PERIPHERAL NERVE SHEATH TUMORS

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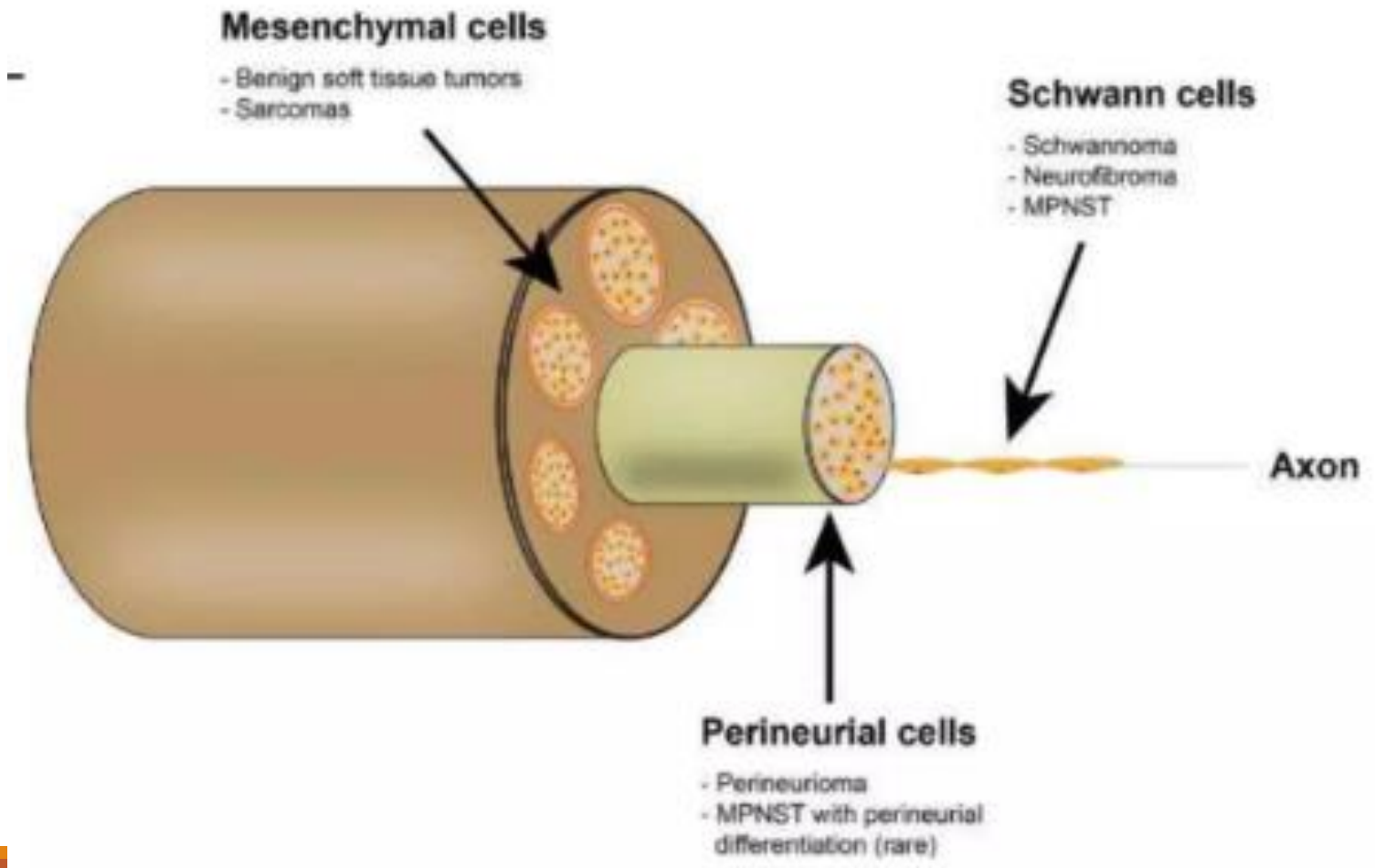


I. Schwannomas

Benign Encapsulated, well circumscribed 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 eighth nerve.

