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SYSTEMIC LUPUS ERYTHEMATOSUS PRESENTING WITH GUILLAIN–BARRE SYNDROME: A CASE REPORT

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ABSTRACT

We present a case of systemic lupus erythematosus (SLE) coexisting with Guillain–Barre syndrome (GBS), where GBS gradually progressed to SLE. A 32-year-old female with previously diagnosed SLE presented with symptoms suggestive of GBS, including leg pain, loss of balance, and lower extremity weakness. Prompt initiation of therapy for GBS and consideration of chronic inflammatory demyelinating polyneuropathy (CIDP) were crucial. This case underscores the significance of early diagnosis of autoimmune disorders to facilitate timely intervention and improve patient outcomes. **Introduction:** The co-occurrence of systemic lupus erythematosus (SLE) and Guillain–Barre syndrome (GBS) is rare. Here, we report a case where GBS preceded the manifestation of SLE, emphasizing the importance of vigilant monitoring and prompt intervention in autoimmune conditions. **Case Presentation:** We present a 32-year-old patient with SLE who presented with GBS. The patient came with progressively worsening neurological symptoms as the first manifestation. Leg pain, loss of balance, and lower extremity weakness were the reasons for her admission to the neurologic ward. The patient staarted the therapy based on the possibility of Guillain– Barre syndrome and chronic inflammatory demyelinating polyneuropathy (CIDP). **Conclusion:** This case highlights the significance of early recognition and management of autoimmune diseases.

Timely intervention not only aids in controlling disease progression but also contributes to better prognostic outcomes. **Data collection:** All the Data was collected from the patient's case sheet and the hospital laboratory medicine database.

KEYWORDS: Systemic Lupus Erythematosus, Guillain-Barre syndrome, chronic inflammatory demyelinating polyneuropathy, autoimmune disease.

INTRODUCTION

Systemic Lupus Erythematosus (SLE) is a multi-organ autoimmune disease, in which the immune system attacks its tissues, leading to widespread inflammation and tissue damage in the affected organs. SLE can affect joints, brain, skin, kidneys, lungs, and blood vessels. The neuropsychiatric syndrome in SLE affects both the central and peripheral nervous systems. The presentation of peripheral nervous system involvement in particular can be in the form of acute demyelinating polyradiculoneuropathy, Guillain–Barré Syndrome (GBS), mononeuropathy, polyneuropathy, myasthenia gravis, and others.[1,2] GBS is considered a rare and one of the least neuropsychiatric syndromes in SLE.[2,3] Guillain-Barré syndrome is sometimes denoted as GBS. It is a rare but serious autoimmune disorder, in which the immune system attacks healthy nerve cells in the peripheral nervous system (PNS). This syndrome leads to weakness, numbness, and tingling. It can eventually result in paralysis.[4] Chronic inflammatory demyelinating polyneuropathy (CIDP) is a rare neurological disorder that leads to inflammation of nerve roots and peripheral nerves and destruction of the fatty protective covering (myelin sheath) of the nerve fibers. Guillain-Barre syndrome (GBS) is a rare neurological disorder where the body's immune system mistakenly attacks its peripheral nerves. It often begins with weakness or tingling sensations in the legs and can progress to paralysis in severe cases. On the other hand, systemic lupus erythematosus (SLE) is an autoimmune disease that can affect various organs and tissues, causing inflammation and damage. While GBS is typically considered an acute and isolated condition, it has been reported as a rare presenting manifestation of SLE in some cases.[5] In most of the cases, Guillain-Barre syndrome (GBS) has rarely been reported with systemic lupus erythematosus (SLE). However, in the last 50 years in a few cases, it has been reported that the initial manifestation of SLE was Gullian–Barre syndrome (GBS). The first case was reported in 1964. 0.6% to 1.7% prevalence of SLE with GBS has been reported. Chronic inflammatory demyelinating polyneuropathy (CIDP) is a classic type of GBS and is very uncommon.[3, 5, 6]

