

## CASE REPORT

# Acute polyneuropathy as the main manifestation of primary Sjogren's syndrome: A case report

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## Abstract

Sjogren's syndrome is an inflammatory disease affecting many systems. We report a Sjogren case with the presenting feature of an acute motor-predominant polyneuropathy resembling Guillain-Barre syndrome. Upon further investigation, it was found that the patient had sicca symptoms for months. Scrupulous history should be taken to prevent a missed diagnosis.

## KEYWORDS

electromyography, nerve conduction, neurological manifestation, polyneuropathies, Sjögren syndrome

## 1 | INTRODUCTION

Sjögren syndrome is a systemic, inflammatory autoimmune disorder characterized by a broad spectrum of manifestations. Clinical presentations vary from single infiltration in salivary or lacrimal glands to extraglandular involvements, such as respiratory, urinary, musculoskeletal, and nervous systems. The prevalence of neurologic manifestations is about 8%–49%, of which about 20%–25% of patients present with peripheral nervous system (PNS) impairments.<sup>1,2</sup> PNS presentations progress at a subacute to chronic rate and vary from multiple mononeuropathies, ganglionopathy, small and large fibers peripheral neuropathy, cranial mononeuropathies, polyradiculoneuropathy, or polyneuropathy.<sup>3,4</sup> Although

the most common form is a sensory predominant, length-dependent, and large fiber peripheral neuropathy, usually it does not present acutely. An acute manifestation is expected to be associated with cranial neuropathies, multiple mononeuropathies, or small fiber neuropathy. In addition to clinical presentation, EMG/NCS (electromyography/nerve conduction studies) studies, muscle and nerve biopsies are helpful in diagnosing different types of peripheral neuropathies. Only 40%–60% of these patients suffer from sicca symptoms at presentation; thus, considering Sjögren as an underlying cause of peripheral neuropathy could improve patient health condition.<sup>5</sup>

We report a rare case of Sjögren disease, primarily characterized by an acute peripheral motor dominant polyneuropathy.

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## 2 | PATIENT INFORMATION

A 45-year-old male patient presented with numbness and acute progressive muscle weakness of bilateral lower extremities to the neurology department. Three days before the visit, his weakness started distally and continued to progress to the proximal parts, so he got difficulty in daily routine activities such as climbing the stairs. Then, it increased in severity, and he had difficulty getting up from a seated position. In addition, the patient suffered from bilateral paresthesias in the lower limbs around the ankle and feet in that period. Over a day, symptoms spread up toward his knees. Finally, he developed numbness in the distal lower limbs up to the knee on the right and up to the mid-calf on the left. The patient did not report any symptoms of musculoskeletal complications such as arthritis and myalgia and we did not detect fever. His past medical history was unremarkable. After visiting a general physician, he was prescribed a trial of Gabapentin. Due to the progression of his symptoms and lack of improvement, he was referred to a neurologist.

## 3 | CLINICAL FINDINGS

The patient's vital signs were stable, and the body temperature was normal. The patient was alert and oriented on the neurological examination with a Glasgow coma score

(GCS): 15/15, and cranial nerves were unremarkable. His upper extremities examination was all normal for sensation, force, and deep tendon reflexes. His lower extremities examination showed moderate superficial sensory loss of pain over the foot up toward mid-calf bilaterally. Joint position perception and vibration sensation were normal in the toes up to the ankle. On motor examination, bilateral foot dorsiflexor, plantar flexor, and toe extensor muscles had decreased force. Moreover, a mild weakness in inversion and eversion of ankle movements was detected. Deep tendon reflexes and tonicity were decreased bilaterally in the lower extremities. Plantar reflexes were downward. His gait was impaired due to the decreased force, and there were no signs in favor of cerebellar dysfunction and ataxia. The neurologist admitted the patient into the Intensive Care Unit (ICU) with the suspicion of Guillain-Barre syndrome.

## 4 | DIAGNOSIS ASSESSMENT

The EMG/ NCS (electromyography/ nerve conduction studies) tests were performed (Tables 1–3), and laboratory tests were requested. Electrophysiologic studies showed an acute mixed-type sensory-motor polyneuropathy in which the motor deficit was more prominent. Laboratory test results were remarkable for ESR (80 mm/h), CRP (+++), AST (623 U/L), ALT (562 U/L), CPK (1025 U/L), and Aldolase (7.6 U/L, normal was up to 3.5).

