Guillain Barré Syndrome as Primary Presentation of Systemic Lupus Erythematosus (SLE-GBS) in a Teenage Girl: A Case Report

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Abstract

Background:

Peripheral nervous system (PNS) involvement, including Guillain Barré Syndrome (GBS), accounts for fewer than 10% of SLE cases with neuropsychiatric manifestations. GBS as the presenting, major manifestation of pediatric SLE is extremely rare, and the best treatment approach is unknown.

Case presentation:

A 14-year-old, previously healthy female Emirati teenager presented with a classic picture of GBS with ascending, progressive bilateral muscle weakness leading to respiratory insufficiency within five weeks of symptom onset, associated with typical protein-cell dissociation in cerebro-spinal fluid and nerve root enhancement demonstrated by spinal MRI. Subsequently, elevated anti-dsDNA and anti-Smith/RNP and anti-SS-A and -B antibody concentrations were detected in serum, suggestive of SLE. GBS treatment was initiated with IVIG and methylprednisolone pulses, with minimal improvement. The patient required endotracheal intubation and ventilation, followed by a second course of IVIG, rituximab, and eventually plasma exchange (PLEX) therapy. The diagnosis of lupus-associated GBS was corroborated by a kidney biopsy demonstrating lupus nephritis WHO class II with “full house” immunofluorescence pattern. After 14 PLEX sessions, her muscle strength and respiratory efforts had improved substantially. Treatment was completed with two more rituximab infusions, followed by mycophenolate mofetil, in addition to HCQ and tapering doses of oral prednisolone. Five weeks after the last PLEX treatment, she had regained her usual strength and achieved full, sustained recovery from GBS. While she continued to demonstrate moderate anti-dsDNA antibodies and high-level anti-Smith and Sjogren antibodies, C3, C4 and urine readings quickly normalized with no other manifestations of lupus or lupus nephritis 11 months after the initial assessment. At this time she was maintained with hydroxychloroquine, with ongoing depletion of circulating B cells. To our knowledge, this is only the third pediatric patient reported with SLE-GBS.

Conclusions:

We report severe GBS as the first, dominant manifestation of pediatric SLE. Our case and a review of the literature reveal that conventional GBS therapy may not be adequate to treat this rare lupus presentation. SLE should be included in the differential diagnosis of GBS. Importantly, treatment experiences and outcomes of such cases need be reported to inform future treatment recommendations.

Background

Systemic lupus erythematosus (SLE) is a systemic autoimmune disease with multi-organ involvement and substantial morbidity. Its etiology involves genetic, epigenetic and environmental factors. (1) The initial diagnosis of SLE can pose diagnostic challenges due to its variable presentation. Reported incidence rates of neuropsychiatric symptoms among children with SLE vary between 14% and 75%.(2) (3) Peripheral nervous system (PNS) involvement, including Guillain Barré Syndrome (GBS), accounts for

less than 10% of SLE cases with neuropsychiatric manifestations. (4)(3) It is rare that CNS symptoms precede the onset of typical features of SLE. (5) Among the few reported cases of SLE with GBS as initial presentation, only two were in the pediatric age group. (6, 7) The exact pathogenic mechanism remains unknown, and a consensus about the treatment of SLE-GBS has still to emerge. Here, we describe a third pediatric patient with such presentation. In this case, progressive, ascending muscle weakness led to the diagnosis of GBS while SLE was recognized later, on the basis of laboratory tests and renal biopsy. We also describe the multidisciplinary diagnostic and therapeutic approach which resulted in the full recovery of the patient.

Case Presentation

The patient, a 14-year-old previously healthy teenager, presented with a 4-week history of numbness and paresthesia in both legs, 10 days of disparate eye movements with subjective rightward-gaze diplopia, and 6–8 days of ascending, bilateral muscle weakness, more pronounced on the left. Two days prior to admission, she developed mild swallowing difficulties, temperatures of 38.1 C, and left-sided knee and hip pain. She had been non-ambulatory for 7 days when she presented to our Emergency Department.

The patient denied fever or immunization preceding the onset of neuromuscular symptoms. Specifically, there was no headache, no seizures, no rash, no diarrhea or vomiting, no signs of upper respiratory infection, rash, edema, joint pain, and no macroscopic hematuria. The family also denied changes in her mood, behavior or academic performance. No constipation or urinary retention was reported. She menstruated regularly, last three weeks prior to the hospitalization.

There was no history of sick contacts, including COVID-19, or recent travel outside the country. She was fully immunized according the national schedule, including polio and quadrivalent *N. meningitidis* vaccines.

Family history

Family history is significant for a paternal aunt with a presumptive diagnosis of SLE. Several members of the paternal family have hypothyroidism and G6PD deficiency. The patient's older sister has been treated for hyperthyroidism. There is type 2 diabetes mellitus and arterial hypertension in elderly members of the family. Consanguinity was denied.