Pathophysiology

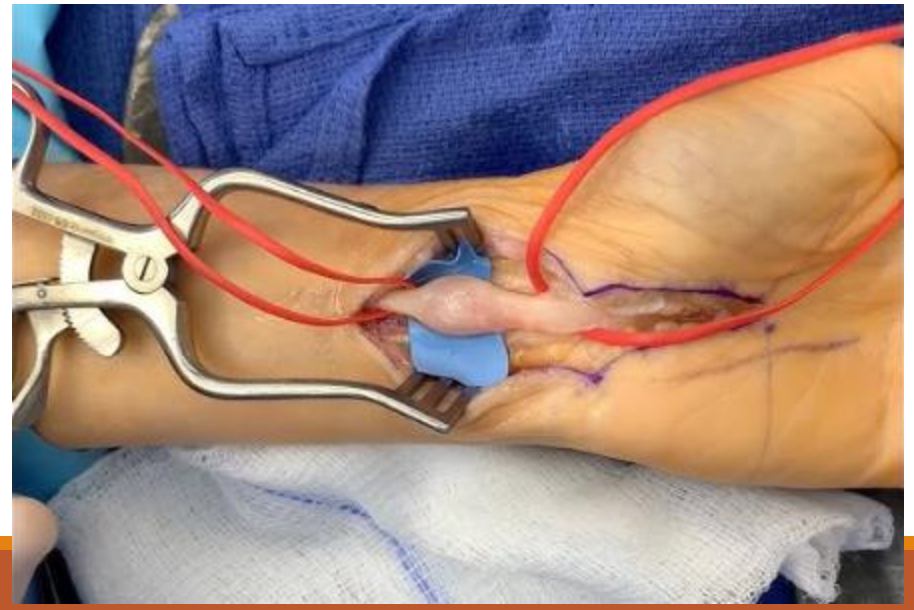
May occur spontaneously.

Can occur in familial tumor syndromes, such as:

