

10 μ m
가
가 150
가
()
가 가

barrier) 가 .^{1,4}
(macrophage), (mast cell) (microvas-
culature) 가
6~14
0.6~1.2mm² .^{1,4}
2.

(teased nerve fiber preparation) , semithin section
가
가 가
가 .^{1,3}

(micro-vascular networks)
가 가 가

1.
가 (epineurium)
(fascicle)
(per-
ineurium) (endo-
neurium)
가 (Fig. 1).^{1,4}
가 (areolar connective
tissue) 30 75%
가 가

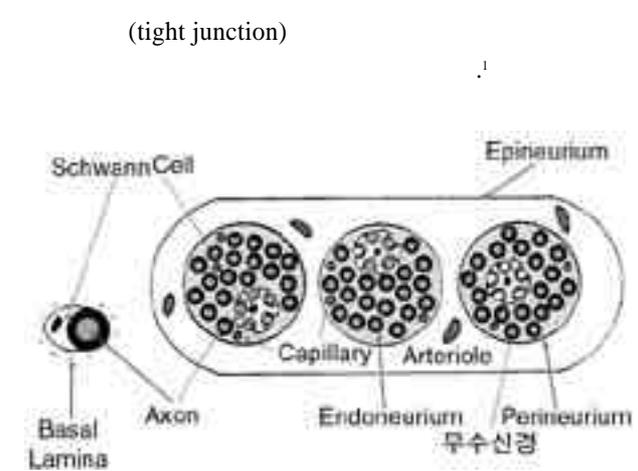


Fig. 1. Diagram of peripheral nerve cross section

(collagen bundle),
(elastic fibers), (fibroblast), (vasa
nervorum) (fat tissue)
(dura mater) .^{1,4}
가
15
junction)
(tight
(perineurial diameter)

3.
(node of Ranvier)
(Fig. 2). (internodal
length) 0.2mm 1.8mm
가 가
가 (intern-
ode) (Schwann cell)가
가
g ratio(axon diameter/total fiber
diameter) g ratio

0.7

g ratio

3).

가

2 μ m

(Fig. 4).^{1,4}



Fig. 2. Electron microscopic finding of a normal sural nerve with myelinated and unmyelinated axons.

4.

10~60
10000/mm²

1 μ m

가
10 μ m
(bimodal curve)

22 μ m

3~4 μ m

가
3:1

7500~

(Fig.

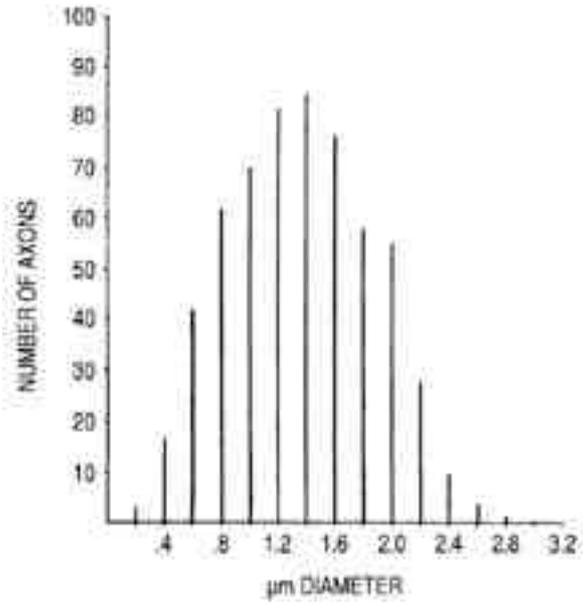


Fig. 4. Frequency distribution of unmyelinated axon diameter in a sural nerve.