CASE REPORT

A 32 yr old female of weight 69 kgs, height 158cm, IBW 56 kg, BMI 27.7 came on 25/1/2021 with complaints of difficulty in breathing for 3 days, low-grade fever, and pedal edema for 6 days. Was admitted to an outside hospital, and came here with a worsening SOB. Medical History: bedridden for 2 months, weakness for 2 months. HTN, Hypothyroidism, GB Syndrome, diffuse sensory-motor neuropathy, CIDP. Medication history: T TELMA AM BD, T THYRONORM 100 microgram OD. On immunosuppressants. Sudden onset of left upper limb weakness on 28/8/2020 with pain and tingling sensation advised HRCT [showed continuous lobular nodules in both lungs – Koch's etiology] after 15 days she noticed weakness in the right lower limb, difficulty in getting up from bed, took steroids and improved after a month, walking with person support. On 3/9/2020 generalized paraesthesia weakness in both distal and proximal Started steroids 500mg for 3 days. On 7/9/2020 partial response to steroids so dose changes to 1 gm for 2 days. Tab. Wysolone 6o mg 1 week then 4 mg 2 days then 20mg. on 29/9/2020 able to walk independently. Took gabapentin 300mg for paraesthesia. In Oct 2020, the patient was diagnosed with S/O AXONAL SENSORY MOTOR NEUROPATHY by ENMG. On 30/10/2020 it was confirmed that the patient has ADP, and CIDP by ENMG [axonal demyelinating sensory and motor radiculoneuropathy]. She experienced weakness again in her right lower limb in December for which she was been bedridden on 11/1/21 worsening of motor weakness took solumedrol 1gm for 3 days. On 11/1/21 prescribed tab. wysolone 60 mg 10 days then 4 mg 10 days sequential tapering. Surgical history: s/p LSCS in 2013.

Table 1: Power based on previous records.

ul	r	l
p	4	4
d	4	3

ll	r	l
p	4	4
d	4	4

On arrival physical examination showed that CNS was conscious, trying to obey commands. E4V4M6. The patient could sit, stand, and walk with assistance. CVS – in shock requiring norad support. Resp – requiring O2 support 15 L of o2 NRBM. Tachypnea RR – 30 /min. P/A – soft. Previous lab data shows Grbs – 236 mg/dl, Sr cr – 3.3, Tlc – 24, Pc – 1.6, Corad – 3, Tsh – 11.29. The patient as advised for HRCT chest, CT abdomen, CBP, RP 2, LFT, PT,

APTT, INR, CUE, Urine C/S, C Profile labs, Ca, Mg, phosphate, ET C/S secretion, RT PCR. The patient was admitted under critical care management, and required MV support, Invasive line placement was done, and cardiology consultation, Nephrology consultation, and neurology consultation were suggested. Stat doses of the following drugs were given Inj pan 40 mg od, Inj thiamine 500 mg iv stat then 200mg bd, Inj optineuron 2 amp iv od, Inj doxy 100 mg iv bd, Inj meropenem 2gm iv stat then 500 mg iv bd, Inj pcm 1gm iv sos, INJ LASIX IV STAT 20 MG, Noradrenaline 5 mcg/ml, Inj heparin 5000 units tid. She was shifted to a medical ICU. Sedation was done by considering her condition by using the following drugs Inj Fentanyl IV 20 microgram, Inj Atracurium, and Inj Noradrenaline.

Table 2: Power on admission includes.

ul	r	l
p	3	3
d	2	2

ll	r	l
p	3	3
d	2	2

HRCT results showed Ground glass opacities in both lung fields. Moderate bilateral pleural effusion with LUL collapse and basal segmental collapse. Cardiomegaly with pericardial effusion. Pulmonary edema. CT ICU B – mild bilateral perinephric standing with thickening of latent condition and posterior paracord fascia likely mild pyelonephritis. Mild ascites. 2D Echo – mild RA RV dilated.