Clinical examination

At the time of admission, the patient was bed-bound due to severe generalized weakness but was conscious and alert. She complained of diplopia and swallowing difficulty. Cranial nerve examination showed right eye convergent squint and bilateral gaze palsy, right more than left. The pupils were equal and reactive. There was no ptosis or facial muscle weakness, but she had bulbar symptoms and a weak gag reflex. Shoulder and arm movements were weak, corresponding to Medical Research Council (MRC) Muscle Scale 1–2/5. (8) Dorsiflexion at the wrist against gravity was 3/5, and the palmar grasp was

weak. The lower limbs also demonstrated significant proximal weakness with MRC scale of 1/5 at hip and knee joint movements. The ankle dorsiflexion and plantar flexion appeared less affected with MRC scale of 3–4/5. Overall, the left side appeared weaker than the right. She was very hypotonic, with total areflexia. There was no sensory impairment, no cerebellar signs, and no meningeal irritation. At presentation, she had no vasculitic lesions, no arthralgia, and no cardiac or other system abnormalities.

Laboratory findings

Initial laboratory findings included a mildly decreased absolute lymphocyte count, normal creatinine clearance (eGFR 144 mL/min/1.73m²), elevated erythrocyte sedimentation rate with only slightly elevated C-reactive protein (CRP), decreased serum levels of C3, C4 and CH50, and elevated antinuclear (ANA) and anti-dsDNA antibodies, and rheumatoid factor. (Table 1) Creatine kinase, acetylcholine receptor antibody, thyroid function and B12 levels were normal.

Table 1
Inflammatory Markers & Lupus Serology ^a

Test	Results	Reference range
Cerebrospinal Fluid Findings ^{b, c}		
Cell count	3 x 10 ⁶ / L	0–7 x 10 ⁶ / L
Total protein	63 mg/dL	0.0–44.0 mg/dL
IgG quant	22.90 mg/dL	0.0–8.6 mg/dL
Gamma globulin	27.5 %	3–13 %
M spike	negative	negative
Inflammatory Markers in Serum		
CRP	11.6 mg/L	< 5 mg/L
ESR	92 mm/h	< 15 mm/h
Fibrinogen	548 mg/dL	212–433 mg/dL
Ferritin	108 ng/mL	12–68 ng/mL
IL-1 beta	< 2.9 pg/mL	< 3.0 pg/mL
IL-10	20.7 pg/mL	3.7–23.3 pg/mL
TNF-alpha	4.0 pg/mL	0.0–2.2 pg/mL
IL-6	14.2 pg/mL	< 7 pg/mL
Lupus Serology		
ANA	1:1280 (speckled)	negative
anti-dsDNA antibody	11 IU/mL	0–4 IU/mL
Rheumatoid factor	79 IU/mL	≤ 13 IU/mL

^a Samples were obtained at presentation or during the first days of illness

^b CSF concentration of IgA was mildly elevated, IgM normal; alpha-1, alpha-2, and beta globulin, albumin and pre-albumin fractions were normal, as were the lactic acid and Cl⁻ concentrations.

^c Anti-MOG (myelin oligodendrocyte glycoprotein), NMDAR (N-methyl D-aspartate receptors) and NMO (Neuromyelitis optica)/AQP4 (aquaporin 4) IgG antibodies were undetectable

^d Urine dipstick

^e Quantitative urine protein creatinine ratio

Test		Results	Reference range
C3		43 mg/dL	83–193 mg/dL
C4		8.7 mg/dL	15–57 mg/dL
CH50		28 U/mL	> 41 U/mL
RNP antibody		> 8.0 AI	0.0–0.9 AI
Smith antibody		> 8.0 AI	0.0–0.9 AI
Scleroderma-70 antibody		< 0.2 AI	0.0–0.9 AI
Sjogren's SS-A (Ro) antibody		> 8.0 AI	0.0–0.9 AI
Sjogren's SS-B (La) antibody		> 8.0 AI	0.0–0.9 AI
Chromatin antibody		> 8.0 AI	0.0–0.9 AI
Ribosomal P antibody		0.6 AI	0.0–0.9 AI
anti-Jo-1		< 0.2 AI	0.0–0.9 AI
anti-GM1 (IgG)		6 %	0–30 %
Urine Studies			
Hematuria	Day 0	0–3 RBC/HPF	0–3 RBC/HPF
RBC microscopy	Day 13	15–20 RBC/HPF	
Proteinuria	Day 0	1+ (0.3 g/L) ^d	negative
	Day 14	2.20 g/g ^e	< 0.2 g/g
^a Samples were obtained at presentation or during the first days of illness			
^b CSF concentration of IgA was mildly elevated, IgM normal; alpha-1, alpha-2, and beta globulin, albumin and pre-albumin fractions were normal, as were the lactic acid and Cl [−] concentrations.			
^c Anti-MOG (myelin oligodendrocyte glycoprotein), NMDAR (N-methyl D-aspartate receptors) and NMO (Neuromyelitis optica)/AQP4 (aquaporin 4) IgG antibodies were undetectable			
^d Urine dipstick			
^e Quantitative urine protein creatinine ratio			

