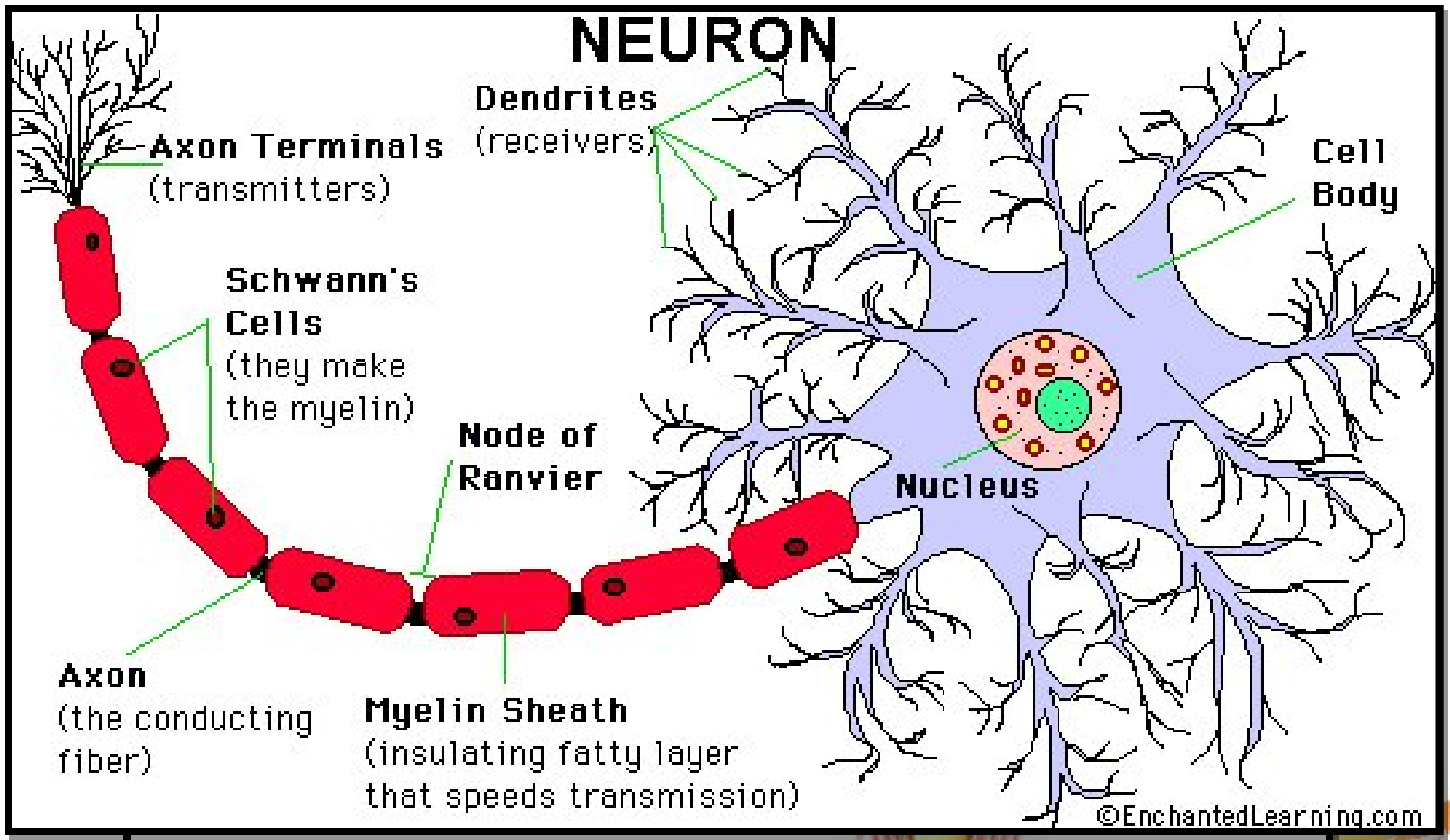
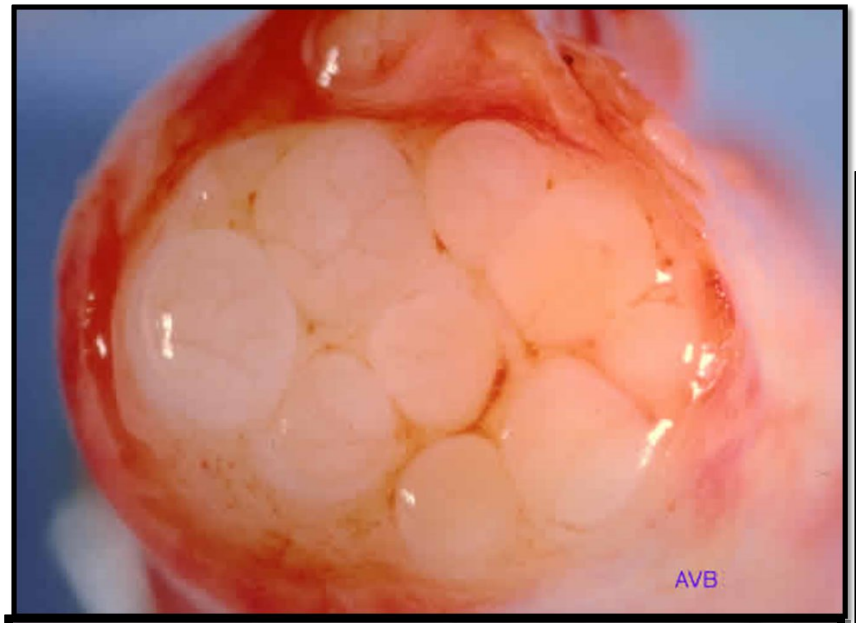
NEURAL TUMORS

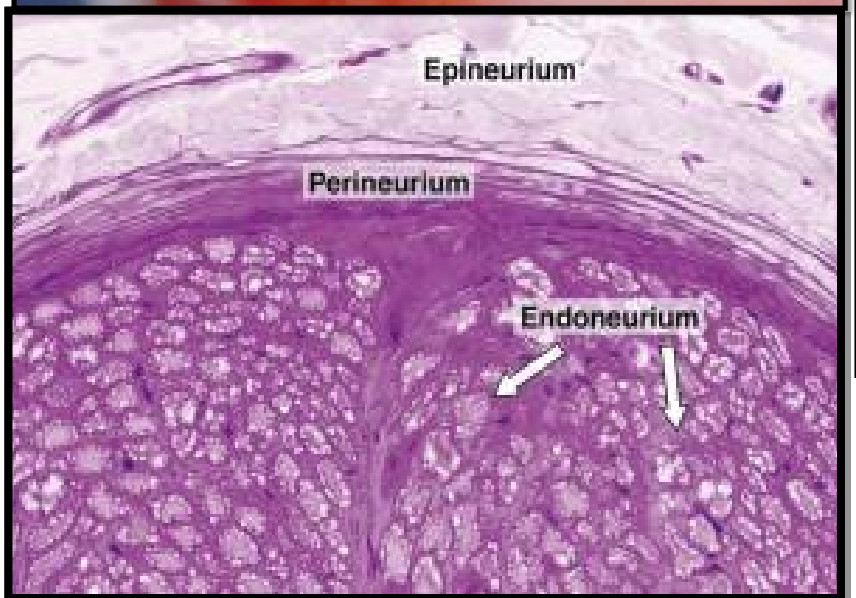
DR. VARSHA SANGLE

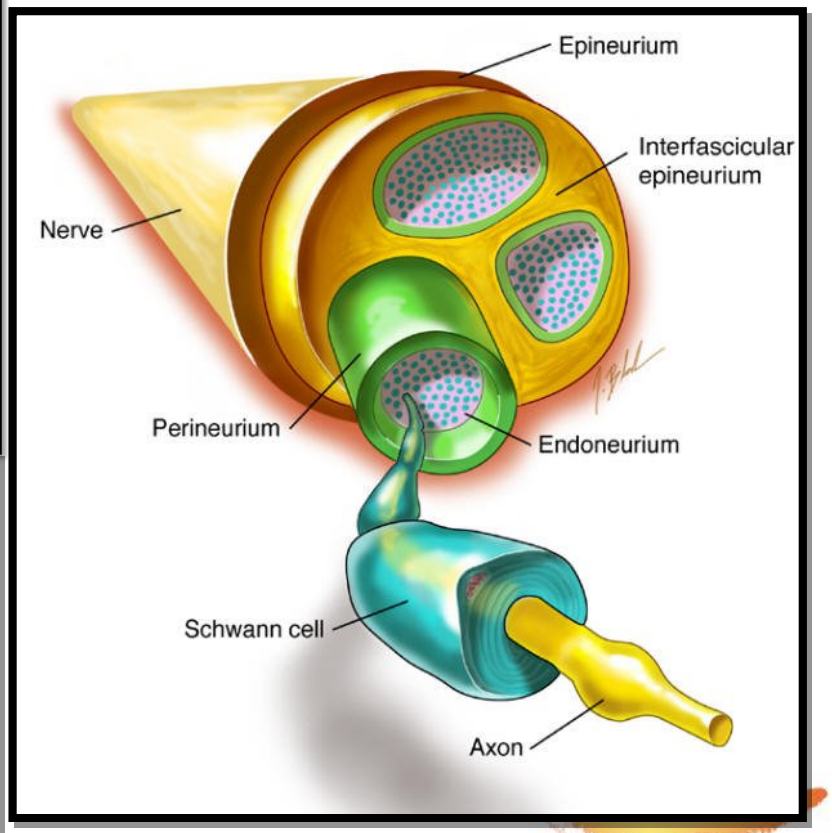
Structure of a neuron



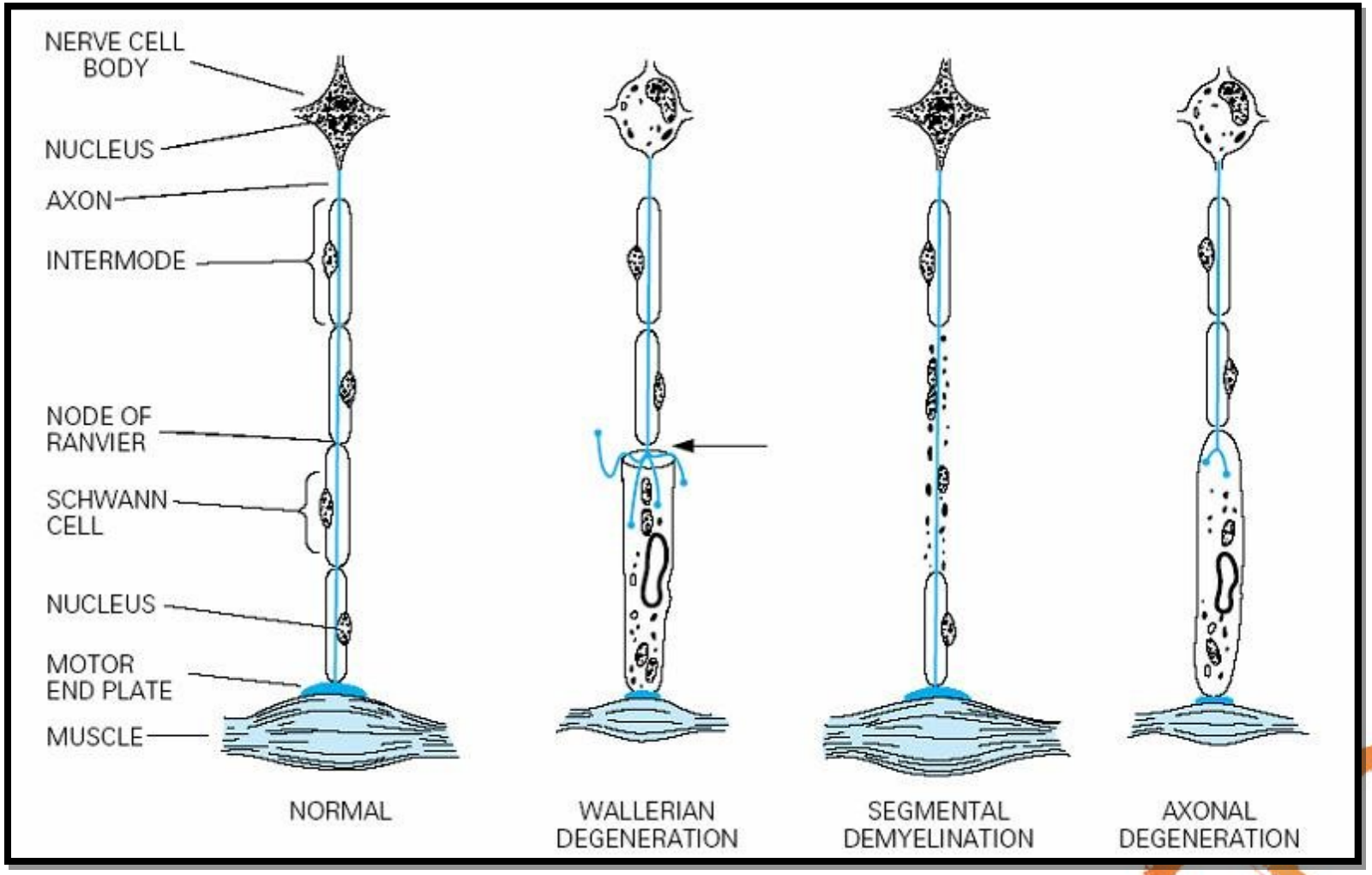
Structure of a nerve







Types of degeneration



BENIGN TUMORS OF NERVE TISSUE ORIGIN

1)Traumatic neuroma

2)Multiple endocrine neoplasia syndrome

3)Neurilemmoma

4)Neurofibroma

5)Neurofibromatosis

Traumatic Neuroma

- Also called Amputation Neuroma. It is not a true tumor but a reactive proliferation of neural tissue after transection or other damage of a nerve bundle.
- H\O of trauma is found like extraction or other surgical procedure.
- Occur at any age but common in middle aged, & females,
- Pain is intermittent or constant & ranges from mild tenderness to burning to severe radiating pain.
- Neuromas of mental nerve are very painful if impinged by denture or palpated.

Traumatic neuroma

- Exuberant attempt to repair damaged nerve trunk.

- Repair of damaged nerve:
- Proliferation of axon, Schwann cells.

- Proliferation meet some obstruction.

- Unorganized bulbous proliferation.

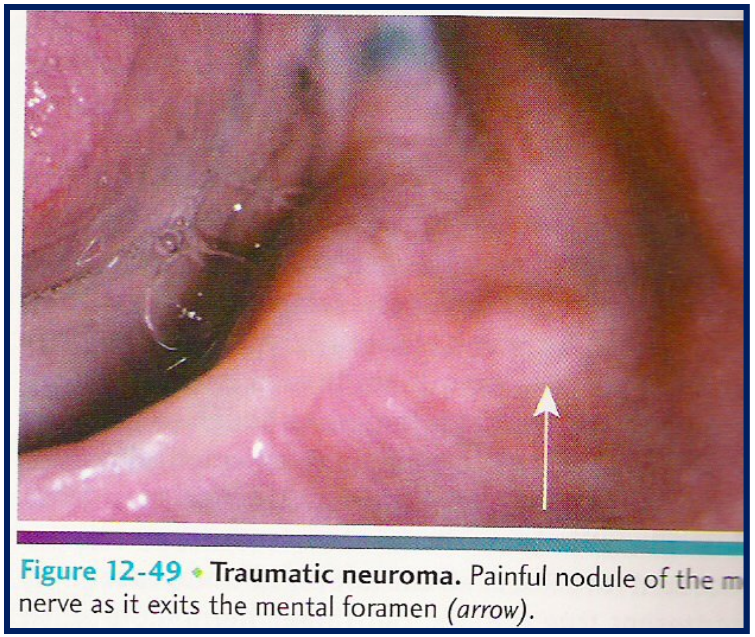


Figure 12-49 • **Traumatic neuroma.** Painful nodule of the m
nerve as it exits the mental foramen *(arrow)*.

