Various Neuropathies

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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 >>