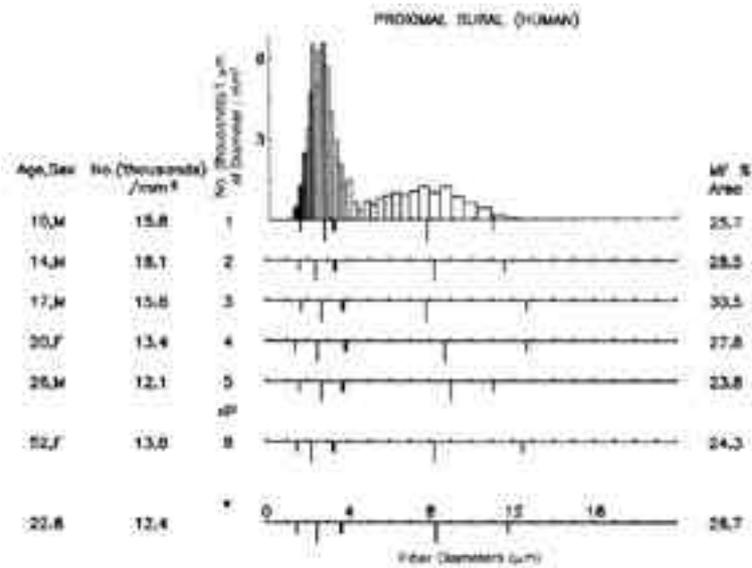


Fig. 3. Histogram and morphometric data of a human proximal sural nerve.

가
glutaraldehyde

1~2
(fascicular biopsy)

20~45
가

3cm

가

가

가

glutaraldehyde fixation 2~4 osmium tetroxide
glycerol 가

1.2

(teased fiber preparation), semithin section

1.2-5

1.

1~2

(metachromatic granules)

가

1.2

hematoxylin-eosin(HE), trichrome, luxol fast blue(LFB), congo red . HE

sil-

conepad 3cm

trichrome LFB 가

epoxy section 가

(Fig. 5). congo red

2% paraformaldehyde buffer

epoxy section

10 2.5% glutaraldehyde in

0.025mol/L cacodylate buffer(PH 7.38)

cacodylate buffer phosphate buffer

10

가 (autolysis)가 4

(microtubule) (depolymerization)

1.2

2. epoxy semithin

glutaraldehyde

0.5cm semithin

1cm

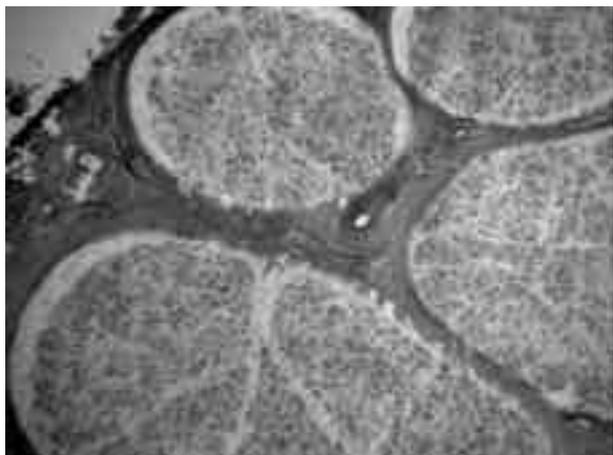


Fig. 5. Frozen section of the sural nerve from a CIDP patient shows markedly decreased density of myelinated nerve fibers(modified Gomori trichrome × 100).

Semithin section (regenerating cluster), (macrophage infiltration) (morphomerty) (Fig. 6).^{1,2}

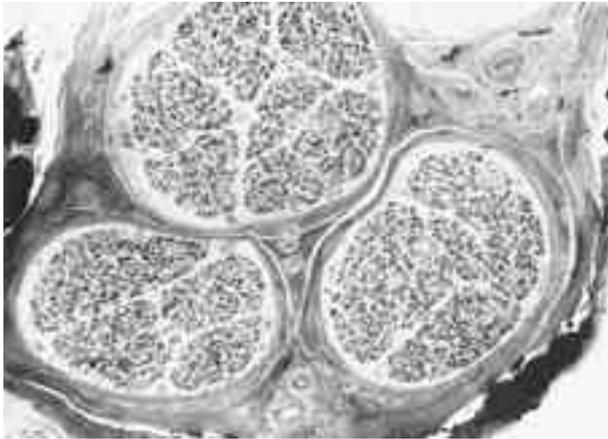


Fig. 6. Cross-section of three fascicles of a normal nerve(plastic embedded section stained with osmium tetroxide and paraphenylenediamine × 25).

