

PERIPHERAL NERVE SHEATH TUMORS

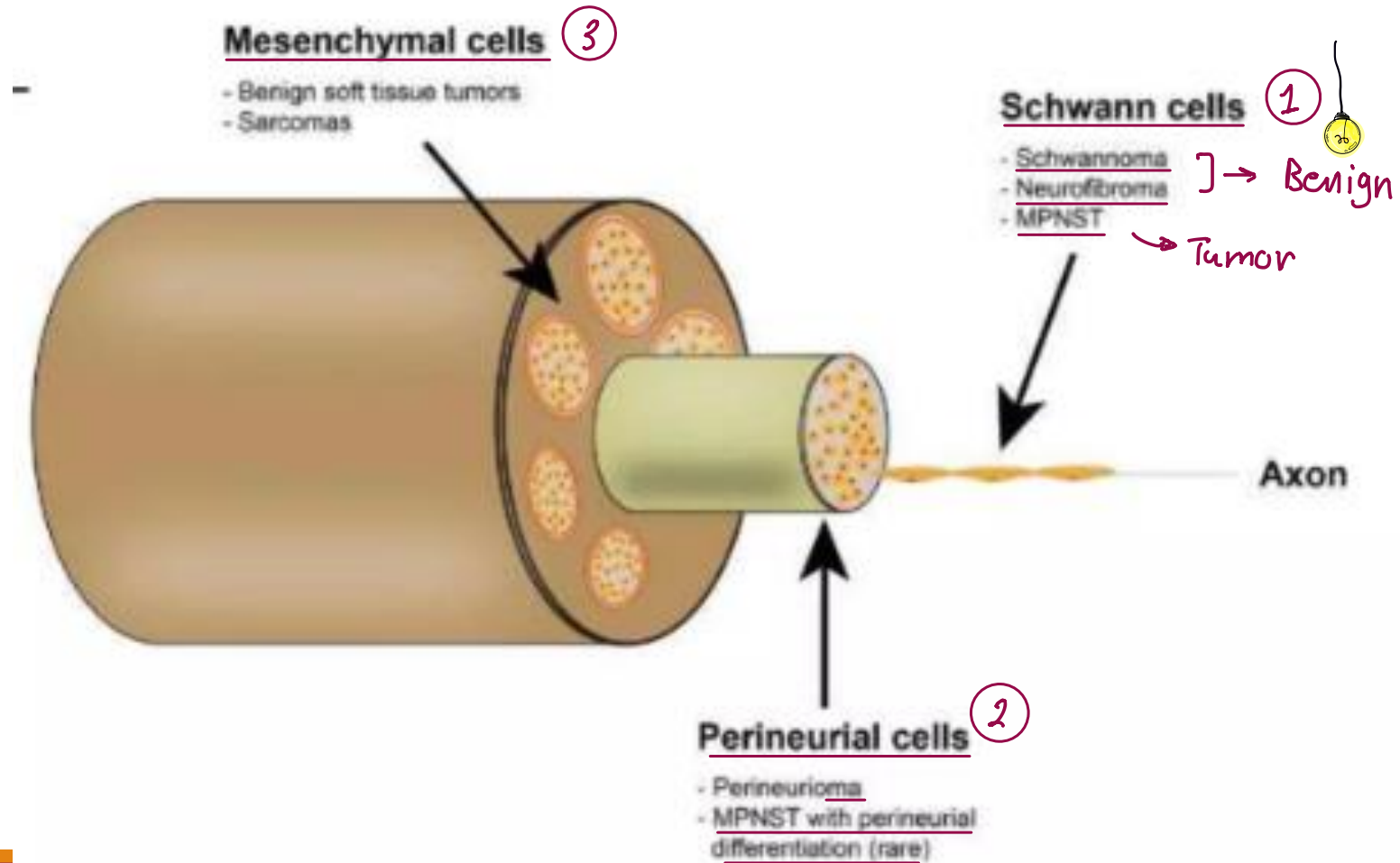
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Comparison of Peripheral Nerve Lesions

