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Clinical Study

Electrophysiological features of POEMS syndrome and chronic inflammatory demyelinating polyneuropathy

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ABSTRACT

Polyneuropathy is often an initial manifestation of polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes (POEMS) syndrome and therefore this disorder is frequently misdiagnosed as chronic inflammatory demyelinating polyneuropathy (CIDP). We reviewed electrophysiological data in 20 patients with POEMS syndrome and 36 matched patients with CIDP to compare the electrophysiological features of POEMS syndrome and CIDP. Compared with CIDP controls, POEMS patients demonstrated (1) less prolonged distal motor latency and less reduced motor nerve and sensory nerve conduction velocities, (2) greater reduction of amplitudes of compound motor action potentials (CMAP) in distal stimulation, and similar reduction of amplitudes of CMAP in proximal stimulation, (3) similar reduction of amplitudes of sensory nerve action potentials (SNAP) in median and ulnar nerves, and a greater reduction of amplitudes of SNAP in tibial and peroneal nerves, (4) less temporal dispersion, (5) less frequent conduction block, (6) more frequent neurogenic injury in the muscles of the upper and lower limbs, and more frequent neurogenic injury in the muscles of the lower than upper limbs, (7) similar F wave and H reflex abnormalities, and (8) less frequent skin sympathetic response abnormalities. We concluded that before development of typical clinical manifestations, POEMS neuropathy can be distinguished from CIDP by neural electrophysiological examination. These electrophysiological features can be used for early diagnosis and initiating correct treatment of POEMS syndrome.

1. Introduction

Polyneuropathy, organomegaly, endocrinopathy, monoclonal gammopathy, and skin changes (POEMS) syndrome is a rare cause of neuropathy associated with plasma cell dyscrasia and multiorgan involvement [1–3]. Chronic inflammatory demyelinating polyneuropathy (CIDP) is a chronic and recurrent neuropathy that is immune-mediated. Patients in the early stage of POEMS syndrome are frequently misdiagnosed with CIDP, as both involve the peripheral nerves and may present with albuminocytologic dissociation in the cerebrospinal fluid (CSF) [4]. As patients with POEMS syndrome and CIDP require different treatments, the differential diagnosis is important. Identification of specific patterns of neural electrophysiological abnormalities is an established approach for distinguishing different types of neuropathies. Thus, we examined whether POEMS syndrome could be differentiated from CIDP by electrophysiological examination.

2. Methods

2.1. Patients

We retrospectively reviewed electrophysiological records of 20 patients (16 males, four females; 36–68 years old; mean age 49.2 years) who met the standard diagnostic criteria of POEMS syndrome, and 36 matched patients (30 males, six females; 13–49 years old; mean age 39.6 years) meeting the standard diagnostic criteria of CIDP. This study was conducted at the Department of Neurology at The First Affiliated Hospital of Chongqing Medical University, China, and was approved by the Ethics Committee of our hospital. All patients gave informed consent.

2.2. Diagnostic criteria

Patients who met the standard diagnostic criteria of POEMS syndrome were included [5]. This included having both major criteria (polyneuropathy and monoclonal plasma cell disorder) and one minor criterion. The minor criteria included a sclerotic bone lesion, Castleman's disease, organomegaly (splenomegaly,

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hepatomegaly, or lymphadenopathy), edema (pleural effusion or ascites), endocrinopathy (adrenal, thyroid, pituitary, gonadal, parathyroid, or pancreatic), skin changes (hyperpigmentation, hypertrichosis, plethora, hemangiomata, or white nails), or papilledema. The clinical criteria for diagnosis of CIDP included symmetric motor weakness in the distal and/or proximal limb, presence of the electrodiagnostic features (fractional conduction block in at least one motor nerve; prolonged distal latency in at least two motor nerves; abnormal F wave in at least two motor nerves), and albuminocytologic dissociation in CSF (CSF protein >45 mg/dl and a cell count of <10) [6].

2.3. Electrophysiology

Conduction studies of the median, ulnar, tibial, peroneal, and sural nerves were performed by conventional procedures using a Keypoint electromyography machine (Dantec, Copenhagen, Denmark). Parameters measured included distal motor latency (DML), motor nerve conduction velocity (MCV), compound motor action potential (CMAP), sensory nerve conduction velocity (SCV), and sensory nerve action potential (SNAP). A measurement deviating from the standard range by ±20% was considered abnormal. Conduction block was defined as a $\geqslant$50% decrease of proximal CMAP amplitude compared with distal CMAP amplitude and a $\leqslant$30% increase of proximal CMAP duration compared with distal CMAP duration. The presence of conduction block and temporal dispersion of the median, ulnar, tibial, and peroneal nerves was recorded.

