CASE REPORT

Myasthenia Gravis: An Unanticipated Cause of Failure to Wean in a Postpartum Patient with Preexisting Systemic Lupus Erythematosus

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Abstract

Sudden onset and *de novo* Myasthenia gravis (MG) in the presence of systemic lupus erythematosus (SLE) is a rare postpartum phenomenon and can easily misguide the treating physician. A known case of SLE, 4 days after an elective cesarean section, presented to the intensive care unit for weaning-off mechanical ventilation after being put on ventilatory support in the emergency room, following acute-onset partial seizures. She was subsequently diagnosed to have new-onset MG, treated for the condition and later successfully extubated.

Keywords: Autoimmune diseases, Myasthenia, Weaning from mechanical ventilation.

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Highlights

Systemic lupus erythematosus (SLE) with no previous history of neuromuscular involvement can also present as weaning failure due to *de novo* MG as an unanticipated and rare finding in a postpartum patient.

Introduction

Both SLE and MG present with organ-specific autoimmune antibodies and exhibit a female preponderance.[1,2] Systemic lupus erythematosus (SLE) and MG have known to either coexist or precede one another, the incidence of MG preceding SLE being common and thus reported more often.[3–6] We report a case of preexisting SLE which later developed MG, to present as an unanticipated reason for failure to wean.

Case Description

A 29-year-old primigravida with a history of SLE since 2 years, with no history of flare in the preceding 12 months, received hydroxychloroquine 8 mg/kg/day, azathioprine 2 mg/kg/day, dexamethasone 40 mg as a single dose, and low-dose aspirin in her antenatal period. She had an uneventful pregnancy with no history or laboratory evidence of pregnancy-induced hypertension throughout her antenatal course. The decision for an elective cesarean section was taken considering a partial placenta previa in addition to her autoimmune disorder, which put her in the high-risk group. She was asymptomatic for the first 3 days of her postpartum period.

On the fourth postpartum day, she complained of drowsiness and difficulty in breathing for which she was brought to the emergency room. In the ER she had one episode of partial seizures, which lasted for 20 minutes, transient loss of consciousness, and sudden onset respiratory failure for a few minutes. She was intubated, and her seizure was controlled by a single dose of 2 mg IV midazolam following which she was spontaneously breathing with adequate respiratory indices half an hour later. In spite of a Glasgow Coma Score (GCS) of 15/15 and stable hemodynamic parameters attempts to wean her off continuous positive airway pressure (CPAP) repeatedly failed. Her arterial blood gas (ABG) analysis and serum electrolytes were normal. A CT scan head was carried out, which was satisfactory. She was put on antiepileptic prophylaxis and shifted to respiratory ICU for weaning after 24 hours of her arrival.

On her arrival to ICU, routine investigations related to hematological indices, renal and liver function parameters, and arterial blood gas analysis were sent and confirmed to be within normal limits. Clinically she was oriented, awake, responding to commands, with clear bilateral lungs and stable hemodynamic variables. A differential diagnosis of flaring of SLE,[7] posterior reversible encephalopathy syndrome,[8] or eclampsia was initially presumed. These were later ruled out after anti-double-stranded DNA, anti Ro/La antibodies, and antiphospholipid antibodies were not seen in plasma, urine spot polymerase chain reaction (PCR), urine proteins were normal, and diffusion weighted imaging- magnetic resonance imaging (DWI MRI) brain with fluid-attenuated inversion recovery (FLAIR) imaging was normal. The treatment for SLE was continued on consultation with the rheumatologist and supportive care was provided.

Weaning attempts with CPAP trials repeatedly failed with desaturation in spite of a good GCS, good motor activity, normal

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chest radiography, and a normal ABG analysis. On the 3rd day of presentation to ICU, a left-sided pleural effusion with collapse was noticed and promptly drained under aseptic precautions. Bronchoscopic removal of a left bronchial mucus plug was carried out, which resolved the collapse. A bronchioalveolar lavage done at the same time revealed the presence of levofloxacin and cotrimoxazole-sensitive *Klebsiella pneumoniae*, following which she was put on levofloxacin in addition to rest of the supportive and SLE-specific treatment.

Unsuccessful attempts of weaning followed for five more days. On the 8th day of presentation, bilateral (L > R) ptosis was noted with a quantitative myasthenia gravis (QMG) score of 14. A diagnosis of MG was made after a positive tensilon test and the presence of AchR antibodies in the serum. Quinolines were promptly omitted in the treatment chart and cotrimoxazole and pyridostigmine 60 mg QID was started. There was a significant improvement in ptosis after 48 hours of starting pyridostigmine. Patient-tolerated weaning trails well thereafter and was weaned of mechanical ventilation after one cycle of plasmapheresis and was discharged from respiratory intensive care unit (RICU) with stable vitals on room air 2 days later.

Discussion and Literature Review

Systemic lupus erythematosus (SLE) and MG coexisting in the same patient is a rare and sporadically reported phenomenon. The incidence of SLE following MG is up to 8% and that of MG following SLE is 1.3%.[3] In most of the available case reports, MG occurred before SLE and thymectomy was a precipitating factor for its appearance.[2–6] Very few cases have reported *de-novo* occurrence of MG in preexisting SLE.[9–11] The co-existence of both the autoimmune diseases has been postulated to be due to (a) loss of central tolerance after thymectomy resulting in T-cell lymphopenia with polyclonal B-cell activation, with production of antibodies; (b) molecular mimicry and structural similarity between main immunogenic region of α 65–80 of AchR and small nuclear ribonucleoprotein; and (c) role of chemokine CXCL13 in pathogenesis of SLE and MG.[11] It has been postulated that B lymphocytes play a major role in SLE, whereas T lymphocytes are critical for MG pathogenesis. CXCL13 is a common activator of B and T lymphocytes.[6]

The long-term use of hydroxychloroquine as an antimalarial agent essentially used in the treatment for SLE has also been associated with myasthenia syndrome like complications or MG itself. Jallouli et al. in 2012[12] studied the association of SLE and MG in a case series of 17 patients to retrospectively find out that 47% patients of SLE (8/17) who were treated with hydroxychloroquine developed MG after initiation of treatment.

Myasthenia gravis (MG) as an independent entity can also present in the postpartum period in the absence of any other autoimmune disease. A cross-sectional population-based cohort study[13] in 2016 revealed that the postpartum period was significantly associated with the onset of symptoms of MG in Norwegian women (relative risk 5.5, 95% CI: 2.6–11.6). The risk was found to be highest after the first childbirth. The possibility of MG occurring as an independent entity in our case therefore cannot be completely ruled out.

We thus conclude that SLE can also present with weaning failure due to MG as an unanticipated and rare finding in a postpartum patient.

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