Feature	Schwannoma	Neurofibroma	MPNST	Traumatic Neuroma
Nature	Benign tumor	Benign tumor	Malignant neoplasm	Non-neoplastic proliferation
Encapsulation	Completely encapsulated	Unencapsulated	Often large with necrosis	Disorganized fascicles
Cell of Origin	Differentiated Schwann cells	Schwann cells & fibroblasts	Peripheral nerve cells	All normal nerve components
Histology	Hypo/hypercellular patterns (Spindle cells)	Spindle cells with wire-like collagen fibrils +Hypocellular	Monotonous spindle cells; fascicular pattern	Axons, Schwann, & perineural cells in stroma
Associations	NF2, Schwannomatosis	Often unknown etiology	NF1 or prior radiation	Prior surgery or inflammation
Common Sites	Soft tissues, spinal roots, 8th cranial nerve	Skin (superficial), Head & Neck (diffuse)	Trunk, extremities, head & neck	Site of nerve damage
Clinical Course	Indolent; symptoms rare unless large	Painless, slowly growing mass	Aggressive; can metastasize	Post-procedural or inflammatory
Treatment	Simple surgical excision (Curative)	Marginal excision or conservative follow-up	Aggressive surgery + Radiation	Not specified (Reactive)

PERIPHERAL NERVE SHEATH TUMORS



I. Schwannomas

Benign ^A Encapsulated, well circumscribed ^B nerve sheath tumor arising from differentiated Schwann cells.

More common in 30 - 60 years of age.

may occur in soft tissues, internal organs, or spinal nerve roots.

The most commonly affected CNS is the  vestibular portion of the ⁸ eighth nerve.

Combination : [Acquired + Inherited]

Pathophysiology

May occur spontaneously. [No genetic Background]

Can occur in familial tumor syndromes, such as:

- neurofibromatosis type 2 (NF2). →
- schwannomatosis.
- Carney complex

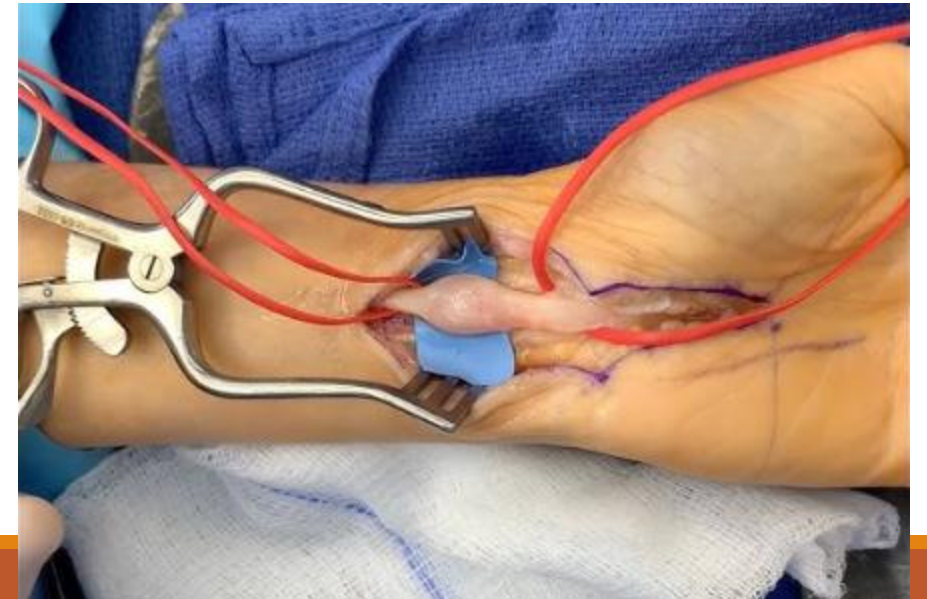


Clinical features

- ❖ Pain and neurological symptoms are uncommon unless the tumor is large.
- ❖ Surgical excision is the treatment of choice, Local recurrence is uncommon
- ❖ Most cases have an indolent course

