

CASE REPORTS

Guillain–Barre Syndrome as Presenting Feature in a Patient with Systemic Lupus Erythematosus

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Abstract

Guillain–Barré syndrome. GBS as initial manifestation of lupus is exceedingly rare and has been reported in a few cases in the literature. We report here a 35 year old woman who presented with 10 day history of progressive muscle weakness and paraesthesias in all four limbs. She was diagnosed as SLE with renal involvement and was treated with steroids and cyclophosphamide.

Introduction

Various neurological features have been reported in association with systemic lupus erythematosus (SLE). However, Guillain–Barré syndrome (GBS) as a presenting feature of SLE appears is rare.¹ We report a patient presenting with GBS, in whom lupus nephritis also diagnosed. The GBS failed to respond to intravenous immunoglobulin treatment, but both GBS and lupus nephritis responded very favourably to intravenous pulses of cyclophosphamide and methyl prednisolone. GBS as initial manifestation of lupus is exceedingly rare and has been reported in a few cases in the literature.^{3,4} GBS in lupus is a complex and poorly understood phenomenon.

Case

A 35-year-old woman was referred to our hospital because of a 10-day history of progressive muscle weakness of arms and legs and paraesthesias in both hands and feet. Her medical history was unremarkable. Physical examination on admission revealed a blood pressure of 130/90 mmHg. Neurological examination showed absent deep tendon reflexes, and severe symmetrical proximal muscle weakness. Laboratory examination revealed haemoglobin 6.5 gram%, an erythrocyte sedimentation rate of 84 mm/h; C-reactive protein 71 mg/l; urea 12.3 mmol/l; creatinine 150 µmol/l; serum albumin 3 gram/dL; LDH 434 U/l; Coombs-positive haemolytic

anaemia; but normal thrombocyte and leukocyte counts. Qualitative urine analysis revealed the presence of protein (3 plus). Later, also active urinary sediment was found (10–25 leukocytes, hyaline and coarse granular casts). The cerebrospinal fluid revealed a normal cell count and total protein. Oligoclonal immunoglobulin bands were absent.

The nerve conduction study (day 1) showed loss of F responses, and decreased amplitudes of sensory nerve action potentials and compound muscle action potentials in arms and legs. Because of the clinical picture of rapid progression of symmetric proximal limb weakness to total paralysis in 2 weeks, associated with areflexia, the diagnosis GBS, was made according to the Asbury criteria.⁴ Retrospectively the patient also fulfilled the American College of Rheumatology (ACR) case definitions for GBS published in 1999.

On day 1 treatment was started with intravenous immunoglobulin G (2 g/kg bodyweight per day, for 5 days. Muscle weakness rapidly progressed to a quadriparesis.

Because of proteinuria with active urinary sediment, additional investigations were performed, including autoimmune serology. This revealed a 3+ positive antinuclear antibody (ANA) test, anti-dsDNA positive, but no antineutrophil cytoplasmic antibodies (ANCA), or anticardiolipin autoantibodies, and decreased C3 and C4 values. Based on the presence of proteinuria with cellular

casts, autoimmune haemolytic anaemia, a positive ANA and the presence of anti-DNA auto-antibodies the diagnosis SLE with renal involvement was made, according to the ACR criteria.

USG guided renal biopsy was done, which showed glomeruli with mesangial and endocapillary proliferation. Focally, glomeruli showed crescent formation and splitting of the glomerular basement membrane. The biopsy findings were classified as a mesangiocapillary lupus nephritis with a membranous component (WHO class IV.C) with a NIH activity index of 10 and a chronicity index of 7.

Because of this severe form of lupus nephritis the patient was treated with intravenous pulses of cyclophosphamide (750 mg/m²) and high oral doses of prednisone (1 mg/kg body weight).² In the first 6 months the patient was treated with monthly pulses of cyclophosphamide 750 mg/m², together with intravenous hydration and MESNA (2-mercapto-ethane-sulphonate) to reduce the urothelial toxicity of cyclophosphamide. In addition, prednisone 60 mg/day was given for 4 weeks and thereafter monthly tapered to a maintenance dose of 10 mg. intravenous immunoglobulin was not given.

This treatment regimen was tolerated well. Muscle strength improved, first proximally, then distally. By that time the proteinuria had decreased to 3.7 g/24 h and renal function had become normal. Six months after the onset of the GBS the patient was in an excellent condition, muscle strength and tendon reflexes were normal without any physical restraints. Twenty-four-hour protein excretion was 150 mg and the complement C3 and

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C4 levels were normal. From then on the patient received cyclophosphamide pulses every 3 months for a total period of 18 months.

Discussion

The signs and symptoms in the presented patient fulfilled the Asbury criteria⁴ and ACR case definitions for the diagnosis of GBS. The patient was treated with cyclophosphamide pulses in combination with oral steroids.

The prevalence of SLE in patients with GBS has been reported to be between 0.6 and 1.7%.³ For the combination of GBS and lupus nephritis the efficacy of prednisone alone for treatment

of the neuropathy is insufficient in about 50% of the cases.² The efficacy of intravenous immunoglobulin for GBS in lupus nephritis is controversial.² Plasma-exchange may have a beneficial effect on the neuropathy⁴ but does not convert an additional effect for lupus nephritis.⁴

In this patient suggest that GBS developed as a feature of lupus. Therefore, the association of GBS with lupus seems to have implications for both treatment and prognosis. Prednisone and cyclophosphamide should be considered in patients with GBS as a feature of lupus.

References

1. Patil AD, Karnik ND, Nadkar MY, et al. Guillain Barré Syndrome, Systemic Lupus Erythematosus and Acute Intermittent Porphyria – A Deadly Trio. *J Assoc Physicians of India* 2015; 63:60-63.
2. Fazio RM, Chen I, Somal N. Guillain-Barré syndrome as first presentation of systemic lupus erythematosus: a rare manifestation complicated by IVIg-induced splenic infarct. *BMJ Case Reports* 2015; doi:10.1136/bcr-2015-211598.
3. Ali N, Rampure R, Malik F, et al. Guillain-Barré Syndrome Occurring Synchronously with Systemic Lupus Erythematosus as Initial Manifestation treated Successfully with Low-Dose Cyclophosphamide. *Journal of Community Hospital Internal Medicine Perspective* 2016; 6:2.
4. Overlap of Bickerstaff Encephalitis and Guillain-Barré Syndrome in a patient with Systemic Lupus - Daniel PA. Santos, Mariana Spitz, Perola Oliveira, Thiago Barcia et al. (Neurology Department, Hospital Servidores do Estado, Rio de Janeiro RJ, Brazil- July 2009.)