Mod Care J. 2021 October; 18(4):e119315. doi: 10.5812/modernc.119315.

Published online 2021 December 5. **Case Report**

A Case of Guillain-Barré Syndrome as the First Presentation of Systemic Lupus Erythematosus

Zeinab Saremi [1], Mahdi Bakhshi Mohammadi [2] and Zahra Ahmadi [3, 2, *]

[1] Valiasr Hospital, Birjand University of Medical Sciences, Birjand, Iran
[2] Clinical Research Development Unit of Razi Hospital, Birjand University of Medical Sciences, Birjand, Iran
[3] Department of Neurology, School of Medicine, Birjand University of Medical Sciences, Birjand, Iran

[*] *Corresponding author*: Department of Neurology, School of Medicine, Birjand University of Medical Sciences, Birjand, Iran. Email: zahmadi062@gmail.com

Received 2021 September 04; **Revised** 2021 November 20; **Accepted** 2021 November 28.

Abstract

Introduction: Systemic lupus erythematosus (SLE) is a chronic autoimmune disease with different clinical manifestations. Acute [resembling Guillain-Barré syndrome (GBS)] or chronic (chronic inflammatory demyelinating polyradiculoneuropathy) inflammatory polyradiculoneuropathy has been reported in rare SLE cases.
Case Presentation: We reported a 39-year-old woman that presented with acute peripheral neuropathy, and she was eventually diagnosed with SLE. She developed distal numbness and paraesthesia followed by progressive upper and lower extremity weakness and difficulty in swallowing and speaking. She had a history of flu-like illness three weeks before to symptoms.
Conclusions: Progressive upper and lower extremity weakness along with areflexia and electrodiagnostic findings suggested the diagnosis of Guillain-Barré syndrome. Over a month, significant neurological recovery occurred, and the patient's function continued to recover.

Keywords: Guillain-Barré Syndrome, Systemic Lupus Erythematosus, Case Report

1. Introduction

Systemic lupus erythematosus (SLE) is a chronic, multisystemic, and autoimmune disease with different clinical symptoms (1). Immunologic, genetic, and environmental factors are involved in developing the disease. The prevalence and incidence of the disease vary around the world. SLE is more prevalent in women than men from 2.0: 1 to 15.1: 1; this ratio varies by age and race (2). Neurologic manifestations of the disease occur in 10 to 80% of patients at baseline or in the course of the disease and are major causes of mortality and morbidity in patients (3). Neuropsychiatric SLE (NPSLE) symptoms are neurologic (both peripheral and central) and psychiatric diseases and include 19 symptoms (4). Acute [like the Guillain-Barré syndrome (GBS)] or chronic (chronic inflammatory demyelinating polyradiculoneuropathy) inflammatory polyradiculoneuropathy has been reported rarely in SLE. GBS is an acute immune-mediated polyneuropathy that is almost provoked by infection. A small percentage of patients develop GBS after another triggering, such as immunization, surgery, and trauma. GBS, in rare cases, can be the first presentation of SLE (4-6). We reported a 39-year-old woman that presented with acute peripheral neuropathy and bell's palsy. She was ultimately diagnosed with SLE and treated with a short course of immunosuppressive therapy.

2. Case Presentation

A 39- year-old female patient presented to the hospital with new-onset upper and lower extremity weakness started seven days before admission. She had developed distal numbness and paranesthesia followed by progressive upper and lower limbs weakness and difficulty in swallowing and speaking. She had a history of flu-like illness three weeks before the symptoms.

On initial physical examination, she had stable vital signs. Oxygen saturation was normal, and she had no respiratory distress. In neurologic assessment, she had left facial muscle paralysis. Motor examination revealed a decrease in muscle strength bilaterally in lower and upper limbs (3/5 distally and 4/5 proximally). She showed a decrease in deep tendon reflexes in both knees and ankles. Plantar reflexes were bilaterally flexor. Electrodiagnostic studies revealed a significant decrease in motor and sensory amplitudes supported the diagnosis of acute motor and sensory polyneuropathy.

Progressive upper and lower extremity weakness along with the findings of flexia and electrodiagnostic studies suggested the diagnosis of Guillen-Barre syndrome; thus, treatment with intravenous immunoglobulin (IVIG) (2 g/kg) was started with bedside physical therapy, but the patient had fever with unknown origin.

