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Electrophysiological and radiological evidence for the multifocal nature of a case of multifocal acquired demyelinating sensory and motor neuropathy

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Multifocal acquired demyelinating sensory and motor (MADSAM) neuropathy is a variant of chronic acquired demyelinating polyneuropathy. A 65-year-old women presented with upper arm weakness. A nerve conduction study showed conduction blocks over intermediate segments with sparing of distal compound action potentials. Magnetic resonance imaging revealed asymmetric hypertrophy of the brachial plexus on the affected side. These findings represent important electrophysiological and radiological evidence of MADSAM neuropathy. The condition of the patient began to improve after starting intravenous immunoglobulin administration.

Key words: Polyneuropathy, Chronic inflammatory demyelinating; Neural conduction; Magnetic resonance imaging

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The traditional concept of classic chronic inflammatory demyelinating polyneuropathy (CIDP) reflects a chronic relapsing or progressive immune-mediated polyneuropathy manifesting with both distal and proximal involvement of the limbs. Recent clinical and laboratory researches have revealed CIDP variants,[1] which have been categorized mainly into a distally dominant subtype (i.e., distal acquired demyelinating symmetric neuropathy) and a multifocal subtype (i.e., multifocal acquired demyelinating sensory and motor [MADSAM] neuropathy, or multifocal motor neuropathy). The spectrum of CIDP has therefore recently been extended as chronic acquired demyelinating polyneuropathy (CADP).

MADSAM neuropathy is one of the subtypes of CADP. This syndrome is characterized by asymmetric involvement and a relapsing nature with demyelinating neuropathy.[2] However, the clinical diagnosis of MADSAM neuropathy can often be challenging because it

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manifests with various patterns of weakness in the limbs.

One patient with relapsing monoparesis of the arm was diagnosed as MADSAM neuropathy in our clinic. Utilizing both nerve conduction study (NCS) and magnetic resonance imaging (MRI), we objectively demonstrated the multifocal nature of this disease and treated the patient successfully.

CASE

A 65-year-old women was admitted to the neurology department due to monoparesis of the right upper limb with an 1-week history. She had no history of chronic diseases such as diabetes or hypertension. About 10 years previously she had visited another clinic due to weakness in the right upper extremity, which was diagnosed as postherpetic paralysis, and corticosteroids were prescribed but stopped due to side effects. Besides she had experienced several episodes of relapsing upper limb weakness of unknown causes. However, she had not sought an appropriate medical consultation because it was not interfere with her activities.

A neurological examination during the first evaluation showed prominent weakness of muscle strength in the right arm, including the shoulder adduction (Medical Research Council [MRC] grade 3), elbow flexion (MRC grade 2), wrist extension (MRC grade 2), and all five fingers flexion (MRC

Table 1. Findings of the nerve conduction study of the patient

Motor (recording site)	Distal motor latency (ms)	Amplitude (mV)	Conduction velocity (m/s)
Right median (APB)			
Wrist	3.7	8.5	
Elbow		NR	
Left median (APB)			
Wrist	3.1	10	
Elbow		5.8	48
Right ulnar (ADM)			
Wrist	2.9	8.8	
Below elbow		7.8	52
Above elbow		1.6	27
Left ulnar (ADM)			
Wrist	2.3	9	
Below elbow		8.1	49
Above elbow		4.6	50
Right radial (EIP)			
8 cm above wrist	3.3	1.7	
Upper arm		NR	
Left radial (EIP)			
8 cm above wrist	2.1	6.9	
Upper arm		2.7	40
Right peroneal (EDB)			
Ankle	4.3	10.4	
Below FH		9.3	48
Left peroneal (EDB)			
Ankle	3.8	5	
Below FH		4.1	43
Right tibial (AH)			
Ankle	3.6	13.1	
Knee		2.5	45
Left tibial(AH)			
Ankle	4.1	9.4	
Knee		0.8	43