Table 3: The lab data was as follows.

Date/Parameters	24/1	26/1	27/1	31/1	6/2	8/2
HB	7.5	7.4	5.9		10	9.5
TLC	27k	16.5k	8.8k		9K	6500
PLT	1.3	1.3	97000		2.1	2.2
ESR	120					
GRBS	183	228	168			81
BLOOD UREA	129	119	77		2.9	21
SR CR	4.19	3.33	2.39		0.34	0.3
CRP	2.4					
PCTQ	95.45					
TEMP		100^F	96.1^F			
Na	138	139	138		139	140
K	4.8	4.7	4.0		3.5	3.0
Cl	105	102	106		102	102
SGOT	21					

SGPT	13					
T PROTEIN	5.6					
APTT	22				25.2	
Ca				8.3		
Mg				2.2		
Phosphate				3.8		

A final diagnosis was made and it was noted that the patient had SLE-> vasculitis + myocarditis + neuropathy + nephropathy(multiorgan involvement), Gullian barre syndrome, CIDP, Urosepsis, AKI, Shock. The CIDP, a variant of GBS, proceeded to SLE. In our patient, the diagnosis of systemic lupus erythematosus (SLE) was confirmed due to renal involvement, anemia, positive ANA, and positive ds-DNA antibodies, fulfilling at least four of the eleven criteria established by the American College of Rheumatology for diagnosing SLE. The patient was advised intravenous Ig 0.4 mg/kg/day maltose-based for 5 days after NCS. On the 3rd day of admission, MRI showed spinal atrophy, TB test was –ve, ET – yeast cells, pseudohyphae candidiasis, Urine culture showed enterococcus, AFB was -ve. Patient undergoing sustained low-efficiency dialysis for AKI. On the 4th day, she was started with IV Ig 30 iv od (33 ml/hr) and after 2 doses planned to taper it to 20 gm for 3 doses of OD. T BETALOC 12.5 mg BD was started as the patient experienced tachycardia. A renal biopsy was done on the 12th day of admission which indicated nephritis. Developed Oral ulcers on the 13th day on admission and was managed with acyclovir ointment (has opportunistic infections oral candidiasis or herpes labialis). Due to the increase in WBC count, the teicoplanin dose was increased from 200mg to 400mg. T BETALOC 12.5 mg BD was added for increased BP and tachycardia. There were fluctuations in potassium levels which were managed with IV KCl and syrup kesol. A Movicol sachet was given for abnormal phosphate levels. As the patient was anemic, a blood transfusion was done. The patient was treated with hydroxychloroquine, methylprednisolone, and IV Ig successfully. The patient was given a regular discharge on the 16th day of admission and was asked for a follow up after 7 days with CBP, renal function, liver function, and lipid profile tests. The following discharge medications were prescribed: SYP KESOL 15ML 3 DAYS BD check sr potassium after 3 days, T THYRONORM PO OD 100MCG, T OPTINEURON 7 DAYS 1 TAB PO OD, T PAN PO OD, OINTMENT ACYCLOVIR TID, T WYSOLONE 60 MG OD, T BETALOC 12.5 MG PO BD, T CINOD 5 MG PO BD, T RIBOFLAVIN 1 TAB FOR 10 DAYS, T HCQ 200 MG OD, ORAL HYDRATION, ENSURE PLUS POWDER 3 scoops in 100ml water TID.