**TABLE 1** Electromyography studies in this patient

EMG	Insertion activity	Spontaneous			Motor unit potential			Recruitment pattern
		Fibrillate	PSW	Other discharges	Amp	Dur	Poly	
deltoid-Lt	NL	0	0	None	NL	NL	NL	NL
deltoid-Rt	NL	0	0	None	NL	NL	NL	NL
FCR-Lt/Rt	NL	0	0	None	NL	NL	NL	NL
Biceps Rt	NL	0	0	None	NL	NL	NL	NL
Brachioradialis-Lt	NL	0	0	None	NL	NL	NL	NL
APB-Rt/Lt	NL	0	0	None	NL	NL	NL	Decreased
FDI-Lt/Rt	NL	0	0	None	NL	NL	NL	Decreased
Vastus lateralis-Lt/Rt	NL	0	0	None	NL	NL	NL	Decreased
TA-Lt/Rt	NL	0	0	None	NL	NL	NL	Decreased
Gastrocnemius-Lt/Rt	NL	0	0	None	NL	NL	NL	Decreased
Paraspinal-L5-Lt	Inc	0	0	None	NL	NL	NL	NL
Paraspinal-L5-Rt	Inc	0	0	None	NL	NL	NL	NL
Iliopsoas-Lt	NL	0	0	None	NL	NL	NL	NL
Iliopsoas-Lt	NL	0	0	None	NL	NL	NL	NL

Abbreviations: Amp, amplitude; APB, abductor pollicis brevis; Dur, duration; FCR, flexor carpi radialis; FDI, first dorsal interosseous; Inc, increase; Lt, left; NL, normal; PSW, poly spike wave; Poly, polyphasic; Rt, right; TA, tibialis anterior.

TABLE 2 Motor nerve conduction studies in this patient

MNCV	Site/Segment	Latency ms		Amplitude mV		NCV m/s	
		Value	Normal	Value	Normal	Value	Normal
Ulnar-Lt	ADM	4.2	≤3.3	4.1	≥6.0	39	≥49
Ulnar-Rt	ADM	4	≤3.3	4.5	≥6.0	41	≥49
Median-Lt	APB	4.5	≤4.4	3.5	≥4.0	40	≥49
Median-Rt	APB	4.6	≤4.4	3.8	≥4.0	41	≥49
Tibial (AHB)-Lt	Medial ankle	6	≤5.8	2	≥4.0	30	≥41
Tibial (AHB)-Rt	Medial ankle	5.9	≤5.8	2.3	≥4.0	31	≥41
DPN(EDB)-Lt	Ankle	7	≤6.5	1	≥2.0	32	≥44
DPN(EDB)-Rt	Ankle	7.1	≤6.5	1.1	≥2.0	30	≥44

Abbreviations: Lt, left; ADM, abductor digiti minimi; Rt, right; DPN, deep peroneal nerve; AHB, abductor hallucis brevis; EDB, extensor digitorum brevis.

TABLE 3 Sensory nerve conduction studies in this patient

SNCV	Site/Segment	Latency ms		Amplitude mV		NCV m/s	
		Value	Normal	Value	Normal	Value	Normal
Median-Lt	Wrist	3.8	≤3.5	13	≥20	46	≥50
Median-Rt	Wrist	4	≤3.5	10	≥20	43	≥50
Ulnar-Lt	Wrist	3.7	≤3.1	11	≥17	45	≥50
Ulnar-Rt	Wrist	3.6	≤3.1	9	≥17	44	≥50
Sural-Lt	Sural-Lt	4.8	≤4.4	3	≥6	33	≥40
Sural-Rt	Sural-Rt	4.7	≤4.4	2	≥6	32	≥40
Superficial Peroneal-Lt	Superficial Peroneal-Lt	4.8	≤4.4	2	≥6	34	≥40
Superficial Peroneal-Rt	Superficial Peroneal-Rt	4.6	≤4.4	3	≥6	35	≥40

Abbreviations: Lt, left; Rt, right.

## 5 | THERAPEUTIC INTERVENTION

The patient received intravenous immunoglobulin (IVIG) 20 g daily for 5 days. No fever or side effects from IVIG administration were reported in the therapy period. Regardless of the therapy and care, there were no significant improvements in neurologic signs and symptoms after this period.

During hospitalization, the patient developed asymmetrical polyarthralgia in small and large joints; therefore, rheumatology consultation was requested. Upon reevaluation of the patient and more considerations in the review of different systems, it was found that the patient had suffered from dry eyes and dry mouth for six months which was in favor of Sjogren's Syndrome. There were no signs in line with vasculitis and arthritis in the rheumatological examinations. Additional laboratory tests were requested,

and the results were positive ANA (titer 1/1250), a positive RF (++++), anti-Ro (183 U), anti-La (176 U), negative anti-CCP and a negative anti-dsDNA. C3, C4, CH50, ANCA, C-ANCA, and P-ANCA were normal.

In suspicion of Sjogren's, a parotid scan was requested, and the result was bilateral decreased uptake which was compatible with Sjogren's syndrome.

