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Motor dominant polyradiculopathy with Primary Sjögren's syndrome mimicking motor neuron disease

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Sjögren's syndrome (SS)-associated polyradiculopathy is rarely reported. A 51-year-old woman presented with a history of gradual weakness in all four extremities for several months. Based on electrophysiological studies, spinal magnetic resonance imaging and cerebrospinal fluid examination, inflammatory polyradiculopathy was confirmed. During a search for the aetiology, the patient was ultimately diagnosed with SS. This study introduces SS-associated polyradiculopathy that primarily presented with motor symptoms, thus mimicking motor neuron disease.

Key words: Polyradiculopathy; Sjögren's syndrome; Motor neuron disease

Sjögren's syndrome (SS) is an autoimmune epithelitis with lymphocytic infiltration of exocrine glands.[1] At least one-third of patients present with systemic extraglandular symptoms.[2] Peripheral nervous system involvement is the most common neurological symptom and may be the first symptom observed before diagnosis of SS. Symmetric sensory polyneuropathy infiltrates are most commonly observed; multiple neuropathy, small fiber neuropathy and cranial neuropathy are also involved.[3] Among these disorders, SS-associated polyradiculopathy is rarely observed. Here, we describe a patient with SS who presented with inflammatory polyradiculopathy mimicking motor neuron disease (MND) as the first symptom, as well as slowly progressive weakness and muscular atrophy.

CASE

A 51-year-old female patient visited the hospital with a main complaint of weakness in both upper limbs for 9 months. The symptoms started with neck stiffness and difficulty

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in lifting her arm over her shoulder. Five months later, the strength in her left arm had decreased, and her legs began to lose strength. She had visited the department of neurology at our hospital due to suspected MND based on extensive denervation changes observed in an electromyography (EMG) examination performed at another hospital. She did not have any past medical history. She complained of stiffness of the neck and a sense of weakness in both shoulders, hands and legs. She showed no fasciculation or atrophy of the tongue. Atrophy was observed in the proximal upper extremities. Regarding the Medical Research Council grade, her bilateral shoulders received grade 3. Her elbows showed an extension of grade 5 and a flexion of grade 3 for the right elbow and grade 4 for the left elbow. In the lower extremities, flexion and extension of the bilateral hips, knees and ankles were grade 4. Her sensory examinations were normal. The deep tendon reflexes of the brachioradialis, biceps brachii, and triceps muscles were hypoactive. Bilateral Hoffman signs were positive. Plantar reflex signs and ankle clonus were negative. Whole spine MRI showed the enhancement of multiple ventral nerve rootlets in the cervical and lumbar spinal cord (Fig. 1). In the nerve conduction test, the compound motor action potential of the right median ulnar, axillary, and musculocutaneous nerves was reduced by approximately 50-70% compared to that of the left side (Table 1). EMG confirmed active denervation and reinnervation changes in the bilateral abductor pollicis brevis; flexor carpi radialis; first dorsal interosseous; biceps brachii; triceps brachii; tibialis anterior; vastus lateralis; and cervical, thoracic and lumbar paraspinal muscles (C6-T1, T6, and L4-5). EMG also revealed giant motor unit potential and delayed recruitment (Table 2). A cerebrospinal fluid (CSF) test showed normal results with red blood cell 17/mm^3, white blood cell 1/mm^3 and glucose 58 mg/dL, whereas CSF protein levels were elevated to 65 mg/dL. No evidence of malignancy was detected in the CSF culture test, and herpes virus, zoster virus and JCV PCR were negative. Antinuclear antibodies were 1:20 speckled, anti-Ro antibodies were positive, and anti-La antibodies and other vasculitis-related test results were negative. Anti-HCV, anti-HIV, anti-GM1, anti-GD1a, anti-GD1b, anti-GT1a and anti-GQ1b were negative. Creatine phosphokinase and VDRL levels were normal. We started steroid pulse therapy (dose: 1 g solumedrol for 5 days) as an empirical treatment based on the possibility of unspecified inflammatory polyradiculopathy, due to spinal MRI findings and elevated CSF protein levels. Three days after starting the steroid, the patient showed improvements in neck stiffness and overall weakness. We confirmed that she had symptoms of dry mouth and dry eyes over the past 2 years. In her salivary gland scan test, the absorption of the contrast agent was decreased in the parotid and submandibular glands. According to SS diagnostic criteria, the patient exhibited 2 symptoms of the eyes and 2 symptoms of the mouth.[4] The non-stimulated salivary flow-rate test showed a rate of 0.31 g/0.3 mL (<1.5 mL), the wafer test result was 8 minutes and 25 seconds (>2 minutes), and the Schirmer test results were 8 mm on the right side and 7 mm on the left side (<5 mm over 5 minutes). Salivary scintigraphy showed delayed uptake, reduced concentration and delayed excretion of the tracer. As she satisfied 4 of the 6 criteria, she was diagnosed with SS.