Table 1. Continued

Sensory (recording method)	Amplitud (μV)	Conduction velocity (m/s)
Right median (orthodromic)		
Wrist	NR	NR
Left median (orthodromic)		
Wrist	NR	NR
Right ulnar (orthodromic)		
Wrist	NR	NR
Left ulnar (orthodromic)		
Wrist	NR	NR
Right radial (antidromic)		
Wrist	NR	NR
Left radial (antidromic)		
Wrist	NR	NR
Right sural (antidromic)		
Ankle	9.6	50
Left sural (antidromic)		
Ankle	6	52
Right superficial peroneal (antidromic)		
Ankle	8.6	48
Left superficial peroneal (antidromic)		
Ankle	14.4	40
F-waves	**Latency (ms)**	
Right median	NR	
Left median	NR	
Right ulnar	NR	
Left ulnar	NR	
Right peroneal	47.99	
Left peroneal	NR	
Right tibial	73.39	
Left tibial	NR	
H-reflex	**Latency (ms)**	
Right H-reflex	NR	
Left h-reflex	NR	

NR, not recordable; APB, abductor pollicis brevis; ADM, abductor digiti minimi; AH, abductor hallucis; EDB, extensor digitorum brevis; EIP, Extensor Indicis Proprius; FH, fibular head.

grade 2). Deep tendon reflexes were absent bilaterally in the upper limbs, except for traces of bilateral biceps brachii jerks. No abnormalities were found in a cranial nerve examination, and there were no upper motor neuron signs and atrophy. The patient did not complain of sensory change but pinprick test showed impairment on distal hand.

On day 10 after admission her motor weakness progressed in the right upper limb, including the shoulder abduction (MRC grade 1), shoulder adduction (MRC grade 1), elbow flexion (MRC grade 0), wrist extension (MRC grade 0), and all five fingers flexion (MRC grade 1).

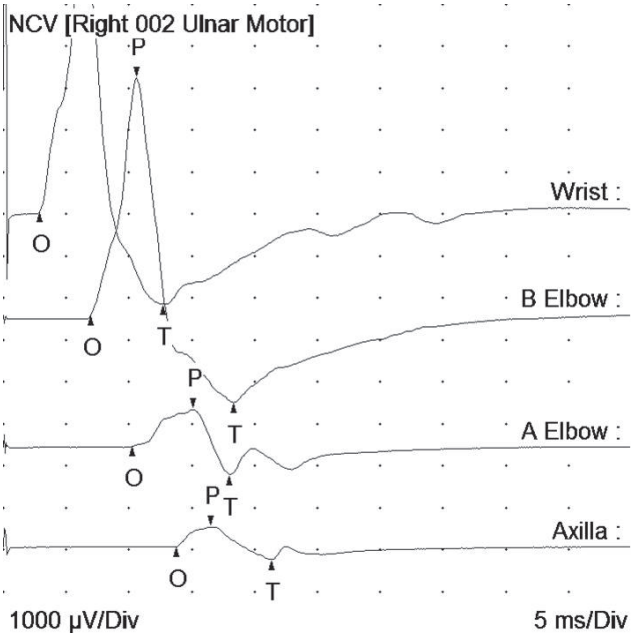

Fig. 1. Findings of the motor nerve conduction study of the right ulnar nerve. The distal motor latency and the compound muscle action potentials were normal at the wrist and below the elbow. Conduction block was observed between above and below the elbow. NCV, nerve conduction velocity.

We performed a cerebrospinal fluid (CSF) study, serologic test for vasculitis, and specific autoantibody assays such as for antiganglioside antibodies and myelin-associated glycoprotein antibodies. No albuminocytological dissociation was found in the CSF analysis (RBC 0/mm^3, WBC 1/mm^3, protein 30 mg/dL, albumin 0.1 mg/dL, glucose 75 mg/dL). All of the applied antibody assays produced negative results.

The detailed NCS findings are presented in Table 1. A particularly notable finding was of prominent complete or partial conduction blocks over the intermediate segments of nerves in the upper limbs (median, ulnar, and radial nerves). These findings were accompanied by relative sparing of both the distal motor latency and motor conduction velocity of the respective nerves (Fig. 1). The other NCS findings for the lower limbs were within the normal ranges. Brachial plexus MRI revealed asymmetric fusiform hypertrophy of the right C5-C8 roots (Fig. 2).

We finally diagnosed her as MADSAM neuropathy, which was treated with a high-dose intravenous methyl prednisolone regimen. However, owing to side effects such as flushing, anxiety, and chest discomfort, we switched her to intravenous immunoglobulin (IVIG) (0.4 g/kg for 5 days). She began to show a remarkable improvement of muscle power (MRC grade 4) by day ┌after starting IVIG. At the last follow-up on 10 months after the symptom onset, her right arm weakness had improved to MRC grade 5-, but NCS and MRI did not show any significant change.

