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Case Report

Systemic lupus erythematosus and myasthenia gravis: More common than we think?

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ABSTRACT

Systemic lupus erythematosus and myasthenia gravis are autoimmune disorders whose association in the same patient has been rarely reported. An account of three cases of SLE and myasthenia coexisting in the same patients is being presented with a review of currently available literature. Case 1 was a 33-year-old female fulfilling 6 SLICC classification criteria for SLE. She developed diplopia, dysphagia, and hoarseness of voice. A CT thorax done for evaluation of pulmonary hypertension showed thymic enlargement and serum anti-acetylcholine receptor antibody was positive. The patient was treated with pyridostigmine for myasthenia gravis with clinical improvement and a subsequent 10-year uneventful follow-up. Case 2 was a 22-year-old girl with new onset proximal muscle weakness after 5 years of immunosuppressive therapy for SLE. Other potential causes of proximal weakness were ruled out. A diagnosis of myasthenia gravis was made based on anti-acetylcholine receptor antibody positivity. She later succumbed to respiratory involvement despite therapy. Case 3 was a case of myasthenia gravis diagnosed at 28 years of age based on decremental response on repetitive nerve stimulation (RNS), who presented 6 months later with pregangrenous changes in her foot. She tested positive for antiphospholipid antibody and satisfied 6 SLICC criteria for classification as SLE. She is now on treatment, well, and on long-term follow-up. In patients with SLE, development of fatigue, neurological manifestations, and muscle weakness requires a high index of suspicion to prompt a search for myasthenia in addition to myositis and drug-induced myopathy.

1. Introduction

Autoimmune disorders have been known to occur concurrently in individuals and families with such a predilection. Three cases of myasthenia gravis (MG) were diagnosed among 500 patients of SLE attending the Immunology Clinic at our center (Table 1). This case report, while highlighting the rarity of such a presentation, attempts to draw attention to variability in clinical manifestations and the difficulties encountered while making the diagnosis.

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Table 1 – Summary of case presentations and diagnostic criteria for SLE[a] And MG.

Case no.	Age in years	Sex	SLE diagnosis clinical	Antibody	Myasthenia diagnosis
1.	33	F	Oral ulcers, Proteinuria, Class V lupus nephritis, hypocomplementemia	ANA Anti-dsDNA	Thymic enlargement, bulbar symptoms, anti-AChR+
2.	22	F	Photosensitivity, oral ulcers, nonerosive arthritis, serositis, hypocomplementemia	ANA Anti-dsDNA	Proximal weakness, anti-AChR+, response to neostigmine
3.	28	F	Immune hemolytic anemia and thrombocytopenia, proteinuria	ANA Anti-dsDNA Anti Cardiolipin	Decremental response on RNS, anti-AChR+

[a] SLICC criteria have been used (ANA, anti-nuclear antibody; dsDNA, double stranded deoxyribonucleic acid; anti-AChR, anti-acetylcholine receptor antibody; RNS, repetitive nerve stimulation).

2. Case report

2.1. *Patient 1*

A previously healthy 33-year-old lady presented with oral ulcers and recent onset vasculitic ulcers on her lower extremities. Her clinical exam was otherwise normal. Indirect immunofluorescence revealed a positive anti-nuclear antibody (ANA) in a titer 1:40 and positive dsDNA. She had low serum complement; C4 – 16.9 mg/dL (Normal 20–40) and significant proteinuria of 1.3 g/day. Renal biopsy showed features suggestive of class V lupus nephritis – according to the International Society of Nephrology/Renal Pathology Society classification. The patient satisfied 6 Systemic Lupus International Collaborating Clinics (SLICC) criteria for classification as SLE. She was treated with 6 cycles of Cyclophosphamide, Prednisolone at a dose of 50 mg/day tapered every 4 weeks, Hydroxychloroquine sulfate (HCQS) (400 mg/day), and Enalapril (5 mg/day). One year later, she complained of exertional dyspnea. On investigation, she was found to have pulmonary arterial systolic pressure (PASP) of 104 mmHg (Normal <35 mmHg) confirming the clinical diagnosis of pulmonary hypertension. The evaluation of pulmonary hypertension entailed a CT scan of the thorax (Fig. 1) which brought to light an enlarged thymus. However, the lungs were normal. Soon after, the patient developed diplopia, hoarseness of voice, and dysphagia. At the time of clinical assessment, the patient had no neurological deficits and MRI brain was normal. Serum anti-acetylcholine receptor (anti-AChR) antibody was positive. A diagnosis of MG was made and the patient was treated with pyridostigmine (60 mg PO thrice daily) and steroids that were tapered over time. She is currently on a 10-year follow-up on therapy with Azathioprine, HCQS, and supplemental levothyroxine for autoimmune thyroiditis. The patient now has no myasthenic symptoms, and proteinuria has improved (30 mg/day) as has pulmonary hypertension (PASP 50 mmHg) with significant relief of symptoms.

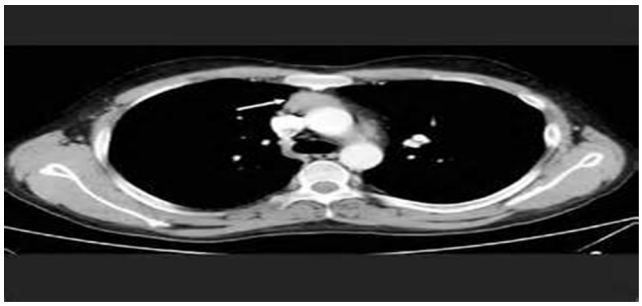

Fig. 1 – Computed tomogram of the thorax showing a prominent anterior mediastinal mass – an enlarged thymus in our first patient.

2.2. *Patient 2*

A 22-year-old girl was admitted with a 1-month history of fever, cough, dyspnea on exertion, and lower limb swelling in her immediate post-partum period. She had large joint polyarthritis, oral ulcers, and photosensitivity 5 years previously. Investigations revealed positive qualitative ANA – 3+ (homogenous pattern), dsDNA, posterior pericardial effusion, and hypocomplementemia (C4 – 14.8 mg/dL). A diagnosis of SLE was made in view of fulfillment of 7 SLICC criteria. The patient was treated with oral prednisolone started at 1 mg/kg and was maintained on 7.5 mg/day, azathioprine and 250 mg chloroquine sulfate. On a routine follow-up, she was diagnosed with autoimmune thyroid disease and treated with thyroxine. The patient was lost to follow-up until, 2 years later, she developed proximal muscle weakness and change in her voice. She had discontinued most of her medications by this time. Routine lab investigations were unrevealing, as were the serum Creatinine Kinase – 20 μg/L (normal – 20–309), Thyroid Stimulating Hormone (TSH), and barium swallow. The repetitive nerve stimulation (RNS) was inconclusive. However, with the background of SLE and autoimmune thyroiditis, there was a strong clinical suspicion of MG. Serum anti-AChR antibody was assayed and the patient was given a therapeutic trial of oral neostigmine (15 mg). She had symptom improvement and subsequently the anti-Ach receptor antibody was found positive. Thereafter, she had a rapid progression of weakness and intravenous immunoglobulin and plasmapheresis were considered. She later succumbed to respiratory muscle weakness.

2.3. *Patient 3*

A 28-year-old software professional, diagnosed case of myasthenia on the basis of >10% decremental response on RNS and a positive anti-Ach receptor antibody test, presented

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