Guillain barre syndrome as initial presentation of systemic lupus erythematosus

Courage U Uhunmwangho[1], Esala A Abene[2], Zumnan M Gimba[2], Godwin Osaigbovo[3], Emmanuel I Agaba[2], Oche O Agbaji[2]

Abstract

Background: Systemic Lupus Erythematosus (SLE) is an autoimmune disease characterized by multiple organ involvement including the peripheral nervous system. Guillan-Barrè syndrome (GBS) has an established association with SLE as one of its neurologic manifestations. However, GBS as an initial manifestation of SLE is only sparingly reported in the literature.
Methods: We present a case of a 26 year old woman who was initially managed by neurology unit for GBS but subsequently developed body swelling, hypertension and polyarthritis. She was evaluated and managed by both rheumatology and nephrology units.
Results: Renal biopsy showed histological features of membranous nephropathy suggestive of stage V lupus nephritis. The patient responded well to high dose corticosteroid, intravenous cyclophosphamide and azathioprine.
Conclusion: This case highlights that GBS can be an initial presentation of SLE and the usefulness of immunosuppressants in the management of such cases.

Keywords: Guillan-Barrè syndrome, Systemic Lupus Erythematosus, Lupus Nephritis, Immunosuppressants

Highland Med Res J 2015;15(2):96-98

Introduction

Guillain Barre Syndrome has an established association with systemic lupus erythematosus (SLE) as one of the neurologic manifestations of lupus, however only in a few cases has it been reported as the initial presenting complaint.[2.] We here report a 26-year-old lady who presented with acute ascending paralysis with clinical features of GBS but in the course of management developed other features of SLE with serological correlates.

Case Report

A 26 year old housewife presented with a three months' history of recurrent fever and two months of lower limb weakness, numbness and inability to walk. Numbness and weakness were said to have started from the toes and progressively involved the entire lower limb till she could not walk. She also had numbness on both hands. There was no history of trauma or fever preceding the onset of the illness. A physical examination revealed symmetrical quadriparesis, with impaired deep tendon reflexes. All other systems were essentially normal. Hepatitis B and C viral screen were negative. She was unreactive to HIV antibodies. She had a random blood glucose of 6. 8mmol/L, hematocrit of 37%, white cell count of 6.54 x 10^9, and an ESR of 26 mm in the first hour. There was no proteinuria on dipstix examination of the urine. A cerebrospinal fluid (CSF) analysis showed less than 5 cells/ml but the protein was 1,080mg/dl. Nerve conduction tests were not available. The diagnosis of Guillain Barre Syndrome was made on the basis of clinical features and CSF albuminocytologic dissociation. She was commenced on physiotherapy, multivitamin containing thiamine, pyridoxine and cyanocobalamin and low dose prednisolone 5 mg later escalated to 40 mg daily and gradually tapered off over six weeks. At discharge she was able to walk without support but had a foot drop having spent a month on admission.

While on outpatient follow up, she complained of recurrent joint pain and stiffness, body swelling with frothy urine. She had facial puffiness, bilateral pitting leg edema, hypertension (BP 160/100mmHg) and polyarthritis. Proteinuria was 3+ on dipstix with a urine protein creatinine ratio (UPCR in mg/mg) of 3.4. Serum creatinine was 39 umol/L and ESR was 145mm/hr. The speckled pattern Anti-nuclear antibodiy (ANA) and anti-double stranded DNA (dsDNA) antibodies were positive with normal C3 and C4 complements. A diagnosis of lupus nephritis was made. Renal biopsy showed histological features of membranous nephropathy suggestive of stage V lupus nephritis.

She had a 3-day course of pulse methyl prednisolone

[1]Rheumatology Division, Department of Medicine, Jos University Teaching Hospital, Jos, Nigeria [2]Nephrology Division, Department of Medicine, Jos University Teaching Hospital, Jos [3]Neurology Division, Department of Medicine, Jos University Teaching Hospital, Jos

All correspondences to:
Dr. Uhunmwangho U. Courage Rheumatology Division, Department of Medicine Jos University Teaching Hospital, P.M.B. 2076, Jos. Plateau State, Nigeria
E-mail: ucourage@yahoo.com

subsequently converted to high dose oral prednisolone 60 mg daily and followed by six courses of intravenous cyclophosphamide over three months and maintained on Azathioprine 50mg daily. She was also commenced on hydroxychloroquine 200mg twice daily and Lisinopril 20 mg daily. She made significant progress, with full recovery of muscle power, gait and renal function. All arthritides and body swelling also resolved completely and ESR dropped to 4 mm in the first hour at the last follow up.

Discussion

Systemic lupus erythematosus is an autoimmune disease characterized by multiple organ involvement. Central nervous system involvement is more common than that of the peripheral nervous system[1]. Peripheral nervous system involvement is dominated by distal symmetric axonal polyneuropathy and multiple mononeuropathy, which usually occurs late in the disease, but rarely present as the initial manifestation. [2,3]

The combination of rapidly progressive symmetrical weakness in the arms and legs with or without sensory disturbances, hyporeflexia or areflexia, in the absence of a CSF cellular reaction (albuminocytologic dissociation) remains the hallmark of the clinical diagnosis of GBS[4], all of which were present in our patient leading to the initial diagnosis of GBS in her. However, in the course of evaluation and management, the patient was found with polyarthritis, antinuclear antibodies and anti-dsDNA antibodies as well as pathological evidence of membranous nephropathy representing conclusive evidence that the patient had SLE.

