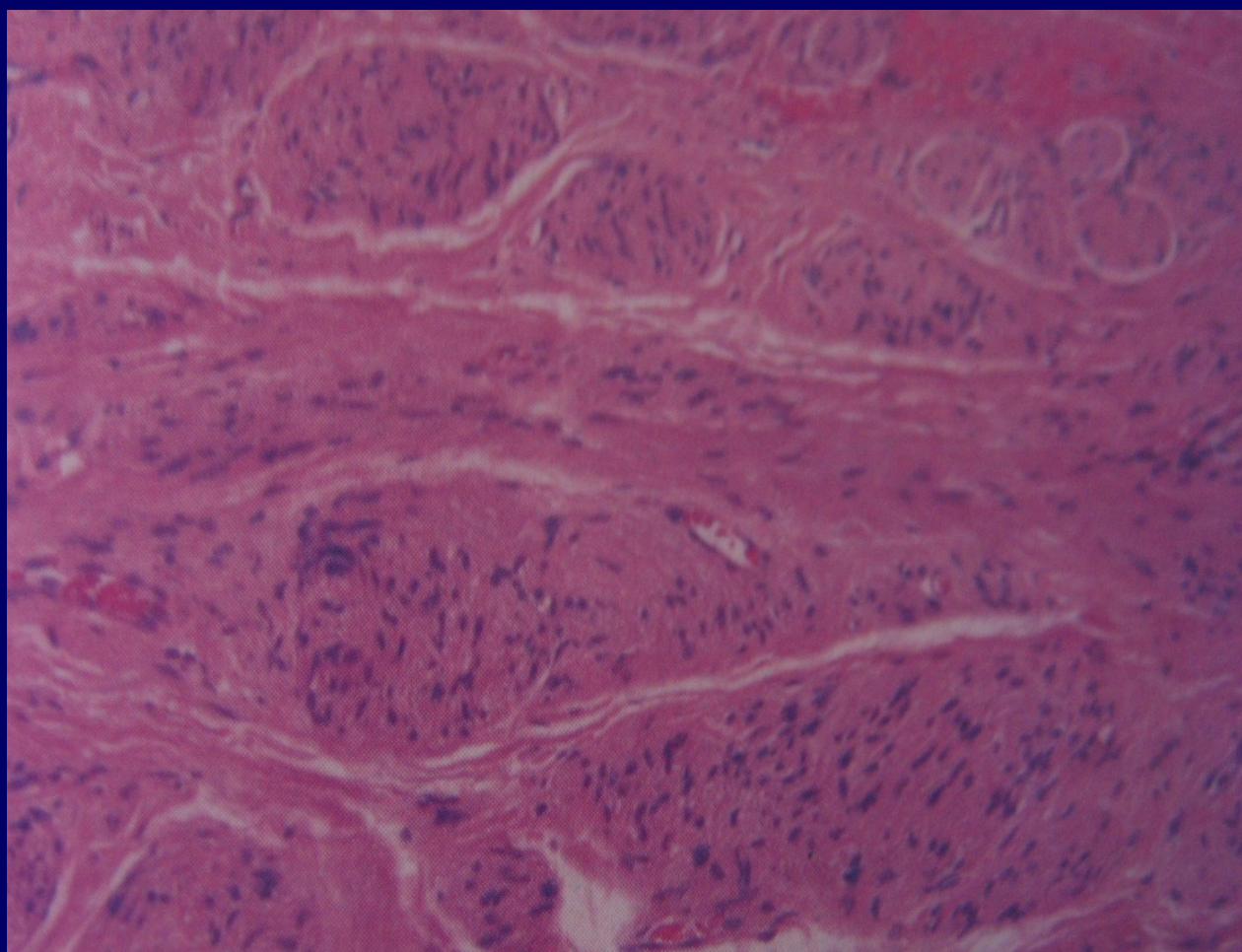
# Traumatic neuroma

- Reactive proliferation of nerve tissue after damage to the nerve bundle,
  - The proximal portion attempts to regenerate and re-establish innervation with the distal segment by growth of axons through tubes of proliferating schwann cells.
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# Clinical features