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

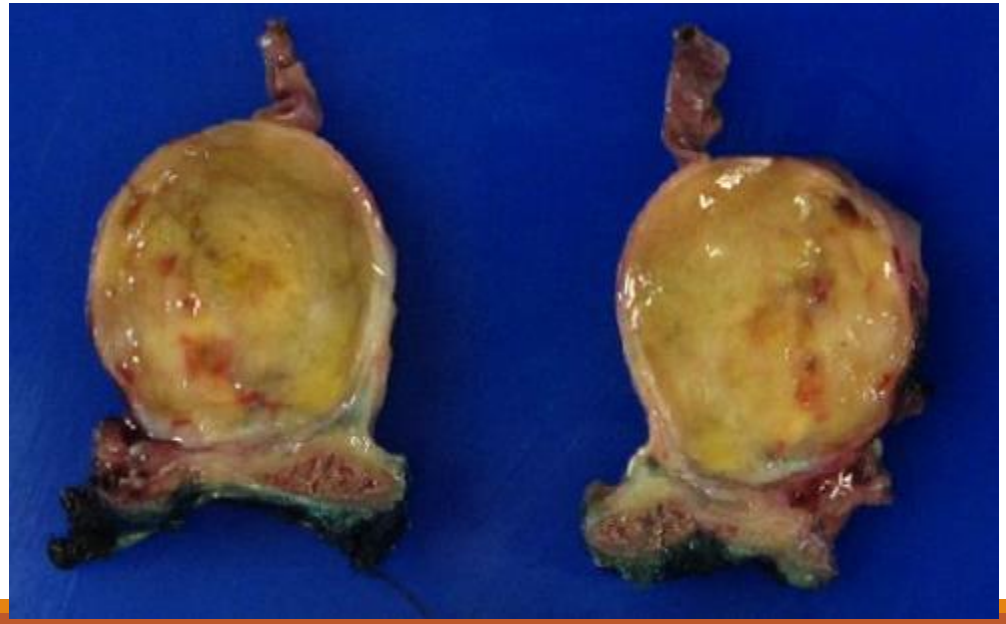
Clinical features

- 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



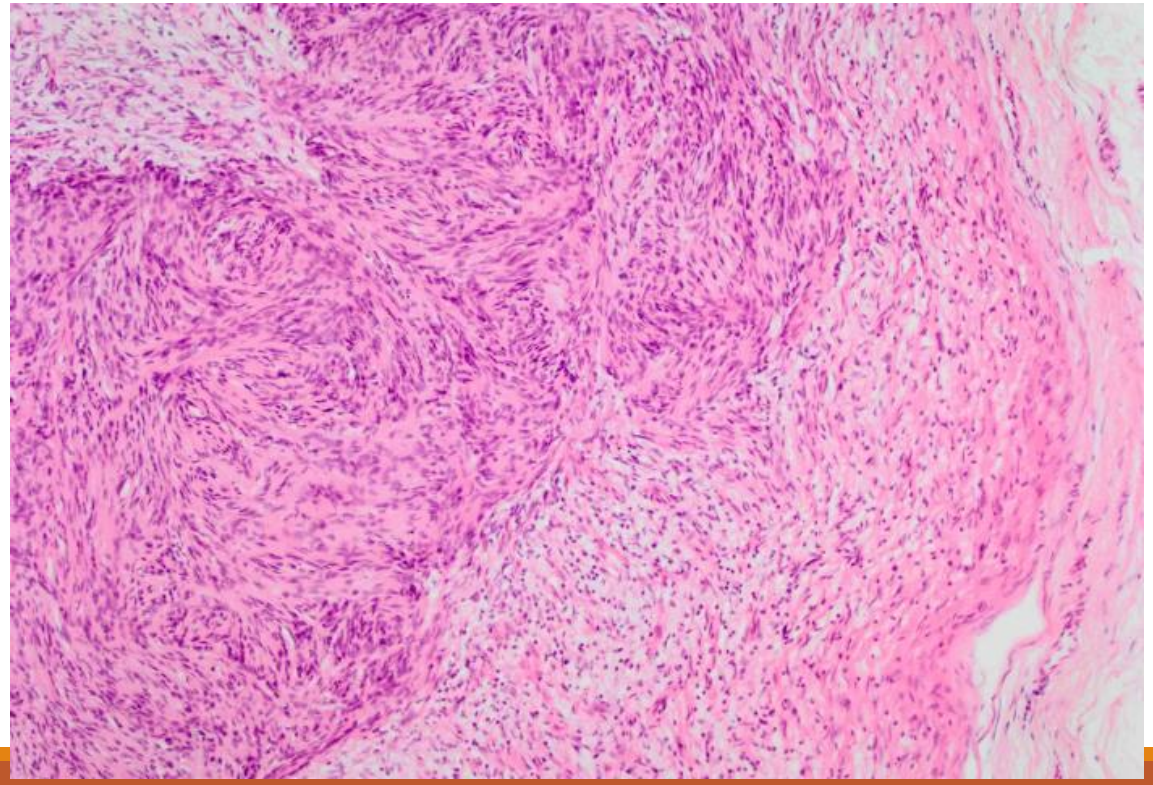
Gross description

Usually solitary and completely encapsulated.



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

Clinical presentation

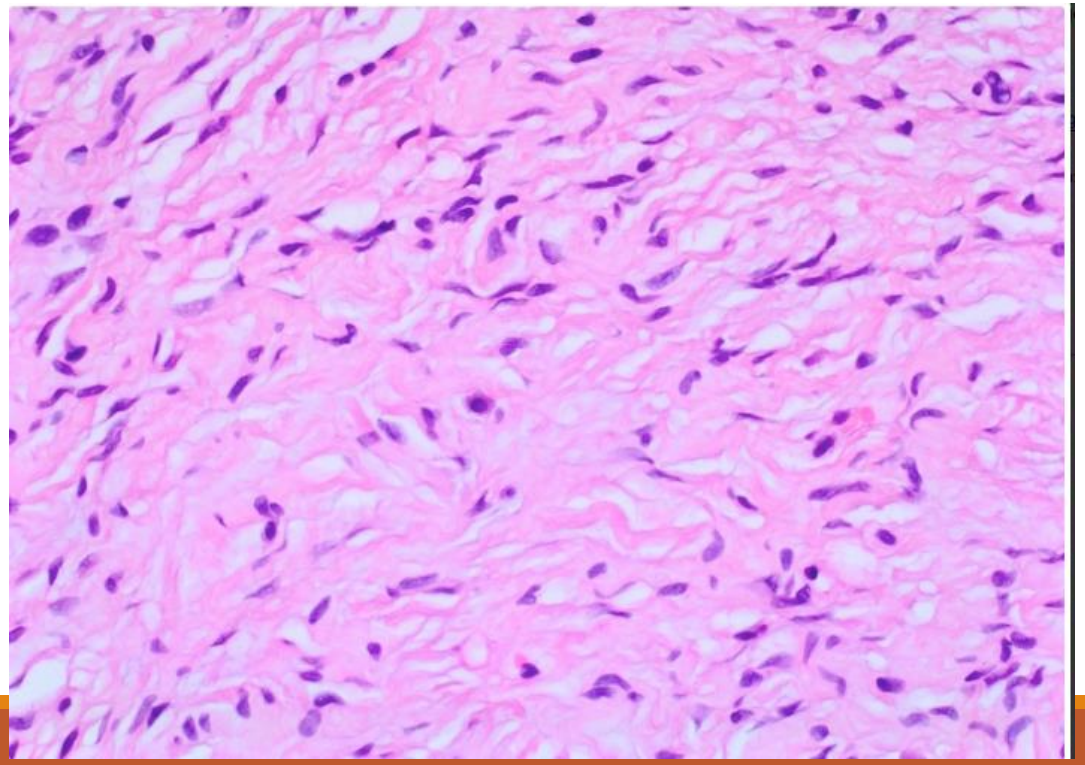
Localized neurofibromas are superficial and evenly disturbed over the body surface.