In laboratory tests, we found leukopenia 2800 per mm3 (neutrophils 35% and lymphocytes 49%). Red blood cells, platelet count, and liver function tests were normal. The erythrocyte sedimentation rate (ESR) and C- reactive protein levels were 92 mm/h and 60 mg/dL, respectively.

Urine and blood culture was negative, and in infectious diseases consultation, there was no infectious etiology for her fever.

Due to a history of hair loss and mouth ulcers and the presence of leucopenia and elevated ESR, more investigation and rheumatology consultation were done. Positive results were obtained for anti-nuclear antibody ANA test, fluorescent anti-nuclear antibody (FANA), anti-Lupus anticoagulant IgG, anti-cardiolipin, anti-Ro, anti-Ro 52, anti-ribosomal protein, and anti- centromere B antibodies, and low levels of serum complement components (C3, C4, and CH50) were observed. Anti- dsDNA was negative. HIV and hepatitis B/C serologic tests were negative. There was no pericardial effusion in echocardiography. Magnetic resonance imaging (MRI) of the brain and cervical spine showed no abnormalities.

In rheumatology consultation, diagnosis of SLE was made because of leukopenia, malar rash, and positive FANA test, anti-cardiolipin, and direct coombs. The patient was treated with prednisolone (30 mg/day), azathioprine (100 mg/day), and hydroxychloroquine (200 mg) daily. After two days, the fever stopped, and leukopenia was eliminated, with a white blood cell (WBC) count of 4300/mm^3. Over the following month, significant neurological recovery occurred, and the patient's function continued to recover. ESR was 16 in two months. After two months, she walked without the help of others, and after six months, neurological examinations were normal, and the strength of all muscle groups was grade 5.

3. Discussion

SLE is a chronic multisystem autoimmune disease characterized by inflammation in various tissues and organs by affecting the innate and adaptive immune systems. Different organs, including the mucous membranes and skin, joints and muscles, kidneys, heart, lungs, and the digestive and neuropsychiatric systems, are affected by SLE (6).

NPSLE can occur primarily due to direct involvement of the central and peripheral nervous system in active SLE or secondary to treatments and complications of the disease, such as infection. The diagnosis of NPSLE is increasing in patients with lupus because of better lab tests and doctors' awareness (7).

Here, we explained a patient that was initially admitted by GBS diagnosis with facial palsy that was eventually diagnosed with SLE. Immediately after admission, the patient underwent electromyography showing demyelination neuropathy, and a diagnosis of GBS was made. She was treated with intravenous immunoglobulin. The patient developed fever and leukopenia during hospitalization. In rheumatology consultation, diagnosis of SLE was made because of leukopenia, malarial rash, hair loss, and positive FANA test and anti-cardiolipin and direct coombs. There was no evidence of renal, brain, cardiac, or lung involvement. Prednisolone (30 mg/day), azathioprine (100 mg/day), and hydroxychloroquine (200 mg/day) were prescribed for the patient, and she was treated.

Neurologic manifestations of NPSLE include central, peripheral, and autonomic nervous system manifestations. Several mechanisms for the pathogenesis of NPSLE in different clinical manifestations have been described: (1) first, damage caused by autoantibodies and cytokine production most commonly seen in diffuse NPSLE symptoms, such as cognitive impairment; (2) second, accelerated atherosclerosis, leading to stroke in patients with SLE; and (3) finally, thrombotic phenomena associated with focal neurological manifestations, such as seizure (8).

Involvement of the peripheral nervous system is a significant component of NPSLE and has a substantial negative effect on the prognosis of the disease. About 10 to 15% of patients with lupus develop peripheral neuropathy, usually due to small artery nerve-feeding vasculitis (8).

Guillain Barre is a rare manifestation of SLE. The incidence of lupus in GBS is 0.6 - 1.7%, but a limited number of cases have been reported indicating Guillen Barre as the first manifestation of SLE (4, 5, 9, 10).