- In middle aged adults
  - Smooth surfaced, non-ulcerated nodule
  - Common in mental foramen area, tongue and lower lip
  - May be associated with pain
-





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# Histopathology

- Haphazard proliferation of mature, myelinated nerve bundles within a fibrous connective tissue
  - With inflammation, it is more likely to be painful.
  - Treated by surgical excision.
-



# Palisaded encapsulated neuroma

- ❑ Benign neural tumor of superficial nerves
  - ❑ Considered to be a reactive lesion
  - ❑ Striking predilection for the face – nose and cheek
  - ❑ In the 5<sup>th</sup> – 7<sup>th</sup> decades
  - ❑ Smooth surfaced, painless, dome shaped nodule
  - ❑ Oral lesions – hard palate and labial mucosa
-



# Histopathology

- Well circumscribed, encapsulated
- Moderately cellular, interlacing fascicles of spindle cells – schwann cells
- Nuclei are wavy and pointed
- S100 positivity
- Treated by local surgical excision.





# Neurilemoma

- Also called schwannoma
  - Benign neural neoplasm of schwann cell origin
  - Slow growing, encapsulated tumor arising within the nerve trunk
  - As it grows, it pushes the nerve aside
  - Usually asymptomatic
  - Tongue – most common location
-



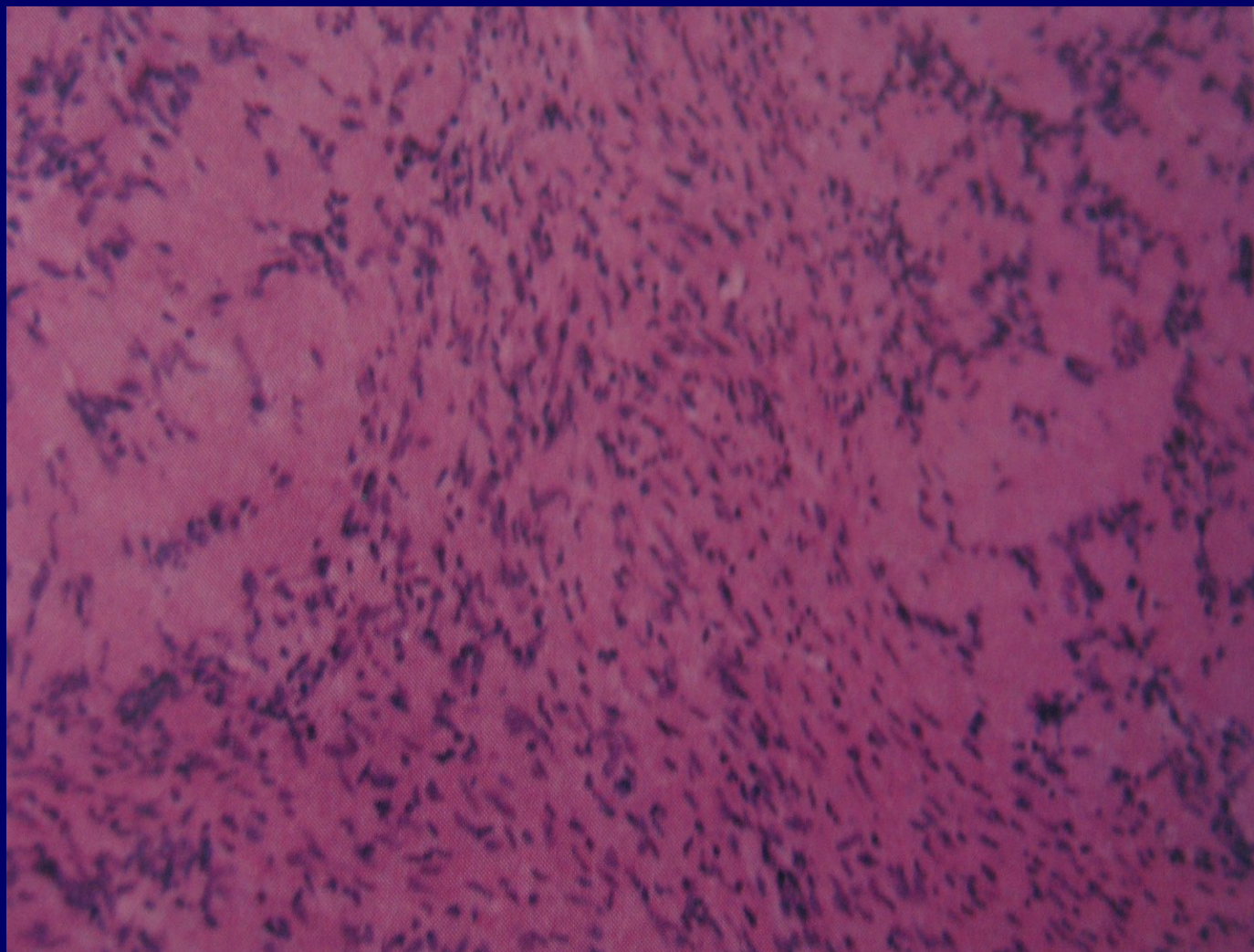
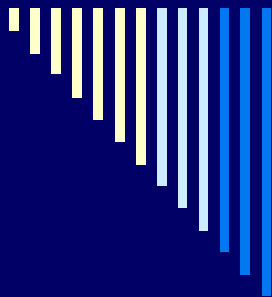
# Histopathology