- Curative
- Simple

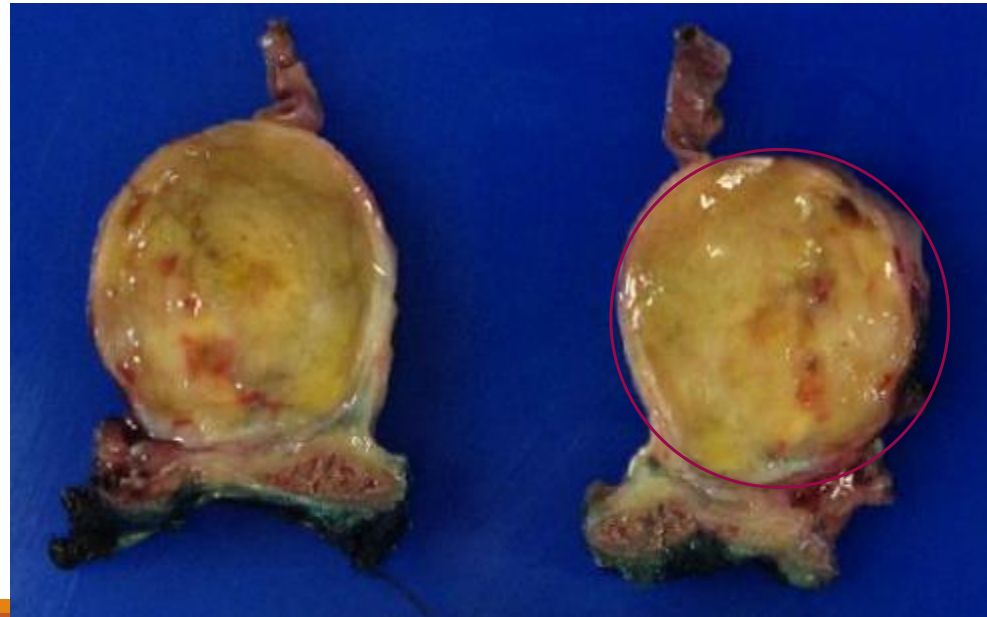
- no infiltration → simple



Gross description

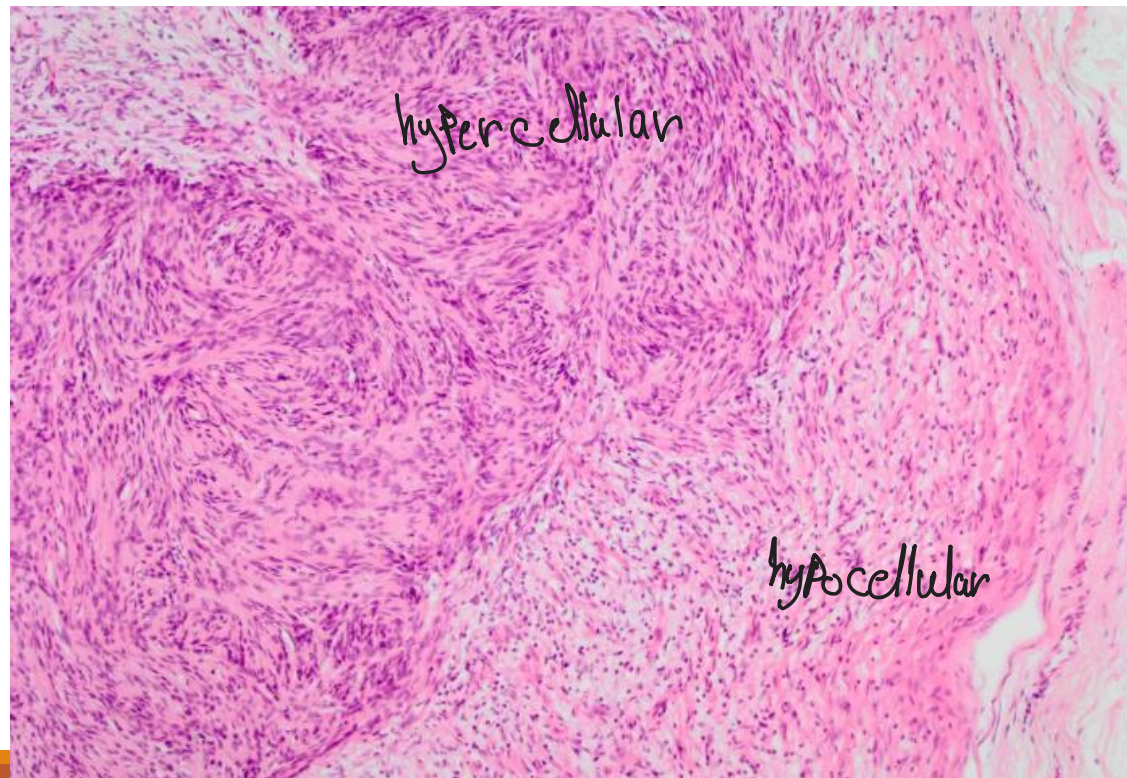
Usually solitary and completely encapsulated.