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



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

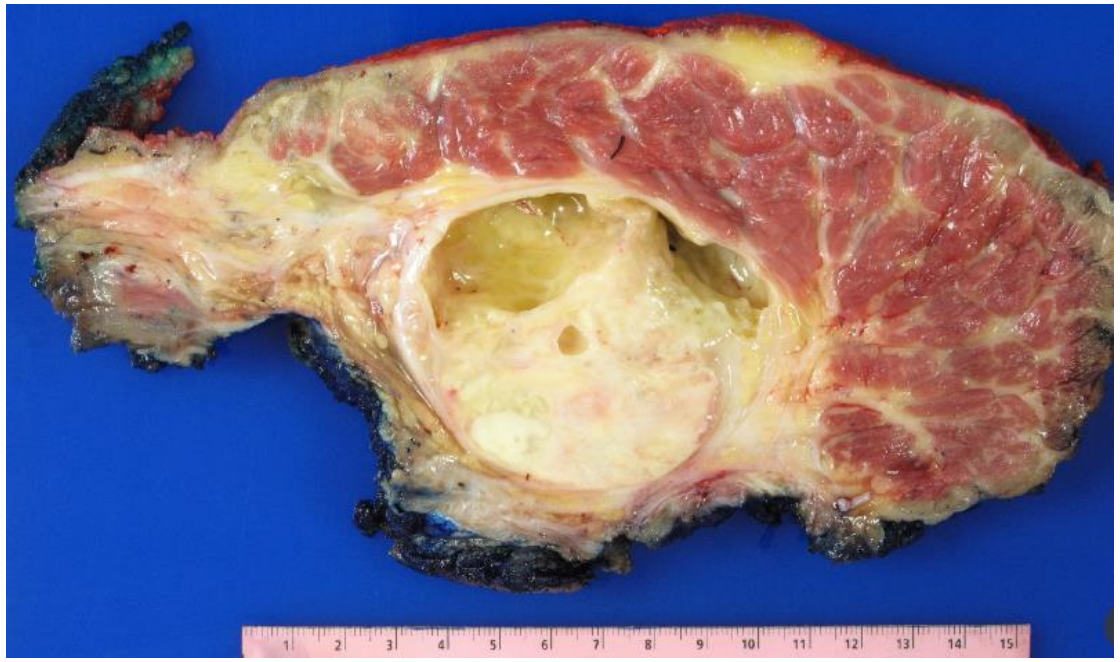
Treatment

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

- Therapeutic options for metastatic MPNST are limited; conventional chemotherapy is usually limited to patients with metastatic disease