- Encapsulated tumor
  - Antoni A –
    - n Streaming fascicles of spindle shaped schwann cells.
    - n Palisaded arrangement around central, acellular, eosinophilic areas – Verocay bodies
    - n Reduplicated basement membrane and cytoplasmic areas
  - Antoni B
    - n Less cellular and less organized spindle cells randomly arranged in a myxomatous stroma
-



# Histopathology

- Diffuse immunoreactivity to S100 protein
  - Older lesions – Ancient neurilemmomas – has hemorrhage, hemosiderin deposits, inflammation, fibrosis and nuclear atypia.
  - Treated by surgical excision
-

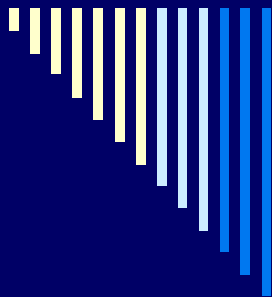




# Neurofibroma

- ❑ Most common peripheral nerve neoplasm
- ❑ Arises from mixture of cell types including schwann cells and perineural fibroblasts.
- ❑ Arise as solitary tumors or **multiple lesions in neurofibromatosis**.
- ❑ Slow growing, soft, painless lesions involving the skin, tongue and buccal mucosa.

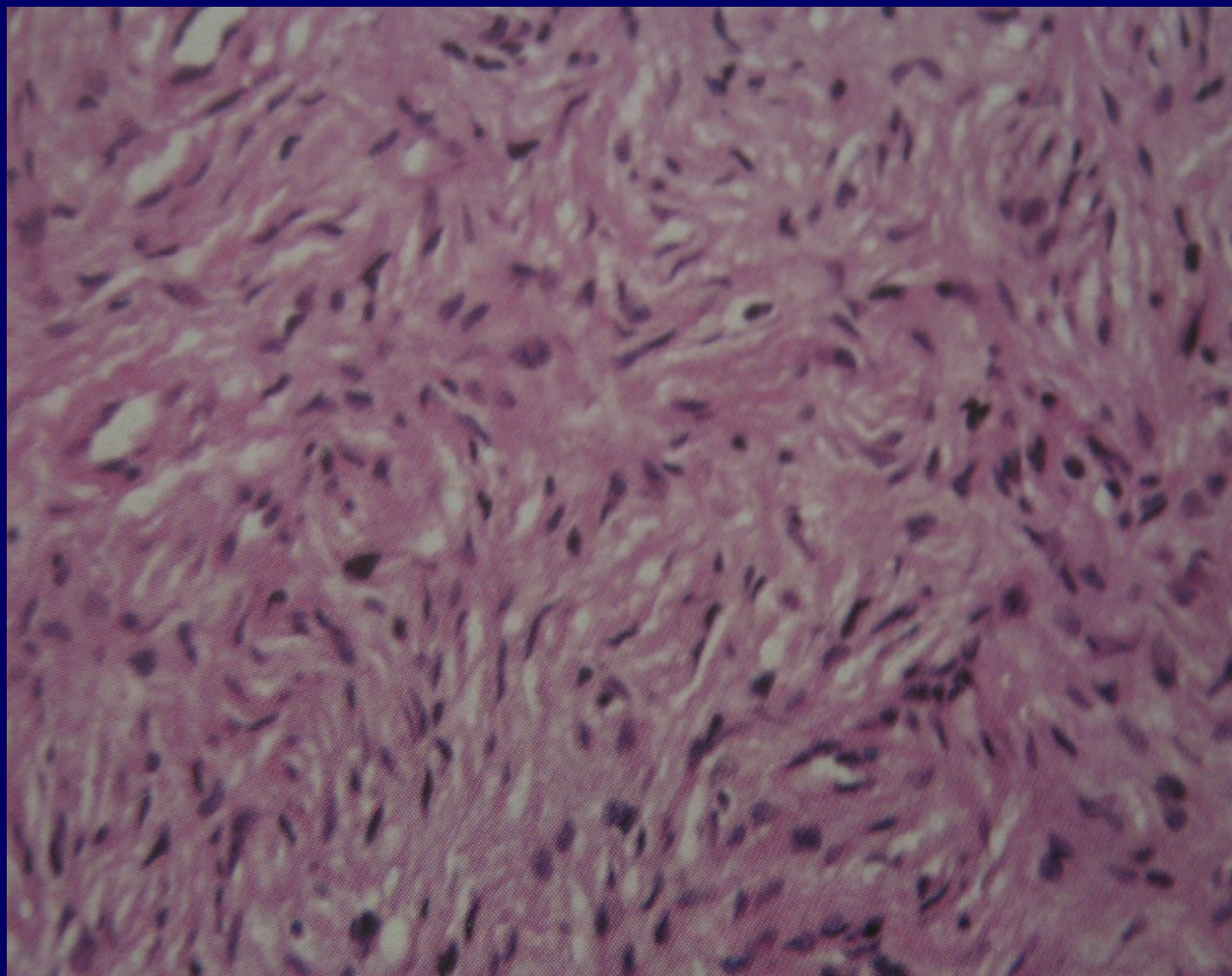
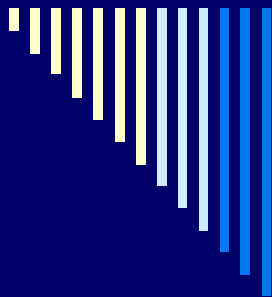






# Histopathology

- Well circumscribed, if proliferates within the perineurium of the involved nerve
- If it is outside the perineurium, it is not well demarcated and blends with the surrounding connective tissue
- Has interlacing bundles of spindle shaped cells with wavy nuclei along with delicate collagen bundles and myxoid matrix.
- Numerous mast cells and scattered S100 positivity.
- Treated with surgical excision.

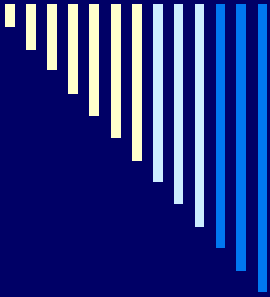






# Neurofibromatosis

- ❑ Also called Von Recklinghausen's disease of the skin
  - ❑ Hereditary condition (autosomal dominant) mapped to chr17– 1:3000 births
  - ❑ 8 forms – Neurofibromatosis Type I is the most common.
  - ❑ These patients have multiple neurofibromas.
  - ❑ Enlargement of fungiform papillae in the tongue
-





