

## Pathology

- ▶ **Schwann cells-** segmental demyelination
- ▶ **Axons** - axonal degeneration either can lead to axonal regeneration or reinervation of the muscle.
- ▶ The process affects some schwann cells sparing others. The disintegrating myelin is engulfed by initially schwann and then by macrophages. The cells in endoneurium replace schwann cells. The new internodes are shorter.

## Segmental demyelination

- ▶ Dysfunction of schwann cells :- syndrome.
- ▶ Damage to myelin sheath:- neuropathy.
- ▶ There is no primary abnormality of axon.

-as in guillain-barre

-e.g. hereditary motor and sensory

- ▶ With sequential episodes of de and remyelination(onion bulbs)

### **Axonal degeneration and muscle fibre atrophy**

- ▶ primary destruction of the axon with secondary loss of its myelin sheath
- ▶ Damage to axon can be local (trauma or ischemia) or more generalised affecting cell body or its axon (neuronopathy or axonopathy).

When axonal injury:-

- ▶ e.g. traumatic transaction of a nerve, the distal portion of fiber undergoes **wallerian degeneration**
- ▶ \_the axon begins to breakdown and the effected schwann cells begin to catabolyze myelin and engulf axon fragments forming small oval compartments (**myelin ovoids**).
- ▶ Macrophages phagocytose axonal and myelin debris.
- ▶ The stump of proximal portion of the nerve shows degenerative changes in distal two or three internodes and then undergoes regenerative activity.

- ▶ When axonal degeneration occurs the muscle fibers within the affected motor unit lose their neuronal input and undergo denervation atrophy.
- ▶ In cross section atrophic fibers are smaller and have a triangular shape.
- ▶ Some muscles cells show disorganized filaments in the center of fiber (target fiber).

# Diseases of peripheral nerves

- ▶ Inflammatory
- ▶ Traumatic
- ▶ Metabolic
- ▶ Toxic
- ▶ Genetic
- ▶ Neoplastic

# Inflammatory neuropathies

They can be either

- 1) Immune mediated or;
- 2) Infectious

## Immune mediated

- Guillain-barre syndrome(acute inflammatory demyelinating polyradiculopathy)
- Life threatening disease
- Weakness in distal limbs(ascending paralysis )
- Histologically --- -inflammation of peripheral nerves
- Demyelination of spinal nerve roots

## Pathogenesis

- ▶ 2/3 of cases are preceded by acute influenza like illness from which patients recover and then symptoms appear.
- ▶ Infections associated with G B;
  - Compylobacter jejuni.
  - Cytomegalo virus.
  - Ebsteinebar virus.
  - Mycoplasma pneumoniae.
- ▶ No consistent demonstration of infectious agent so immunological reaction is favoured as the underlying cause.
- ▶ Experimental proof of its being immunological reaction are there.