- * encapsulated
- * uniform shape
- * No hemorrhage



Histological features

Spindle cell proliferation, arranged in hypo/hypercellular pattern.



II. Neurofibroma

❖ Benign, unencapsulated, low cellularity, spindle cell neoplasm

❖ Benign peripheral nerve sheath tumor with classic identifiable features including the presence of a neuronal component comprising transformed Schwann cells and a nonneoplastic fibrous component that includes fibroblasts.

❖ According to WHO, etiology is unknown

❖ Superficial neurofibromas respond well to marginal excision and deep-seated neurofibromas are treated conservatively

① neck head
② Need to follow-up
③ خيار الجراحة غير محبب إلا عند زيادة الأعراض

Ⓐ
Ⓑ[Ⓜ]
[capsule مائي]

Clinical presentation

Localized neurofibromas are superficial and evenly distributed over the body surface.

Diffuse neurofibromas are usually in the head and neck region. ✖

Presented as Painless, slowly growing, solitary, skin colored, soft mass.

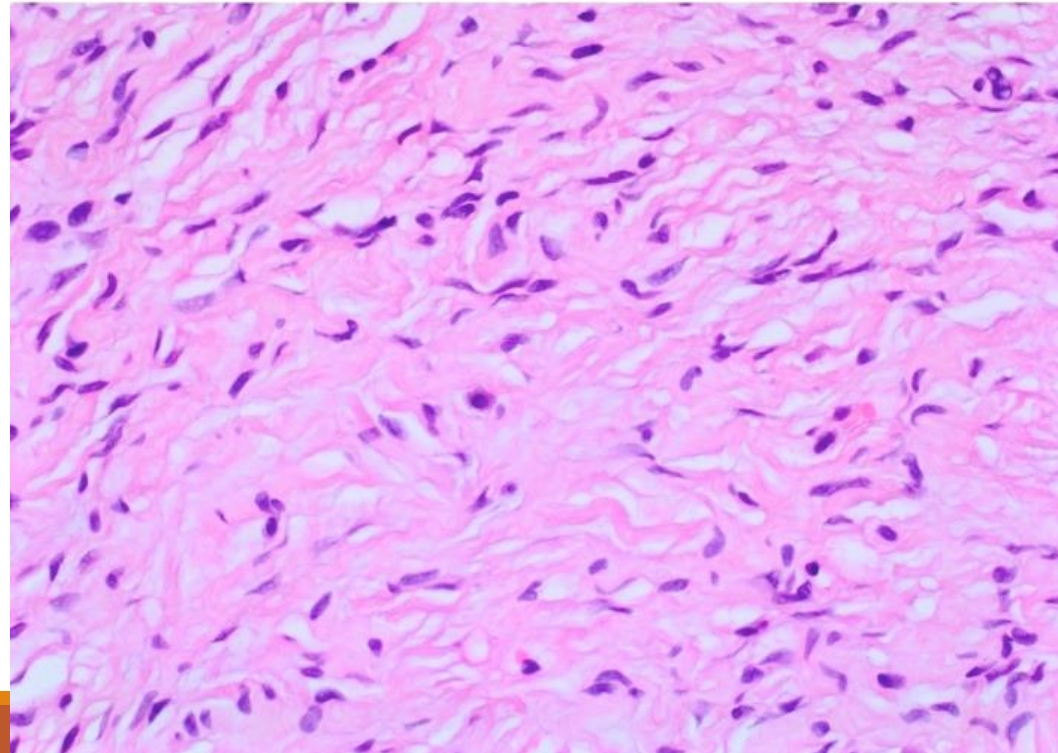


Histological features

proliferation of all elements of peripheral nerves including Schwann cells with **wire-like collagen fibrils** and fibroblasts



-H.E Stain -



ما بقدر أميِّز الصورة شو هي بال

H.E stain

لذلك لازم أستعمل صبغة ثانية


بتوضح التفاصيل وتدعى ← Immune stain

III. Malignant Peripheral Nerve Sheath Tumors

- ❖ Malignant neoplasm arising from peripheral nerve.
- ❖ May arise from a preexisting nerve sheath tumor in neurofibromatosis type 1 (NF1) or in the setting of prior radiation therapy

Clinical features

❖ No sex predilection

❖ Can arise in virtually any anatomic location. 
Most common sites are the trunk and extremities, followed by head and neck.

