

POEMS

Clinical and Electrophysiological Features of the Patients with POEMS Syndrome

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Backgrounds and objectives: POEMS (polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes) syndrome is the rare cause of polyneuropathy. Although the polyneuropathy is essential for the diagnosis of the disease, the pattern of electrodiagnostic abnormalities has not been characterized in detail. The purpose of this study was to elucidate the features of nerve conduction abnormalities in POEMS syndrome. Methods: We reviewed the medical records and nerve conduction studies (NCS) of 12 consecutive patients with POEMS. Results: A total of 68 motor and 46 sensory nerves were examined. Compound muscle action potentials (CMAPs) and sensory nerve action potentials were abnormally attenuated or not elicited in majority of motor and sensory nerves (80.88% in motor, and 82.6% in sensory nerves). Frequency of the nerves with no potential was significantly higher in lower limbs than in upper limbs ($p < 0.01$ in both motor and sensory nerves), and CMAP amplitude was more reduced in lower limbs than in upper limbs ($p < 0.01$). Conduction slowing was very frequently observed with 95% and 76% of motor and sensory nerves, respectively, having the abnormally reduced values of conduction velocity. Distal motor latencies were abnormally prolonged in 75% of motor nerves, and terminal latency indices were significantly higher in patients than in normal controls ($p < 0.05$). Conduction block was observed only in 5% of motor nerves. Conclusions: NCS in POEMS syndrome showed characteristic patterns, in which conduction abnormalities were more frequently and severely affected in the lower limbs, and more predominantly in the intermediate nerve segments than in the distal portions. The recognition of these characteristic patterns may be helpful in early diagnosis of polyneuropathy in POEMS syndrome.

Key Words: POEMS syndrome (Polyneuropathy, Organomegaly, Endocrinopathy, M-spike and skin changes), Nerve conduction study

POEMS
가
POEMS 1956 Crow
Fukase
POEMS
Polyneuropathy, organomegaly, endocrinopathy,
M-spike skin changes

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POEMS
12 (Age, 52.9 ± 11.1 years,

Female:Male, 8:4) (1998. 5 - 31.0°C
 2002. 12). POEMS Angelã (median nerve), (ulnar nerve),
 (peroneal nerve), (posterior tibial nerve)
 (sensorimotor ; DML) (MNCV)
 polyneuropathy) (mon- (distal stimulation distance)
 oclonal plasmaproliferative disorder) 5 cm 8
 (osteosclerotic bone lesion), cm, 10 cm
 Castleman (motor nerve conduction velocity, MNCV)
), (head of fibula),
 (popliteal fossa)
 (NCS) 가
 (standard techniques of surface stimu-
 lation and recording (Nicolet Viking IV)) (conduction

Table 1. The clinical and laboratory characteristics of POEMS syndrome patients. (N=12)

Characteristics	Frequency (%)
Polyneuropathy	100
Organomegaly	100
Splenomegaly	75
Lymphadenopathy	66.7
Hepatomegaly	50
Castleman disease	16.7
Endocrinopathy	100
Adrenal insufficiency	58.3
Hypothyroidism	58.3
Hypogonadism	41.7
Gynecomastia	12.5 (of men)
Diabetes mellitus	8.3
Hyperparathyroidism	8.3
Hyperthyroidism	8.3
Monoclonal plasma cell dyscrasia	100
M component on serum protein electrophoresis	100
Bone lesions	41.7
Osteosclerotic only	66.7
Mixed sclerotic and lytic	16.7
Lytic only	16.7
Skin changes	100
Hyperpigmentation	91.7
Hypertrichosis	41.7
Thickening	25
Hemangioma/telangiectasia	16.7
Papilledema	8.3
Extravascular volume overload	91.67
Peripheral edema	83.3
Ascites	41.7
Pericardial effusion	41.7
Pleural effusion	25
Other features	66.7
Thrombocytosis: platelet count $450 \times 10^3/\mu\text{L}$	33.3
Polycythemia: hemoglobin 15 g/dL in women and 17 g/dL in men	8.3
Clubbing	8.3

