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Case Report

Guillain-Barre Syndrome as the Presenting Feature in a Patient with SLE

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Abstract

Various neurological features have been reported in association with Systemic Lupus Erythematosus (SLE) however, Guillaine Barre Syndrome (GBS) as a presenting feature of SLE appears to be rare. We report a patient presenting with GBS in whom lupus nephritis was subsequently diagnosed. Patient was treated with steroids and cyclophosphamide which resulted in improvement of both her neurological status and proteinuria.

Keywords: Guillaine barre syndrome; Systemic lupus erythematosus; Lupus nephritis

Introduction

The involvement of the peripheral nervous system in systemic lupus erythematosus is rare and is dominated by symmetric axonal polyneuropathy and multiple mononeuropathies [1].

Acute inflammatory demyelinating polyneuropathy presenting as the initial manifestation of SLE is rather rare. The precise mechanism of SLE related AIDP remains unclear but is probably immune related. Although steroids are not recommended in the management of AIDP or GBS, patients with SLE related AIDP may benefit from steroid therapy [2].

Case Presentation

A 17 year old female, resident of Bara Kahu, Islamabad presented with history of tingling and numbness of feet 4 months back with progressively increasing weakness of lower limbs followed by upper limbs. 2 months later, she developed generalized body swelling. There was no other significant past medical history. Physical examination on admission showed BP of 130/85 mmHg and edema. Neurological examination revealed power of 1/5 in lower limbs and 3/5 in upper limbs. Tone decreased bilaterally with absent reflexes.

Laboratory examination showed; urinalysis: +++, Hb: 11 g/dl, Platelets: 130,000, ESR: 42 mm/hr, 24 hrs proteinuria: 3126 mg, albumin: 1.8 g/dl, cholesterol 369 mg/dl, triglycerides 285 mg/dl. On chest X-ray, bilateral pleural effusions with cardiac enlargement were seen. Echocardiogram showed pericardial effusion. Lumbar puncture revealed normal cell count with raised protein levels. Oligoclonal immunoglobulin bands were absent. Nerve conduction studies showed the findings of acute demyelinating polyneuropathy. Because of the clinical picture of rapid progression of symmetric proximal limb weakness to total paralysis in 2 weeks, associated with Areflexia, diagnosis of GBS was made. Retrospectively patient also fulfilled the American college of rheumatology case definitions for GBS.

Because of proteinuria with active urinary sediment, additional investigations were performed, including autoimmune serology. This revealed a 3+ Antinuclear Antibody (ANA) test and positive anti-dsDNA, titer 72 (pos>25), ANCA and anti-cardiolipin antibodies were negative and low C3 0.69 (0.83 g/l to 1.93 g/l) and C4 0.07 (0.15 g/l to 0.57 g/l).

Based on the presence of pleural, pericardial effusion and ascites, proteinuria with cellular casts, thrombocytopenia, positive ANA and anti dsDNA, the diagnosis of SLE with renal involvement was made, according to the ACR criteria. Renal biopsy was not performed as patient was not stable enough and attendants did not give consent.

During hospital stay, patient was treated with IV diuretics and Methyl prednisolone 60 mg followed by oral steroids (1 mg/kg body weight). Intravenous pulse of cyclophosphamide 750 mg/m^2 given. Prednisolone 60 mg per day given for 4 weeks and thereafter monthly tapered off to a maintenance dose of 10 mg.

On follow-up, patient's condition improved with decreasing body swelling, proteinuria and improved muscle strength, with power of 3/5 in upper limbs and 4/5 in lower limbs.

Discussion

We shall discuss whether there is any evidence for a true association between GBS and SLE and consider possible immunopathological mechanisms by which the two conditions might be linked.

The prevalence of SLE in patients with GBS has been reported to be between 0.6% to 1.7% [3]. The clinical course may be acute, resembling GBS [4] or chronic, resembling chronic inflammatory demyelinating polyneuropathy [5]. Prospective series of 50 patients with SLE showed only 1 to have GBS [6]. A retrospective series of 11,000 patients of GBS showed seven patients to have SLE, and a total of 14 to have connective tissue disease [7]. These data strongly suggest that there is a 'real' association between SLE and inflammatory demyelinating polyneuropathy, although the two conditions co-exist only rarely.

Possible mechanisms of an association

There is evidence for both cell-mediated and humoral processed

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in inflammatory demyelinating polyneuropathies [8]. One possible explanation for development of inflammatory polyneuropathy in SLE, is that the disease develops as a consequence of concurrent infection. Patients with SLE are immunocompromised by both their disease and its treatment and are thus susceptible to infection.

There are two other possible mechanism which might explain the association of the two condition: (1) auto antibodies which react specifically with neural tissue (directed, e.g., against myelin components, gangliosides or neuro filaments) may be produced as part of the wide spectrum of auto reactive antibodies which typify the condition; (2) immunological cross reactivity between one of the many other auto antibodies to cell surface or nuclear antigens occurring in SLE, and antigens present on neural tissue. Antibodies showing 90% cross reactivity with lymphocytes and culture neuronal cells occur in cerebral lupus [9].

Several observations in this patient suggest that GBS developed as a feature of lupus; the synchronicity of clinical symptoms of GBS and lupus nephritis, the positive effect of treatment of lupus also on GBS.

Therefore, the association of GBS with lupus seems to have implications for both treatment and prognosis. Prednisolone and cyclophosphamide should be considered in patients with GBS as a feature of lupus. Furthermore, we propose that in patients with GBS, not responding to intravenous gamma globulins, lupus should exclude.

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