# Diagnostic criteria

- ❑ 6 or more café au lait macules – Coffee with milk pigmentation of skin, smooth edges, yellowish to dark brown macules
- ❑ Two or more NF or one plexiform NF (feels like bag of worms)
- ❑ Freckling in axilla (Crowe's sign) or inguinal regions
- ❑ Two or more lisch nodules (pigmented spots on the iris)
- ❑ Osseous lesion
- ❑ First degree relative (parent, sibling or offspring) with NF-1, based on the above criteria



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# Complications

- As multiple lesions – no specific treatment
  - High risk for malignant transformation – Malignant peripheral nerve sheath tumor (Neurofibrosarcoma, Malignant Schwannoma)
-



# Multiple endocrine neoplasia

- Rare condition with 3 forms,
    - n Type I – benign tumors of pancreas, adrenal cortex, parathyroid and pituitary glands
    - n Type 2A – Sipple syndrome – Adrenal pheochromocytomas and medullary thyroid carcinomas
    - n Type 2B – along with 2A, mucosal neuromas
-



# MEN type 2B

- ❑ Inherited as autosomal dominant
  - ❑ Mutation in RET gene in Chr 10
  - ❑ Marfanoid body build – thin, elongated limbs with muscle wasting
  - ❑ Face is narrow, lips are thick and protruberant – proliferation of nerve bundles
  - ❑ Oral mucosal neuromas – soft, painless papules of lips and anterior tongue
-



# Lab investigations

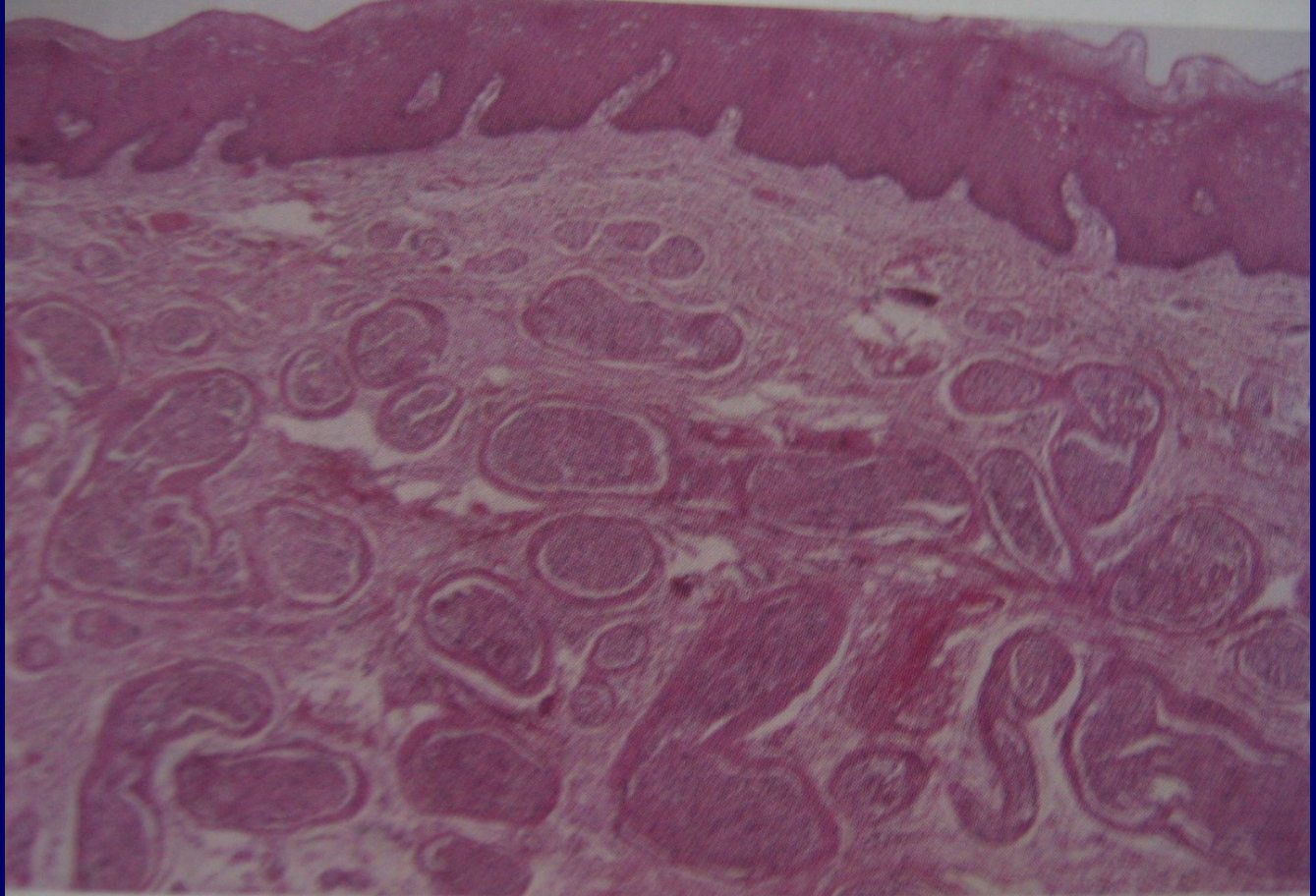
- Medullary carcinoma of thyroid – increase in serum and urinary levels of calcitonin
  - Pheochromocytomas – Increase in Vanillyl Mandelic Acid (VMA), alterations in epinephrine to norepinephrine ratios
-



