NERVE BIOPSY D32 (1)

Nerve Biopsy

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Nerve biopsy has high chance of being NONINFORMATIVE:

- 1. Limited repertoire of pathological findings
- 2. Limited amount of nervous tissue available for examination

Nerve biopsies are generally useful only in:

- 1) differentiation *segmental demyelination* vs. *axonal degeneration* (when clinical, laboratory, and electrophysiological examinations are nondiagnostic).
- 2) inflammatory neuropathies
- 3) *vascular* conditions (affecting blood supply of nerve) vasculitis, cholesterol emboli, malignant angioendotheliomatosis (intravascular lymphoma).
- 4) amyloidosis
- 5) some *neoplasms*
- 6) some *genetic* disorders (e.g. metachromatic leukodystrophy, Krabbe's disease, adrenoleukodystrophy, giant axonal neuropathy, infantile neuroaxonal dystrophy, neuronal ceroid lipofuscinosis, Lafora disease) both CNS and PNS are affected.
- N.B. generally biopsy is performed in *mononeuropathy multiplex* (vs. *distal symmetric polyneuropathy* biopsy is often uninformative), *palpably enlarged nerves*.

TECHNIQUES

- A) **FULL-THICKNESS biopsy** complete transection of nerve to remove segment
 - technically easier to perform.
 - preferable when pathological evaluation should include both *nerve fibers* and surrounding *connective tissue* and *vascular structures*.
- B) <u>FASCICULAR biopsy</u> longitudinal dissection of nerve to remove segments of only one or several fascicles (sparing at least portion of nerve) favored when larger nerves are biopsied.

Sample amount is varied (2-3 cm segment of full-thickness nerve or fascicles is adequate).

N.B. deficits arising from nerve transection will not necessarily be increased by removing extra centimeter or two that transforms nondiagnostic biopsy into useful diagnostic tool.

TESTING METHODS

Technique	Fixation	Use
Routine light microscopy	Formalin, paraffin	Survey (vasculitis, amyloidosis)
	Glutaraldehyde, paraffin, resin	Survey
Frozen specimen light microscopy	None	Special stains (immunohistochemistry metachromasia)
Teased nerve examination (dissection of single nerve fibers from fascicles)*	Glutaraldehyde, osmium	Myelin internodes, thickening of myelin sheaths (tomacula)
Electron microscopy (should be performed on most nerve biopsies!)	Glutaraldehyde, osmium, resin	Fine structure (most important morphologic alterations!)

^{*} time-consuming procedure not done routinely

- cross sections for *morphometric* studies (i.e. scoring of abnormalities).
 longitudinal sections for *focal* processes (irregularly distributed may
- **longitudinal sections** for *focal* processes (irregularly distributed may be missed in cross section).

PATHOLOGICAL FINDINGS

<u>AXONAL neuropathy</u> - marked depletion of fibers, interstitial fibrosis, ± myelin debris or regeneration of axons.
most likely caused by *toxic* or *metabolic* disorder.

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<u>Segmental DEMYELINATION & REMYELINATION</u> - thinly myelinated fibers and onion bulbs.

• most often in *immunologically* mediated or *hereditary* neuropathy.

- most often in *immunologically* mediated of *nerealitary* neuropatry.
 may be proved by **electron microscopy** or analysis of **teased** myelinated nerve fibers.

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