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Guillain-Barré Syndrome as the Initial Presentation of Systemic Lupus Erythematosus: A Case Report

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ABSTRACT

Guillain-Barré Syndrome (GBS) can be the initial presentation of Systemic Lupus Erythematosus (SLE). Neurologists should consider SLE as a differential diagnosis in patients with limb tingling and weakness, adapting treatment accordingly for optimal management.

1 | Introduction

Guillain-Barré Syndrome (GBS) is a rare acute autoimmune polyradiculoneuritis that affects the peripheral nervous system [1]. The incidence of GBS ranges from 1.1 to 1.7 per 100,000 individuals [2].

Systemic Lupus Erythematosus (SLE) is a chronic autoimmune disease with diverse clinical manifestations, influenced by various immunological, genetic, and environmental factors [3, 4]. Neuropsychiatric manifestations occur in 39% to 50% of SLE patients. Involvement of the peripheral nervous system, including conditions such as GBS, accounts for less than 10% of Neuropsychiatric Systemic Lupus Erythematosus (NPSLE) cases [5]. The reported prevalence of SLE occurring alongside GBS ranges from 0.6% to 1.7% [6].

This report details the complex case of a 42-year-old female with a recent history of GBS, for which she received 1 cycle of intravenous immunoglobulin (IVIg) over a span of 5 consecutive days, now presenting with a severe SLE flare involving multiple organ systems and an intraparenchymal bleed.

2 | Case History

A 42-year-old married woman with a known history of hypothyroidism, hypertension, type 2 diabetes mellitus (T2DM), and depressive disorder was previously diagnosed with GBS 5 months ago and treated with 1 cycle of IVIg over 5 days.

Five months post-treatment for GBS, she presented with joint pain affecting large and small joints (thoracolumbar spine, hips, knees, ankles, shoulders, elbows, metacarpophalangeal, and proximal interphalangeal joints) for 10 days, early morning stiffness for 7 days, generalized body aches for 10 days, progressive pitting-type bilateral lower limb swelling for 8 days, scaly non-itchy rashes on the trunk, arms, and forearms for 6 days (Figure 1), and hair loss for 15 days.

Examination showed no pallor, icterus, lymphadenopathy, clubbing, cyanosis, edema, or dehydration. Her vital signs were: BP 140/80 mmHg, PR 98/min, RR 20/min, afebrile, no jugular venous distension, and SpO2 94% on room air. However, she exhibited musculoskeletal complaints, hematological issues (pancytopenia), serositis, ascites, pleural effusion,

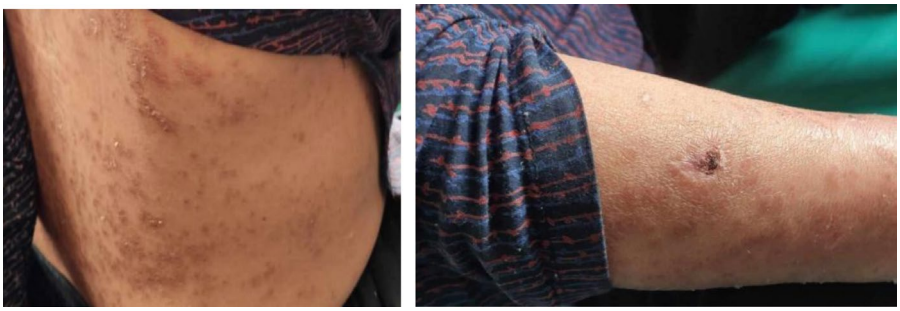

FIGURE 1 | Scaly non-itchy rashes on the trunk and forearms at the time of the patient's presentation.

mucocutaneous involvement, and renal involvement with subnephrotic proteinuria.

3 | Investigations and Treatment

3.1 | Initial Presentation and Treatment of GBS

Her initial presentation at the hospital included a tingling sensation in the upper limbs for 12 days and in the lower limbs for 10 days, followed by weakness in the lower limbs for 10 days and the upper limbs, difficulty swallowing, slurred speech, and facial deviation for 6 days. Physical examination revealed a Glasgow Coma Scale (GCS) score of 15, indicating that the patient was conscious and oriented, with normal verbal responses and appropriate motor responses to commands. The patient had intact higher mental functions, bilateral facial nerve weakness, absence of meningeal signs, decreased muscle tone in all limbs, normal muscle bulk, and a motor power of 4/5 in all limbs.