# Histopathology

- Marked hyperplasia of nerve bundles in a normal connective tissue background
  - Prominent thickening of perineurium
  - Prognosis depends on the severity of the associated diseases.
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# Melanotic neuroectodermal tumor of infancy

- Rare pigmented neoplasm in the first year of life.
  - Of neural crest origin
  - Older terms - pigmented ameloblastoma, retinal anlage tumor, melanotic progonoma
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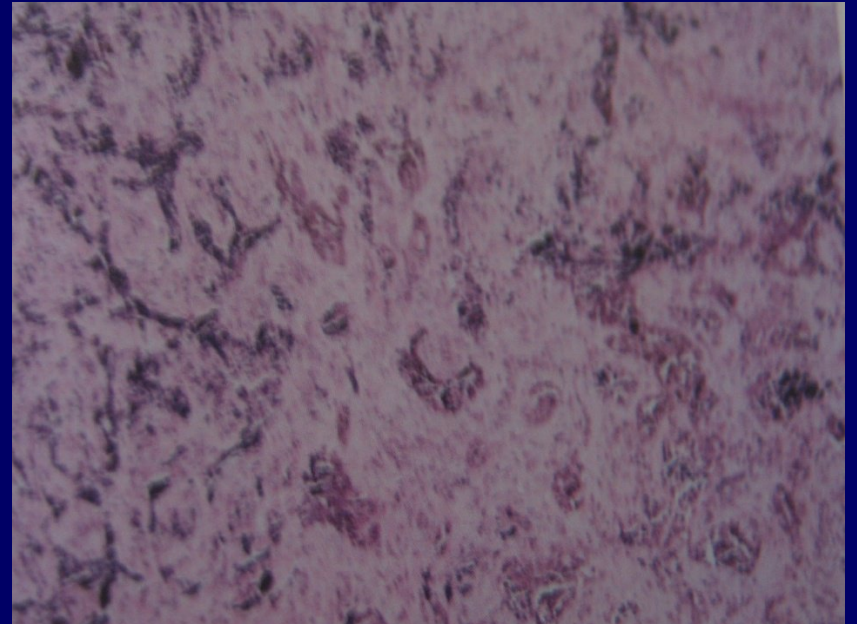
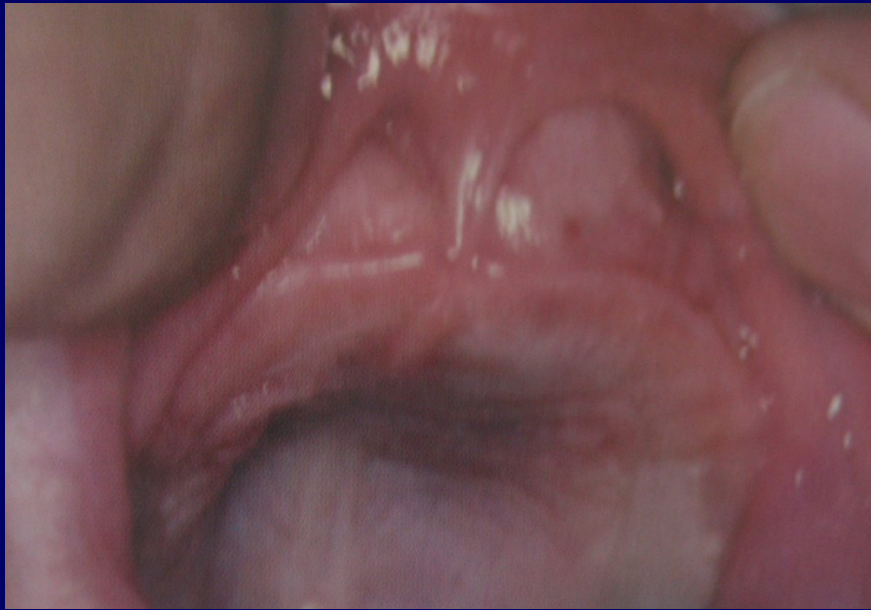
# Clinical features