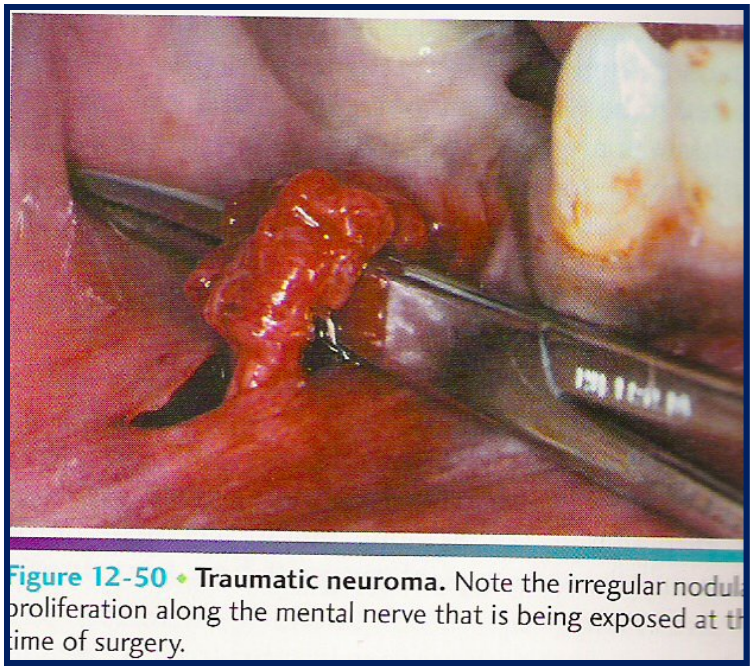
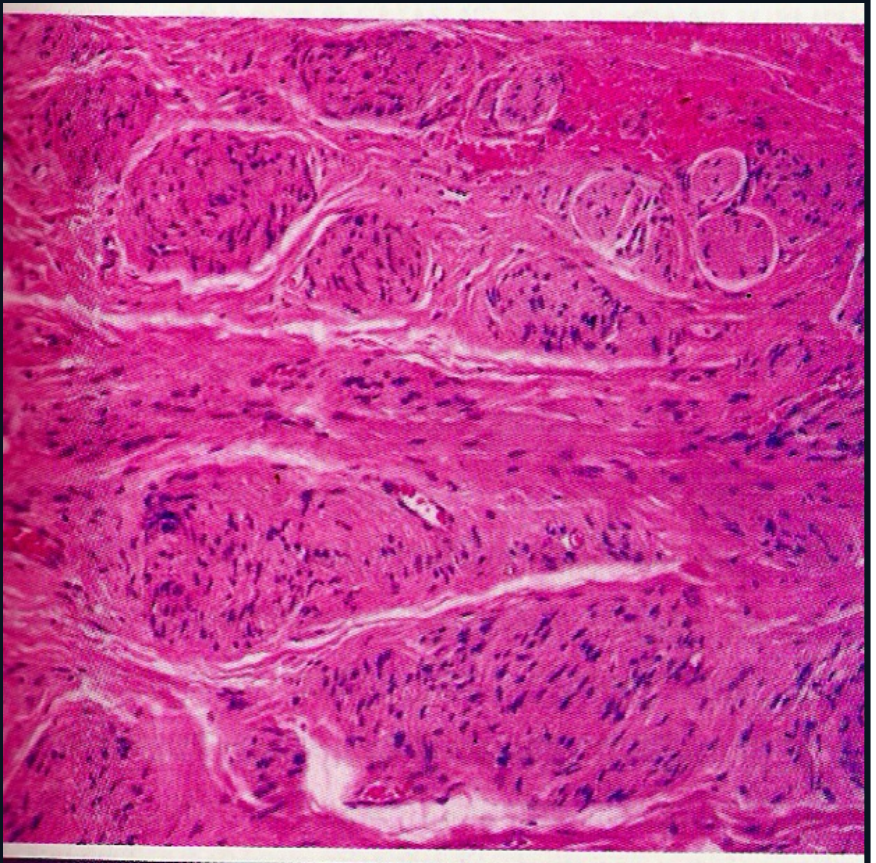
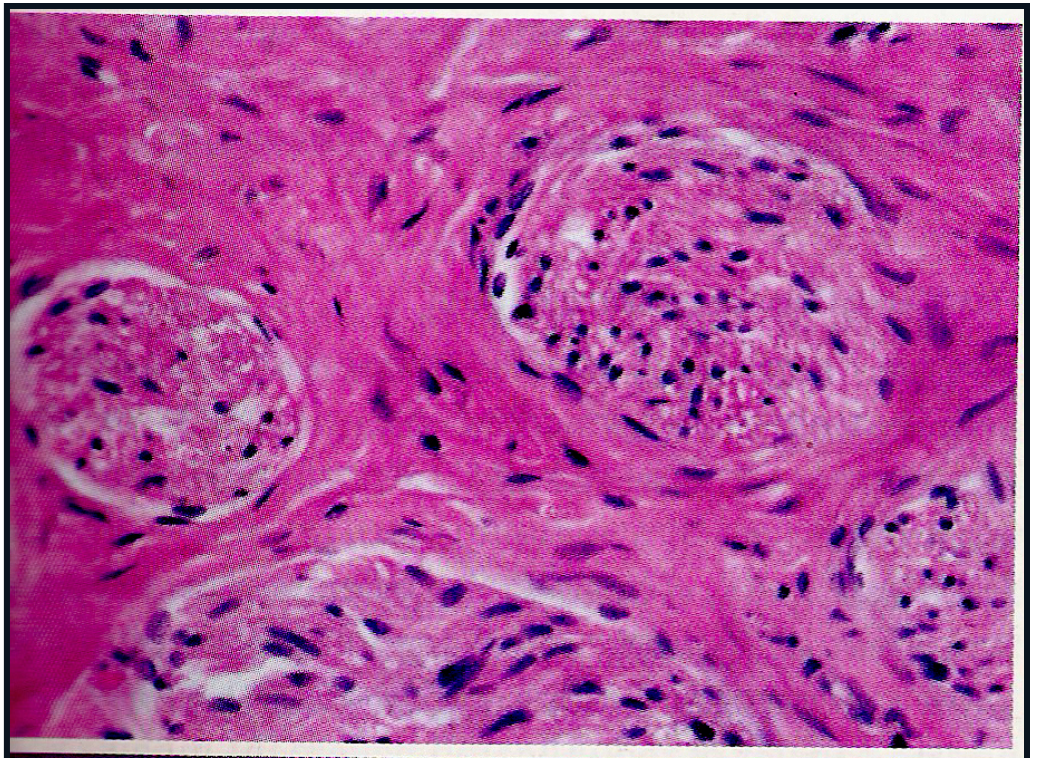


Figure 12-50 • **Traumatic neuroma.** Note the irregular nodul
proliferation along the mental nerve that is being exposed at t
time of surgery.

Clinical Features– are typically smooth nodules, common at mental foramen area, tongue, lower lip. Other sites like T\N of greater auricular nerve in 5-10% of pt's undergoing surgery for Pleomorphic Adenoma.



e 12-51 • **Traumatic neuroma.** Low-power view showing aphazard arrangement of nerve bundles within the back- nd fibrous connective tissue.



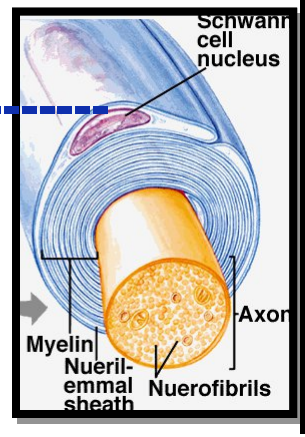
12-52 • **Traumatic neuroma.** High-power view showing sectioned nerve bundles within dense fibrous connective

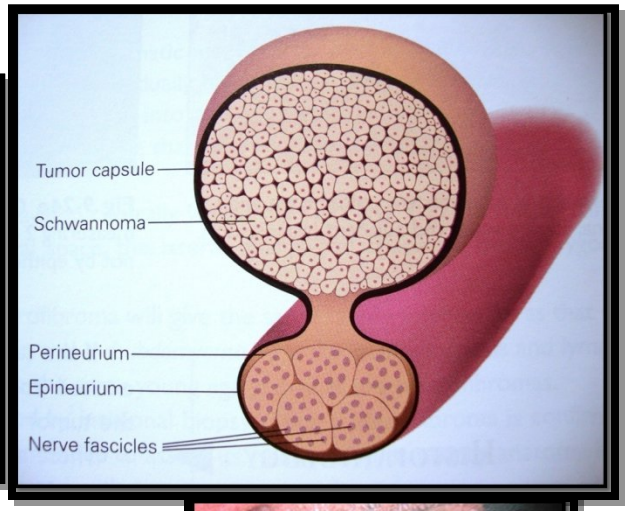
Schwannoma

- Synonyms: Neurilemmoma
  Perineural Fibroblastoma
  Neurinoma
  Lemmoma

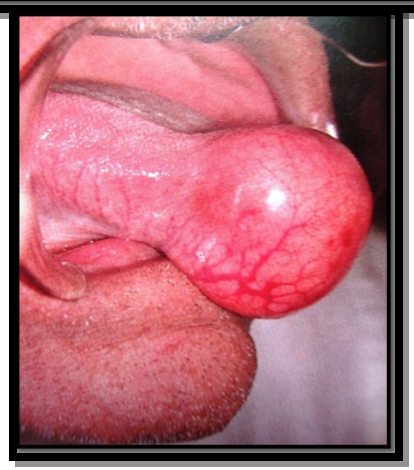
Schwannoma

Composed exclusively of----------



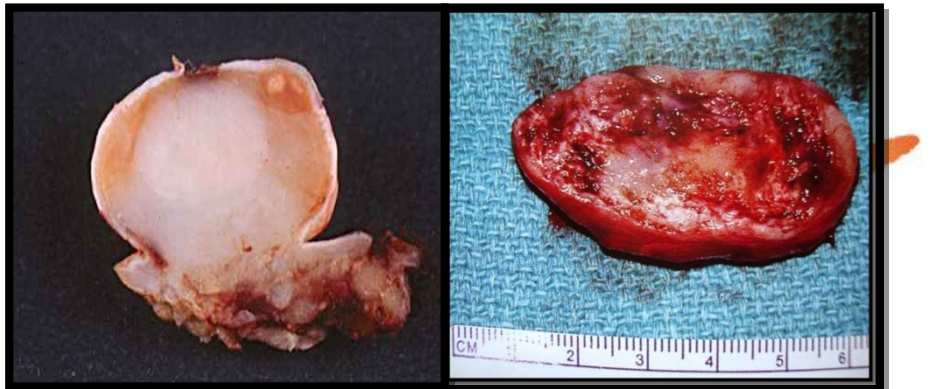


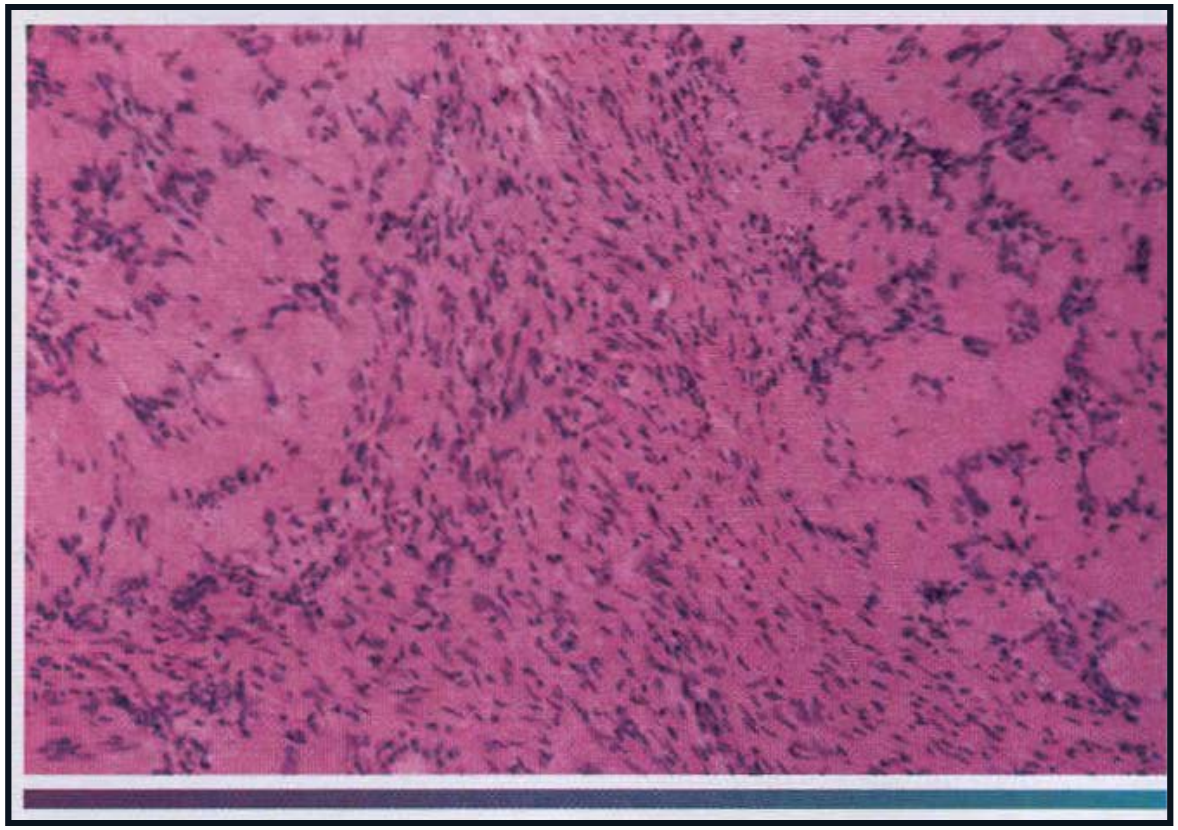
Age predilection: 30-50 years



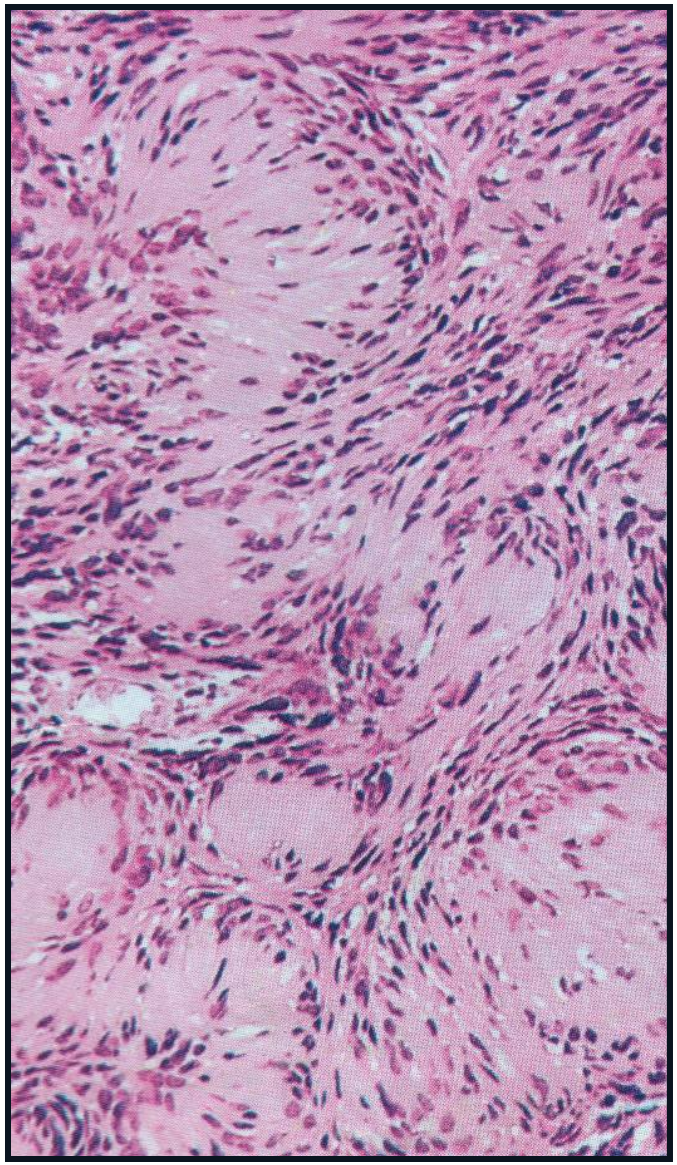
Site: Tongue, floor of the mouth----------