DISCUSSION

Guillain-Barré syndrome (GBS) is characterized by an acute peripheral neuropathy that causes limb weakness, progressing over days to a maximum of four weeks. It is often triggered by bacterial or viral infections.[7,8] The exact cause of Guillain-Barré syndrome (GBS) is unknown. Diagnosis is based on clinical and electrophysiological criteria, including limb weakness that progresses symmetrically, flaccid paralysis, sensory loss in a "socks and gloves" distribution, cerebrospinal fluid analysis showing elevated protein levels without an increase in cell count, and electromyography revealing signs of demyelinating polyneuropathy.[6] Other diagnostic features are signs of polyneuropathy or polyradiculopathy in nerve conduction studies.[24,25] This patient showed the presence of renal involvement, anemia, positive ANA, and positive ds-DNA antibodies, meeting the criteria for a diagnosis of SLE. In addition to these findings, the renal biopsy identified Lupus nephritis (LN). The subtypes of GBS include Miller-Fisher, acute inflammatory demyelinating polyneuropathy (AIDP), acute motor and sensory axonal neuropathy (AMSAN), and acute motor axonal neuropathy (AMAN). AIDP is the most common form of GBS in Western countries and it accounts for 85–90% of the patients with GBS.[9] Only a few case reports presented Guillain-Barré syndrome as the initial presentation of SLE.[10,11] Systemic Lupus Erythematosus is a multifactorial autoimmune systemic disease. Causal factors of SLE include genetics, sex hormones, environment, infection, drugs, and food. It is seen most commonly in females.[12,13,14] Nervous system involvement in SLE is known as neuropsychiatric lupus erythematosus. As in this patient, we have chronic inflammatory demyelinating polyneuropathy, a variant of GBS, which involves central and peripheral nervous system involvement. Common presentations include cerebrovascular accidents, seizures, headaches, psychosis, and depression. Neuropsychiatric manifestations of lupus may precede the onset of lupus and may occur at the time of diagnosis or later in the course of the disease.[13,15]

Peripheral nervous system involvement is observed in over 10% of all nervous system manifestations in systemic lupus erythematosus (SLE).[16] The prevalence of SLE with Guillain-Barré syndrome (GBS) has been reported to be between 0.6% and 1.7%.[17,18] The pathogenesis of GBS as a manifestation of active SLE is idiopathic, but it is believed that both cell-mediated and humoral processes may play a significant role.[19] Studies suggest that vascular occlusion in systemic lupus erythematosus (SLE) may result from vascular endothelial hyperplasia, occlusive endometrial fibrosis, thrombosis, and other non-inflammatory vascular lesions. Vascular occlusion is primarily caused by damage to the lupus

erythematosus mental system in focal regions, involving autoantibodies. There are four main types of anti-neuronal antibodies: anti-lymphocyte antibodies, antiphospholipid antibodies (including cardiolipin antibodies and lupus anticoagulants), and anti-ribosomal P protein antibodies. These antibodies are often found in the plasma and cerebrospinal fluid, leading to extensive neurological damage. Additionally, cytokines such as α-interferon and interleukin-6 are believed to play a significant role in the pathogenesis.[6] According to previous literature cyclophosphamide, plasmapheresis, corticosteroid, and immunoglobulin have been considered to be useful in CIDP associated with SLE. Anyhow the combination of corticosteroids and cyclophosphamide is considered as the first-line therapy.[20,21,22,24,25] In many cases the treatment of GBS in SLE was effective using IV immunoglobulin and plasmapheresis.[16,23]

CONCLUSION

Guillain-Barre syndrome (GBS) as a rare manifestation of systemic lupus erythematosus (SLE) presents a clinical challenge, as evidenced by the limited supporting clinical trials. Despite this, literature reports offer substantial evidence for such cases. Here, we present a case of a patient with concurrent SLE and chronic inflammatory demyelinating polyneuropathy (CIDP) [GBS], whose condition significantly improved with intravenous immunoglobulin (IVIG), hydroxychloroquine, and corticosteroids. Our study suggests that early administration of hormones and immunosuppressants can be effective in treating these conditions. Successful management of autoimmune diseases hinges on appropriate therapy. However, further clinical research is needed to fully understand the pathogenesis of both conditions.

In summary, although rare, GBS as a manifestation of SLE warrants clinical attention, supported by existing literature. The successful management of such cases underscores the need for tailored treatment strategies and points to avenues for future research to deepen our understanding of these complex autoimmune diseases and their complications.

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