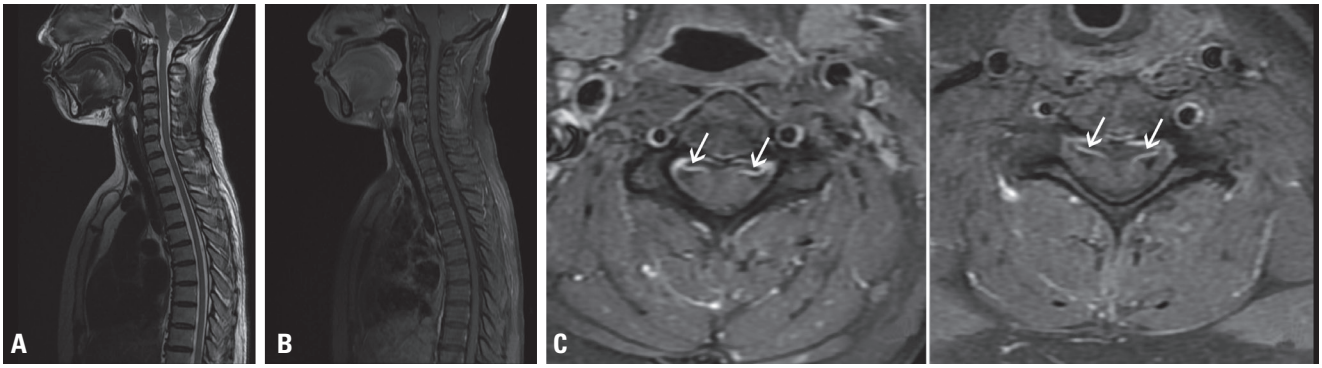

Fig. 1. Cervical and thoracic spinal MRI. A T2 sagittal image (A) and T1 gadolinium-enhanced sagittal image (B) show unremarkable findings. T1 gadolinium-enhanced axial images at the (C) C4 level and (D) C6 level show enhancement of the ventral nerve rootletsof the ventral nerve rootlets (white arrows). MRI, magnetic resonance imaging.

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Table 1. The results of nerve conduction study

Nerve	Stimulation	Latency (msec)	Amp.	Velocity (m/sec)	F-latency (msec)
Motor					
Lt. median	Wrist	3.13 (<3.6)	11.3 (>5.0)		24.7
	Elbow		10.7	58.5 (>50.0)	
	Axilla		10.4	60.1 (>56.0)	
Rt. median	Wrist	3.17 (<3.6)	5.4 (>5.0)		26.6
	Elbow		5.3	53.7 (>50.0)	
	Axilla		5.3	64.5 (>50.0)	
Lt. ulnar	Wrist	2.46 (<2.5)	8.8 (>5.0)		21.4
	Elbow		8.8	62.7 (>50.6)	
	Axilla		8.5	75.2 (>52.7)	
Rt. ulnar	Wrist	2.29 (<2.5)	6.1 (>5.0)		20.8
	Elbow		5.6	54.9 (>50.6)	
	Axilla		5.4	59.9 (>52.7)	
Lt. Axillary	Erb	3.05 (<5.4)	8.9 (>4.6)		
Rt. Axillary	Erb	4.01 (<5.4)	5.4 (>4.6)		
Lt. MC	Erb	4.11 (<5.6)	5.4 (>4.0)		
Rt. MC	Erb	4.23 (<5.6)	4.2 (>4.0)		
Lt. peroneal	Ankle	3.25 (<4.8)	5.1 (>4.0)		42.5
	Popliteal fossa		5.1	46.4 (>41.9)	
Rt. peroneal	Ankle	4.04 (<4.8)	5.3 (>4.0)		45.0
	Popliteal fossa		4.4	45.9 (>41.9)	
Lt. tibial	Ankle	3.21 (<5.1)	12.9 (>5.0)		46.1
	Fibular neck		8.7	45.3 (>40.6)	
Rt. tibial	Ankle	3.96 (<5.1)	10.9 (>5.0)		45.0
	Fibular neck		6.9	46.2 (>40.6)	
Sensory					
Lt. median	Digit 2	2.48	27.2 (>10.0)	44.4 (>41.3)	
Rt. median	Digit 2	2.54	21.3 (>10.0)	43.3 (>41.3)	
Lt. ulnar	Digit 5	1.98	10.4 (>10.0)	45.5 (>39.3)	
Rt. ulnar	Digit 5	1.85	34.3 (>10.0)	48.6 (>39.3)	
Lt. sural	Calf	2.71	9.9 (>6.0)	51.7 (>34.7)	
Rt. sural	Calf	2.85	21.0 (>6.0)	49.1 (>34.7)	
H reflex Lt		26.6			
Rt		29.3			

Latencies are in milliseconds, amplitudes of compound muscle action potentials in millivolts. amplitudes of sensory nerve action potentials in microvolts. velocities in m/sec.
Amp, amplitude; Lt, left; Rt, right; MC, musculocutaneous.