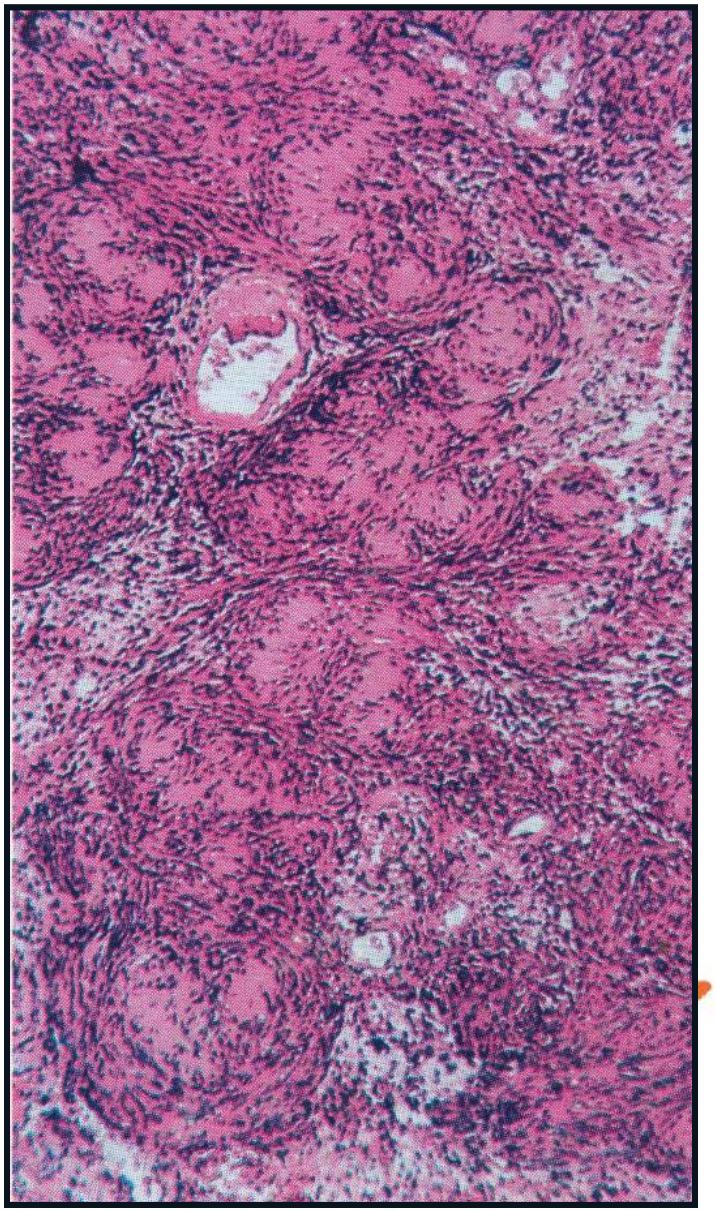
Gross Pathology





- Neurilemoma. showing a central zone of Antoni B tissue that is bordered on both sides by better-organized Antoni A tissue. The Schwann cells of the Antoni A tissue form a palisaded arrangement around acellular zones known as Verocay bodies.





Clinical Differential Diagnosis

- Granular cell tumor
- Lipoma
- Neurofibroma

Neurofibroma

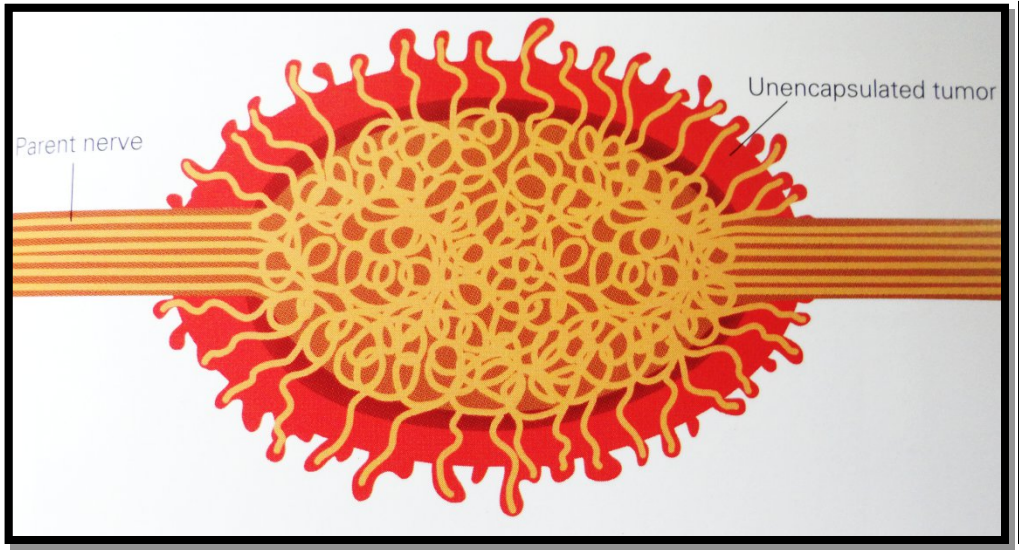
•Common type of peripheral nerve neoplasm .
•Arises from a mixture of cell types like Schwann cell, perineural fibroblasts.

•Malignant transformation can occur but risks are high in neurofibromatosis

Solitary Neurofibroma

- **Account for 90% cases of neurofibroma**
- **Asymptomatic, diffuse mass within subcutaneous or sub-mucosal tissue**

Characteristic bag or worms appearance



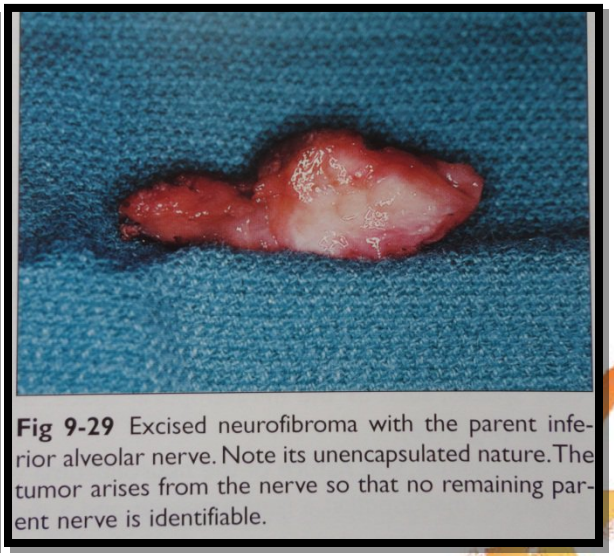


Fig 9-29 Excised neurofibroma with the parent inferior alveolar nerve. Note its unencapsulated nature. The tumor arises from the nerve so that no remaining parent nerve is identifiable.

- Clinical Features– arises as solitary tumor or component of neurofibromatosis.
- Solitary – common in young adults, slow growing soft nodular mass
- Common on skin, oral lesions are not uncommon.
- Tongue, buccal mucosa, bone

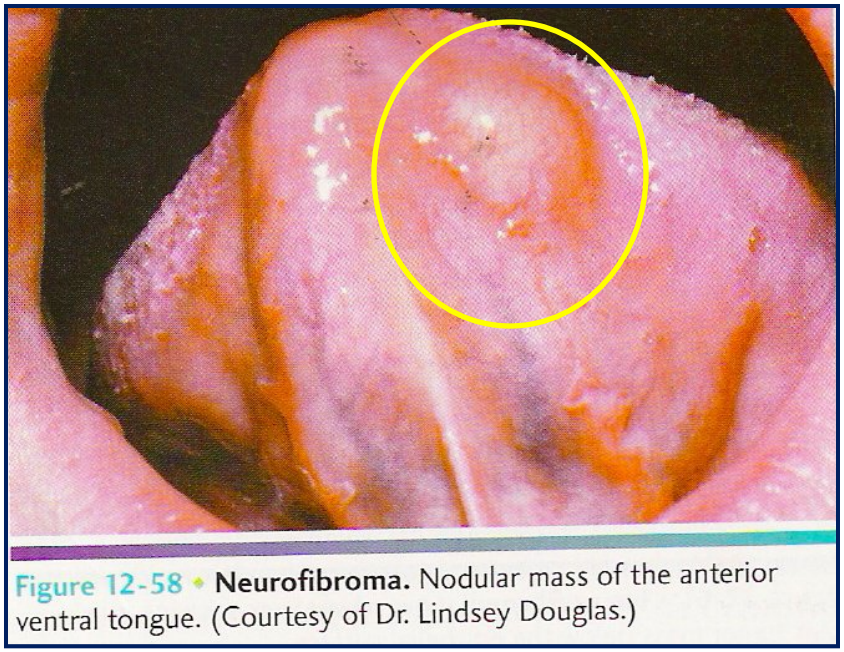
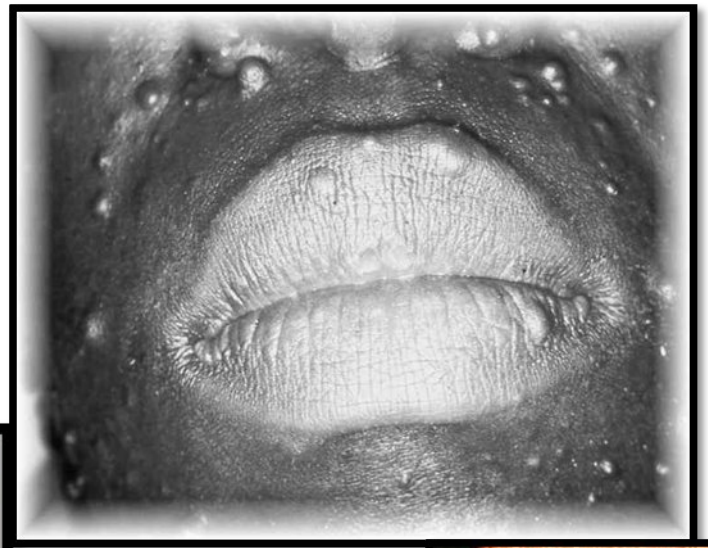


Figure 12-58 • **Neurofibroma.** Nodular mass of the anterior ventral tongue. (Courtesy of Dr. Lindsey Douglas.)



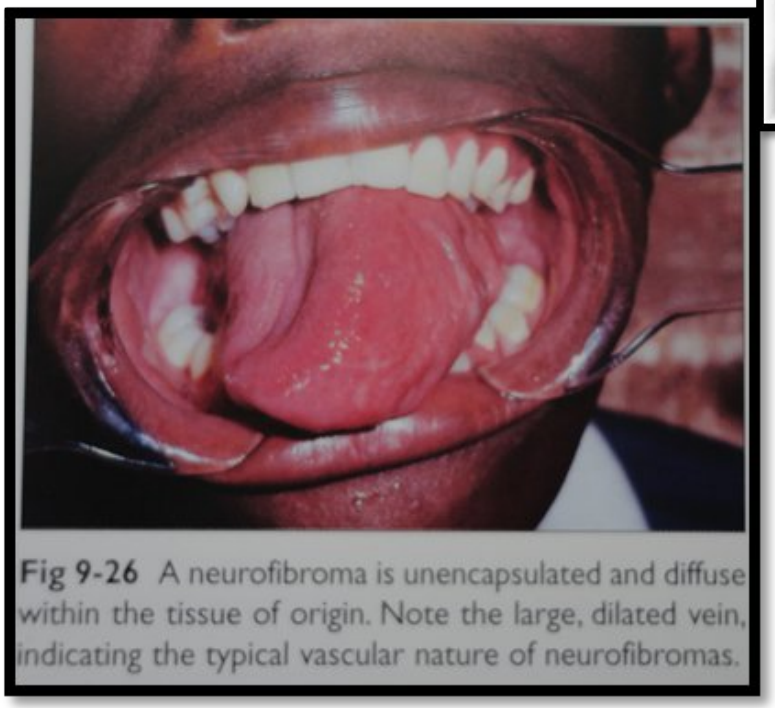


Fig 9-26 A neurofibroma is unencapsulated and diffuse within the tissue of origin. Note the large, dilated vein, indicating the typical vascular nature of neurofibromas.

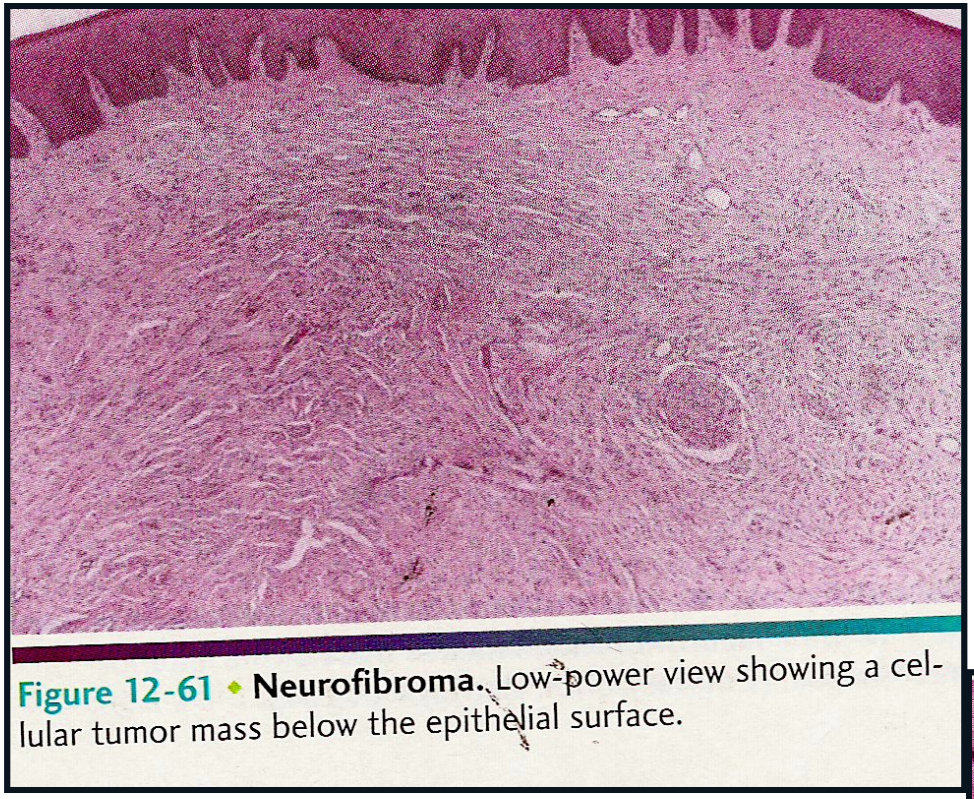


Figure 12-61 • Neurofibroma. Low-power view showing a cellular tumor mass below the epithelial surface.