Laboratory results showed a total leukocyte count (TLC) of $2.9 \times 10^3/\mu L$, hemoglobin (Hb) of 11.3 g/dL, platelets of $259 \times 10^3/\mu L$, sodium (Na) at 140 mmol/L, and potassium (K) at 4.9 mmol/L. Renal function tests (RFT) showed urea at 48 mg/dL and creatinine at 2.9 mg/dL. Cerebrospinal fluid (CSF) analysis revealed a total cell count of less than 5 cells/μL, a differential count (DC) of 30%, and a protein level of 300 mg/dL. A CT scan suggested mixed multiple sensorimotor neuropathy with predominant demyelination. She was diagnosed with GBS (Acute Inflammatory Demyelinating Polyneuropathy) and treated with 1 cycle of IVIg over 5 days, alongside physiotherapy for residual weakness, achieving a modified Rankin scale score indicating moderate disability.

3.2 | Diagnostic Assessment After Initial Management of GBS

Further investigations revealed positive anti-dsDNA (1:160), anti-Smith (strong positive), anti-Ro (positive), anti-La (positive), and anti-U1RNP antibodies (positive) with features of myositis and neuropsychiatric symptoms, decreased C3 (35 mg/dL) and C4 (8 mg/dL) levels, a positive direct Coombs test, and positive antiphospholipid antibodies (lupus anticoagulant, IgG antiphospholipid).

Electroneuromyography showed electrophysiological disturbances, with conduction velocities reduced to 30 m/s (normal range: 40–60 m/s) and amplitudes of 2.1 mV (normal range: 5–10 mV), consistent with acute inflammatory demyelinating polyneuropathy. Cerebrospinal Fluid (CSF) analysis revealed a total white blood cell count of 4 cells/μL (normal range: 0–5 cells/μL), a differential count of 30% (normal: 0%), and a protein level of 300 mg/dL (normal: 15–45 mg/dL), supporting the diagnosis of albuminocytological dissociation, which is characteristic of GBS.

Autoimmune panel results confirmed positive antinuclear antibodies (ANA) with a titer of 1:1280 (normal: <1:80), positive anti-dsDNA at a titer of 1:320 (normal: negative), anti-Smith antibodies (strong positive), along with anti-Ro (positive), anti-La (positive), and anti-U1RNP antibodies (positive).

Computed Tomography (CT) scan of the head showed intraparenchymal hemorrhage. (Figure 2) An MRI of the head showed left parietal and occipital lobe intraparenchymal hemorrhage (Figure 3). Echocardiography revealed mild mitral regurgitation (MR), mild tricuspid regurgitation (TR), and a left ventricular ejection fraction (LVEF) of 60%.

Elevated vitamin B12 levels (1701 pg/mL), folic acid levels (19.93 ng/mL), increased lactate dehydrogenase (LDH) levels (333 U/L, normal range <247 U/L), decreased C3 (17.6 mg/dL, normal range 90–180 mg/dL) and C4 (8.9 mg/dL, normal range 10–40 mg/dL) levels were noted, along with normocytic normochromic cells, microcytes, ovalocytes, and elliptocytes on peripheral blood smear (PBS). A thorough rheumatological and autoimmune workup led to a diagnosis of SLE.

3.3 | Treatment

She was treated with intravenous methylprednisolone 1 g once daily for 3 days and cyclophosphamide 750 mg according to the NIH protocol. Following treatment, her joint symptoms improved, and her platelet count increased to 98,000/μL. For thrombocytopenia with intracerebral hemorrhage (ICH), neurology advised continuing aspirin due to her positive antiphospholipid antibodies (IgG). She showed improvement during her hospital stay and was discharged after 30 days with a plan for outpatient follow-up.