❖ Patients with NF1 are typically younger than their sporadic and radiation associated counterparts

↓
→ Any tumor with genetic Background → usually occur at younger Age

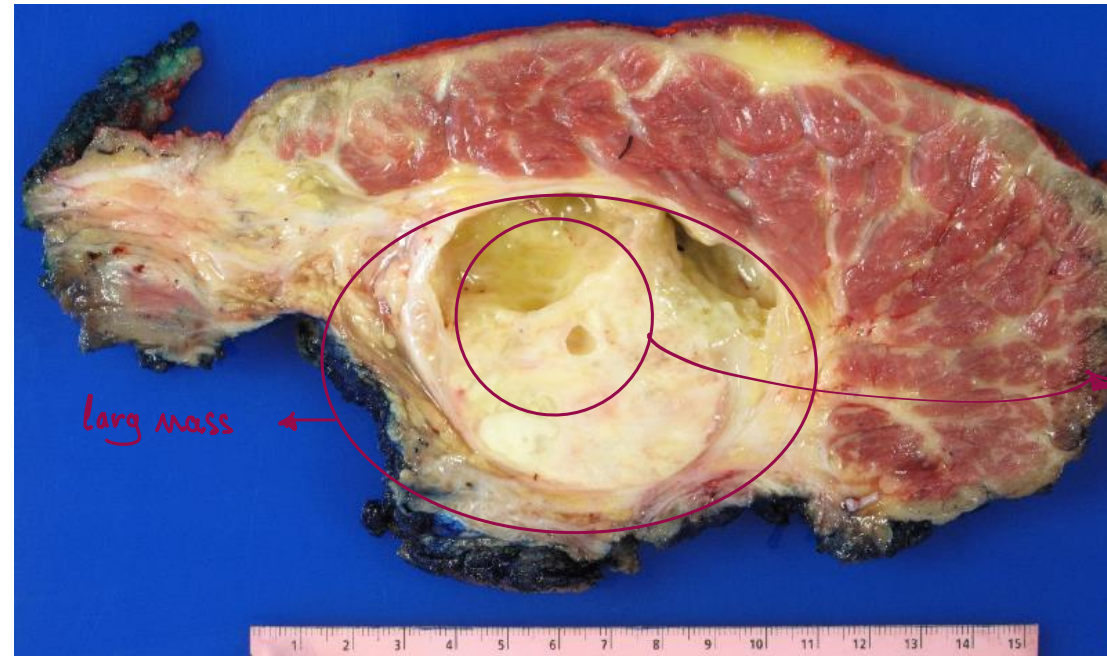
Treatment

- ⊛ *- clearance -* Aggressive surgical resection followed by radiation therapy to achieve local control .

- Late stage:* ⊛ Therapeutic options for metastatic MPNST are limited; conventional chemotherapy is usually limited to patients with metastatic disease

Gross features

Large gluteal mass with areas of **necrosis** and **cavitation**.

