Case Report

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Multifocal acquired demyelinating sensory and motor neuropathy misdiagnosed as carpal tunnel syndrome: a case report

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Abstract

Multifocal acquired demyelinating sensory and motor neuropathy (MADSAM), a subtype of chronic inflammatory demyelinating polyneuropathy, is a non-compressive peripheral nerve disorder. Symptoms of MADSAM include asymmetrical weakness and sensory deficits in the distribution of individual peripheral nerves, which are frequently noted in the distal portion of peripheral nerves. MADSAM can be easily misdiagnosed as any of the various compressive peripheral neuropathies. Here, we present a case of MADSAM misdiagnosed as carpal tunnel syndrome (CTS). A 53-year-old woman had bilateral asymmetrical hand weakness (left hand: significant weakness, right hand: slight motor weakness) and a slight weakness of her bilateral lower extremities. Sensory deficit was found on the volar side of her left hand. She had visited many clinics previously and was diagnosed with CTS. However, an electrodiagnostic study performed in our hospital did not identify CTS but indicated a demyelinating peripheral neuropathy in all limbs. On the basis of the patient's clinical symptoms and laboratory findings, she was diagnosed with MADSAM. When patients exhibit progressive aggravating motor weakness and sensory deficits in more than one distal limb without a specific finding of compressive neuropathy in electrodiagnostic studies, clinicians should consider the possibility of MADSAM.

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Introduction

In clinical practice, clinicians frequently encounter patients with motor weakness and sensory deficit caused by peripheral nerve disorders. The most common cause of these neurological deficits is compressive peripheral neuropathy such as carpal tunnel syndrome (CTS), Guyon's canal syndrome, anterior and posterior interosseous syndrome, and tardy ulnar neuropathy. However, in some cases of non-compressive peripheral neuropathy, motor weakness and sensory deficit can develop.[1] As non-compressive peripheral neural disorders are less common than compressive neural disorders, cases of non-compressive peripheral neuropathy are frequently undiagnosed or misdiagnosed.

Multifocal acquired demyelinating sensory and motor neuropathy (MADSAM) is a non-compressive peripheral nerve disorder that is a clinical subtype of chronic inflammatory demyelinating polyneuropathy.[2] It causes demyelination of the distal portion of multiple peripheral nerves and involves the sensory and motor components of the peripheral nerves. Accordingly, its diagnosis could be confused with several peripheral neural disorders caused by compression in distal portions of the limbs.

Here, we present a case of MADSAM misdiagnosed as CTS. Through this case, we present important knowledge on MADSAM and its possible presence in patients with neurological deficits in more than one distal limb without a specific finding of compressive neuropathy on electrodiagnostic studies.

Case presentation

We present a case of a 53-year-old woman with no specific medical history who visited the physical medicine and rehabilitation department of our hospital for bilateral asymmetric hand weakness of her upper extremities (left hand: significant weakness, right hand: slight motor weakness) and a slight weakness of her bilateral lower extremities. Her symptoms started 2 years prior and had slowly progressed. Prior to visiting our hospital, the patient had visited several clinics with her symptoms and had been diagnosed with bilateral or left CTS by clinicians. Our patient was misdiagnosed with CTS because of her clinical symptoms (hand weakness and sensory deficit) and abnormal bilateral median nerve findings on an electrodiagnostic study. The weakness in the lower extremities was probably neglected as it was only minor. This study was approved by the institutional review board of Yeungnam University Hospital, and written informed consent for the case to be published was obtained from the patient. In addition, we obtained informed consent from the patient to treat her disorder. This report followed the CARE guidelines checklist.

The physical examination in our hospital revealed that the degree of the patient's motor weakness was Medical Research Council scale 3/5 in her left wrist and left