There is no randomized clinical trial, especially investigating the treatment of GBS in SLE because of the rarity of GBS in patients with SLE; however, neuropathy in these patients usually responds well to corticosteroids. Our case, unlike other studies, was treated with azathioprine (100 mg/day) for three months (10). After three months, azathioprine was discontinued because of nausea and vomiting, and treatment with prednisolone and hydroxychloroquine continued. After a one-year follow-up, there was no neurologic problem in the patient. In SLE patients, GBS can be treated with a short course of azathioprine, and there is no need for aggressive immunosuppression.

3.1. Conclusion

Guillen Barre can be the first presentation of SLE. Neurologists must consider SLE if young women have GBS to choose the best treatment for the patient.

Acknowledgments

Special thanks to the patient for permitting us to report his illness.

Footnotes

Authors' Contribution: All authors contributed equally to the preparation of the manuscript for publication, and they read and confirmed the final manuscript for publication.

Conflict of Interests: The authors declared that they have no conflicts of interest.

Ethical Approval: This case was approved by the Ethics Committee of Brijand University of Medical Sciences (IR.BUMS.REC. 1400.009).

Funding/Support: The authors received no specific funding for this report.

Informed Consent: Informed written consent was obtained from the patient.

References

1. Pons-Estel GJ, Alarcon GS, Scofield L, Reinlib L, Cooper GS. Understanding the epidemiology and progression of systemic lupus erythematosus. *Semin Arthritis Rheum*. 2010;**39**(4):257–68. doi: 10.1016/j.semarthrit.2008.10.007. [PubMed: 19136143]. [PubMed Central: PMC2813992].
2. Ugarte-Gil MF, Gonzalez LA, Alarcon GS. Lupus: the new epidemic. *Lupus*. 2019;**28**(9):1031–50. doi: 10.1177/0961203319860907. [PubMed: 31299878].
3. Unterman A, Nolte JE, Boaz M, Abady M, Shoenfeld Y, Zandman-Goddard G. Neuropsychiatric syndromes in systemic lupus erythematosus: A meta-analysis. *Semin Arthritis Rheum*. 2011;**41**(1):1–11. doi: 10.1016/j.semarthrit.2010.08.001. [PubMed: 20965549].
4. Okoh HC, Lubana SS, Langevin S, Sanelli-Russo S, Abrudescu A. A Case of Systemic Lupus Erythematosus Presenting as Guillain-Barre Syndrome. *Case Rep Rheumatol*. 2015;**2015**:528026. doi: 10.1155/2015/528026. [PubMed: 26366317]. [PubMed Central: PMC4553270].
5. Chandrashekhar S, De Sousa E. Guillain-Barre Syndrome as a Rare Initial Presentation of Systemic Lupus Erythematosus.(P2. 447). *Neurology*. 2018;**90**(Suppl 15).
6. Genga EK, Otieno FO, Mativo PM, Oyoo GO. Systemic lupus erythematosus with acute inflammatory demyelinating polyneuropathy: A case report. *Afr J Rheumatol*. 2018;**6**(1):15–7.
7. Schwartz N, Stock AD, Putterman C. Neuropsychiatric lupus: new mechanistic insights and future treatment directions. *Nat Rev Rheumatol*. 2019;**15**(3):137–52. doi: 10.1038/s41584-018-0156-8. [PubMed: 30659245]. [PubMed Central: PMC8023338].
8. Bertsias GK, Boumpas DT. Pathogenesis, diagnosis and management of neuropsychiatric SLE manifestations. *Nat Rev Rheumatol*. 2010;**6**(6):358–67. doi: 10.1038/nrrheum.2010.62. [PubMed: 20458332].
9. Gao Z, Li X, Peng T, Hu Z, Liu J, Zhen J, et al. Systemic lupus erythematosus with Guillian-Barre syndrome: A case report and literature review. *Medicine (Baltimore)*. 2018;**97**(25). e11160. doi: 10.1097/MD.0000000000011160. [PubMed: 29924024]. [PubMed Central: PMC6034562].
10. Li X, Wang Y. Systemic Lupus Erythematosus With Acute Inflammatory Demyelinating Polyneuropathy: A Case Report and Review of the Literature. *J Clin Med Res*. 2016;**8**(7):555–9. doi: 10.14740/jocmr2550w. [PubMed: 27298667]. [PubMed Central: PMC4894028].