She was discharged with a maintenance treatment of oral steroids. Based on the results of the muscle strength assessment performed at the 2-week follow-up visit, an improvement in overall strength was observed. According to the fol-

Table 2. The findings of electromyography

Muscle	Spontaneous activity				Voluntary contraction		
	IA	Fibrillation	PSWs	Amplitude	Duration	Recruitment	IP
Lt. ABP	↑	+1	+1	NL	NL	NL	NL
Rt. ABP	↑	0	0	↑	↑	↓	↓
Lt. FDI	↑	+1	+1	NL	NL	NL	NL
Rt. FDI	↑	+1	+1	NL	NL	NL	NL
Lt. FCR	↑	0	0	NL	NL	NL	NL
Rt. FCR	↑	+1	+1	↑	↑	↓	↓
Lt. biceps brachii	↑	+1	+2	↑	↑	↓	↓
Rt. biceps barchii	↑	+2	+2	↑	↑	↓	↓
Lt. triceps brahii	↑	+1	+2	↑	↑	↓	↓
Rt. triceps brachii	↑	+2	+2	↑	↑	↓	↓
Lt. vastus lateralis	↑	+1	+1	↑	↑	↓	↓
Rt. vastus lateralis	↑	0	+1	↑	↑	↓	↓
Lt. TA	↑	0	0	↑	↑	↓	↓
Rt. TA	↑	0	0	↑	↑	↓	↓
Lt. GCM	↑	0	0	↑	↑	↓	↓
Rt. GCM	↑	0	0	↑	↑	↓	↓
Lt. paraspinal C5-C8, T1, T6	↑	+1	+1				
Rt. paraspinal C5-C8, T1, T6	↑	+1	+1				
Lt. paraspinal L4-5	↑	+1	+1				
Rt. paraspinal L4-5	↑	+1	+1				
Lt. paraspinal S1	NL	0	0				
Rt. paraspinal S1	NL	0	0				
Lt. masseter	NL	0	0	NL	NL	NL	NL
Lt. tongue	NL	0	0	NL	NL	NL	NL

IA, insertional activity; PSW, positive sharp wave; IP, interference pattern; Lt, left; ABP, abductor pollicis brevis; NL, normal; Rt, right; FDI, first dorsal interosseous; FCR, flexor carpi radialis; TA, tibialis anterior; GCM, gastrocnemius medialis.

low-up spinal MRI results, the initially observed ventral nerve root enhancement had disappeared. After 2 years, the patient was under prognostic observation without recurrence and had maintained a low-dose prednisolone treatment.

DISSCUSION

Our patient showed a pattern of gradual progression of weakness and atrophy with no definite sensory symptoms over 9 months. In addition, because of EMG findings with a widespread denervation pattern, she was referred to our hospital with suspicion of early-stage MND. We focused on the discrimination of MND. In her spinal MRI, only the ventral roots were contrast enhanced, and the dorsal roots were not enhanced. Spinal MRI results and increased CSF protein levels were more consistent with polyradiculopathy than with MND. In the differential diagnosis, we also considered myopathies. As observed in this patient, if sensory symptoms are rarely accompanied by muscle weakness, we should consider myopathy, such as inflammatory myositis and inclusion body myopathy. However, we considered the possibility of myopathies low and did not perform muscle biopsy because the patient had normal creatine phosphokinase, and

the EMG finding was compatible with neuropathy.

Peripheral nervous system involvement in SS varies in the literature, ranging from 2% to 23% and 60%.[3,5,6] In two large-scale studies, four of 92 patients and six of 54 patients showed symptoms of polyradiculopathy.[7,8] Most of these patients exhibited a subacute pattern (2 weeks to a few months) and mainly described sensory symptoms, and dorsal root involvement was identified upon neuroimaging.[7] A case of subacute inflammatory polyradiculopathy associated with SS was previously reported in 2008.[9] The patient complained of simultaneous sensory discomfort and weakness. However, in contrast to our patient, this patient exhibited sensory dominant symptoms.

Inflammatory polyradiculopathy and SS may be comorbid conditions. However, SS can share a pathological mechanism as it causes autoimmune inflammatory responses, such as lymphocytic infiltration of the self-organ. If an autoimmune response occurs in the spinal nerve, specifically in the motor root, then it may cause the same symptoms reported in the present case. Moreover, steroid use improved our patient's condition; thus, our patient likely had an inflammatory lesion.

Based on the results of this case study, primary SS may induce inflammatory polyradiculopathy, primarily presenting with motor rather than sensory symptoms. A rapid, accurate diagnosis is needed for proper treatment with steroids and an appropriate immunosuppressive agent.

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