3. (Fig. 7) (Fig. 8, 9), (Fig. 10), (Fig. 11) 가

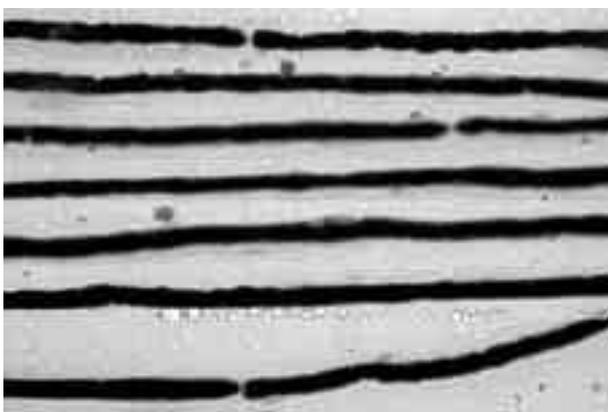


Fig. 7. Consecutive length of a teased nerve fiber without specific abnormality(osmium tetroxide. × 400).

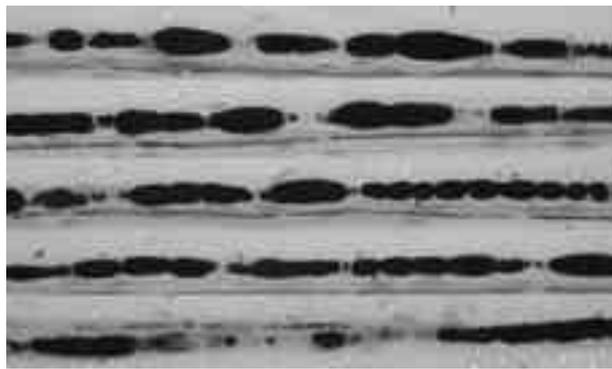


Fig. 8. Consecutive length along one teased nerve fiber from the sural nerve of a patient with the vasculitis shows very early axonal degeneration(osmium tetroxide × 400).

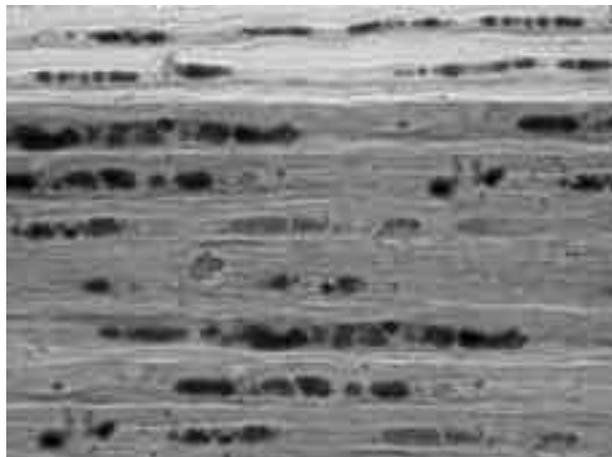


Fig. 9. Consecutive length along one teased nerve fiber from the sural nerve of a patient with the vasculitis demonstrates early axonal degeneration(osmium tetroxide × 400).

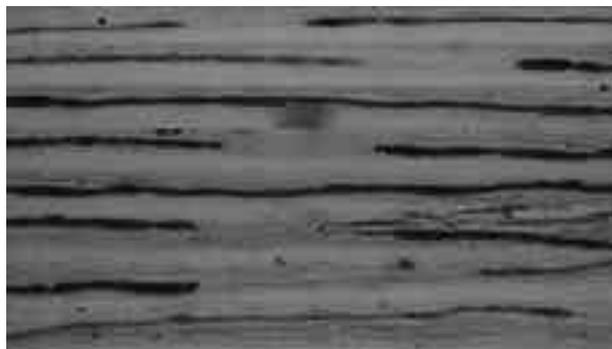


Fig. 10. Consecutive length along one teased nerve fiber from the sural nerve of a patient with CIDP demonstrates segmental demyelination and remyelination(osmium tetroxide × 250).