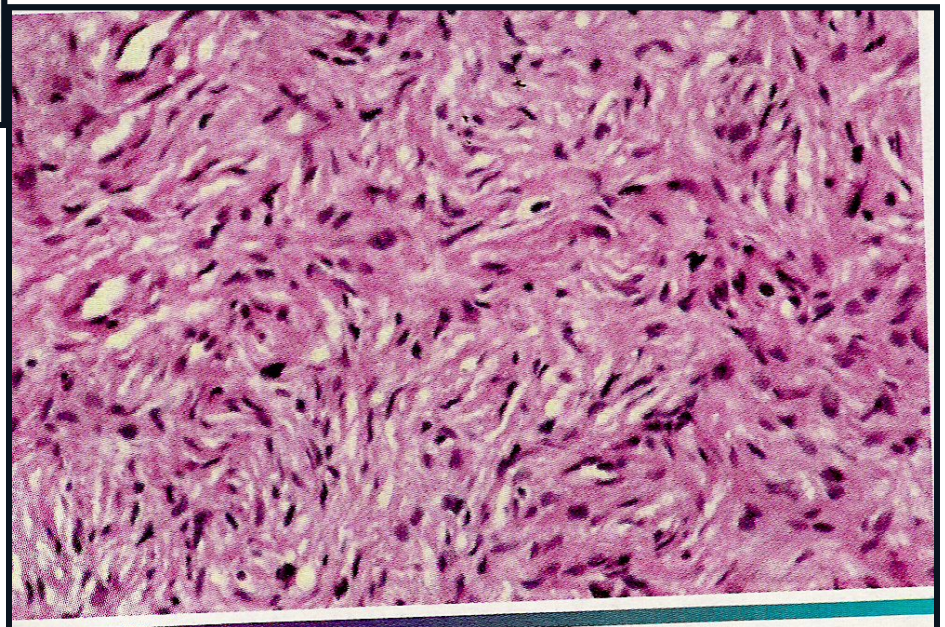
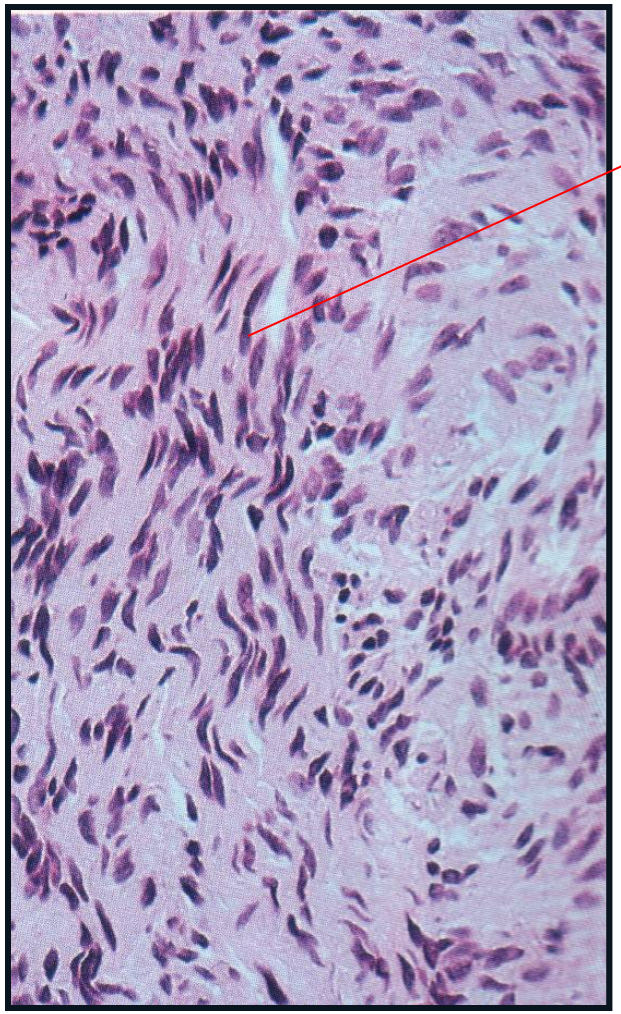
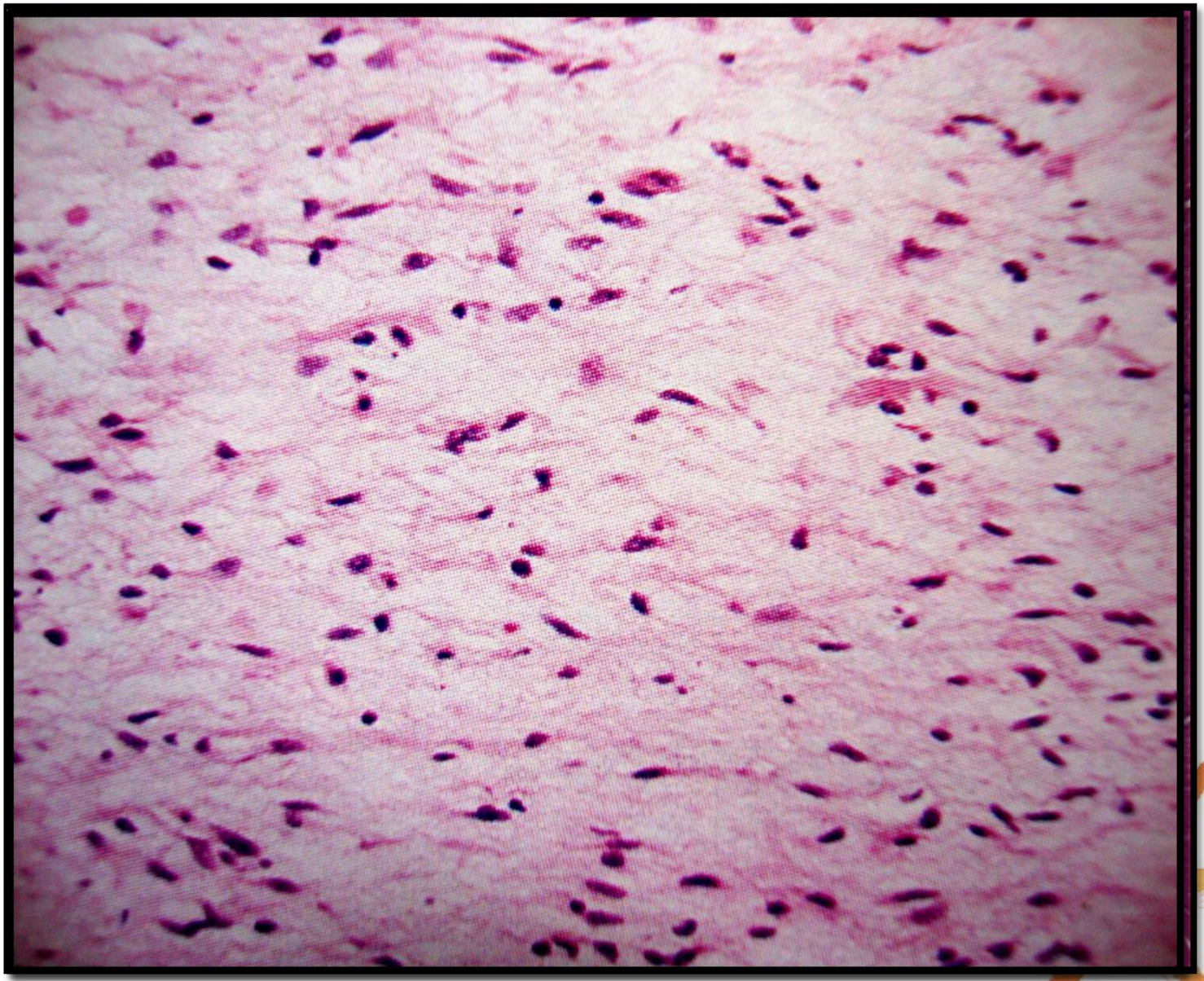


Figure 12-62 • Neurofibroma. High-power view showing spindle-shaped cells with wavy nuclei.



- Composed of interlacing bundles of spindle shaped cells with wavy nuclei, along with collagen bundles, myxoid matrix, mast cells are numerous .

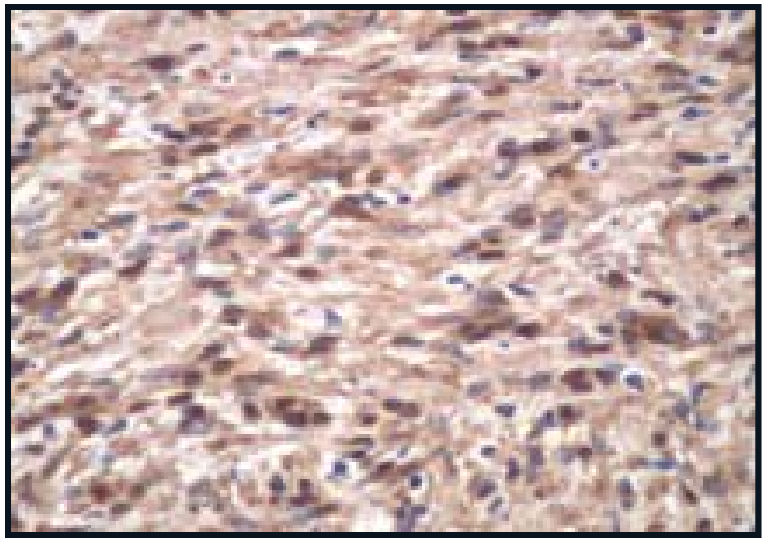


Clinical Differential Diagnosis

- Vascular malformation
- Rhabdomyoma
- Lipoma

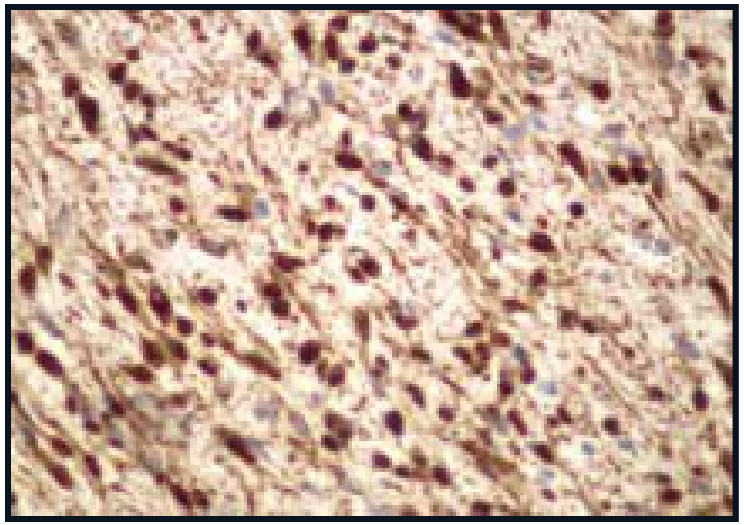
	Neurilemoma	Neurofibroma
Peak age	20-50 years	20-40 years
Location	Any, especially tongue	Any. Especially tongue, buccal mucosa
Cell origin	Schwann cell	Schwann cell and peripheral fibroblast
Histological appearence	Encapsulated tumor composed of Antoni A and B; rarely plexiform growth	Localized , diffuse, or plexiform tumor that is usallly not encapsulated
Degenerative changes	common	occasional
S-100 protein immunostaining	Intense and relatively uniform	Variable staining of cells
Occurrence in NF-I	Uncommon	Plexiform neurofibroma or multiple neurofibromas are characteristics
Malignant trasformation	Extremely rare	Rare in solitary form; more common in neurofibromatosis

Neurilemoma



S-100 protein ++++
(nearly every cell +)

Neurofibroma



S-100 protein ++ to ++++

Immunohistochemistry in the Differential Diagnosis of Schwannoma and Neurofibroma; 2004

Neurofibromatosis

- Peripheral form-type I / Neurofibromatosis Type-I (Von Recklinghausen's Neurofibromatosis) associated with changes in chromosome 17, accounts for 90% of the cases
- Central form-type II / Neurofibromatosis Type-II - long arm of chromosome 22, accounts for bilateral acoustic neuromas

Figs 9-35a to 9-35d In these facial photographs, taken about 10 years apart, the gradually advancing progress and number of cutaneous neurofibromas can be seen. (Reprinted from Reynolds RL, Pineda CA. Neurofibromatosis: Review and report of a case. JADA 1988;117:735–737. Copyright 1988 American Dental Association. Reprinted by permission of ADA Publishing, a Division of ADA Business Enterprises, Inc.)

Neurofibromatoses type 1
Von Recklinghausen's Neurofibromatoses

- Inherited as an autosomal dominant trait.
- Gene responsible for neurofibromatosis type I has been mapped to chromosome 17.
- Multiple neurofibromas and Café au lait spots-PRIME CHARACTERISTICS

- Clinical Features—

- 1)multiple neurofibromas, anywhere on skin. Appearance varies from small papules to large nodules or massive baggy pendulous masses called Elephantiasis Neuromatosa.
- Presents at birth or later in life.
- 2)Café au lait (coffee with milk) spots- smooth yellow to brown macules vary in size from 1-2 mm or cms. Present at birth or develop during 1st year of life.
- 3)Axillary freckling (Crowe's sign)
- 4)Lisch nodules translucent brown pigmented spots on iris, in nearly , all patients.
- Other abnormalities– CNS tumors, mental retardation, seizures, short stature, scoliosis
- Oral lesions-- common - enlargement of fungiform papillae(50%)

Clinical presentation

Development of café au lait spots, years before neurofibromas

Occur on areas not exposed to sun- differentiated from freckle

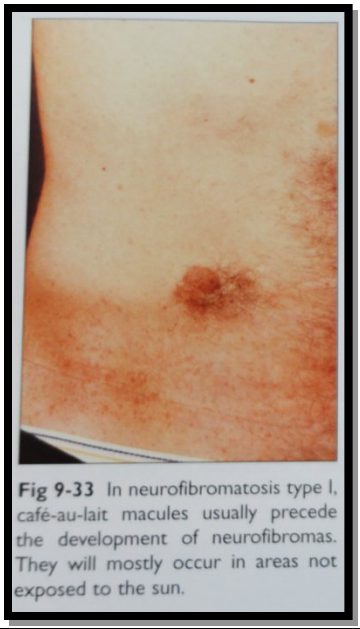
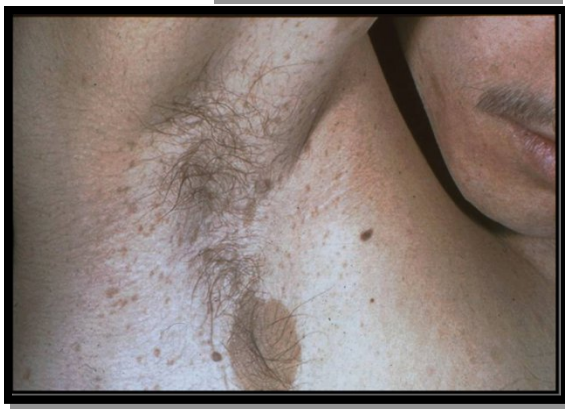


Fig 9-33 In neurofibromatosis type I, café-au-lait macules usually precede the development of neurofibromas. They will mostly occur in areas not exposed to the sun.