block) CMAP (negative amplitude), Student's t-test
 50% (negative area) Mann-Whitney U test (SPSS
 40% 가 CMAP (duration) 30% 10.0).
 (Sensory nerve conduction velocity, SNCV)
 가 (orthodromically) POEMS
 가 (mid-calf) 가 Table 1 가
 (lateral malleolus) (66.7%),
 (antidromically) (terminal seg- (16.7 %) (8.3%)
 ment) (intermediate segment) (25%) 가 (pares-
 (Terminal latency indices; TLI) thesia) 가 (62.5 %),
 : TLI=terminal distance (mm)/ (25%) 가
 [(DML (ms) × MNCV (m/second))].⁵ TLI NCS 68 46
 24 ⁶ TLI Table 2 CMAP
 80.9 % (55/68)
 가 SNAP 82.6 % (38/46)
 p value 가 0.05
 chi-square test

Table 2. The NCS results in patients

	N	Mean ± SD	Range
DML (% of ULN)			
Median	12	146.97 ± 35.66	95.10-198.85
Ulnar	12	141.41 ± 42.13	94.7-242.42
Posterior tibial	6	126.55 ± 61.43	76.49-238.81
Peroneal	10	101.84 ± 21.6	65.3-128.57
MNCV (% of LLN)			
Median	12	60.63 ± 16.57	35.09-92.35
Ulnar	12	61.07 ± 19.57	29.75-89.25
Posterior tibial	6	78.42 ± 30.98	34.4-111.14
Peroneal	10	67.97 ± 19.56	42.41-99.1
TLI			
Median	12	0.32 ± 0.06	0.23-0.45
Ulnar	12	0.45 ± 0.07	0.34-0.6
Posterior tibial	6	0.59 ± 0.03	0.53-0.63
peroneal	10	0.58 ± 0.83	0.43-0.7
CMAP (mV)			
Median	12	5.84 ± 3.74	0.6-10.8
Ulnar	12	6.68 ± 3.1	0.5-10.2
Posterior tibial	6	5.18 ± 6.54	0.4-14.1
Peroneal	10	1.38 ± 1.97	0.1-5.3
SNCV (% of LLN)			
Median	8	98.82 ± 14.45	85.13-127.7
Ulnar	9	87.57 ± 13.03	73.81-116.13
Sural	8	93.52 ± 9.89	82.19-107.84
SNAP (µV)			
Median	8	9.9 ± 6.31	3.8-24.5
Ulnar	9	10.29 ± 2.81	6.0-14.9
Sural	8	9.69 ± 6.8	4.7-22.0

N, number of examined nerves; DML, distal motor latency; MNCV, motor nerve conduction velocity; TLI, terminal latency index; CMAP, compound muscle action potential; SNCV, sensory nerve conduction velocity; SNAP, sensory nerve action potential

가 41.2 % (28/68),
 45.7 % (21/46) 가
 (63.6%, 28/44) (0/24)
 (p<0.01, chi-square

segments)
 (TLI)
 (p<0.01)(Fig. 2).
 4.2 % (1/24), 6.3% (1/16)

test). 가
 (63.6%, 14/22) (29.2%, 7/24)
 (p<0.01, chi-square test).

가 , CMAP
 (p<0.01)

, SNAP
 (Fig. 1A).

가 ,

(p<0.01)가 (Fig. 1B). DML
 75% (30/40) (p<0.05) ,
 (Fig. 1C). (p<0.05)
 (intermediated

POEMS 가
 가 18 (±14.3
 Mon, Range 4~48 months)

가
 (75%)

(51~84%)