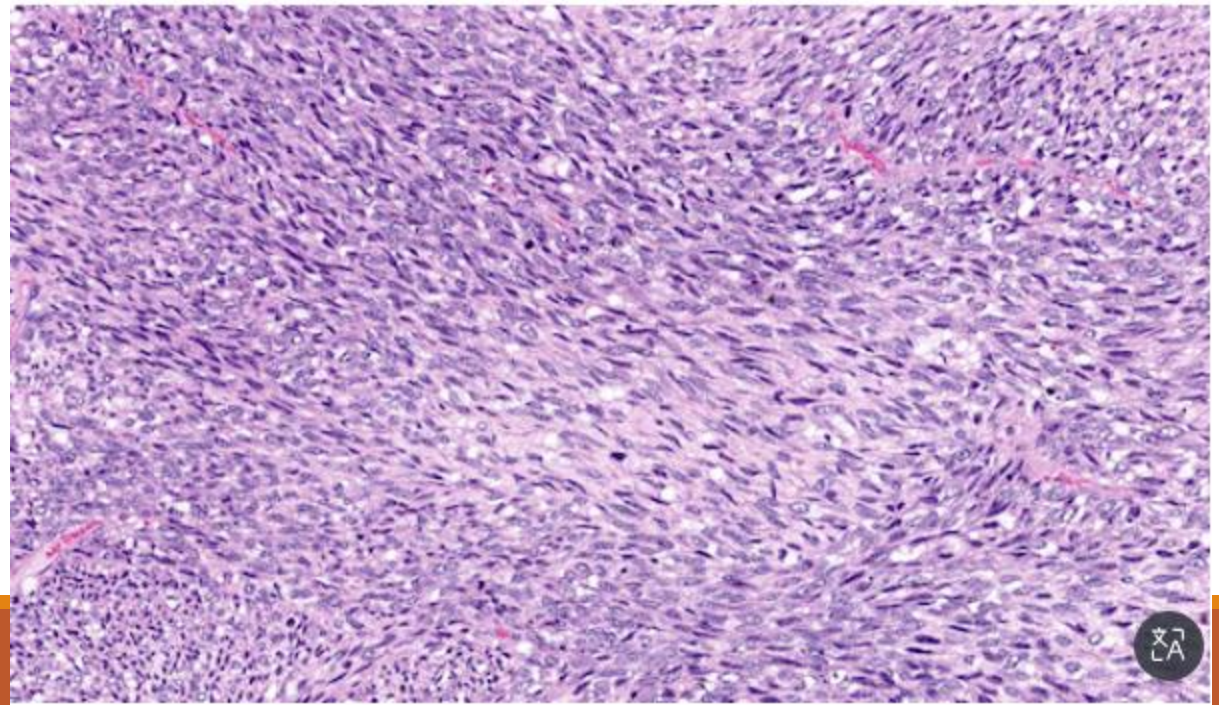
Gross features

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.



Neurofibromatosis

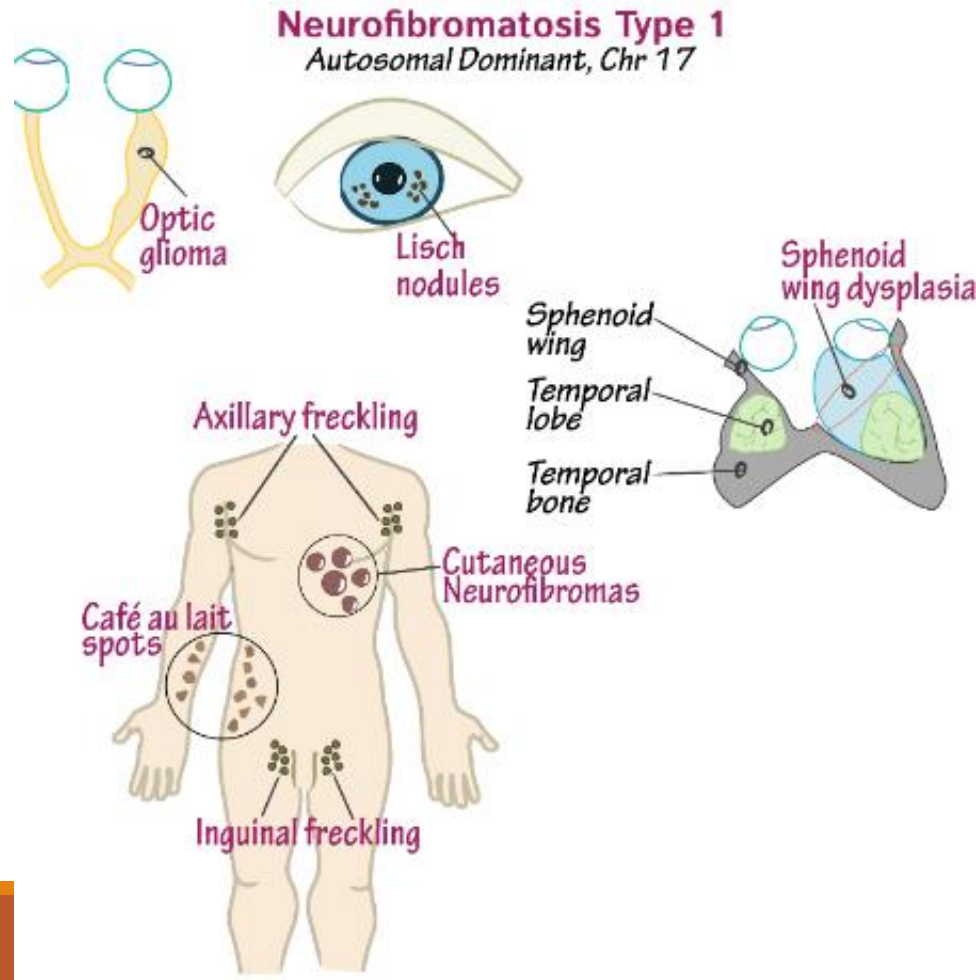
- Neurofibromatosis (NF) refers to a group of genetic conditions in which tumors grow in the nervous system.

