Systemic lupus erythematosus with acute inflammatory demyelinating polyneuropathy: a case report

Genga EK¹, ², Otieno FO³, Mativo PM⁴, Oyoo GO¹, ²

Abstract

We recently managed a case of acute inflammatory demyelinating polyneuropathy associated with SLE. A 20-year-old newly diagnosed SLE patient presented with a three-week history of acute bilateral ascending weakness associated with inability to walk. Physical examination revealed muscle strength in the legs with graded 2/5 proximally and 2/5 distally bilaterally and absence of deep tendon reflex in both knees and ankles. The muscle strength in upper limb was 3/5 proximally and 3/5 distally bilaterally. Paresthesia was observed in distal limbs with glove and stocking distribution. Cerebrospinal fluid analysis was normal. Electrophysiologic survey indicated asymmetrical mixed sensory motor demyelination and radiculopathy. The diagnosis of SLE was established based on her initial symptoms including fevers, fatigue, malar rash, myalgia, and positive ANA. Treatment with intravenous immunoglobulin and methylprednisolone resulted in clinical improvement.

Key words: Systemic lupus erythematosus, Acute inflammatory demyelinating polyneuropathy, Guillain-Barre syndrome

Introduction

Systemic Lupus Erythematous (SLE) is a chronic, inflammatory, autoimmune disease characterized by multisystemic involvement with a myriad of clinical presentations. Neurologic complications are common and frequent in SLE. Central Nervous System (CNS) involvement is one of the more common complications that can occur at any stage of the SLE. These symptoms may precede the onset of SLE or can occur at any time during the course of SLE¹. Peripheral nervous system involvement occurs in 3–18%². Here we report a patient with AIDP that was associated with SLE.
sensory motor demyelination and radiculopathy. A pulse-dose methylprednisolone was initiated while hydroxychloroquine was continued while awaiting the CSF and EMG results. Guillain Barre Syndrome (GBS) was diagnosed on the basis of clinical symptoms, EMG, and lumbar puncture. Further GBS workup, such as antiganglioside antibodies were not performed. The patient was started on intravenous immunoglobulins (IVIG) for 5 days. Motor strength and facial weakness improved during the course of therapy. From SLE standpoint, she was maintained on hydroxychloroquine and azathioprine and remained in clinical and serological remission.

Discussion

Neuropsychiatric lupus (NPSLE) is one of the least understood yet possibly could be one of the most prevalent manifestations of lupus. It can occur independently of active systemic disease and without serologic activity. The numbers affected range from 14% to over 80% in adults and 22% to 95% in children. The American College of Rheumatology established 19 specific neuropsychiatric syndromes case definitions from two broad categories: central and peripheral manifestations. Common presentations include seizures, depression, and psychosis, headaches and cerebrovascular accidents. Peripheral neuropathy is an often-underestimated complication in SLE. The incidences range from 1.5% to 27.8%. Guillain Barre Syndrome which is a manifestation peripheral neuropathy in SLE is rare with incidences reported to be 0.6-1.7%.

Li et al reported that GBS with SLE was more common in females (73.3%) than males (26.7%). Our patient was female. GBS manifested early in the course of lupus which was consistent with the previously reported case reports. We suspect the trigger for the GBS was the flu-like symptoms she had prior to the onset of the illness. This is consistent with what is reported in literature that up to two thirds of cases are preceded by symptoms of upper respiratory tract infection or diarrhoea. The most frequently associated infectious agent being Campylobacter jejuni (30%). Others include cytomegalovirus, Epstein–Barr virus, varicella–zoster virus, and Mycoplasma pneumoniae. Autoantibody formation against gangliosides as part of immunological response in SLE can potentially elicit demyelinating polyneuropathy such as GBS. Elevated proinflammatory cytokines such as interleukin-6 and interleukin-8 have been found in patients with SLE with neurological symptoms. We did not perform antiganglioside antibodies in our patient due to cost implications. The third potential trigger of GBS like response in SLE is vascular including vasculitis, microangiopathy, and premature atherosclerosis leading to ischemic demyelination. The b2microglobulin, antiphospholipid and lupus anti-coagulant were negative in this case. The last triggers involve host specific factors such as genetics or ethnicity and environmental. Further research is required to elucidate the underlying reasons for GBS with SLE.

As GBS in SLE is rare controlled clinical trials are largely lacking which results in various non-standardized treatment regimens. The treatment options available for GBS with SLE, include corticosteroids, cyclophosphamide, plasmapheresis and immunoglobulin. Although clinical trials have demonstrated no benefits of corticosteroids in GBS, it’s still the most frequent treatment option for neuropsychiatric manifestations of SLE. This regime wasn’t successful in our patient. Combination with cyclophosphamide may have had better results. This regime has been shown to have improved the overall outcome in patients with SLE where GBS was the initial presentation. Due to her being in the reproductive age we opted for IVIG which has demonstrated efficacy against GBS and is the first line therapy along with plasmapheresis. The exact mechanism of action of IVIG in GBS is unknown. Its proposed mechanism involves antagonization of circulating pathological antibodies by anti-idiotypic antibodies, modulation of cell-mediated immunity and complement pathways. The present patient received IVIG for treatment of GBS in the background of SLE and responded well.

Conclusion

There is a rare association between GBS with lupus probably due to an immunological aetiology. This may have an impact on both treatment and prognosis. This is strongly evidenced by reported literatures, which is translated into decisions for their management and impact on long-term outcomes. Our case suggests prompt diagnosis and treatment, early in the course of illness can result in a positive clinical outcome. It also supports corticosteroids alone does not alter the course of GBS and that IVIG should be considered as first line therapy for GBS associated with SLE.

Conflicts of Interest

No potential conflict of interest relevant to this article was reported.

References