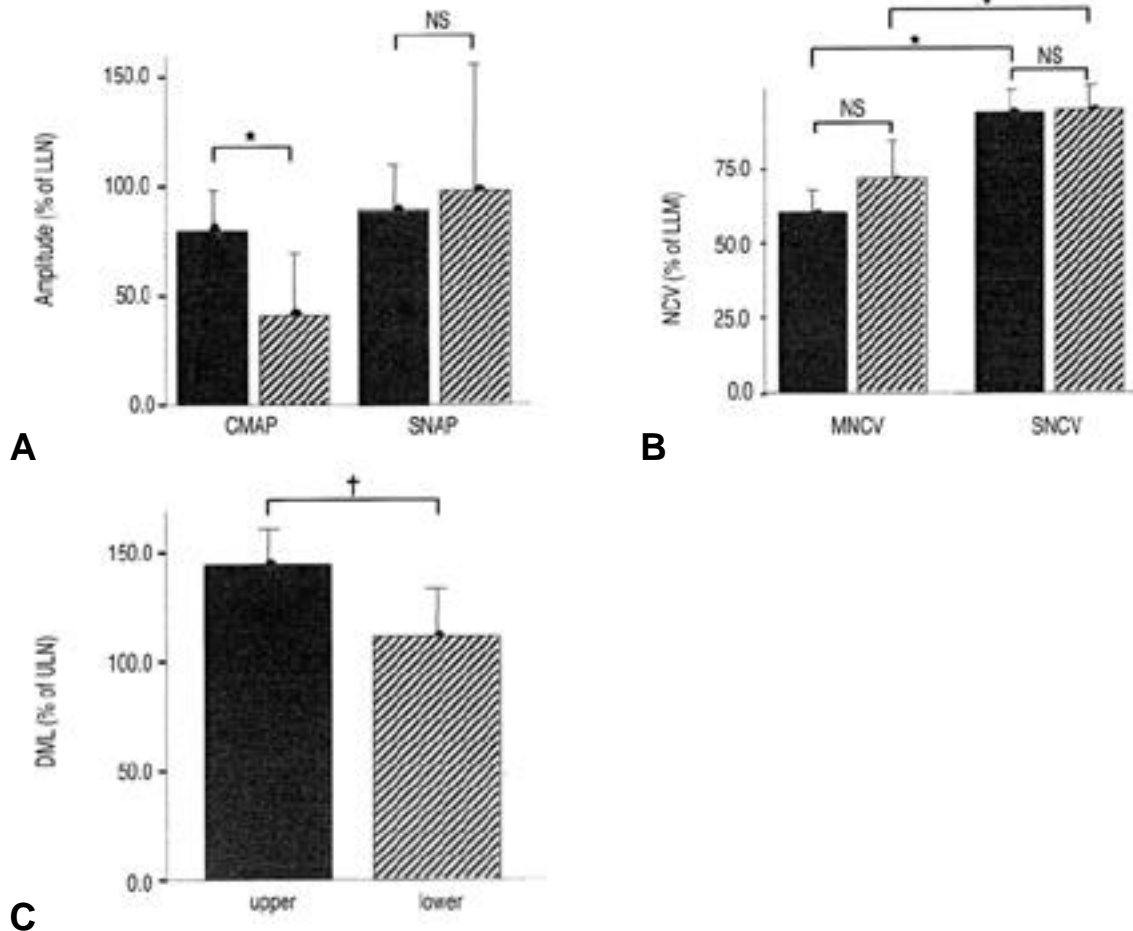


Figure 1. Comparisons of NCS parameters between upper and lower limbs in patients with POEMS syndrome. Compound muscle action potential (CMAP) amplitude is significantly reduced in lower limbs than in upper limbs, whereas sensory nerve action potential (SNAP) amplitude is not different between upper and lower limbs (A). There is no difference in conduction velocity between upper and lower limbs for both motor and sensory nerves, whereas conduction velocity is significantly lower in motor than in sensory nerves (B). Distal motor latency (DML) is more predominantly prolonged in upper limbs than in lower limbs. All parameters are expressed as the percentage of the relevant upper (ULN) or lower (LLN) limit of the normative values(C). Bars show means. Error bars show 95.0% confidence interval (CI) of mean. *: p<0.01, † : p<0.05 using Mann-Whitney U test.

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