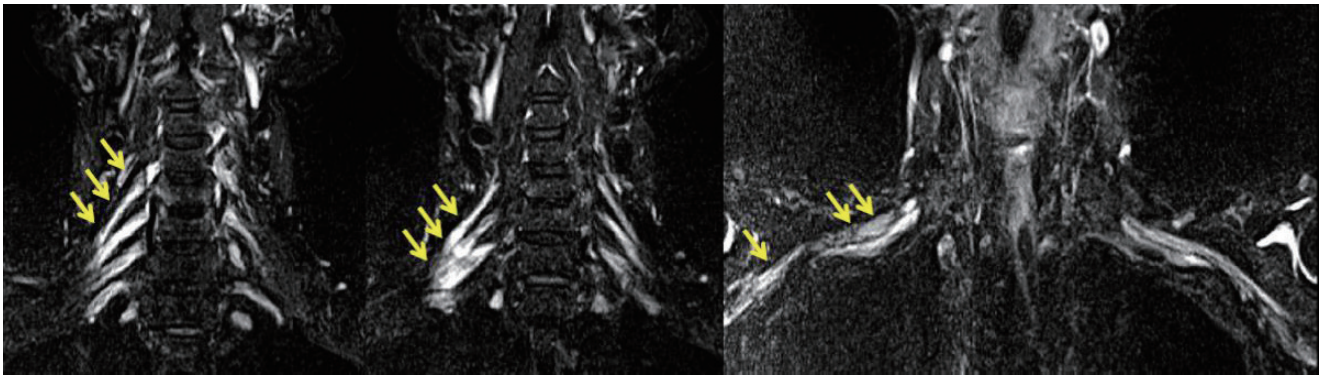

Fig. 2. Brachial plexus magnetic resonance imaging (Short T1 Inversion Recovery Image, non-contrast enhanced) revealed fusiform hypertrophy of the right C5-C8 roots, and plexus was observed (arrows). No hypertrophy was observed on the left side.

DISCUSSION

Classic CIDP is clinically defined as a symmetric polyneuropathy involving both proximal and distal muscles, and affects a relatively uniform cohort of patients. According to the American Academy of Neurology guideline, CIDP is defined by progressive (more than 2 months) or relapsing motor and sensory dysfunction of a peripheral nerve in more than one limb.[3,4]

MADSAM neuropathy shows quite distinctive clinical features compared to typical CIDP. While typical CIDP shows symmetric involvement, MADSAM neuropathy exhibits an asymmetric mononeuritis multiplex, usually affecting an upper limb with a distal topography.[1] Electrophysiological findings suggest that typical CIDP shows a prolonged distal motor latency with conduction block by invading the nerve terminal and spinal root. On the other hand, since MADSAM neuropathy invades the intermediate segment of the nerves, the distal motor latency and the most-distal compound muscle action potential (CMAP) are nearly normal, while the intermediate segments show increased latency and conduction block.[1]

In our case, MADSAM neuropathy showed conduction blocks in the intermediate segment with preserved distal CMAPs. Asymmetric fusiform hypertrophy in the right cervical spinal roots and brachial plexus has also been identified in MADSAM neuropathy.[2] The reported responses to treatments of MADSAM neuropathy have been somewhat inconsistent. MADSAM neuropathy is sometimes unresponsive to primary treatment with steroids or IVIG, but is known to respond well to repeated treatment.[5] Plasma exchange can also reportedly be effective when there is no response to repeated treatment.[6]

Neither a triggering factor nor a specific target for MASAM neuropathy has been well elucidated. Aside from a response to immunological treatment, the evidence for an immunological mechanism-either a cellular or humoral component-is needed for a better understanding of this unique syndrome. These intermediate segment invasion may be helpful in understanding the pathophysiology of MADSAM neuropathy and the diagnostic challenge presented by there being various subtypes of CADP can be overcome by correctly interpreting NCS abnormalities combined with performing adequate imaging investigations.

The main presentation of this patient was relapsing right arm weakness, with the focal nature of this manifestation being related to the delayed diagnosis and treatment. We could diagnosis her as MADSAM neuropathy by applying careful history-taking for relapse, identification of atypical features of demyelinating neuropathy by NCS, and finally the asymmetric involvement of proximal nerves and roots by MRI.

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