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Case report

Precise correlation between structural and electrophysiological disturbances in MADSAM neuropathy

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Abstract

Multifocal acquired demyelinating sensory and motor neuropathy is characterised by multifocal clinical deficits. Imaging studies have identified multifocal enlargements of nerve trunks, but a precise correlation between structural abnormalities and electrophysiological dysfunction has not been elucidated. Two patients diagnosed with multifocal acquired demyelinating sensory and motor neuropathy were evaluated with nerve conduction studies, including short segment nerve conduction studies to precisely localise motor conduction block, and ultrasound studies of corresponding nerve trunks. Motor conduction block was identified in each patient (upper limb nerves in two patients), superimposed on additional demyelinating neurophysiological features. Upper limb ultrasound studies demonstrated focal nerve enlargement that precisely correlated with neurophysiological conduction block. The results of this study suggest that factors contributing to focal structural abnormalities in multifocal acquired demyelinating sensory and motor neuropathy are also those that produce conduction block.

Keywords: MADSAM; Chronic inflammatory demyelinating polyradiculoneuropathy; Ultrasound; Conduction block; Pathophysiology

1. Introduction

Patients with multifocal acquired demyelinating sensory and motor (MADSAM) neuropathy present with multifocal motor and sensory deficits, and this may represent an asymmetric variant of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) [1]. Nerve conduction studies (NCS) are characterised by persistent conduction block and other demyelinating features, detection of which is important to distinguish MADSAM neuropathy from related neuropathies with different treatment approaches [1,2].

Nerve ultrasound is emerging as a possible additional diagnostic tool in the work-up of patients with peripheral neuropathy, in particular those patients with suspected inflammatory neuropathy [3]. However, there are few reports of the ultrasound findings in MADSAM neuropathy [4], and the correlation between ultrasound, clinical and electrodiagnostic findings in typical CIDP remains incompletely elucidated. We report two consecutively studied patients with MADSAM neuropathy defined on clinical and neurophysiological grounds [1], where there was precise correlation between nerve ultrasound and clinical and electrophysiological dysfunctions.

2. Patients and methods

Patients were included in this study if they fulfilled clinical and electrodiagnostic criteria for MADSAM neuropathy defined by Saperstein et al. [1]. Detailed clinical, electrodiagnostic and ultrasound evaluations were performed on each patient on the same visit to a specialised neuromuscular clinic. Each patient underwent standardised nerve conduction studies. At a minimum, these included bilateral median and ulnar motor NCS including stimulation at the axilla and Erb's point. Median and ulnar sensory NCS were performed using antidromic stimulation techniques. Short segment NCS (SSNCS) were performed to further localise the motor nerve conduction block [5], with stimulation points 2.5 cm apart. Nerve ultrasound studies were performed using a MyLab Alpha System (Esaote, Genoa, Italy) with a 6–18 MHz linear array probe. The median and ulnar nerves between the wrist and axilla, the brachial plexus and spinal nerves were imaged. Alternative diagnoses were excluded with additional laboratory and imaging studies as necessary. Ultrasound 'inching' studies were performed in nerve segments corresponding to the locations of the SSNCS [6].

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The Sydney University Human Research Ethics Committee approved the study and written consent was obtained from all subjects.

2.1. Case report 1

Patient 1 was a 71-year-old woman who developed right hand sensory disturbance and hand grip weakness 3 years prior to review. One year prior to review she developed left hand numbness and weakness. The patient had received no prior treatment. Examination identified mild proximal muscle weakness, severe weakness of right median-innervated muscles, and mild weakness of left median-innervated muscles. Sensation was reduced in the median-innervated digits and palm.

NCS detected motor nerve conduction block in the forearm segment of the right median nerve (Table 1). Median motor nerve conduction block was localised to the cubital fossa using median nerve short segment nerve conduction studies (SSNCS, 'inching studies'; Fig. 1A). Additional asymmetric demyelinating electrodiagnostic features were identified with slowing of motor nerve conduction in the right ulnar nerve and left median nerve, and prolongation of left median nerve minimum F-wave latencies suggesting proximal left median nerve involvement.