Axillary freckle's sign or Crowe's sign



Can be diagnosed early from a biopsy of the café au lait macule

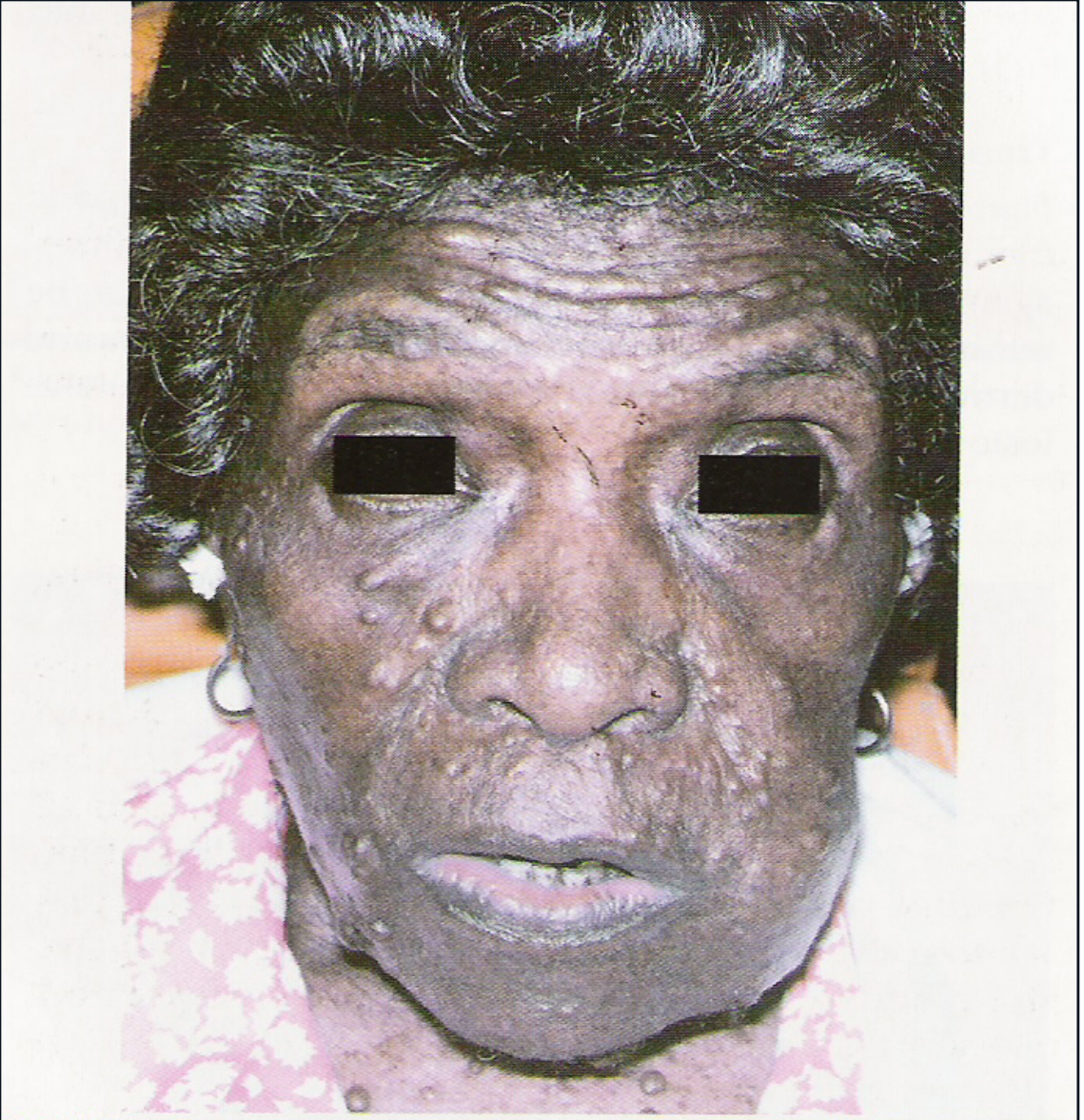
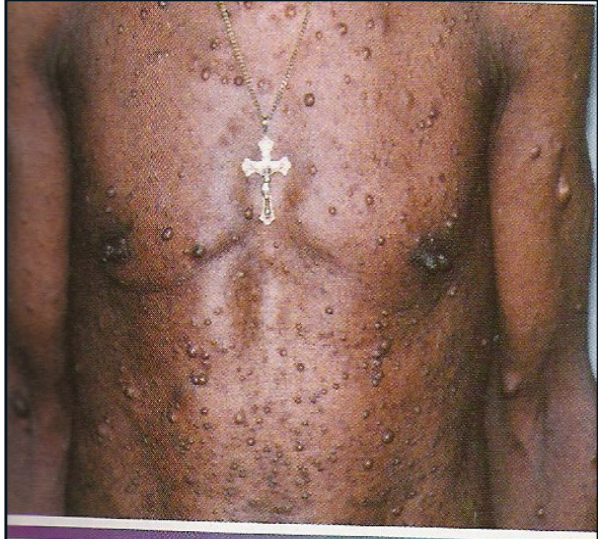
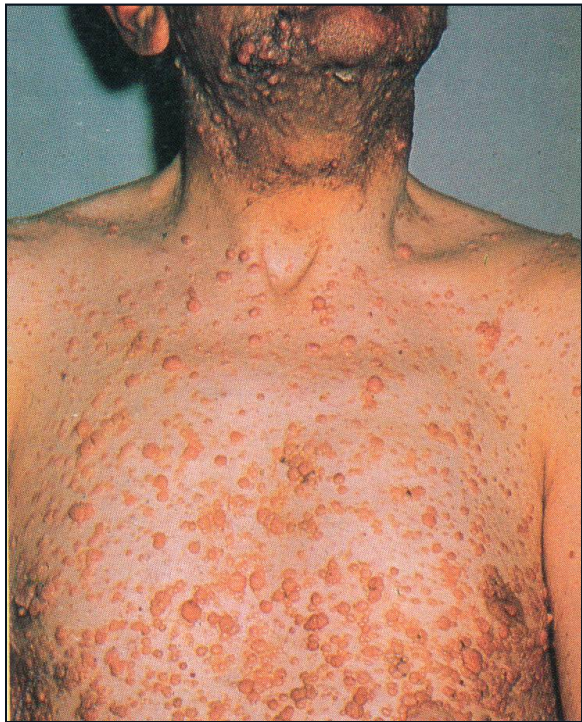


Figure 12-67 • **Neurofibromatosis.** Neurofibrosarcoma of the left cheek in a patient with type I neurofibromatosis. (From Neville BW, Hann J, Narang R, et al: Oral neurofibrosarcoma associated with neurofibromatosis type I, *Oral Surg Oral Med Oral Pathol* 72:456-461, 1991.)



12-63 • **Neurofibromatosis.** Multiple tumors of the trunk
is.



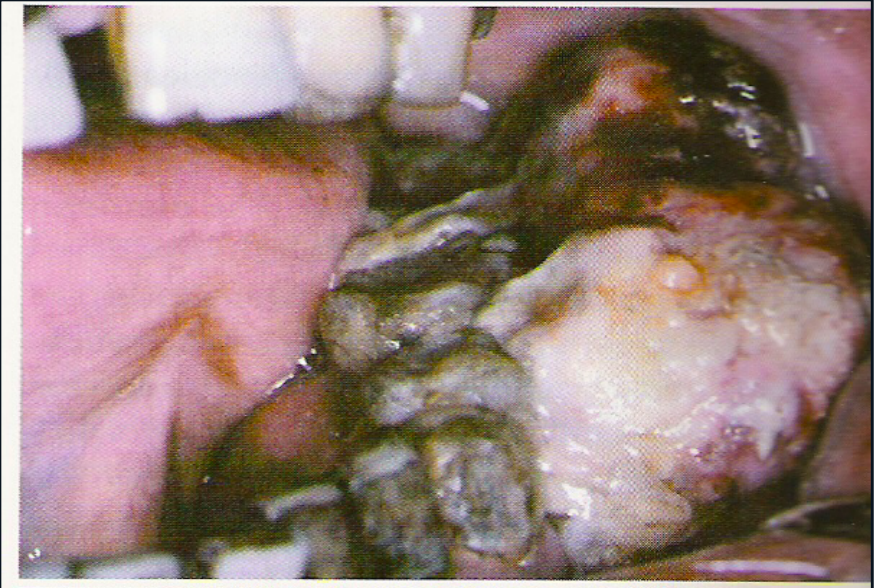


Figure 12-68 • **Neurofibromatosis.** Same patient as depic
Figure 12-67. Note the intraoral appearance of neurofibrosa
of the mandibular buccal vestibule. The patient eventually d
this tumor. (From Neville BW, Hann J, Narang R, et al: Oral n
brosarcoma associated with neurofibromatosis type I, *Oral S*
Oral Med Oral Pathol 72:456-461, 1991.)

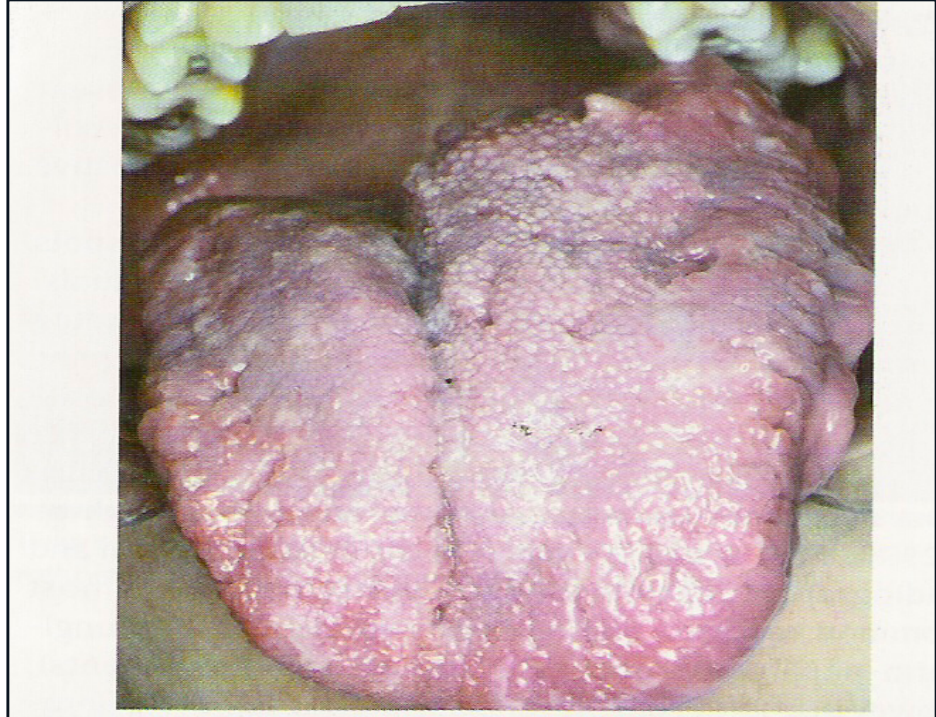


Figure 12-66 • **Neurofibromatosis.** Intraoral involvement char-
acterized by unilateral enlargement of the tongue.

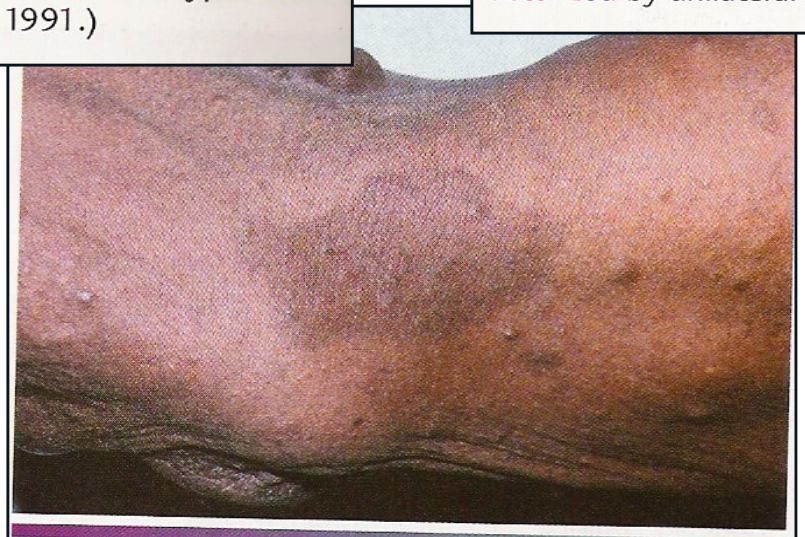
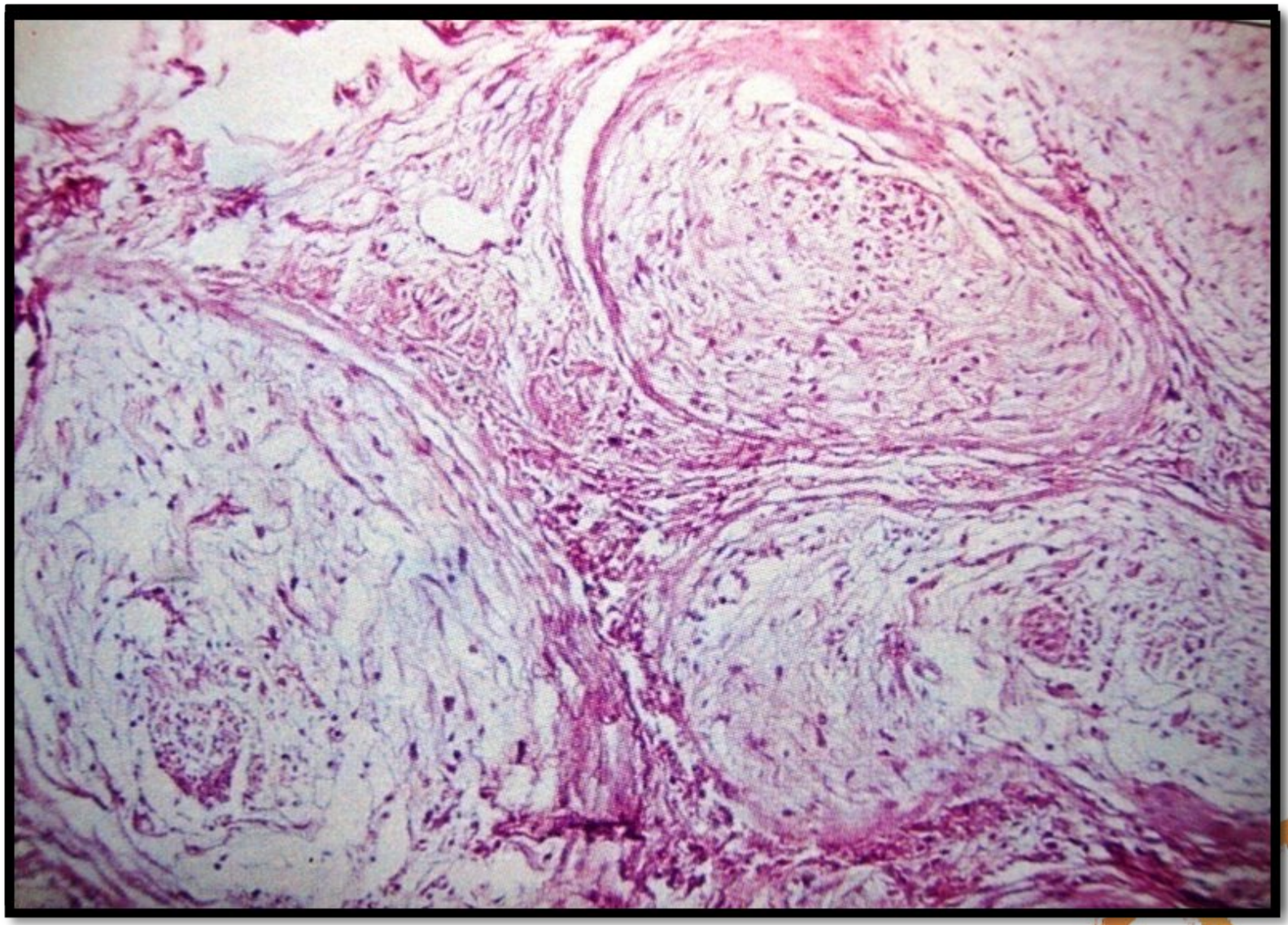


Figure 12-65 • **Neurofibromatosis.** Same patient as depicted in
Figure 12-63. Note the *café au lait* pigmentation on the arm.