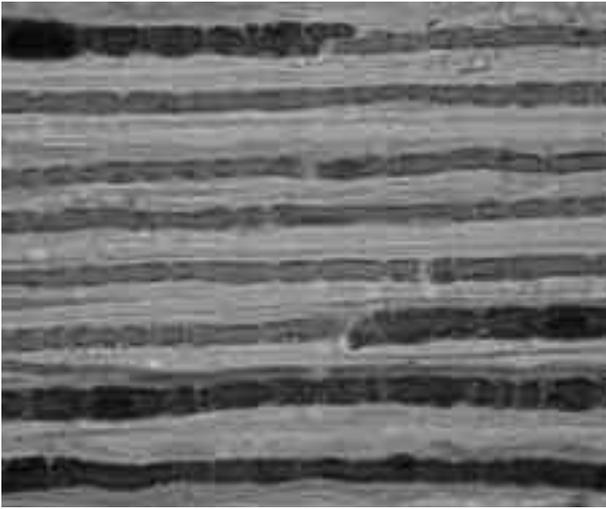


Fig. 11. Consecutive length of a teased nerve fiber from the sural nerve of a patient with CIDP demonstrates remyelination(osmium tetroxide ×400).

4.

Krabbes disease, Fabry disease, Tangier's disease (inclusion body) neuroaxonal dystrophy(giant axon (neurofilament) (Fig. 12).⁶

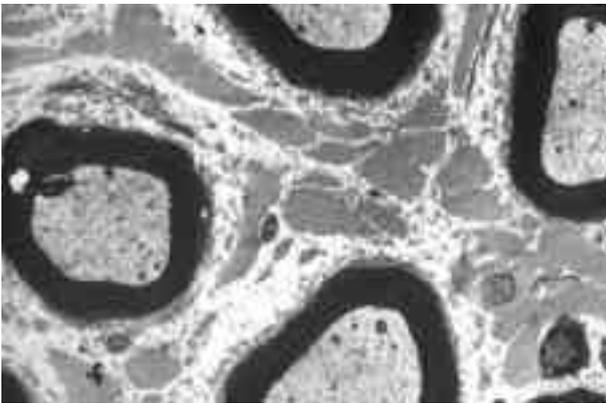


Fig. 12. Electron microscopic finding of the sural nerve with hereditary sensory autonomic neuropathy type IV shows characteristic absence of unmyelinated nerve fibers.

1.

() semithin section

(multifocal fiber loss pattern)가 가

(central fascicular fiber loss).

(perivascular cuffing) fibrinoid necrosis,

, , recanalization, (Fig. 13).⁷⁻¹⁰

hemosiderin laden macrophages가 가

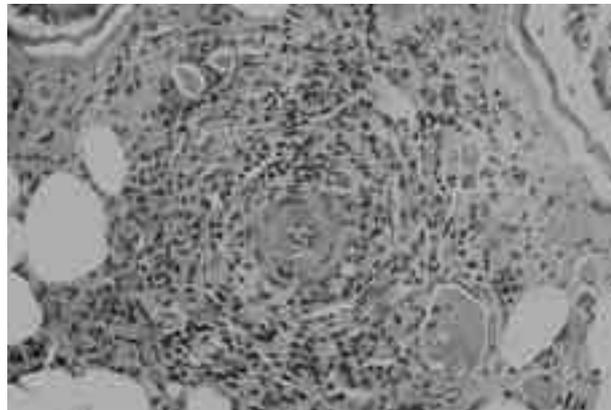


Fig. 13. Paraffin section of the sural nerve from a patient with polyarteritis nodosa shows fibrinoid necrosis and prominent infiltration of inflammatory cells around a small epineurial vessel(H & E × 250).

(multiple mononeuropathy)

(conduction block)

(Fig. 14).