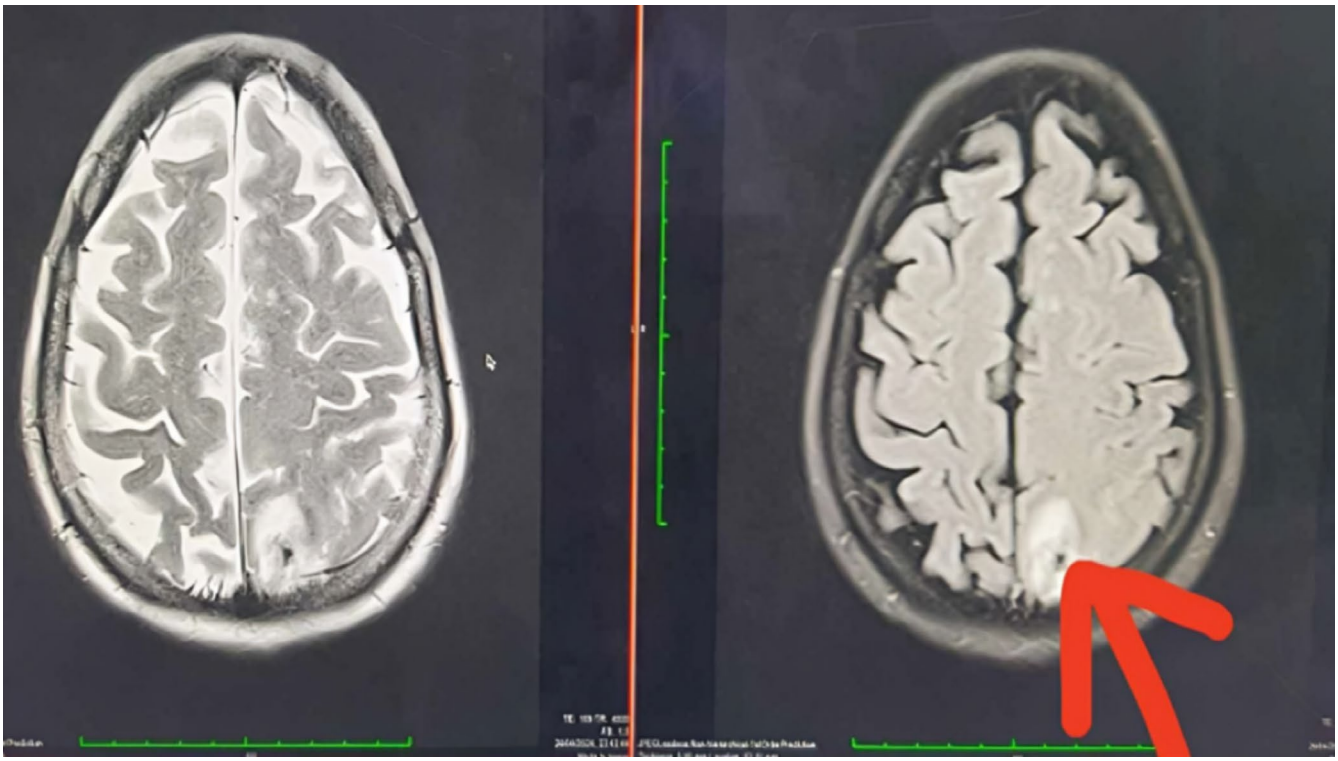

FIGURE 2 | CT scan of the head showing intraparenchymal hemorrhage.

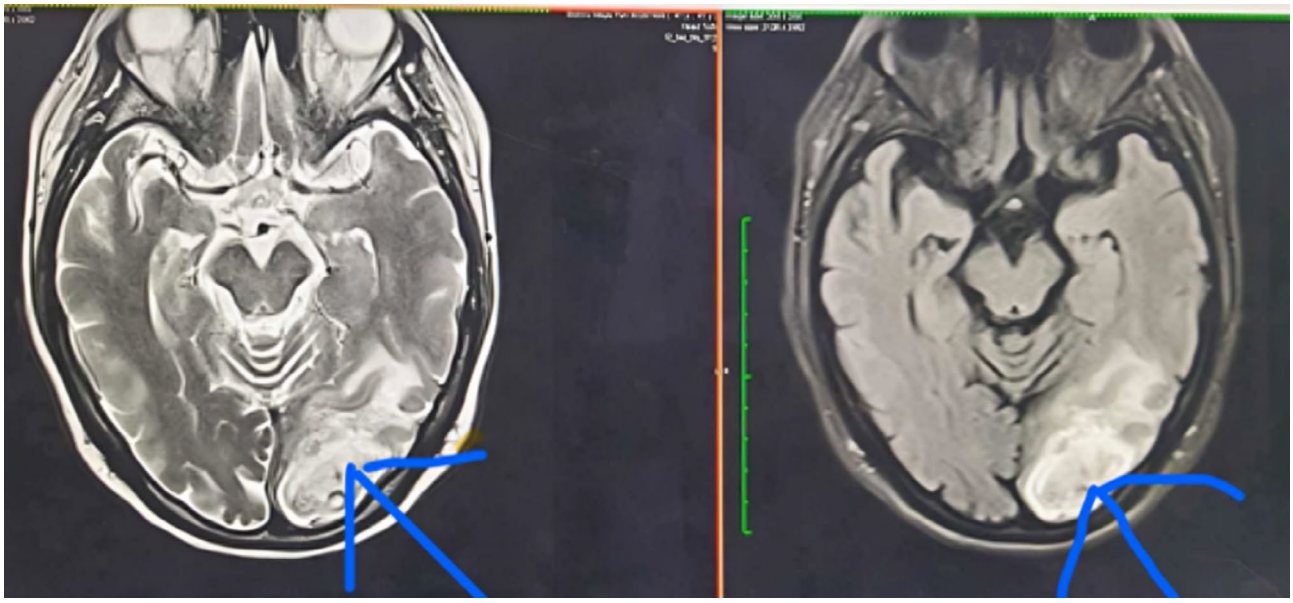

FIGURE 3 | MRI of the head showing areas of T1-weighted imaging (T1WI), T2-weighted imaging (T2WI), and Short Tau Inversion Recovery (STIR) hyperintensities with surrounding edema are seen in the left occipital lobe, likely representing a hemorrhagic infarct along the left Posterior Cerebral Artery (PCA) territory.