The patient received a course of glucocorticoids (1000 mg methylprednisolone daily) for three days, followed by oral prednisolone (1 mg/kg) and cyclophosphamide (1000 mg daily). After this period of therapy, the patient's neurological symptoms were resolved, and the impaired muscle force reached the normal level after two weeks; so, the prednisolone was tapered after one month. Cyclophosphamide was continued for three months. His medication was changed upon improving his symptoms, and he continued with azathioprine (100 mg daily) for maintenance therapy.

## 6 | FOLLOW-UP AND OUTCOMES

The patient was put on a schedule for appointment follow-up every three months. The patient had not reported any adverse effects from his treatment until now.

## 7 | DISCUSSION

This was a Sjogren case in which the primary presentation was acute polyneuropathy, more pronounced in the lower extremities. The patient whom we present was finally diagnosed with Sjogren syndrome by meeting the minimum required score (four out of six) for the diagnosis according to 2016 ACR-EULAR classification criteria for Sjogren's syndrome.<sup>6</sup> The criteria which were qualified in our patient were dry eyes and mouth for more than three months, compatible parotid sialography, and positive anti-RO/La antibodies. The important point is that the patients may not inform the physicians about their sicca symptoms without directly asking them about these conditions. Therefore, screening precisely about these symptoms is crucial for clinicians for an accurate diagnosis.

Naddaf et al. reported a similar Sjogren case in which the patient (an elderly female) presented with acute peripheral neuropathy.<sup>5</sup> The difference between that case and the one we report is that their case presented mainly sensory deficits, whereas in our case, the prominent neurologic deficit was motor. In another case reported by Mochizuki et al,<sup>7</sup> the patient was an elderly female with predominant motor deficit presentation who responded well to the treatment with IVIG, whereas in the case, we reported IVIG did not elicit an appropriate response in the patient.

The main organs involved in primary Sjogren's syndrome are exocrine glands. Yet, extraglandular manifestations are a significant long-term prognostic factor for the disease.<sup>8</sup> In a large cohort study conducted by Cafaro et al,<sup>9</sup> the most common extra-glandular manifestation was found to be Raynaud's phenomenon. Our patient did not have a compatible history for the phenomenon. One-third of the patients had a history of parotid swelling, and one-third showed leukopenia in the laboratory studies. None of these were present in our case. Heterogeneous neurological manifestations associated with Sjogren's syndrome pose a diagnostic challenge.<sup>10</sup> Both central and peripheral nervous systems can be affected, though central involvement is more uncommon than peripheral nervous system involvement.<sup>9</sup> Our patient did not have central nervous system involvement. Although peripheral neuropathy is not an uncommon presentation of Sjogren syndrome, it is rarely acute; so, that it could lead to a missed diagnosis,

in this case, Guillain–Barre syndrome instead of Sjogren's associated polyneuropathy.

In the mentioned cohort study,<sup>9</sup> comparing patients with and without a peripheral nervous system reported that purpura, other organ involvement, low complement level (C4), and cryoglobulinemia were meaningfully more frequent in patients in the former group. However, none of these signs were present in our patient. They also reported that the two most common peripheral nervous system involvement in primary Sjogren's syndrome are pure sensory neuropathy and sensory-motor polyneuropathy. Only 9% of the patients with sensory-motor polyneuropathy had neurological manifestation onset before the diagnosis of Sjogren's syndrome.<sup>9</sup>

In previous reports of cases with a similar presentation,<sup>7,11</sup> patients usually characterized by severe quadriparesis dramatically responded to treatment with IVIG. In contrast, our patient presented with paraparesis and paresthesia, which did not respond well to IVIG. This unresponsiveness to IVIG makes us suspicious of another underlying cause that describes the patient's condition, which finally guides the diagnosis to Sjogren syndrome. After initiating the specific therapy for Sjogren syndrome by the rheumatologist, the neurological deficits resolved.

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### AUTHOR CONTRIBUTIONS

Arsh Haj Mohamad Ebrahim Ketabforoush, Nahid Abbasi Khoshshirat, Arman Maghoul, Matineh Nirouei, and Elahe Dolatshahi involved in study concept and design, acquisition of data, drafting of the manuscript, critical revision of the manuscript for important intellectual content, administrative, technical, and material support, and study supervision.

### CONFLICTS OF INTEREST

The authors declare that they have no conflicts of interest.

### DATA AVAILABILITY STATEMENT

Data sharing is not applicable to this article as no new data were created or analyzed in this study.

### ETHICAL APPROVAL

None.

### CONSENT

Written informed consent was obtained from the patient.


## PATIENT PERSPECTIVE

The prognosis of Sjogren's syndrome was discussed with the patient. It was explained that it is an inflammatory autoimmune disorder. The patient returned to normal life, and he was very satisfied with the outcome of his treatment.

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