Diagnostic Criteria for NF-I

Individual with any of these two criteria

- 6 café au lait macules, >5mm diameter in pre-pubertal and >15mm diameter in post-pubertal patients
- At least 2 neurofibromas of any type, or one plexiform neurofibroma
- Axillary or inguinal freckling
- Optic nerve glioma
- A first degree relative of NF- I
- Lisch nodules-2 or more
- Distinct osseous lesion



Neurofibromatoses type II

- Less common than NF- I
- Intracranial or intra spinal neural tumors

- Most frequently produce Schwannoma – as acoustic neuroma

- Hearing loss

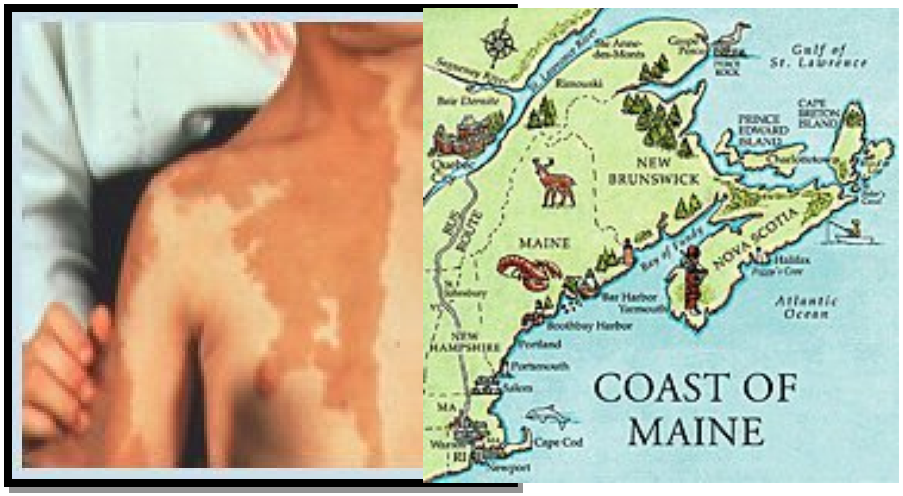
Diagnostic Criteria for NF-II

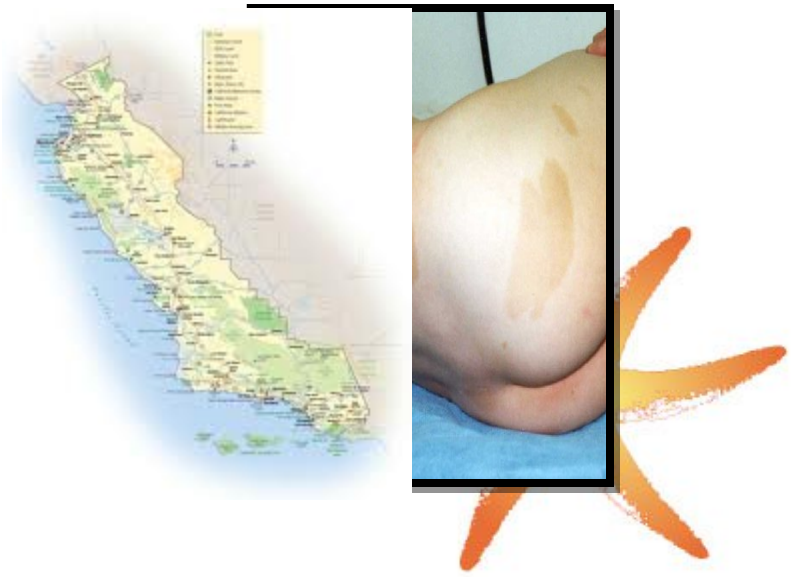
- Bilateral 8th nerve tumor or First degree relative of NF-II

- Unilateral 8th nerve tumor, or two of the following
- Dermal or subcutaneous neurofibromas
- Plexiform neurofibromas
- Schwannoma
- Meningioma
- Juvenile posterior capsular lenticular opacity

Clinical Differential Diagnosis

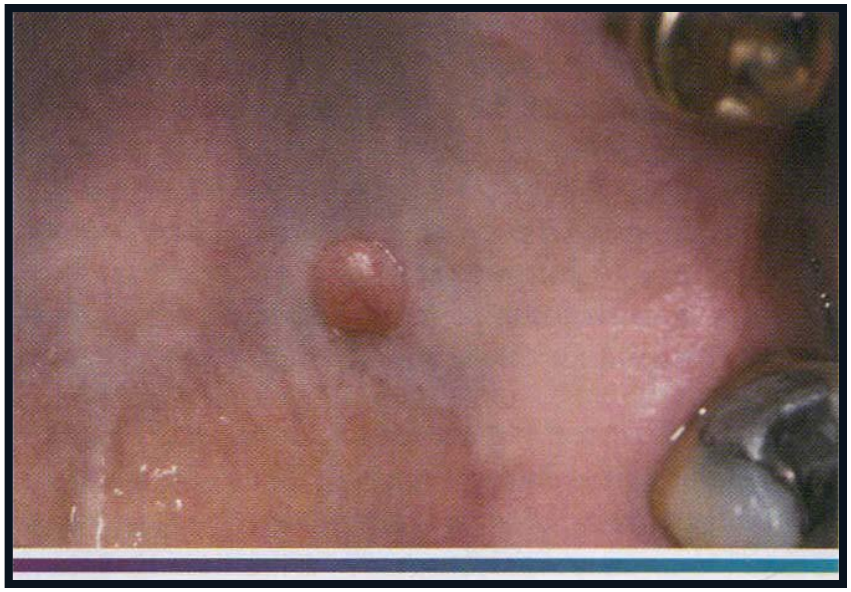
- Jaffe type of fibrous dysplasia
- Albright syndrome





Palisaded Encapsulated Neuroma

- Predilection for peri-oral facial skin and palate
- Non-painful sub-mucosal or intradermal mass
- Result of reactive axonal regeneration attempts
- Considered a hyperplasia rather than a neoplasm



Palisaded encapsulated neuroma. Small. Painless nodule of the lateral hard palate.

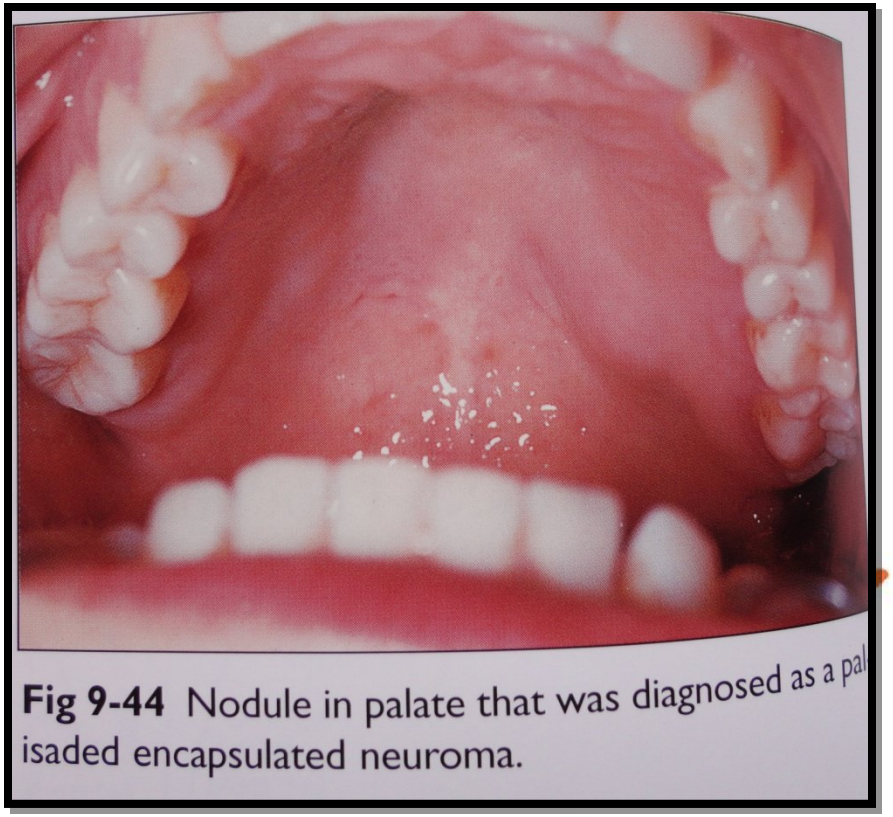
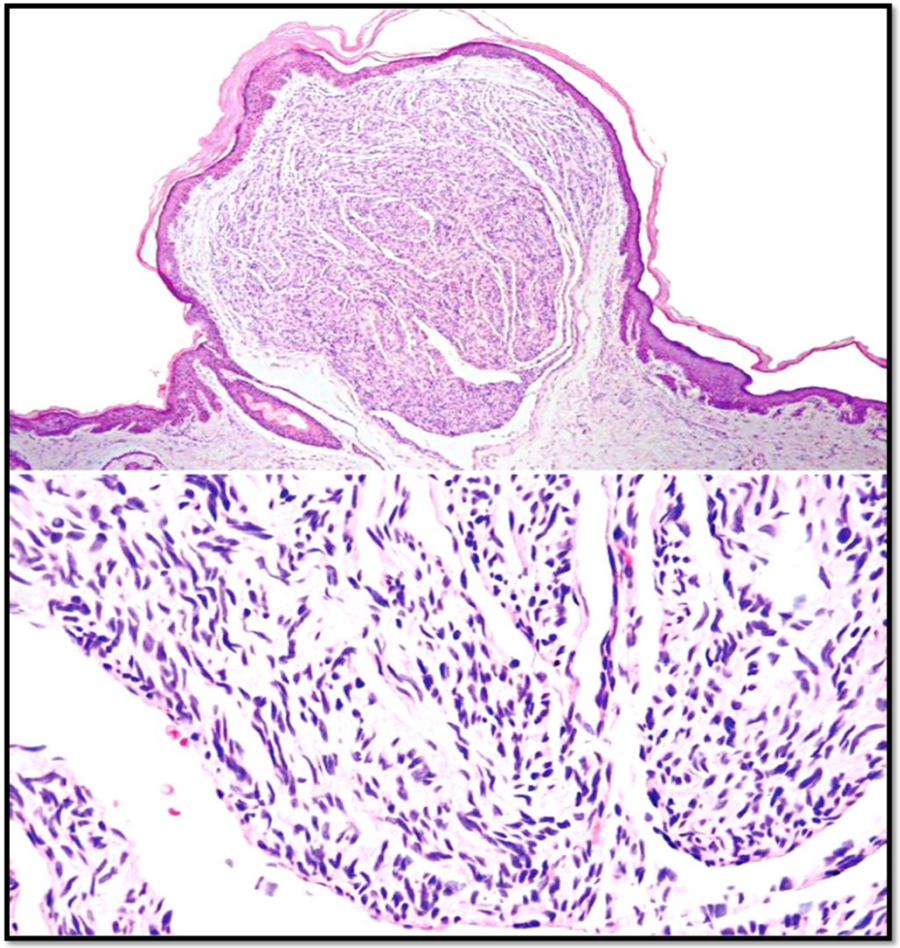
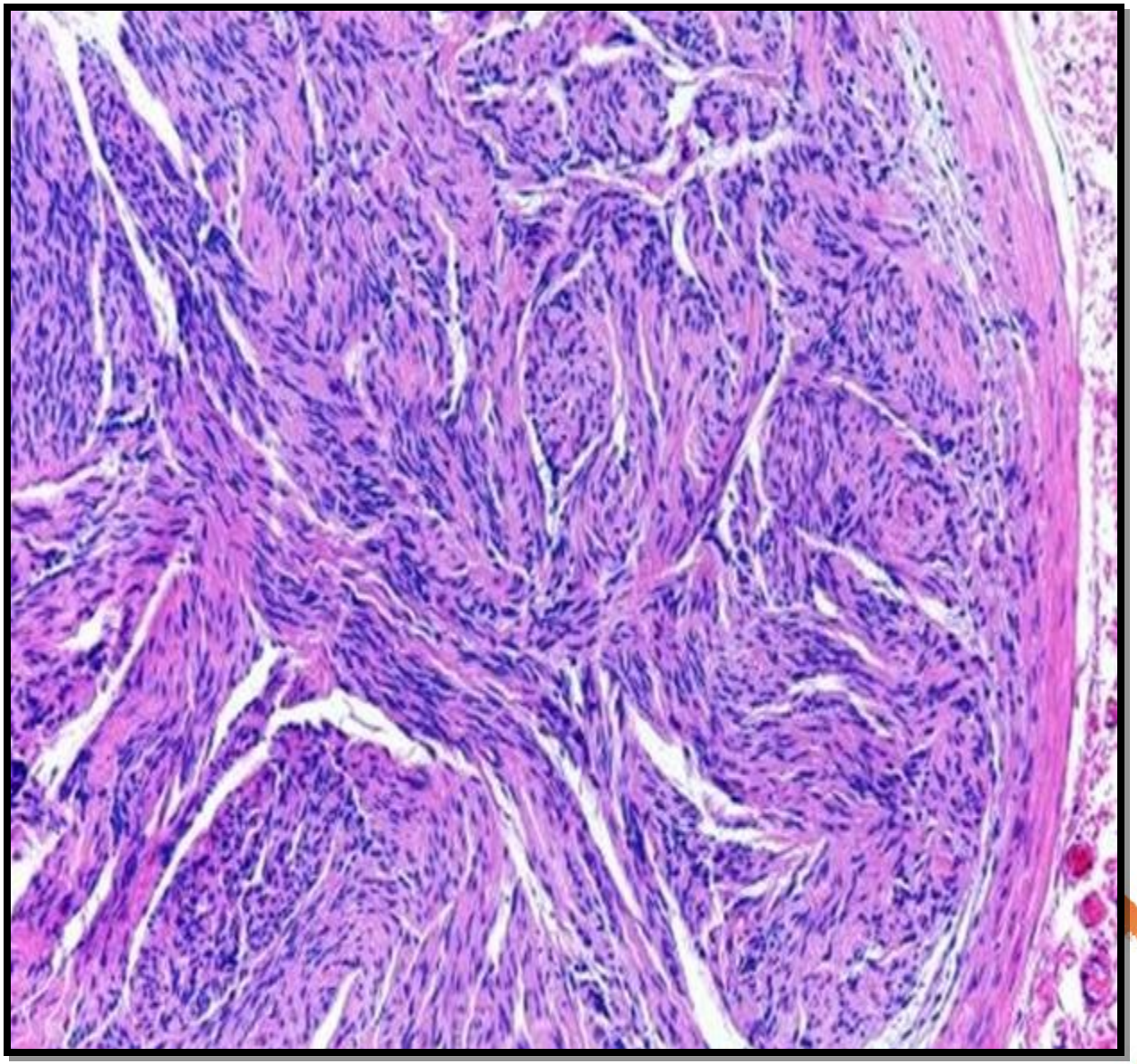


Fig 9-44 Nodule in palate that was diagnosed as a pal
isaded encapsulated neuroma.



Palisaded encapsulated neuroma. low-power view showinga well-circumscribed, nodular proliferation of neural tissue.