Subsequently, serum antibodies to extractable nuclear antigens (ENA), including ribonucleoprotein (RNP), chromatin, Smith and SS-A (Ro)/SS-B (La), were detected in high concentrations. The initial urine sample showed a small amount of protein. Hematuria and proteinuria peaked about 2 weeks after admission. (Table 1)

The laboratory results were suggestive of SLE. A kidney biopsy was performed on day 8 of admission; results were consistent with lupus nephritis WHO class II, with a “full house” immunofluorescence pattern. (9) (Fig. 1)

Cerebrospinal fluid analysis prior to treatment revealed protein-cell dissociation with an increased total protein concentration, mainly due to IgG and IgA and only 3×10^6 /L mononuclear cells (lymphocytes and monocytes). (Table 1) Sequential COVID-19 PCR tests over the first 2 ½ weeks and SARS-CoV-2 specific IgG antibody assays remained negative. CMV IgG was elevated, but CMV IgM, EBV serology and Mycoplasma IgM results were negative.

Imaging studies

MRI of the spine showed normal cord width, signal morphology and intensity, and no syrinx or central spinal canal widening. There was mild enhancement of nerve roots in the cervical and thoracic regions, and mild thickening of the cauda equina nerve roots. (Fig. 2)

EMG and nerve conduction velocity

The nerve conduction study from Day 25 of admission showed significant reduction in compound muscle action potential (CMAP) amplitude in all four limbs with mild reduction in conduction velocity. Sensory conduction was normal. Overall, the findings were suggestive of acute motor axonal neuropathy typical of Guillain-Barré Syndrome.

Clinical evolution and therapy

Based on the Brighton criteria, the patient had the highest level of diagnostic certainty for acute inflammatory demyelinating polyneuropathy (GBS). (10) Treatment was initiated with 2 g IVIG per kg body weight over two days without clinical improvement, followed by three methylprednisolone pulses. However, her muscle weakness progressed, leading to hypoventilation. Spirometry showed a decline in vital capacity (VC) over the preceding 48 h from 25 to about 15 mL/kg, prompting elective endotracheal intubation and ventilation on day 7 of admission. Due to the lack of improvement following methylprednisolone pulses, IVIG, and subsequently rituximab, we started plasma exchange (PLEX) therapy on day 14 of admission. (Fig. 3)

Tracheostomy was performed on day 20 of admission in anticipation of prolonged invasive ventilation. Post tracheostomy analgesedation was weaned completely allowing neurological assessment of the wake patient and initiation of rehabilitative therapies. Of note, the patient experienced a brief episode of hyperactive delirium post tracheostomy.

The disease course was complicated by labile blood pressure, mostly hypertension, likely due to central autonomous dysregulation. While intubated, she developed an uncomplicated, ventilator-associated *S. pneumoniae* pneumonia, diagnosed on day 9 of admission. The patient suffered from painful dysesthesia which improved with high-dose gabapentin and fentanyl boluses. Hair loss was noted during

the early course of the disease. She received low molecular weight heparin (enoxaparin) during the period of paralysis.

PLEX treatment consisted of 14 sessions over 22 days, where 1-1.5 times the plasma volume was replaced with 5% albumin and 0.5 L fresh frozen plasma to maintain physiological coagulation factor concentrations. After about 10 exchanges, the patient's muscle strength and respiratory efforts improved, and after the 12th session, we were able to gradually wean ventilator support.

The treatment was completed with two additional doses of rituximab on days 39 and 46 of admission. Peripheral CD19⁺ B cells was undetectable following the first rituximab infusion.

The patient was discharged home on Day 70 of admission with continued physiotherapy. At her first follow-up visit four days after discharge, she ambulated freely and performed all daily activities without assistance. Eleven months after discharge, the patient maintained normal muscle strength. She had been able to wean immunosuppression to hydroxychloroquine only. Peripheral CD19/20⁺ B cells were undetectable, and serum levels of C3, C4 and anti-dsDNA antibodies, renal function and urinalysis were all normal.