가

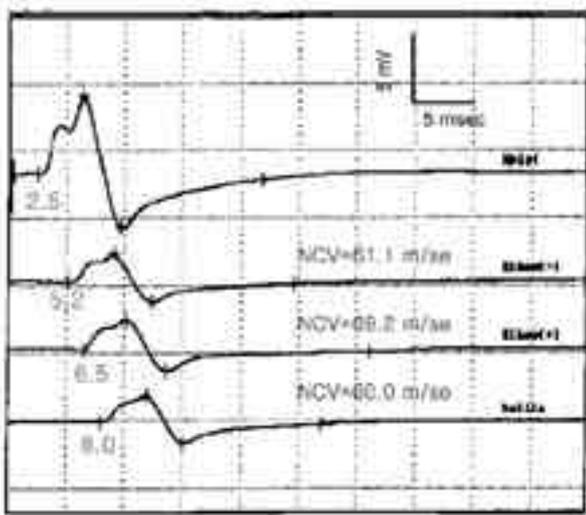


Fig. 14. Ulnar motor nerve conduction study in a patient with Churg-Strauss syndrome (eosinophilic vasculitis) shows definite conduction block but without changes of CMAP duration.

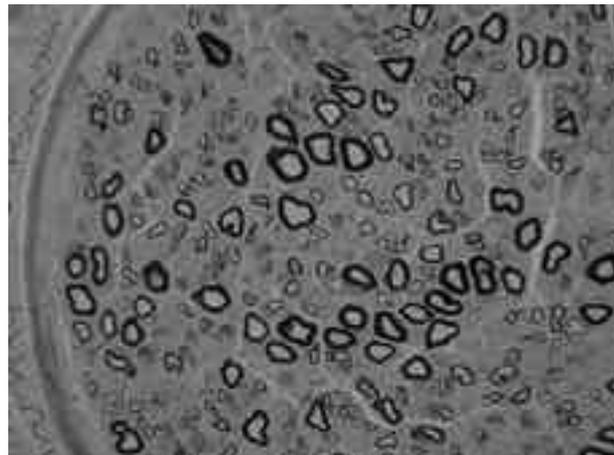


Fig. 15. Semithin section of the sural nerve from a patient with CIDP shows relatively thin myelin sheath comparing to the axon diameter (toluidine blue $\times 400$).

(dispersion)

(Fig. 16).

2. 가 /

(acute inflammatory demyelinating polyneuropathy; AIDP),
 (chronic inflammatory demyelinating polyneuropathy; CIDP),
 (neuropathy associated with plasma cell dyscrasia),
 (HIV neuropathy) Lyme's disease

AIDP CIDP

(Fig. 10, 11) semithin

section

가 (Fig. 15).

T-

..

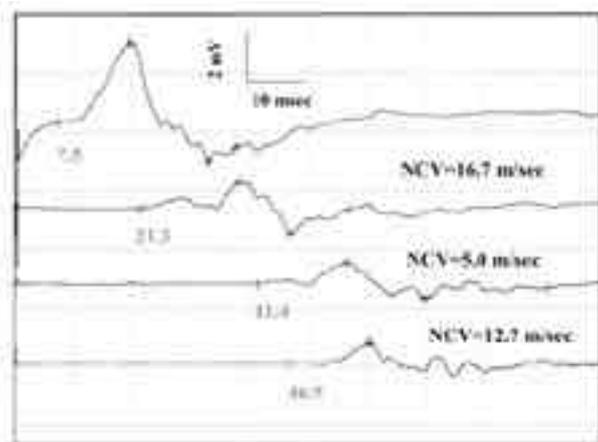


Fig. 16. Ulnar motor nerve conduction study of a patient with AIDP shows markedly slow NCV with conduction block and dispersed CMAPs.

3.

가 . CIDP

onion bulb formation (subperineurial edema)

가

가

가

가

가

(hereditary motor sensory neuropathy; HMSN) type I, type III, Refsum's disease

onion bulb formation
 (Fig. 17, 18).^{1,4} onion bulb formation 가
 Schwann cell process 가

.¹²
 가 가 가
 가 가 가
 ,
 (focal hyper-
 trophic neuropathy)¹³ (neurofibroma)
 onion bulb like structure, microfasciculation