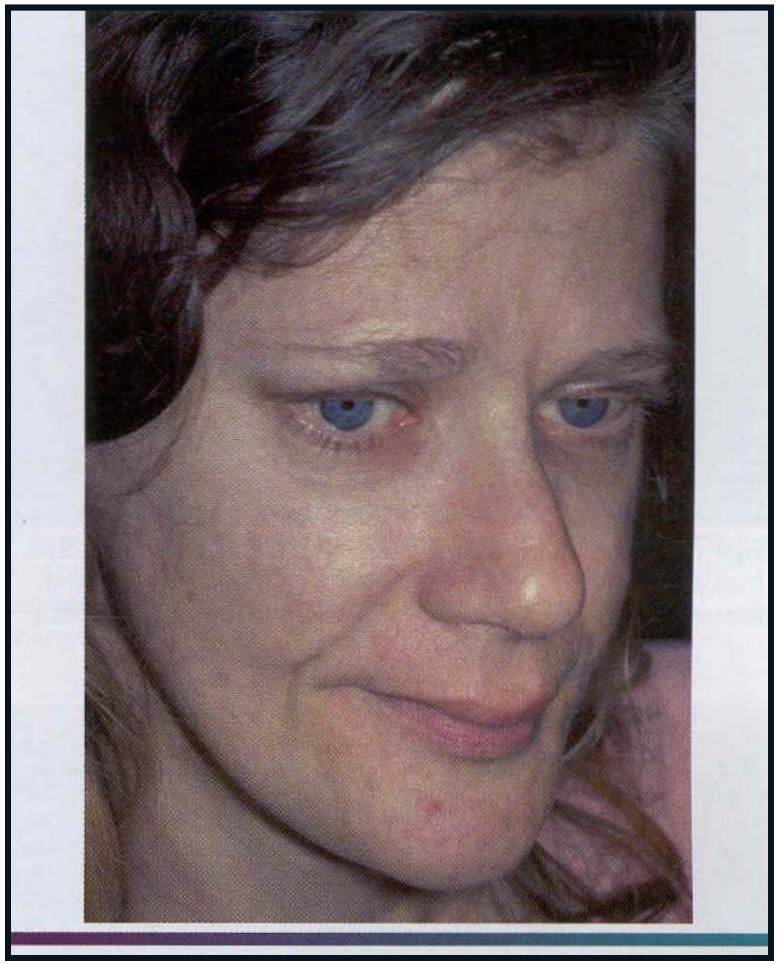


Clinical Differential Diagnosis

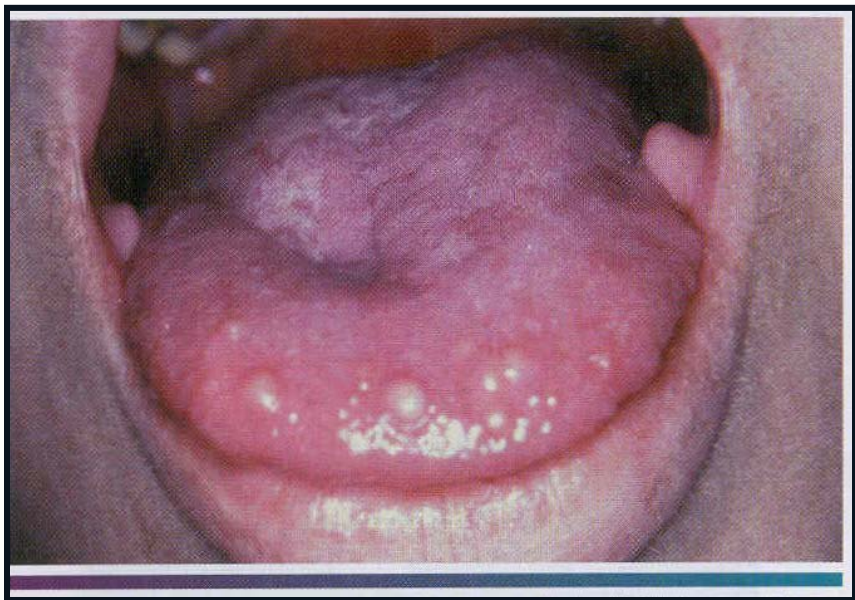
- Schwannoma
- Sebaceous or epidermal cyst
- Nasolabial cyst

MUITIPLE ENDOCRINE NEOPLASIA TYPE 2B

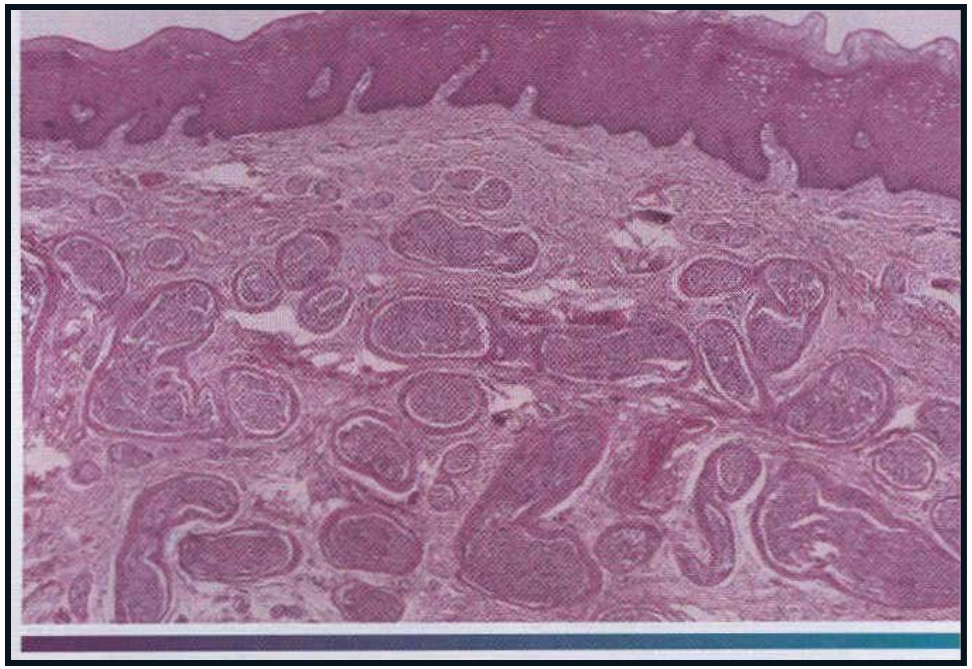
- The multiple endocrine neoplasia (MEN) syndromes are a group of rare conditions characterized by tumors or hyperplasias of neuroendocrine tissues.
- MEN type 2B- pheochromocytomas and medullary thyroid carcinoma
- Patients with MEN type 2B have mucosal neuromas that especially involve the oral mucous membranes



Multiple endocrine neoplasia (MEN) type 2B. Note the narrow face and eversion of the upper eyelids.

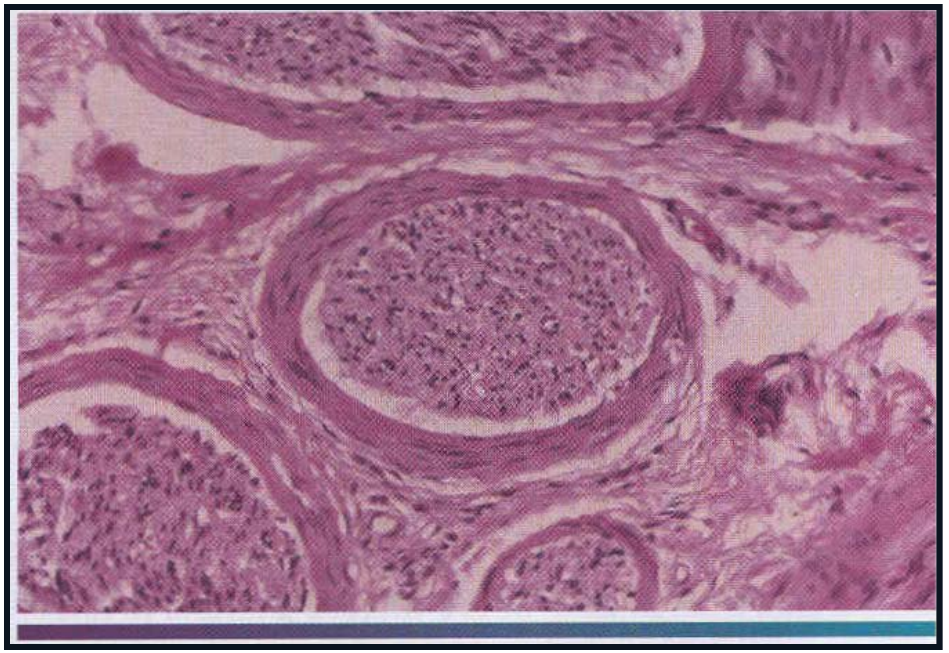


Multiple endocrine neoplasia (MEN) type 2B. Multiple neuromas along the anterior margin of the tongue and bilaterally at the commissures



Multiple endocrine neoplasia (MEN)type 2B.
low-power view of an oral mucosal neuroma showing marked hyperplasia of nerve bundles.

Multiple endocrine neoplasia (MEN) type 2B. Note the prominent thickening of the perineurium.



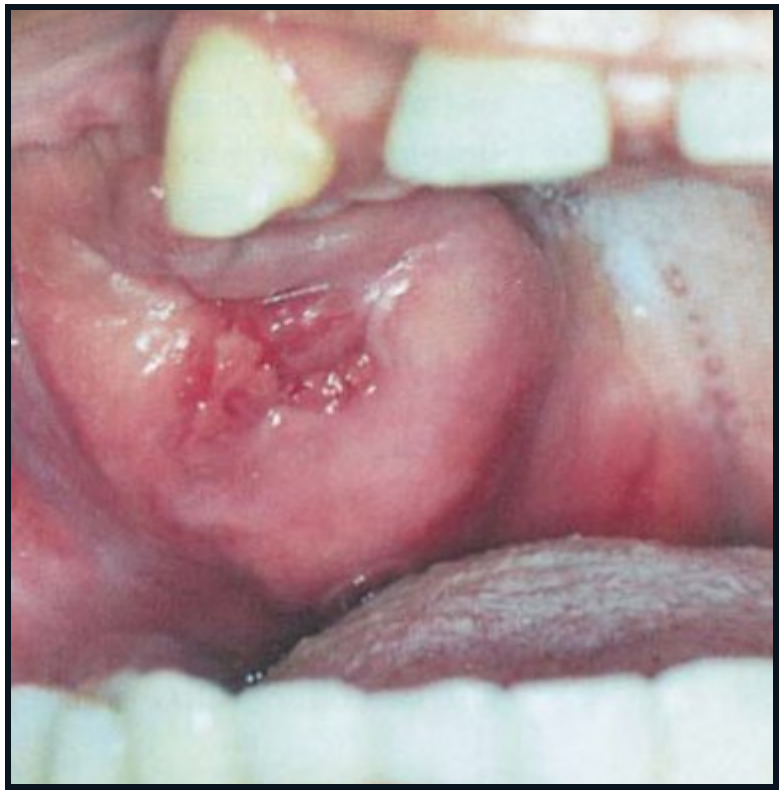
MALIGNANT TUMORS OF NERVE TISSUE ORIGIN

1. MALIGNANT PERIPHERAL NERVE SHEATH TUMOR
2. OLFACTORY NEUROBLASTOMA

Malignant Peripheral Nerve Sheath Tumor

- Also called as neurofibrosarcoma; malignant neurilemmoma; malignant schwannoma; neurogenic sarcoma.
- Malignant transformation of a neuro-fibroma in hereditary neurofibromatosis with a latency of 15 to 20 years
- Centrally within the jaws or as a deep soft tissue malignancy
- M: F- (3:2) , 20 to 60 years

- Spindle cell malignancy of Schwann cells.
- Half of the cases are associate with neurofibromatosis-I.
- More common in lower extremities.
- Involves neck, tongue, soft palate.
- Central lesion involves mandibular nerve.
- Manifest diffuse radiolucency.



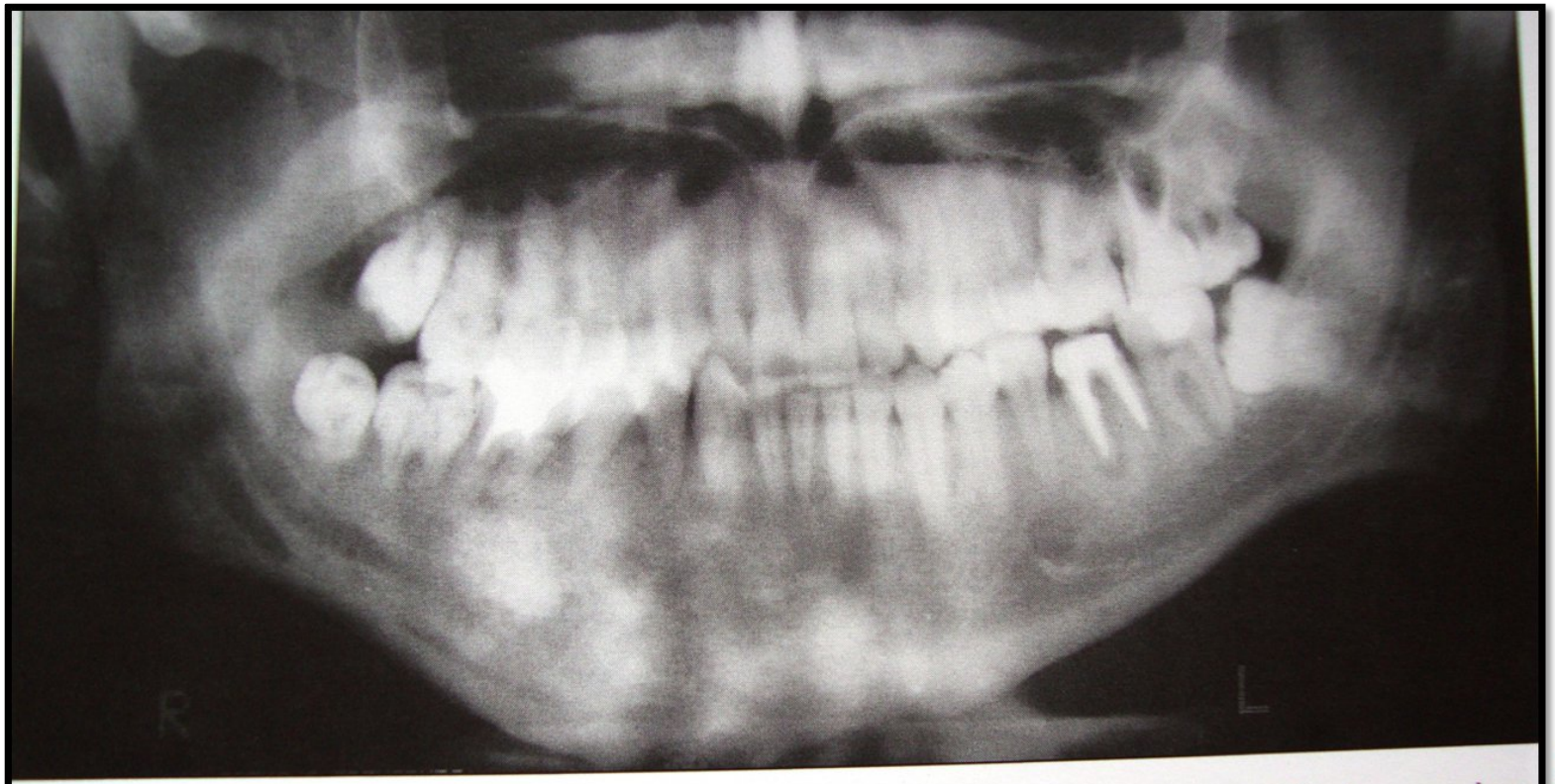
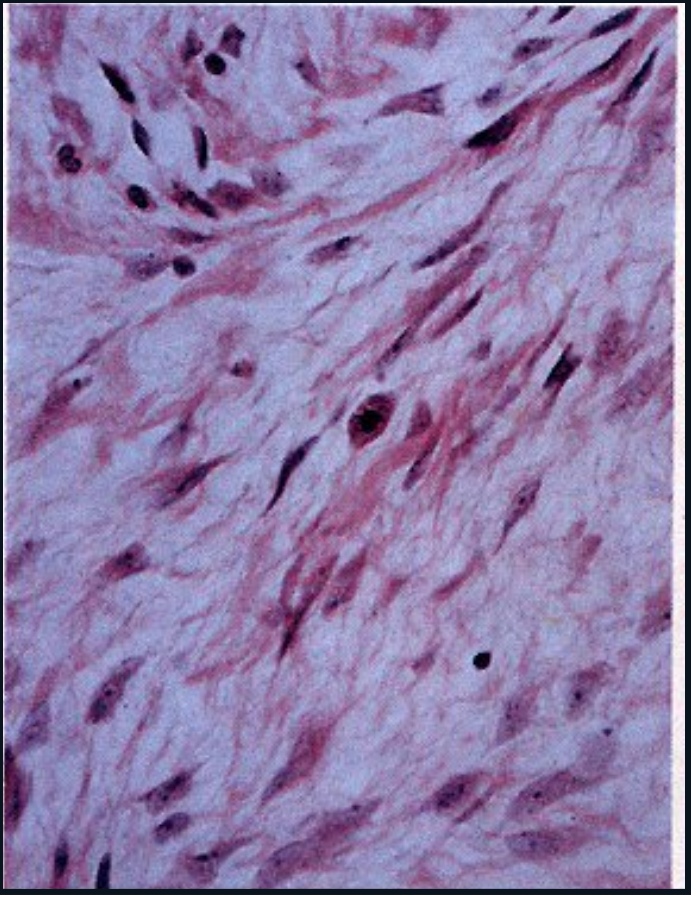