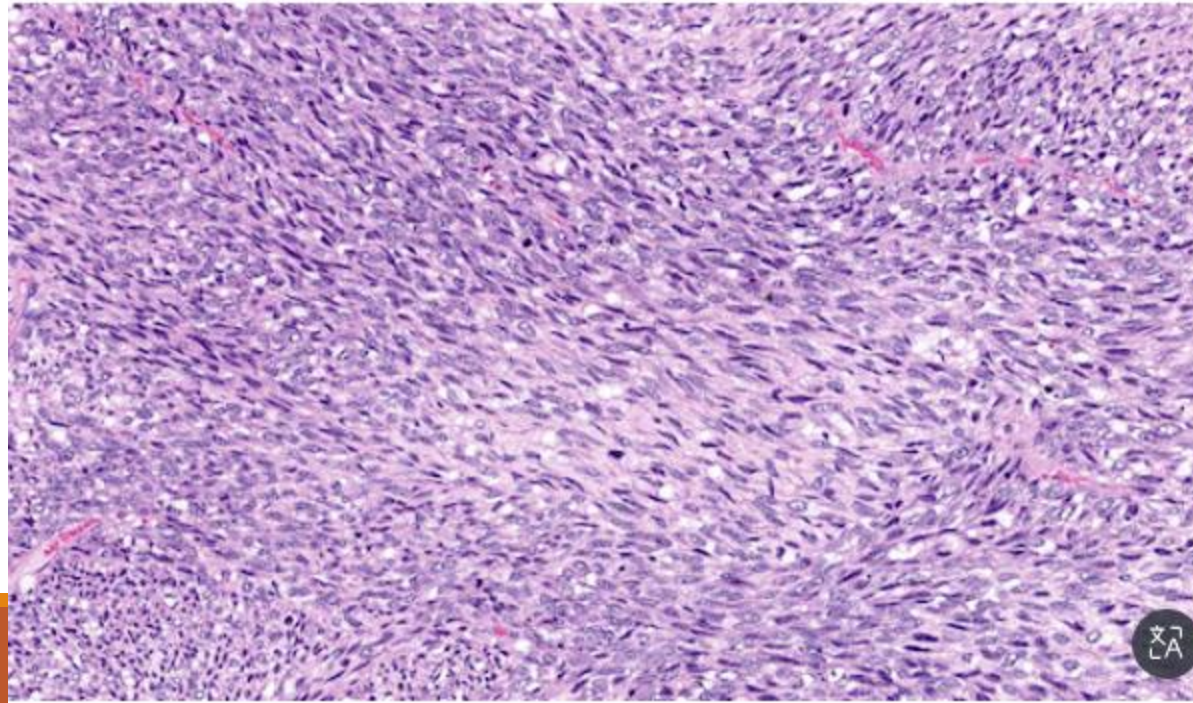


Microscopic features

Typical morphologic features of MPNST include a fairly monotonous spindle cell morphology with a fascicular growth pattern.

↪ growth like fascicles [Bundles]