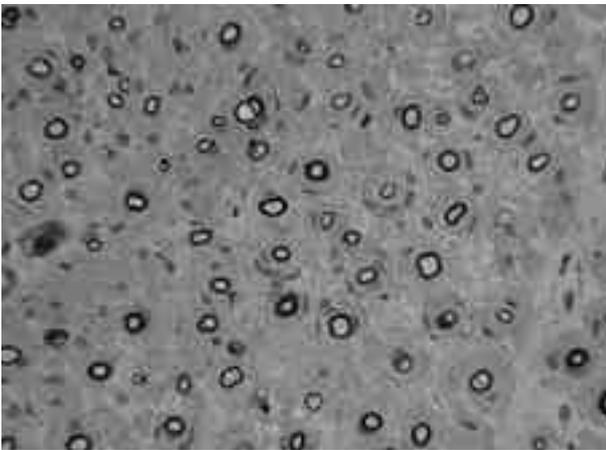


Fig. 17. Semithin section of the sural nerve from a patient with HMSN type 1A shows numerous onion bulb formations (toluidine blue × 400).

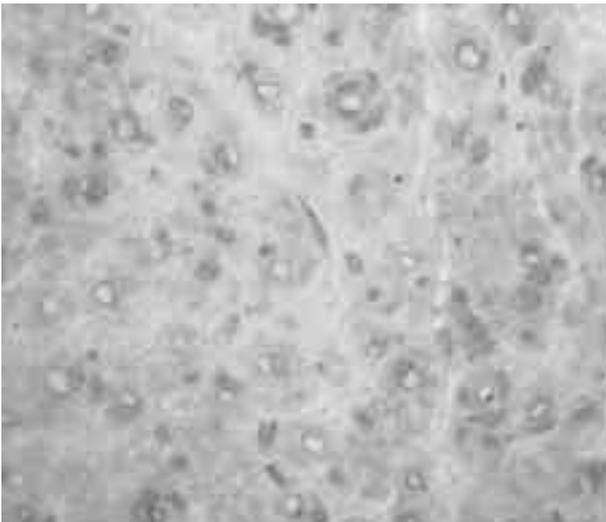


Fig. 18. Semithin section of the sural nerve from a patient with Dejerine-Sottas disease shows numerous onion bulb formations and demyelinated fibers (methylene blue × 400).

onion bulb formation 가 . HMSN
 type I type III semithin 가
 가

가
 가
 Hereditary neuropathy with liability to pressure palsies
 (HNPP)
 가 tomacula가
 (Fig. 19, 20).

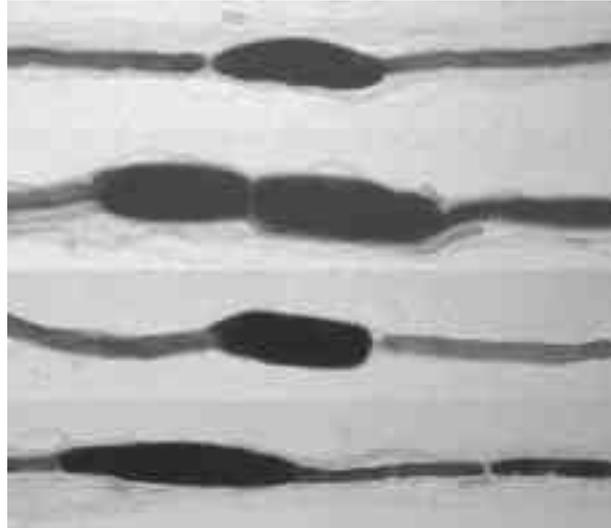


Fig. 19. Teased myelinated nerve fibers of the sural nerve from a patient with HNPP shows typical tomacular changes (toluidine blue × 400).