finger flexors; however, weakness of the abductor of the fifth left finger was not observed. Weakness of her right hand and lower extremities was subjective; hence, it was not assessed during the physical examination. The patient complained of having difficulty in walking up and down stairs and in walking for long distances. Mild hypoalgesia and hypoesthesia were found on the volar side of the left thumb to the index finger, middle finger, and thumb side of the ring finger. However, no sensory deficit was observed in other areas. Tinel's test and Phalen's test were negative. The Hoffman sign was also negative on both sides. The patient had normal bilateral bicep and triceps reflexes. No abnormality was observed on cervical magnetic resonance imaging. Compound motor action potentials were observed to have a decreased conduction velocity in the left median nerve and a decreased amplitude in the left median and left peroneal nerves during an electrophysiological examination (Table 1). A conduction block was also found on the left median and left tibial nerves during electrophysiological examination (Figure 1). Sensory nerve action potentials in the left median and bilateral superficial peroneal nerves exhibited a low amplitude. While evaluating the presence of CTS, the distal median sensory latencies checked at the wrist level were observed to be 2.4 ms and 2.2 ms on the left and right sides (cut-off value: 3.5 ms), respectively. The wrist-palm median sensory conduction velocities were also observed to be 59 m/s and 63 m/s on the left and right sides (cut-off value: 50 m/s), respectively. These results indicated the absence of CTS. Electromyography-positive sharp waves (1+) were observed in the left abductor pollicis brevis, bilateral tibialis anterior, right peroneus longus, and bilateral gastrocnemius. Anti-GD1b immunoglobulin M (IgM), anti-GM1 immunoglobulin G, and anti-GM1 IgM antibodies were not detected. A cerebrospinal fluid examination revealed no abnormalities. The patient was diagnosed with MADSAM accordingly, based on the established diagnostic criteria.[3–5] A dose of 120 g (2 g/kg) of intravenous immunoglobulin (IVIg) was administered over 5 days. The patient mildly recovered from her motor weakness at 2 weeks after completion of the 5-day IVIg treatment. One month later, a dose of 60 g (1 g/kg) of IVIg was infused over 2 days. Two weeks after the 60 g IVIg infusion, the patient had nearly completely recovered from her motor weakness. A monthly infusion of 60 g of IVIg was subsequently administered to maintain the patient's motor function.

Discussion

In the present study, we describe a patient with motor weakness in the bilateral upper and lower extremities caused by MADSAM. Because the motor weakness in the lower extremities was only subjective and not checked in a physical examination, the case was misdiagnosed as CTS.

MADSAM is a chronic acquired demyelinating neuropathy.[6] Symptoms of MADSAM include asymmetric weakness in the distribution of individual peripheral nerves combined with sensory deficit.[6] MADSAM is caused by autoimmune mediated inflammation and has an onset age in the early 50s.[7] Patients with MADSAM frequently present with weakness in the hand or wrist. However, because MADSAM is relatively rare, it is often misdiagnosed as compressive neuropathy such as CTS, Guyon's canal syndrome, anterior and posterior interosseous syndrome, or tardy ulnar neuropathy. In addition, it is challenging for clinicians to recognize MADSAM; hence, its diagnosis is often delayed for years. Likewise, although our patient had visited several clinics prior to visiting our hospital, she did not receive an accurate diagnosis. Because MADSAM

Table 1. Nerve conduction study results.

Nerve (normal values)	Results	Nerve (normal values)	Results
CMAPs			
Rt. median		**Lt. median**	
MNDL (<4.2), ms	3.0	MNDL (<4.2), ms	**5.6**
CMAP amp D/E (>5.0), mV	10.0/9.0	CMAP amp D/E (>5.0), mV	**0.4/0.1**
Percentage of conduction block E-D	–	Percentage of conduction block E-D	**75**
MNCV (>50), m/s	56	MNCV (>50), m/s	**22**
Rt. ulnar		**Lt. ulnar**	
MNDL (<4.2), ms	2.7	MNDL (<4.2), ms	2.7
CMAP amp D/E (>5.0), mV	14.7/14.0	CMAP amp D/E (>5.0), mV	10.9/10.5
Percentage of conduction block E-D	–	Percentage of conduction block E-D	–
MNCV (>50), m/s	66	MNCV (>50), m/s	63
Rt. radial		**Lt. radial**	
MNDL (<4.2), ms	1.5	MNDL (<4.2), ms	1.6
CMAP amp D/E (>5.0), mV	8.7/8.1	CMAP amp D/E (>5.0), mV	10.0/9.1
Percentage of conduction block E-D	–	Percentage of conduction block E-D	–
MNCV (>50), m/s	65	MNCV (>50), m/s	55
Rt. peroneal		**Lt. peroneal**	
MNDL (<6.0), ms	4.7	MNDL (<6.0), ms	4.7
CMAP amp D/K (>2.0), mV	2.5/2.5	CMAP amp D/K (>2.0), mV	**1.2/1.1**
Percentage of conduction block K-D	–	Percentage of conduction block K-D	–
MNCV (>40), m/s	47	MNCV (>40), m/s	48
Rt. tibial		**Lt. tibial**	
MNDL (<6.4), ms	3.9	MNDL (<6.4), ms	3.8
CMAP amp D/K (>2.6), mV	16.8/10.8	CMAP amp D/K (>2.6), mV	9.0/4.1
Percentage of conduction block K-D	–	Percentage of conduction block K-D	**54.4**
MNCV (>40), m/s	47	MNCV (>40), m/s	45
SNAPs			
Rt. median SNAP amp (>20), μV	40	**Lt. median** SNAP amp (>20), μV	**9**
Rt. ulnar SNAP amp (>20), μV	40	**Lt. ulnar** SNAP amp (>20), μV	28
Rt. supf. radial SNAP amp (>20), μV	28	**Lt. supf. radial** SNAP amp (>20), μV	27
Rt. supf. peroneal SNAP amp (>10), μV	**5**	**Lt. supf. peroneal** SNAP amp (>10), μV	**4**
Rt. sural SNAP amp (>15), μV	23	**Lt. sural** SNAP amp (>15), μV	19