- 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

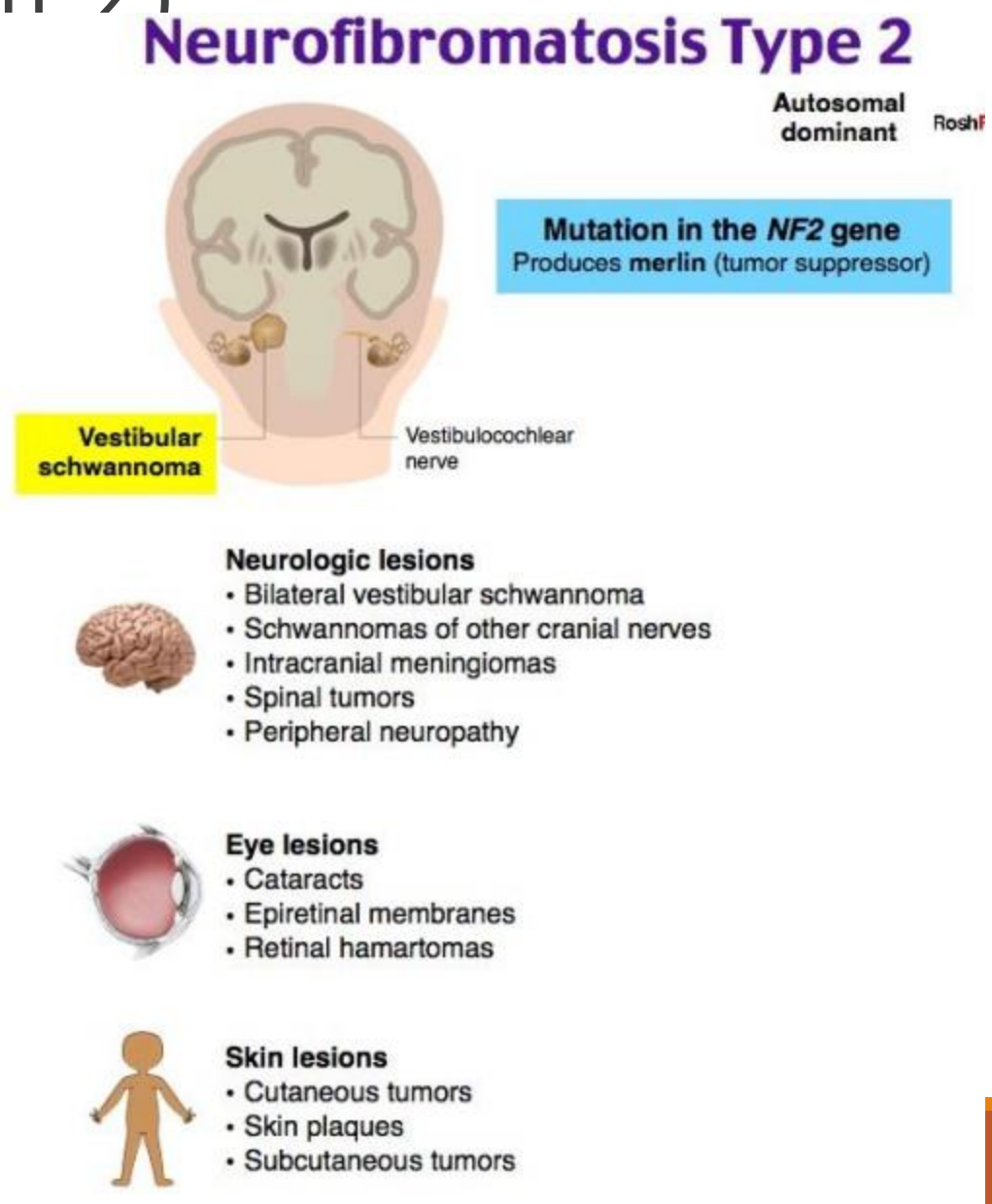


Neurofibromatosis type 2 (NF2)

Is a rare genetic disorder causing

noncancerous tumors on:

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



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