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A RARE PRESENTATION OF ACUTE MOTOR-SENSORY AXONAL NEUROPATHY ASSOCIATED WITH SYSTEMIC LUPUS ERYTHEMATOSUS

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ABSTRACT Systemic lupus erythematosus (SLE) is a chronic, multisystem autoimmune disorder that typically requires immunosuppressive therapy for disease control. Acute motor-sensory axonal neuropathy (AMSAN), a rare and severe variant of Guillain-Barré syndrome (GBS), is usually managed with intravenous immunoglobulin (IVIG) or plasmapheresis. The co-occurrence of AMSAN as an initial presentation of SLE is exceedingly uncommon and poses significant diagnostic and therapeutic challenges. We report the case of a 22-year-old female who presented with rapidly progressive lower limb weakness (paraparesis), facial deviation, dysarthria, change in voice tone, difficulty in swallowing foods, dysesthesia, generalized fatigue, and joint pain associated with morning stiffness. Based on the acute neurological presentation, a diagnosis of AMSAN was initially made, and the patient was treated with IVIG. However, there was minimal clinical improvement following therapy. Subsequent serological investigations revealed positive autoimmune markers consistent with a diagnosis of SLE. The patient was commenced on high-dose intravenous methylprednisolone followed by cyclophosphamide. Over the course of three months, her neurological status improved significantly, transitioning from a bedridden state to being wheelchair-dependent. This case underscores the importance of considering systemic autoimmune diseases such as SLE in patients presenting with atypical or treatment-refractory neuropathies. AMSAN may represent a rare neurological manifestation of SLE and should be included in the differential diagnosis, particularly in young female patients with systemic features.

KEYWORDS : *Acute motor-sensory axonal neuropathy, AMSAN, Guillain-Barré syndrome, systemic lupus erythematosus*

CASE REPORT

A 22-year-old Assamese woman who had been in good health before developed rapidly worsening bilateral ascending flaccid weakness and tingling in all four extremities. Within three days, she developed quadriplegia. In addition, both the finger and knee joints experienced morning stiffness, pain, tenderness, and swelling. She gave history of hair loss, easy fatigue for the past 8 months. Additionally, there was difficulty swallowing, slurred speech, and a change in mouth angle. She had no recent history of vaccinations, fever, or diarrhoea. She was normotensive (126/65 mm Hg) and afebrile (98.8°F) at presentation. At the time of presentation, she did not have an oral ulcer,or rash. No superficial lymphadenopathy was present. Both knees and finger joints showed tenderness and pain when moving passively. A neurological examination revealed areflexia, hypotonia, and complete quadriplegia. All four extremities showed signs of hyperesthesia.

Initial blood work revealed positive results for ds-DNA, histones, Rib-P protein, Ro 52, SS-A, SM, nRNP/Sm, and Ku, as well as elevated erythrocyte sedimentation rate, C-reactive protein, and antinuclear antibody. Protein was detected by urinalysis. Urine protein was 3.37 grams per 24 hours. Rheumatoid factor- 11IU/ml, X ray of the joint shows no erosions or deformity. A cerebrospinal fluid analysis revealed a white blood cell count of 4/μL, protein of 100.02 mg/dL, and sugar of 58 mg/dL. Acute diffuse axonal sensorimotor polyneuropathy (motor predominant) was suggested by a nerve conduction study (Table 1). Multiple episodes of lymphopenia (<1000/μL) were revealed by serial complete blood counts.

Sensory NCS

Nerve/sites	Rec. Sited	Onset Lat ms	Peak Lat ms	NP Amp uV	PP Amp uV	Segments	Distance mm	Velocity m/s
L Median –drug III (Antidromic)								
Wrist	Index	2.79	3.38	5.4	3.6	Wrist-Index	140	50
R Median- Dig III (Antidromic)								
Wrist	Index	2.77	3.25	4.2	4.5	Wrist-Index	140	51
R Ulnar-Dig V (Antidromic)								
Wrist	Dig V	2.40	3.17	5.8	7.7	Wrist- Dig V	120	50
L Ulnar-Dig V (Antidromic)								
Wrist	Dig V	2.48	3.23	5.1	6.0	Wrist- Dig V	120	48