Discussion & Conclusions

SLE is a chronic autoimmune disease with multiple organ system involvement. The clinical spectrum is variable and includes cutaneous, musculoskeletal, renal and neuropsychiatric manifestations. The American College of Rheumatology (ACR) research committee described twelve central (CNS) and seven peripheral nervous system (PNS) manifestations (including GBS) of neuropsychiatric systemic lupus erythematosus (NPSLE) where other causes have been excluded.(11) GBS has been rarely reported as the initial presentation of SLE.(2, 7) For example, a large study from China comprising 4924 SLE patients identified a subset of 73 patients (1.5%) with peripheral neuropathy (SLE-PN), of whom a single patient presented with acute inflammatory demyelinating polyradiculoneuropathy (GBS).(12) A systematic review, summarizing data of 1463 children and adolescents with SLE, identified 351 patients with NPSLE, two of whom presented with GBS (0.6% of all NPSLE patients).(3, 5) No clinical details are available of those two cases.(5)

The pathophysiology of GBS in SLE remains speculative and is thought to involve lupus-induced auto-antibodies against myelin tissue.(13) In the absence of recommendations for the treatment of patients with SLE-GBS, glucocorticoids, cyclophosphamide, PLEX and IVIG have been used in different combinations. (14, 15) Current guidelines for classical pediatric (non-SLE) GBS emphasize the use of IVIG over PLEX. (16, 17) However, this approach may be inadequate for patients with SLE-GBS as inferred from available experience, including our patient.(4, 5)

The decision for tracheostomy in patients with severe GBS depends on the expected duration of respiratory failure, which may range from a few days to more than 6 months.(18) Early tracheostomy (19)

helps avoiding the drawbacks of long-term oral or nasal intubation and allows the patient to cooperate with functional assessment and physiotherapy.

We found only two previous descriptions of pediatric patients with SLE-GBS. Reddy et al reported a 9-year-old girl from India who presented in a manner similar to our patient. Treatment with IVIG, methylprednisolone pulses and IV cyclophosphamide failed to prevent progression to complete paralysis and intubation. She eventually improved after two doses of rituximab.(5) Javadi Parvaneh et al reported a 12-year-old Iranian boy with GBS symptoms, associated with the detection of anti-dsDNA antibodies.(6) He received high-dose IVIG with little improvement. Symptoms only resolved a few months after adequate lupus therapy with methylprednisolone pulses, hydroxychloroquine and IV cyclophosphamide was initiated. (6)

Our case highlights the importance of prompt recognition of SLE as a trigger of GBS, which changes conventional GBS management. Due to its rarity, however, no treatment recommendations are available for SLE-GBS. A rationale approach would target the underlying autoimmune disorder (SLE) and the removal of presumed pathological antibodies and/or suppression of the pathophysiological, inflammatory effects. B cell depleting agents and plasma exchange are potentially effective strategies. The dissemination of further experiences and studies is warranted to establish therapeutic efficacies and outcomes for children presenting with GBS in the context of SLE.

List Of Abbreviations

CNS	Central nervous system
CSF	Cerebrospinal fluid
GBS	Guillain Barré Syndrome
IVIG	Intravenous immunoglobulin
NPSLE	Neuropsychiatric systemic lupus erythematosus)
PLEX	Plasma exchange
PNS	Peripheral nervous system
SLE	Systemic lupus erythematosus
SLE-GBS	GBS due to SLE

Declarations

Ethics approval and consent to participate

Consent was obtained from the patient and her parents.

Availability of data and materials

The datasets generated and analysed during the current study are not publicly available due patient confidentiality reasons but are available from the corresponding author in anonymized form on reasonable request.

Competing interests

The authors declare that they have no competing interests

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Authors' contributions

E. Beshir participated in the care of the patient, collected data, reviewed the literature and wrote the manuscript. E. Belt supervised the medical treatment and revised the manuscript; NC and AA supervised and interpreted the neurological aspects of the diagnosis and treatment, and contributed to the manuscript; AI, MP and UT supervised the patient's treatment in the critical care unit and contributed to the manuscript; LH read and interpreted the kidney biopsy and provided the photographs; ES provided expert rheumatological advice; MB contributed to the patient's medical and renal care, reviewed and interpreted the clinical and laboratory data, and wrote and edited the manuscript. All authors critically reviewed the manuscript and agreed to the final version.

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Figures

Figure 1

Renal biopsy. A (Periodic Acid Schiff, 400x magnification) shows segmental mesangial hypercellularity (orange arrow) with greater than 3 mesangial cells in a mesangial area. Peripheral capillaries are patent and no lesions of endocapillary hypercellularity or crescent formation was seen in the biopsy. B shows the granular mesangial immune complex deposition that stained 2+ for IgG, IgA, C3, C1q, kappa and lambda (IgG is shown, 400x magnification). C and D highlight the electron microscopy findings. Electron dense deposits (red arrows) are present globally in mesangial areas but are not seen to involve the subepithelial or subendothelial distributions. Podocyte foot process effacement is mild.

Figure 2

Sagittal post-contrast T1 weighted MRI image of lumbo-sacral spine demonstrating cauda equina root enhancement (arrow)

Figure 3

Disease evolution and management. Abbreviations: ET endotracheal tube