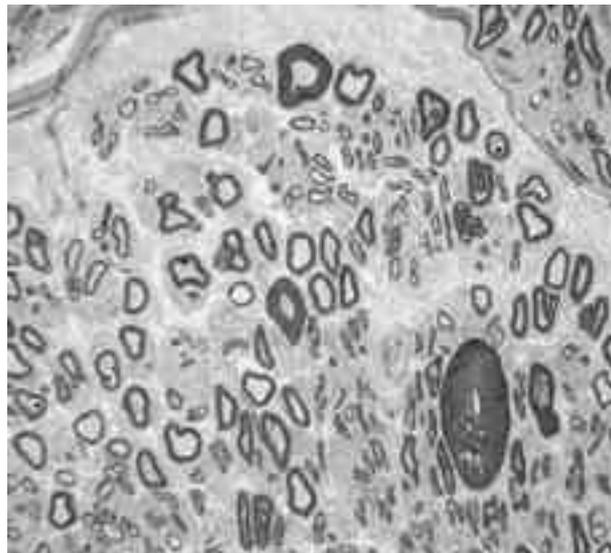


Fig. 20. Semithin section of the sural nerve from a patient with HNPP shows a large nerve fiber with loose myelin sheath (osmium tetroxide × 400).

(deletion) PMP22 (mutation)가 . HNPP
가

(entrapment neuropathy)

(Fig. 21).¹⁴

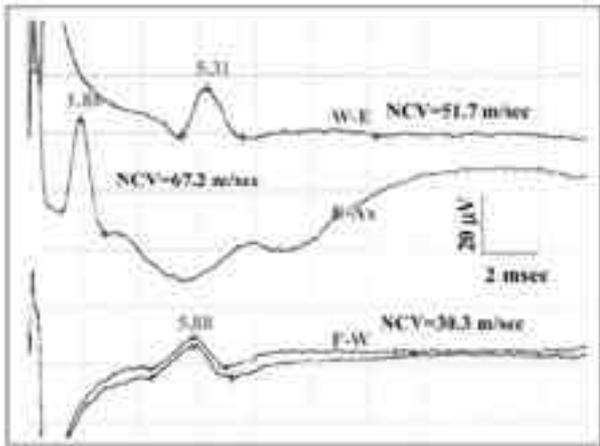


Fig. 21. Median sensory and mixed nerve conduction study of a patient with HNPP shows prolonged duration of CNAPs with slow NCV over transcarpal tunnel.

HMSN type II onion bulb 가
가

4.

HE
methyl violet congo red
(metachromasia)
congo red
polarized microscope apple green bi-refringence
(Fig. 22).

가

amyloid P protein
and chain
, transthyretin

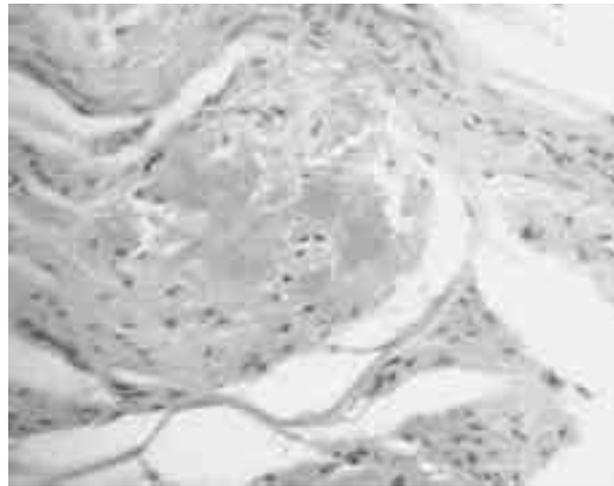


Fig. 22. Congophilic amorphous infiltrations are seen in endoneurium of the sural nerve from a patient with primary amyloidosis (congo red $\times 250$).

가 .^{1,4,15}

16

5.

가 . (sarcoid granuloma) non-caseating granuloma epithelioid cell, lymphocyte, Langerhan's giant cell
granuloma-
tous periangitis가 (Fig. 23).^{1,4,17,18}

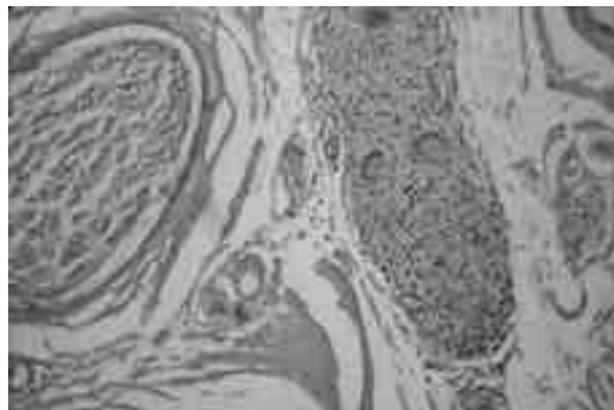


Fig. 23. Noncaseating granulomas with giant cells are noted in epineurium in a patient with sarcoid neuropathy (H & E $\times 250$).