- Develops during the first year of life
  - Predilection for maxilla anterior region
  - Rapidly expanding mass – blue or black
  - Often destroys the underlying bone
  - Can occur in skull, mandible, brain and testes
  - Increase in VMA – neuro-crestal origin
-



# Histopathology

- Biphasic population in the form of nests, tubules or alveolar structures within a dense, collagenous stroma
  - 2 cell groups
    - n Cuboidal cells with vesicular nuclei and granules of melanin
    - n Small, round cells with hyperchromatic nuclei and little cytoplasm – neuroblastic appearance
  - Round cells in loose nests surrounded by larger, pigmented cells.
-





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# Ttt and prognosis

- Benign – inspite of their rapid growth and involvement of bone
  - Surgical removal
  - Recurrence can occur.
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# Leiomyoma

- Benign tumors of smooth muscle
  - Seen in uterus, GIT and skin.
  - Oral presentation is rare
  - Arise from vascular smooth muscle.
-



# Clinical features

- Three types
    - n Solid – normal in color
    - n Vascular / angiomyomas / angioleiomyoma – bluish hue
    - n Epithelioid (leiomyoblastomas)
  - Slow growing, asymptomatic, firm, mucosal nodule
  - Involve tongue, palate, cheek
-





# H/P

## □ Solid –

- n Well circumscribed tumors with interlacing bundles of spindle shaped smooth muscle cells
- n Nuclei are elongated, pale staining and blunt ended.

## □ Angio

- n Multiple, tortuous blood vessels
- n Thickened walls – hyperplasia of their smooth muscle coats.

## □ Epithelioid type – has epithelioid cells.



# Special stains

- Demonstrated with
  - n Masson trichrome
  - n PTAH
  - n Vimentin
  - n Smooth muscle actin
  - n Muscle specific actin
- Local surgical excision



# Rhabdomyoma

- Benign neoplasm of skeletal muscle
- Extremely rare
- Cardiac – seen in tuberous sclerosis
- Extra-cardiac – present in head and neck
- Two types
  - n Adult
  - n Fetal



# C/F

## □ Fetal

- n In young children
- n In face and peri-auricular region

## □ H/P

- n Less mature appearance
- n Haphazard arrangement of spindle shaped cells

## □ Treated by local surgical excision.



# C/F

## □ Adult

- n In middle aged and older men
- n Pharynx, oral cavity and larynx
- n In floor of mouth, soft palate and base of tongue
- n Appears as a nodule or mass

## □ H/P

- n Well circumscribed lobules of large, polygonal cells  
– granular, eosinophilic cytoplasm
  - n Peripheral vacuolization – spider web appearance
  - n Focal cells with cross striations can be seen
-