

Various Neuropathies

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VASCULITIC NEUROPATHIES.....	1
CRITICAL ILLNESS POLYNEUROPATHY	1
TOXIC NEUROPATHIES	1

VASCULITIC NEUROPATHIES

Peripheral nerves have *low metabolic demands* + *extensive collateral circulation*:

- invulnerable to occlusion of *large peripheral arteries*;
- susceptible to *small blood vessel* diseases (focal circulation interruption in vasa nervorum - individual nerve fascicles) - many types of systemic vasculitis affect peripheral nerves!

ETIOLOGY

- 1) polyarteritis nodosa (nerves are most frequently damaged organs!)
- 2) RA, SLE, Sjögren syndrome, systemic sclerosis
- 3) vasculitides associated with infections (hepatitis B, Lyme disease, HIV).
- 4) Churg-Strauss syndrome
- 5) Wegener granulomatosis
- 6) VASCULITIS RESTRICTED TO PNS - special diagnostic challenge, because footprints of systemic inflammatory disease (e.g. ESR↑) are often absent.

CLINICAL FEATURES

- reflect patchiness of underlying disease; characteristically – **MONONEUROPATHY MULTIPLEX**:
- **asymmetry & length-independence**.
- evolves in stepwise fashion (e.g. wristdrop → contralateral footdrop → patchy areas of subjective numbness or sensory loss elsewhere on extremities).
- cranial nerve involvement, respiratory complications, and sphincter dysfunction are uncommon.

DIAGNOSIS

In absence of diabetes mellitus, vasculitis becomes prime diagnostic consideration!

- screening to detect systemic vasculitis.
- vasculitis is histologic diagnosis - if no other organ involvement is identified → combined nerve and muscle **biopsy** (*axon loss*).
- **CSF** typically is normal (except with SLE).

TREATMENT

- treatment of underlying vasculitis.

- VASCULITIS RESTRICTED TO PNS - **corticosteroids**, but most patients require **cytotoxic therapy** (as in polyarteritis).

CRITICAL ILLNESS POLYNEUROPATHY

- occurs in critically ill patients (sepsis, multiple organ failure, etc).
- pathophysiology **unknown** (dietary deficiency is not considered candidate).
- severe **SENSORIMOTOR NEUROPATHY** (*axon loss*).
- patients experience difficulty being weaned from ventilators.
- complete recovery may occur if underlying cause of multiple organ failure is successfully treated.

TOXIC NEUROPATHIES

- persons with *pre-existing nerve disease* are unusually susceptible to neurotoxins!
- most, although not all, neurotoxins produce **distal axonal degeneration** – distal sensory loss, loss of ankle tendon reflexes, distal weakness.
- **sensory** component suffers most;
toxins that produce predominantly **motor** neuropathy:
 - 1) lead
 - 2) *n*-hexane (glue sniffer's neuropathy)
 - 3) tri-ortho-cresyl phosphate ("ginger jake") - adulterant in illegal liquor (moonshine)
 - 4) dapsone (leprosy treatment)
- with continued exposure, symptoms may progress **proximally**.
COASTING - continuing progression even after offending agent is withdrawn.
- key to **treatment** - prompt recognition and withdrawal.
specific therapy for metal poisoning - **D-PENICILLAMINE**.

BIBLIOGRAPHY for ch. "Peripheral Neuropathies" → follow this [LINK >>](#)