6. (acid fast bacilli; AFB) Mycobacterium leprae (granuloma) (caseation)가 . AFB cocco-bacilli가 formy cell (Fig. 24).^{1,4}

가 가 가 .^{1,4,19,20}
(large myelinated fibers) 가



Fig. 24. Panfascicular infiltration of inflammatory cells and hypertrophied endoneurium are noted in paraffin section of a patient with lepromatous leprosy(H & E ×250).

7. metachromatic leukodystrophy sulfatase galactosyl-3-sulfate가 Cresyl violet (metachromatic granule) .^{1,4}

가 가

가

REFERENCES

1. Dyck PJ, Thomas PK, Griffin JW, Low PA, Podulso JF: Peripheral neuropathy. 3rd ed. Philadelphia: Saunders, 1993
2. : Diagnostic value of nerve biopsy. 1994; 12(sup): 15-24
3. . 1990; 33: 1288-1292
4. Richardson EP, Girolami U: Pathology of the peripheral nerves. Philadelphia: Saunders, 1995
5. Asubury AK, Connolly ES: Sural nerve biopsy: Technical note. J Neurosurg 1973; 38: 391-392
6. , , , , : 4 1 . 1997; 15: 211-215
7. Chalk CH, Dyck PJ, Conn DL: Vasculitic neuropathy. In: Dyck PJ, Thomas PK, Griffin JW, Low PA, Podulso JF, eds. Peripheral neuropathy. 3rd ed. Philadelphia: Saunders, 1993: 1424-1436
8. Cohen RD, Conn DL, Ilsturp DM: Clinical features prognosis, and response to treatment in polyarteritis. Mayo Clin Proc 1980; 55: 146-155

9. Dyck PJ, Conn DJ, Okasaki H: Necrotizing angiopathic neuropathy. Three-dimensional morphology of fiber degeneration related to sites of occluded vessels. *Mayo Clin Proc* 1972; 47: 461-475
10. Fauci AS, Katz P, Haynes BF, Wolff SM: Cyclophosphamide therapy of severe systemic necrotizing vasculitis. *N Eng J Med* 1979; 301: 235-238
11. Dyck PJ, Lais AC, Ohta M, Baston JA, Okasaki H, Grover RV: Chronic inflammatory polyradiculoneuropathy. *Mayo Clin Proc* 1975; 50: 621-637
12. Dyck PJ: Experimental hypertrophic neuropathy. Pathogenesis of onion-bulb formations produced by repeated tourniquet application. *Arch Neurol* 1969; 21: 73-95
13. Mitsumoto H, Wilbourn AJ, Goren H: Perineurioma as the cause of localized hypertrophic neuropathy. *Muscle Nerve* 1980; 3: 403-405
14. , , , : 17p11.2-hereditary neuropathy with liability to pressure palsies . 1999; 1: 13-18
15. Cohens AS: The diagnosis of amyloidosis. In: *Laboratory diagnostic methods in the rheumatic disease*. 2nd ed. Boston: Little & Brown, 1975: 375-412
16. , , , : 1 . 1995; 13: 987-995
17. Nemi R, Galassi G, Cohen M, et al: Symmetric sarcoid polyneuropathy: analysis of a sural nerve biopsy. *Neurology* 1981; 31: 1217-1223
18. Oh SJ: Sarcoid polyneuropathy: a histologic proven case. *Ann Neurol* 1980; 7: 178-181
19. Dyck PJ, Logfren EP: Nerve biopsy. Choice of nerve, method, symptoms and usefulness. *Med Clin North Am* 1968; 52: 885-893
20. Oh SJ: Diagnostic usefulness and limitations of the sural nerve biopsy. *Yonsei Med J* 1990; 31: 1-26