MOTOR NCS

Nerve/sites	Muscle	Latency ms	Amplitude mV	Rel Amp %	Segments	Distance mm	Lat diff ms	Velocity	Rel vel %
L Median- APB									
Wrist	APB	3.71	4.1		Wrist-APB	80			
Elbow	APB	7.56	3.7	90.6	Elbow- Wrist	190	3.85	49	100
R Median- APB									
Wrist	APB	3.56	4.4		Wrist-APB	80			
Elbow	APB				Elbow- Wrist	190	4.02	47	100
R Ulnar- ADM									

Wrist	ADM	3.65	4.2	100	Wrist-ADM	80			
Elbow	ADM				A. Elbow- B. Elbow	190	4.63	48	100
L Ulnar- ADM									
Wrist	ADM	3.65	4.2	100	Wrist-ADM	80			
Elbow	ADM	7.56	3.7	87.7	A. Elbow- B. Elbow	190	4.21	46	1000
L- Peroneal-EDB									

At level 1 of diagnostic certainty, the patient was initially diagnosed with GBS based on the Brighton Collaboration diagnostic criteria. Consequently, intravenous immunoglobulin was used as her initial treatment. But there was no discernible clinical improvement. Both the 1997 American College of Rheumatology criteria and the 2012 Systemic Lupus International Collaborating Clinics criteria were used to diagnose SLE because of the patient's history of arthritis, lymphopenia, and the seropositivity of antinuclear antibody and anti-double-stranded DNA. Cyclophosphamide and methylprednisolone were administered intravenously. The patient's neurological deficiencies like slurring of speech, facial deviation, difficulty in swallowing gradually improved but her motor strength did not improve. She was given low-dose prednisolone and sent to an inpatient rehabilitation center. At discharge, She remained bedridden. Even after at the end of three months, the patient needed help to carry out everyday tasks.

DISCUSSION

The pathophysiology of SLE, an immune-mediated illness, depends on persistent autoantibody production and tolerance loss. In contrast, SLE is a chronic illness, whereas GBS is typically a self-limited monophasic condition. GBS can occasionally be a sign of SLE. According to a meta-analysis of several cohort studies, the prevalence of GBS is less than 0.1% of patients with SLE, while the prevalence of polyneuropathy and mononeuropathy (single or multiplex) is 1.5% and 0.9%, respectively.6. The axonal variants of GBS are AMSAN and acute motor axonal neuropathy. Seldom has it been documented that the axonal variants of GBS and SLE are related. Furthermore, there are currently no accepted treatment guidelines for this condition.

The substantial advantages of intravenous immunoglobulin and plasma exchange in the treatment of GBS were shown by meta-analyses of several clinical trials.7, 8 On the other hand, a 1993 GBS steroid trial and a Dutch study demonstrated that high-dose intravenous methylprednisolone was not helpful in treating GBS.[9,10] While the production of antibodies that cross-react with self-ganglioside is the primary cause of GBS, the immunopathogenesis of SLE includes numerous additional innate and adaptive immune system mechanisms.11,12 Thus, in this situation, glucocorticoids and other immunosuppressants might be helpful.

Furthermore, we conducted a thorough search for prior GBS case reports involving SLE. Demyelinating variants were found in most cases. Table 2 provides a summary of our case as well as the seven axonal variant cases from earlier reports13–18. Numerous immunotherapy regimens were used to treat the majority of GBS cases, which happened in the early stages of SLE. With treatment, the prognosis was generally favorable.

AMAN indicates acute motor axonal neuropathy; AMSAN, acute motor-sensory axonal neuropathy; CP, cyclophosphamide; CS, corticosteroids; F, female; GBS, Guillain-Barré syndrome; HCQ, hydroxychloroquine; IVIG, intravenous immunoglobulin; M, male; MMF, mycophenolate mofetil; PCB variant, pharyngeal-cervical-brachial variant; PE, plasma exchange; SLE, systemic lupus erythematosus.

[a]The patient was known to have SLE of unknown duration.

In conclusion, AMSAN can be a rare first manifestation of SLE. Furthermore, regimens other than intravenous immunoglobulin and plasma exchange should be considered in patients with GBS or its variant who also have supportive finding of SLE.

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