4 | Outcome and Follow-Up

No adverse events were encountered after the discharge of the patient. The patient was regular in her follow-ups and showed improvement.

5 | Discussion

SLE is a chronic multisystem autoimmune disease characterized by the involvement of both the adaptive and innate immune systems [7]. It affects various organs, including the mucous membranes

and skin, joints and muscles, kidneys, heart, lungs, and digestive system, and can cause neuropsychiatric symptoms [5]. The presence of autoantibodies in SLE can affect multiple organs, leading to complex and diverse clinical manifestations of the disease.

GBS consists of acute immune-mediated polyneuropathies and is one of the most common causes of acute, acquired weakness, often triggered by a preceding infection [8]. Complications of GBS can include respiratory failure or dysfunction. Although GBS is a rare manifestation of SLE, with an incidence of lupus in GBS ranging from 0.6% to 1.7%, there are limited reports of GBS as the initial manifestation of SLE [6]. In our case, bilateral tingling sensations in the hands and legs, progressive upper and lower extremity weakness, and areflexia, along with albuminocytological dissociation, suggested the diagnosis of GBS. The patient was treated with IVIg. However, the subsequent development of progressive joint, hematological, skin, and renal involvement indicated a diagnosis of SLE.

The exact pathogenesis of GBS as a manifestation of active SLE is unclear, but both cell-mediated and humoral mechanisms are thought to significantly contribute [9]. Autoimmune antibodies induced by lupus targeting myelin tissue are believed to play a role [10]. Several anti-neuronal antibodies such as anticardiolipin antibodies, anti-lymphocytic antibodies, and lupus anticoagulants are known to damage the myelin components of nerves [11]. Two large studies reported that among 1463 and 4924 SLE patients, GBS was observed in only 1 and 2 individuals, respectively [12, 13].

At present, there is no established treatment combination proven to effectively manage GBS in the context of SLE [14]. For GBS, IVIG and plasma exchange have been considered effective therapies [15]. Various treatment modalities for GBS in the context of SLE include corticosteroids, cyclophosphamide, plasmapheresis, and immunoglobulin therapy [16]. Gao et al. reported that the patient received intravenous pulses of methylprednisolone for 5 days along with monthly pulses of cyclophosphamide [11]. In another report, the patient was treated with a single intravenous pulse of methylprednisolone and underwent plasmapheresis, resulting in significant improvement [17]. Vaidya et al. reported that plasmapheresis, corticosteroids, IVIG, and cyclophosphamide were administered with favorable outcomes in the patient [18].

Due to the lack of randomized clinical trials, there are few established guidelines for treating GBS in the context of SLE. However, neuropathy and musculoskeletal symptoms have been shown to improve with the addition of corticosteroids. While rare, GBS as an initial presentation in SLE patients should always be considered.

6 | Conclusion

GBS can be the initial manifestation of SLE. This case underscores the importance for neurologists to consider SLE as a potential differential diagnosis in patients presenting with limb tingling and weakness. Early recognition and appropriate management are crucial, as SLE can lead to multisystem involvement and severe complications. Comprehensive diagnostic evaluation is essential for optimizing patient outcomes in such complex presentations.

Author Contributions

Rishi Raj Sharma: conceptualization, data curation, supervision, validation, visualization, writing – original draft, writing – review and editing. **Pukar Ghimire:** conceptualization, data curation, validation, visualization, writing – original draft, writing – review and editing. **Nebula Devkota:** conceptualization, data curation, validation, visualization, writing – original draft, writing – review and editing. **Prashant Pant:** conceptualization, data curation, validation, visualization, writing – original draft, writing – review and editing. **Sunil Gyawali:** conceptualization, data curation, validation, visualization, writing – original draft, writing – review and editing. **Suchit Thapa Chhetri:** conceptualization, data curation, validation, visualization, writing – original draft, writing – review and editing.

Consent

Written informed consent form was obtained from the patient to publish this report in accordance with the journal's consent policy.

Conflicts of Interest

The authors declare no conflicts of interest.

Data Availability Statement

All the findings are present within the manuscript.

Patient Perspective

The patient and her family members were anxious about her condition. They were properly counseled and assured that she would get better.

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