The electrical potentials of the abductor digiti minimi, the deltoid muscle, the anterior tibial muscle, and the quadriceps femoris muscles were also examined using concentric needle electrodes. Spontaneous potentials (fibrillation potential, positive sharp wave) were monitored when the muscles were relaxed. The muscle was regarded as abnormal if >2 spontaneous potentials were detected. Next, 20 motor unit potentials were monitored when the muscles were slightly contracted. The mean duration was considered prolonged if it exceeded the reference value of the age group by $\geqslant$20%. Reference values were obtained from data collected at our department and the laboratory of Peking Union Medical College Hospital (Beijing, China), which were determined from age- and sex-matched healthy adult volunteers using the same protocols. Finally, recruitment order was measured when the muscles were contracted to the maximum extent. A recruitment pattern was considered abnormal if it showed a simple phase.

The presence and conduction velocity of an F wave and the latency and the absence of an H reflex were recorded. The normal

Table 1
Nerve conduction study data for patients with POEMS syndrome and CIDP

Nerve conduction study	POEMS			CIDP			*p* value
	Median	Range	% abnormal	Median	Range	% abnormal	
Median nerve							
DML (wrist–APB) (ms)	4.0	2.8–6.9	49.2	3.9	3.0–7.3	62.1	0.042
CMAP (wrist–APB) (mV)	3.5	0.0–27	50.1	5.1	4.5–26	30.2	0.037
Temporal dispersion			4.1			15.2	0.01
CMAP (elbow–APB) (mV)	3.2	0.0–22	62.7	4.3	3.9–23.2	60.5	0.14
Temporal dispersion			3.9			30.3	0.005
MCV (elbow–wrist) (ms)	35.9	18.1–62.8	56.2	30.3	17.3–65.8	75.1	0.031
Ulnar nerve							
DML (wrist–ADM) (ms)	2.6	2.1–5.7	44.5	2.2	1.9–6.7	60.2	0.022
CMAP (wrist–ADM) (mV)	5.2	0.0–26	76.3	6.0	0.5–27	33.5	0.029
Temporal dispersion			2.3			10.6	0.028
CMAP (elbow–ADM) (mV)	5.0	0.0–25.9	68.8	4.3	0.5–26.7	72.5	0.43
Temporal dispersion			4.2			33.2	< 0.005
MCV (elbow–wrist) (ms)	41.5	15.8–61.2	58.3	34.7	15.3–60.5	70.5	0.027
Tibial nerve							
DML (ankle–AH) (ms)	3.9	3.5–8.9	53.4	4.0	3.3–9.5	75.8	0.01
CMAP (ankle–AH) (mV)	3.5	0.0–16	88.5	4.0	0.0–24	35.3	0.005
Temporal dispersion			2.8			12.4	0.015
CMAP (popliteal–AH) (mV)	3.5	0.0–14.9	90.1	3.8	0.0–24	84.2	0.15
Temporal dispersion			3.5			29.7	< 0.005
MCV (popliteal–ankle) (m/s)	38.7	14.6–50.2	64.3	32.5	15.3–48.1	77.2	0.014
Peroneal nerve							
DML (ankle–EDB) (ms)	3.1	2.5–7.2	55.6	2.8	2.3–7.0	80.3	0.005
CMAP (ankle–EDB) (mV)	4.0	0.0–18	93.3	4.0	0.0–23	42.6	< 0.005
Temporal dispersion			3.7			16.3	0.01
CMAP (capitulum fibulae–EDB) (mV)	3.9	0.0–19	88.3	3.8	0.0–22	79.2	0.18
Temporal dispersion			3.9			31.8	< 0.005
MCV (capitulum fibulae–ankle) (ms)	40.7	13.3–57.2	60.2	38.4	12.1–58.4	75.5	0.01
Median nerve							
SCV (ms)	40.1	20.5–64.2	60.1	42.6	25–67.3	78.9	0.017
SNAP (mV)	6.5	0.0–30.5	71.2	7.5	0.0–32.2	65.6	0.74
Ulnar nerve							
SCV (ms)	45.0	22–57.2	52.1	45.8	20–58.2	70.7	0.043
SNAP (mV)	7.3	0.0–29.0	72.7	7.1	0.0–31.6	71.3	0.57
Sural nerve							
SCV (ms)	40.5	23.5–61.3	65.7	35.4	21.8–60.9	83.7	0.005
SNAP (mV)	2.5	0.0–8.5	90.8	2.6	0.0–8.4	67.9	0.017
Tibial nerve							
SCV (ms)	35.1	23.3–46.7	70.8	30.3	20.5–45.8	74.3	0.05
SNAP (mV)	2.3	0.0–7.0	91.2	2.8	0.0–8.9	70.2	0.009

ADM = adductor minimi, AH = abductor halluces, APB = abductor pollicis brevis, CIDP = chronic inflammatory demyelinating polyneuropathy, CMAP = compound muscle action potential, DML = distal motor latency, EDB = extensor digitorum brevis, MCV = motor conduction velocity, POEMS syndrome = polyneuropathy, organomegaly, endocrinopathy, M protein and skin changes, SCV = sensory conduction velocity, SNAP = sensory nerve action potential.