Neurological manifestations are well recognized classification features of SLE. However most of the common features are of the central nervous system found in 24-51% of cases whereas peripheral neuropathy is only recognized in 10-21% of cases[3]. The pattern of peripheral nervous system involvement seen in SLE patients can be grouped into three viz: symmetrical distal sensorimotor neuropathy; Guillain Barré syndrome (GBS) and mononeuritis multiplex[5].

GBS patients have been reported to have an SLE prevalence rate of 0.6-1.7%, described mainly in the course of systemic illness[6] while the global incidence of GBS is put at about 0.6-4 cases per 100,000. Most of these cases are related to a preceding history of infection with Campylobacter jejuni, while their association with active SLE is considered to be extremely rare[6]

The mechanism of GBS in patients with SLE is not well understood. It has been proposed that autoantibodies that react with specific neural tissues, such as myelin components, may be produced as part of the wide spectrum of autoantibodies found in SLE[7]. Complement fixing antibodies to nerve and kidney in patients with GBS and membranous glomerulonephritis, point to a common pathogenic relationship. The increased susceptibility to infection due to immunosuppression resulting from both the activity of their disease and the immunosuppressive therapy may render SLE patients susceptible to GBS[7]. However, in our patient, GBS predated the diagnosis of SLE.

The main treatment modalities with proven efficacy in the management of GBS include plasma exchange and intravenous immunoglobulin (IVIG) [8]. The optimal treatment modality for GBS in the setting of SLE has not been clearly established[9]. However, high dose corticosteroid have been found to be very useful in the treatment of SLE associated GBS even in cases where traditional GBS treatments like plasmapheresis and intravenous immunoglobulin (IVIG) failed[7,9]. Our patient showed significant improvement in neuropathy to high doses of corticosteroid, the effect was further enhanced by the addition of other lupus treatments including hydroxychloroquine, intravenous cyclophosphamide and oral azathioprine. The extent to which these drugs may have contributed to her complete neurological recovery is uncertain. Even though glucocorticoids are not recommended for treatment of adults with GBS, it is well recognized as a standard therapeutic modality in patients with SLE who present with neuropsychiatric manifestations[10]. The association between SLE and GBS is well established in the literature; however the rare occurrence of GBS as an initial presentation of SLE which has been sparingly reported[2] should be borne in mind in the evaluation of patients with GBS.

References

1. Yildiz OK, Balaban H, Senel S, Cevik S. Acute lumbosacral polyradiculopathy heralding transformation to systemic lupus erythematosus in a patient with discoid lupus. Lupus 2011; 20: 972-974.
2. Ait Benhaddou E, Birouk N, El Alaoui-Faris M, et al. Acute Guillain-Barre-like polyradiculoneuritis revealing acute systemic lupus erythematosus: two case studies and review of the literature. Rev Neurol (Paris). 2003;159:300-306.
3. Matsuki Y, Hidaka T, Matsumoto M, Fukushima K, Suzuki K. Systemic lupus erythematosus demonstrating serum anti-GM1 antibody, with sudden onset of drop foot as the initial presentation. Intern Med. 1999;38:729-732.
4. van Doorn PA, Ruts L, Jacobs BC. Clinical features, pathogenesis, and treatment of Guillain-Barre syndrome. Lancet Neurol. 2008;7:939-950.
5. Baharadwaj A, Shenoy D, Aggarwal A. Polyradiculopathy in a patient with Systemic lupus erythematosus. J Assoc Physician India 2003; 51:304-305.
6. Korn-Lubetzki I, Abramsky O. acute and chronic demyelinating inflammatory polyradiculopathy. Association with autoimmune diseases and Lymphocyte response to human neuritogenic protein. Arch Neurol

1986; 43: 604-608.
7. van Laarhoven HWM, Rooyer Fergus A, van Englen BGM, van Dalen R, Berden JHM. Guillain-Barré syndrome as a feature in a patient with lupus nephritis, with complete resolution after cyclophosphamide treatment. Nephrol Dial Transplant 2001; 16: 840-842.
8. Hughes RA, Wijdick EF, Barohn R, et al. Quality Standards Subcommittee of the American Academy of Neurology. Practice parameter: Immunotherapy for Guillain-Barré syndrome; Report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology 2003; 61: 736-740
9. Santiago-Casas Y, Peredo RA, Vila LM. Efficacy of low-dose intravenous cyclophosphamide in systemic lupus erythematosus presenting with Guillaine-Barré syndrome-like acute axonal neuropathies: report of two cases. Lupus 2013; 22: 324-327.
10. Aranow C, Ginzler EM. Treatment of Non-renal lupus. In: Hochberg MC, Silman AJ, Smolen JS, Weinblatt ME, Weisman MH (eds), Rheumatology, 4th ed. Philadephia: Mosby/Elsevier, 2008. P.1309-1318.