The Rare Coexistence of Systemic Lupus Erythematosus and Myasthenia Gravis in a 17-Year-Old Female

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Systemic Lupus Erythematosus (SLE) and Myasthenia Gravis (MG) are two distinct diseases, which can coexist or precede each other; however, their occurrence in the same patient is rare. This rare phenomenon has already been observed; however, only sporadic clinical cases have been described.

A seventeen-year-old female who is a known case of SLE developed MG years later. Nerve conduction studies showed the decremental response over facial muscles and right median nerve. Anti-acetylcholine receptor (AChR) antibodies were positive. She was treated with oral pyridostigmine 60 mg three times a day. Clinical improvement and resolve of muscular weakness was seen within a couple of days. The patient was discharged on prednisolone 10 mg OD and pyridostigmine 60 mg TDS.

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Systemic Lupus Erythematosus (SLE) is an autoimmune disease with multi-organ involvement of unknown etiology, in which there is an inter-play of genetic, epigenetic and environmental factors[1]. It is not uncommon to find more than one autoimmune disease in the same patient[2]. It has been described that SLE patients can have Primary Sjogren's Syndrome (PSS), Systemic Sclerosis (SS), Hashimoto's Thyroiditis (HT), Myasthenia Gravis (MG), Polymyositis (PM) and many other autoimmune diseases. The combination of SLE and MG is rare in the literature[3].

The aim of this case report is to present a case of SLE in a 17-year-old female who was found to have MG many years after the initial diagnosis of SLE.

THE CASE

A seventeen-year-old female suffered from SLE. She was maintained on Prednisone 10 mg daily and Naproxen 250 mg twice daily. She was advised to reduce Prednisone to 7.5 mg daily. One month after reducing steroids, she presented to her local hospital with progressive muscle weakness and dysphagia. She was treated with high dose of steroids with significant improvement. She was discharged on tapering dose of steroids.

Three months later, she had a relapse with muscle weakness and dysphagia. She was unable to swallow both liquids and solids. The patient was treated with pulse-steroids. Two days later, she was in better condition. She was started on Imuran (azathioprine) and advised to taper steroids slowly. She continued to do well until she presented with one week history of post-nasal drip and progressive difficulty in swallowing for both solids and liquids but with no odynophagia. The clinical examination was unremarkable except for evidence of progressive proximal muscle weakness (power-grade IV/V), more with exertion. She was having difficulty in getting up from a sitting position but with no muscle pain. The gait was normal. No cerebellar signs. Extra-ocular muscles are all normal. Bilateral ptosis was noticed sometimes, especially on repeated movements but there was no diplopia.

Her investigations revealed normal CBC, ESR, RFT, LFT, CK, LDH and TSH. Thyroglobulin and smooth muscle antibodies were negative. CXR and ECG were normal. HRCT was normal with no evidence of interstitial lung disease and no thymoma. Pulmonary Function Test was normal. ANA +ve was 1:640 (Speckled), Anti-RNP +ve was 54 (Normal Range: 0-16), Anti-Ds DNA +ve was 74 (Normal Range: 0-35), anti-cardiolipin antibody IgG was 20 (Normal Range 0-15) and anti-acetylcholine receptor (anti-AChR) was 0.63 (Normal Range < 0.25 nmol/L). Other investigations included endoscopy revealed gastritis. Barium swallow revealed hypo-motility of the esophagus. X-ray sinuses showed moderate mucosal swelling in both maxillary sinuses. She was evaluated by an ENT surgeon and was diagnosed to have rhino-sinusitis for which she was prescribed Flixonase (fluticasone propionate). The patient improved upon increasing the dose of Prednisone, see table 1.

Table 1: Auto-Antibodies Profile of the Patient

Antibody	Value
ANA	1 : 640 (Speckled)
Anti-Ds DNA	74 IU/mL (Normal Range : 0-35 IU/mL)
Anti-RNP	54U (Normal Range: 0-16 U)
Anti-AChR	0.63nmol/L (Normal Range< 0.25 nmol/L)
ACA IgG	20 U/ml (Normal range 0-15 U/ml)

ANA: Anti-Nuclear Antibody
Anti-Ds DNA: Anti-Double Stranded DNA
Anti-RNP: Anti- RibonucleoProtein
Anti-AChR: Anti-Acetylcholine receptor
ACA IgG: Anti-Cardiolipin Antibody IgG

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The nerve conduction study showed significant decrement pattern on repetitive stimulation of both right median and facial nerves which is consistent with neuromuscular function disorder such as seen in MG. Needle examination showed no evidence of myopathic or neuropathic changes. Ultimately, the diagnosis of overlap syndrome with SLE and MG was established. She was started on pyridostigmine 60 mg TDS.

The patient was feeling very well with complete resolution of the dysphagia and muscle weakness (Power grade V/V). She was discharged on Imuran 75 mg OD, with a tapering dose of prednisolone, pyridostigmine 60 mg TDS, lansoprazole 30 mg OD, calcium and vitamin D supplements.

DISCUSSION

SLE is an idiopathic autoimmune disorder that can affect any organ system in the body. It can present with a wide variety of different clinical and laboratory manifestations. It has a variable course and prognosis.

In a study of SLE which included 215 patients, 30% had another autoimmune disease, Sjogren's syndrome being the most common[2,4]. MG is a chronic autoimmune disease characterized by varying degree of weakness of the voluntary muscles as a result of immune-mediated destruction of acetylcholine receptors at the neuromuscular junctions. SLE can precede or follow the development of MG. The latter scenario is more rare, as the prevalence of SLE in MG was 2.2%–8.3% and the prevalence of MG in SLE was 1.3%[5].

So far, there had been a total of 17 previously published cases of SLE and MG occurring in the same patients[6]. In addition, many of these patients had abnormally high titres of various autoantibodies, including anti-ds DNA and anti-acetylcholine receptor antibodies such as in this case[7].

It has been observed that some MG patients have developed very aggressive SLE after undergoing thymectomy[8-11]. In addition, special care should be given to any SLE patient taken hydroxychloroquine, as this medication can trigger any undiagnosed MG.

In our case, the patient had a very significant clinical improvement after oral acetylcholinesterase inhibitor (pyridostigmine).

The association of SLE and MG is rare. Early detection, diagnosis, management would minimize the complications[12].

CONCLUSION

The association between SLE and MG is rare and complex, but the possibility of co-existing MG in lupus patients should be considered, especially when they are complaining of fatigue and decrease in muscle strength.

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