Nerve ultrasound demonstrated fusiform enlargement of a 5 cm segment of the right median nerve in the cubital fossa (maximum cross-sectional area (CSA) 91 mm^2; laboratory normal <8 mm^2), with disruption of the normal fascicular architecture within the lesion (Fig. 1A). The location of the segment of nerve enlargement exactly matched the region of conduction block identified on SSNCS (Fig. 1A). Ultrasound of other upper limb nerve regions identified mild diffuse enlargement of proximal median and ulnar nerves, and left more than right brachial plexus and spinal nerves.

2.2. Case report 2

Patient 2 was a 57-year-old man with an 11-year history of progressive right hand weakness. This initially involved isolated thumb weakness, followed by the development of more diffuse weakness of the intrinsic hand muscles 1 year after onset. 10 years prior to our review, intravenous immunoglobulin was initiated on the suspicion of an immune-mediated neuropathy, resulting in partial clinical improvement and then stability of motor deficits. The patient represented to our institution with deteriorating right hand weakness and sensory impairment. Examination identified weakness of right hand intrinsic muscles, more severe in median- than ulnar-innervated groups. Sensation was reduced in the right median and ulnar nerve distributions. Right upper limb reflexes were absent.

NCS identified conduction block of right median motor nerve conduction in the forearm segment (Table 1). Median nerve SSNCS localised conduction block to the segments 2.5 cm and 5 cm proximal to the wrist crease (Fig. 1B). The right median sensory amplitude was reduced. Additional mild demyelinating neurophysiological features were identified (Table 1) on right ulnar nerve conduction studies with prolonged minimum F-wave latency suggesting proximal involvement. The right ulnar sensory amplitude was reduced.

Table 1
Results of nerve conduction studies in three patients with MADSAM neuropathy.

	Nerve segment	Latency (ms)	Conduction velocity (m/s)	Amplitude
Patient 1	*R) median motor nerve*			
	Wrist	5.6	41	7.3 mV
	Elbow	10.9	46	2.4 mV
	Axilla	13.0		2.3 mV
	Erb's point			1.8 mV
	F-wave	37.3		
	Sensory nerve			NR
	R) ulnar motor nerve			
	Wrist	3.8	40	5.1 mV
	Below elbow	8.6	40	4.6 mV
	Above elbow	11.1	41	4.4 mV
	Axilla	13.5		4.6 mV
	Erb's point			4.5 mV
	F-wave	33.1		
	Sensory nerve			NR
	L) median motor nerve			
	Wrist	6.0	49	3.8 mV
	Elbow	9.9	51	3.6 mV
	Axilla	11.8		3.7 mV
	Erb's point			2.9 mV
	F-wave	31.2		
	Sensory nerve		30	6 μV
	L) ulnar nerve			
	Wrist	3.8	45	4.2 mV
	Elbow	8.0	42	4.0 mV
	Axilla	10.2	46	3.3 mV
	Erb's point			2.6 mV
	F-wave	35.2		
	Sensory nerve		49	7 μV
Patient 2	*R) median nerve*			
	Wrist	4.2	19	7.3 mV
	Elbow	14.1	36	2.1 mV
	Axilla	16.8		3.2 mV
	Erb's point			3.2 mV
	F-wave	39.3		
	Sensory nerve		38	2.6 μV
	R) ulnar nerve			
	Wrist	3.0	48	8.0 mV
	Below elbow	6.5	53	7.9 mV
	Above elbow	8.3	51	7.7 mV
	Axilla	10.3		7.4 mV
	Erb's point			6.5 mV
	F-wave	40.1		
	Sensory nerve		49	3.8 μV
	L) median nerve			
	Wrist	3.4	52	9.2 mV
	Elbow	7.5	52	8.6 mV
	Axilla	9.4		7.3 mV
	Erb's point			6.4 mV
	F-wave	27.9		
	Sensory nerve		53	17.2 μV
	L) ulnar nerve			
	Wrist	2.8	52	8.2 mV
	Below elbow	6.4	46	7.9 mV
	Above elbow	8.5	49	7.6 mV
	Axilla	10.5		7.6 mV
	Erb's point			6.0 mV
	F-wave	32.3		
	Sensory nerve		56	13.0 μV