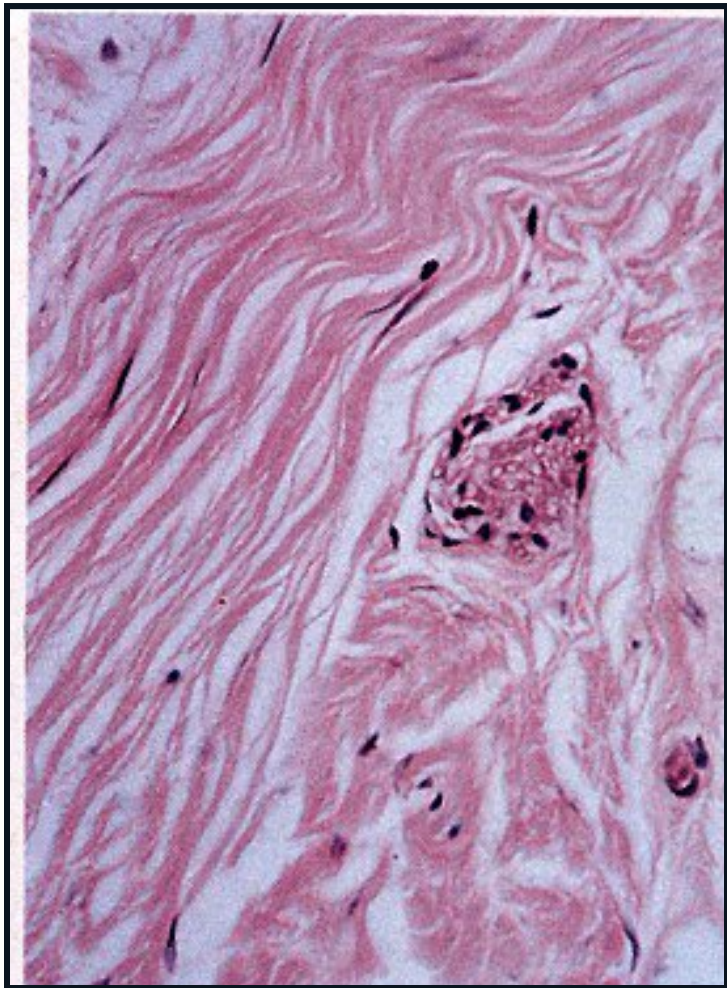
Fig 10-15a Diffuse irregular radiolucency with expansion of the mandibular canal related to a malignant peripheral nerve sheath tumor of the mandible.



Spindle cells arranged in fascicles.

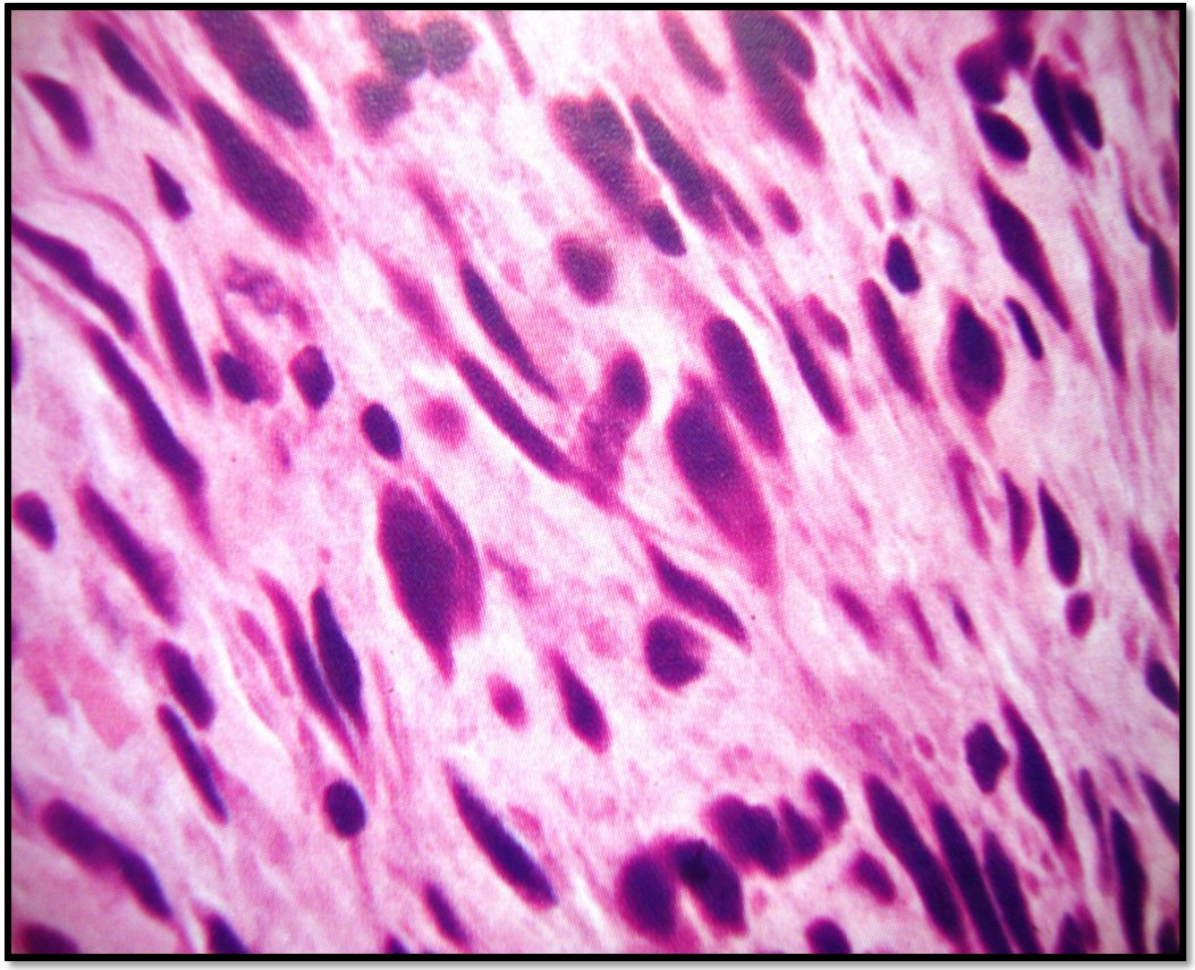
myxoid regions.

Cellular and nuclear pleomorphism



Eosinophilic cytoplasm.

High mitotic activity.



Spindle cells arranged in fascicles.
and myxoid regions.
Cellular and nuclear pleomorphism
Eosinophilic cytoplasm.
High mitotic activity.

- Three categories:

1) Epitheloid: plump, rounded or ovoid epitheloid cells scattered throughout the spindled lesional cells
2) Mesenchymal
3) Glandular: contains areas with usually well differentiated ductal structures lined by simple, stratified, cuboidal or columnar epithelial cells with occasional goblet cells.

- Malignant Triton Tumor: Malignant nerve sheath tumors with rhabdo-myoblastic differentiation

- .

Differential diagnosis:

- Leiomyosarcoma
- Fibrosarcoma
- Benign nerve sheath tumor- neurofibroma

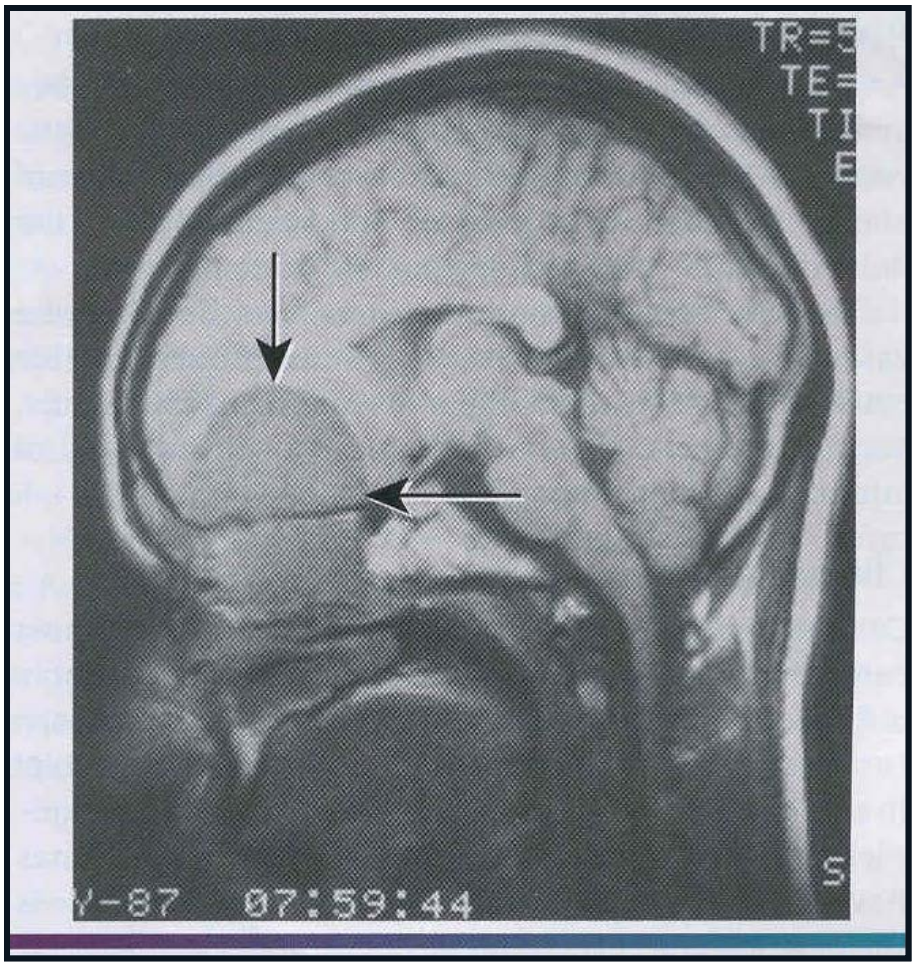
Olfactory Neuroblastoma or Esthesioneuroblastoma

- Malignant tumors of neuroectodermal cell origin arising from olfactory placode high in the nasal cavity
- Production of norepinephrine and dopamine beta-hydroxylase, the enzyme that catalyzes the conversion of dopamine to norepinephrine
- Infancy up to the age of 3 years and are a common malignancy in children, 90% occur after the age of 10 years.

- Lesion- unilateral nasal obstruction and epistaxis

- Large lesions that erode into paranasal sinuses or the orbits may obstruct the nasolacrimal duct - nasal speculum examination will identify a gray fleshy mass in the superior meatus of the nasal cavity

- Symptoms -epiphora, headaches, or anosmia



Olfactory neuroblastoma.
A TI-weighted sagittal magnetic resonance image (MRI) showing a tumor filling the superior nasal cavity and ethmoid sinus, with extension into the anterior cranial fossa

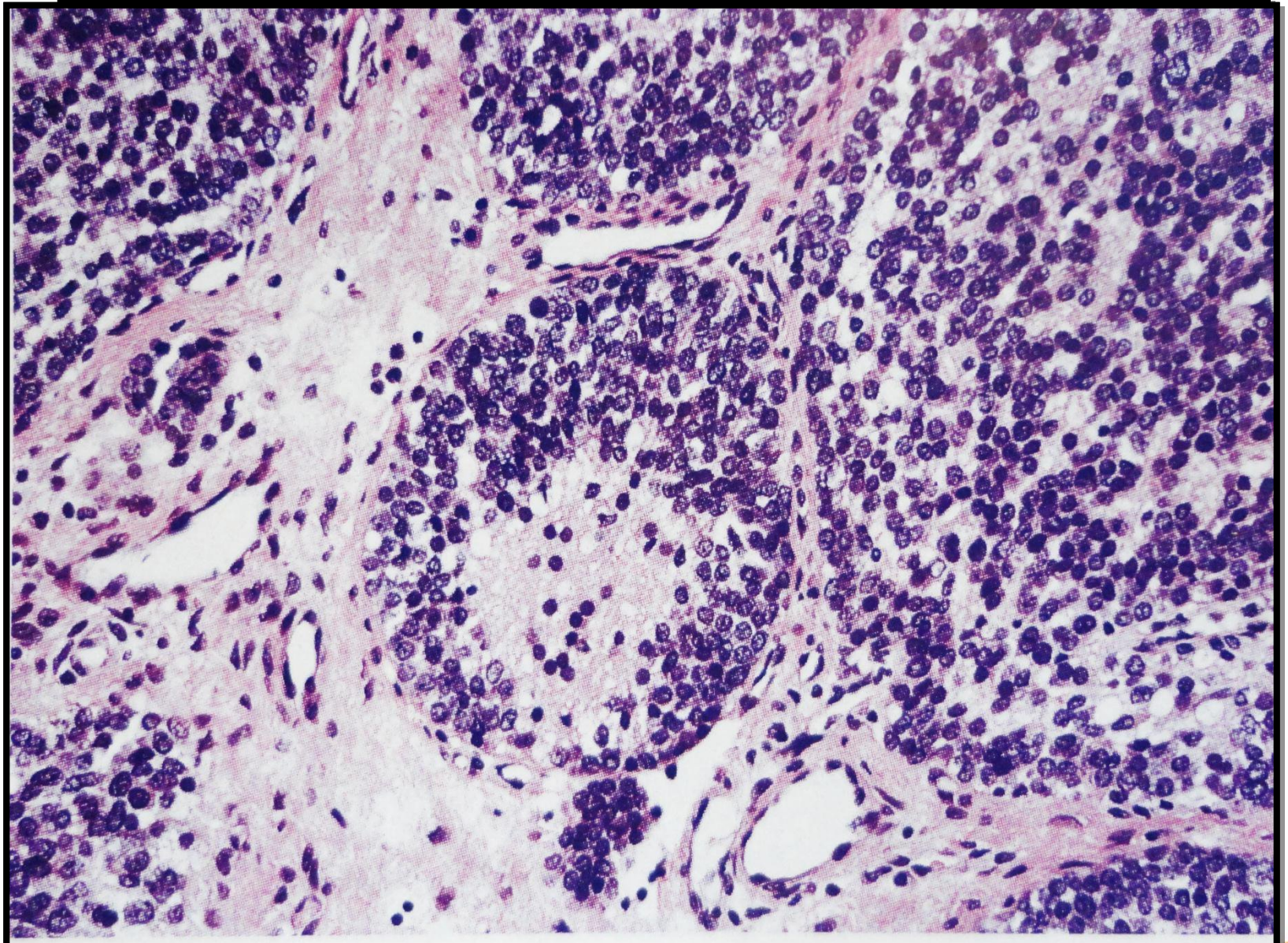


Fig. 41.3 Olfactory neuroblastoma. Higher power view shows the lobules of round cells in a fine fibrillary matrix.

Differential Diagnosis

- Nasal obstruction, epistaxis, and periorbital expansion in a teenager or young adult suggestive of a nasopharyngeal angiofibroma or a rhabdomyosarcoma
- Malignant fibrous histiocytoma or fibrosarcoma- A transnasal incisional biopsy done
- Meningioceoles that have perforated the cribriform plate to present in the superior aspect of the nasal cavity- Meningioceole would leak cerebrospinal fluid if it were biopsied


THANK
YOU