↑ mitosis



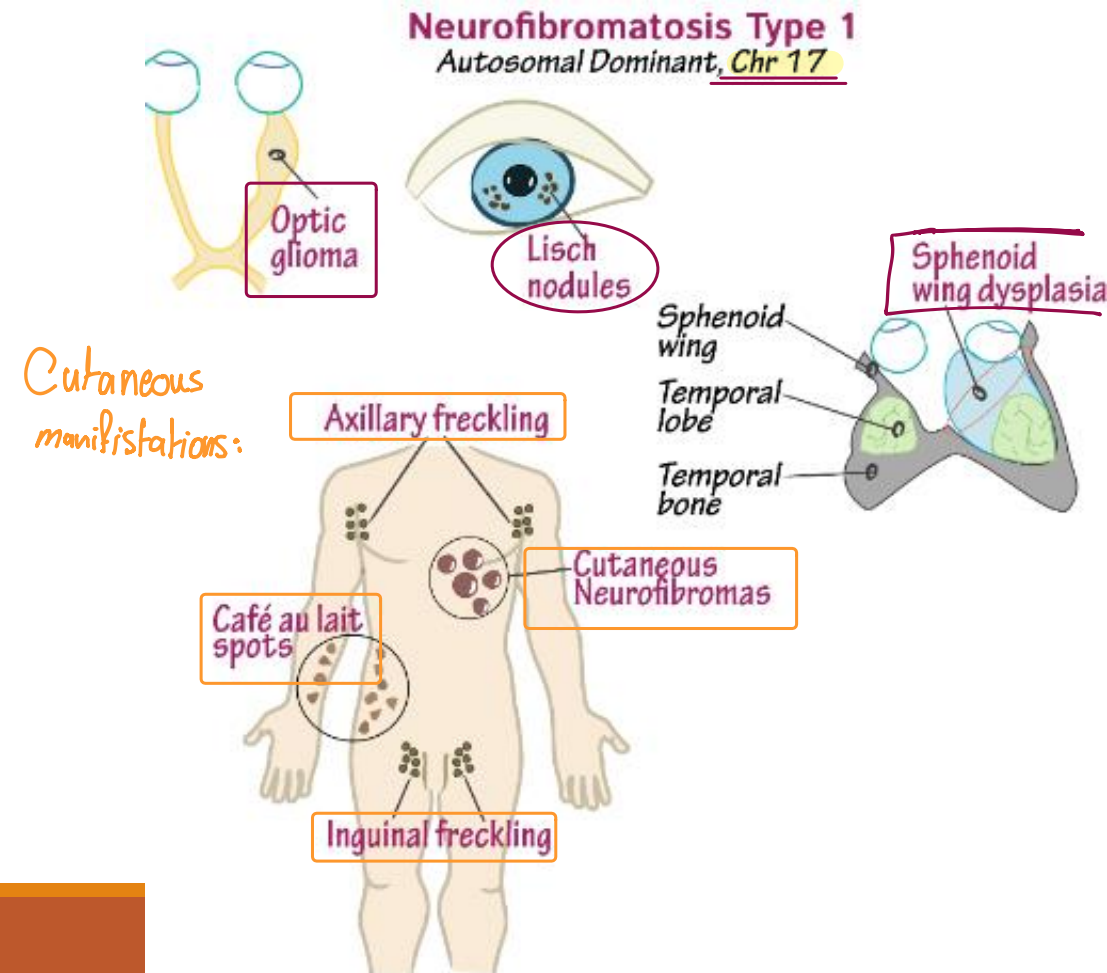
Neurofibromatosis

- ❖ Neurofibromatosis (NF) refers to a group of genetic conditions in which tumors grow in the nervous system.
- ❖ The tumors are non-cancerous (benign) and often involve the skin or surrounding bone. symptoms are often mild and each condition presents differently.

Neurofibromatosis type 1 (NF1)

Is a multisystem genetic disorder that commonly is associated with:

- ✓ cutaneous.
- ✓ neurologic.
- ✓ Orthopedic e.g scoliosis



Neurofibromatosis type 2 (NF2)

Is a rare genetic disorder causing noncancerous tumors on:

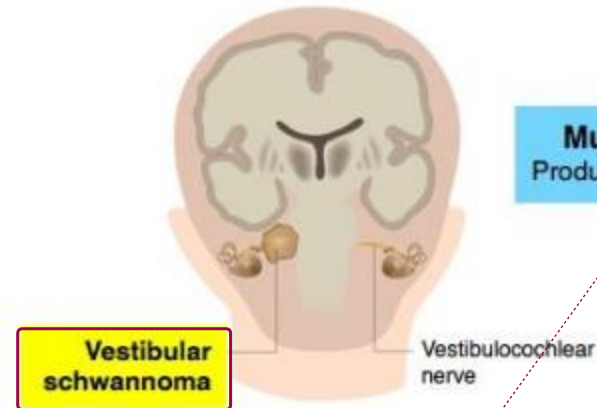
- ✓ the brain.
- ✓ spinal cord.
- ✓ cranial nerves

Neurofibromatosis Type 2

Autosomal dominant Roshif

غير مطلوب ↑

Mutation in the *NF2* gene
Produces merlin (tumor suppressor)



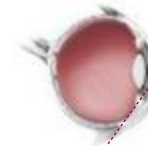
Neurologic lesions

- Bilateral vestibular schwannoma
- Schwannomas of other cranial nerves
- Intracranial meningiomas
- Spinal tumors
- Peripheral neuropathy



Eye lesions

- Cataracts
- Epiretinal membranes
- Retinal hamartomas



Skin lesions

- Cutaneous tumors
- Skin plaques
- Subcutaneous tumors



Traumatic neuroma

- ❖ Nonneoplastic, disorganized proliferation of normal nerve components at the site of previously damaged peripheral nerves.
- ❖ Presents after surgery or procedure, Can arise without prior surgery (e.g., inflammation).

Microscopic features

Disordered proliferation of nerve fascicles composed of axons, surrounded by Schwann cells and perineural cells within collagenous stroma