Normal values are presented in parentheses.
Abnormal values are presented in bold.
CMAP, compound motor action potential; amp, amplitude; D, distal; E, elbow; E-D, elbow to distal segment; MNCV, motor nerve conduction velocity; MNDL, motor nerve distal latency; K, knee; K-D, knee to distal segment; SNAP, sensory nerve action potential; supf., superficial; Rt., right; Lt., Left.

usually shows an excellent response to IVIg or corticosteroid treatment, early treatment tends to result in better outcomes.[8]

Regarding the difference in electrodiagnostic features between MADSAM and CTS, in MADSAM, conduction block is frequently observed,[9] while this is only rarely observed in CTS.[10] In addition, patients with MADSAM often present with asymmetrical demyelinating features of multiple peripheral nerves on nerve conduction studies. Additionally, ultrasound is

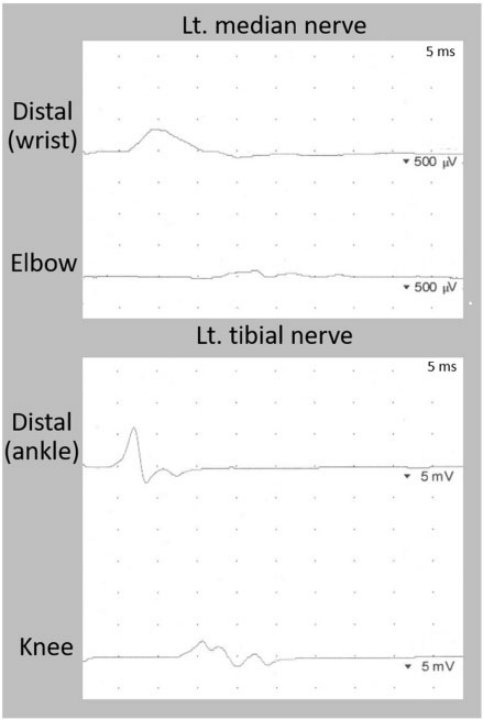

Figure 1. Compound motor action potential testing of the left median and left tibial nerves showed a conduction block.
Lt., left.

useful for differentiating MADSAM from CTS. In MADSAM, multifocal nerve enlargements can be detected at sites at which conduction blocks are present.[11] In contrast, in CTS, the median nerve becomes flattened under the site of maximum nerve compression, and the nerve is swollen proximal to this region.[12]

Many clinicians do not have adequate knowledge of MADSAM, and its diagnosis is often delayed as a result. Thus, it is essential for clinicians to have sufficient knowledge of this disorder to avoid misdiagnosing it as compressive neuropathy. When patients show progressive aggravating motor weakness and sensory deficit in more than one distal limb without a specific finding of compressive neuropathy on electrodiagnostic studies, clinicians should consider the possibility of MADSAM.

Ethics statement

The authors are accountable for all aspects of the work in ensuring that questions related to the accuracy or integrity of any part of the work are appropriately investigated and resolved. This study was approved by the institutional review board of Yeungnam University Hospital, and written informed consent was obtained from the patient for publication of this study and any accompanying images.

Declaration of conflicting interests

All authors have completed the ICMJE uniform disclosure form. The authors have no conflicts of interest to declare.

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