CASE REPORT

JIACM 2022; 23 (3-4): 143-145

A Rare Case of Minimal Change Disease in a Patient of Myasthenia Gravis

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Abstract

Myasthenia gravis is a chronic autoimmune neuromuscular junction disorder characterised by weakness and fatiguability. It is associated with thymoma in 10% of cases. Paraneoplastic glomerulonephritis is a rare clinical presentation of the same. We report the case of a 27-year-old male who presented with complaints of fluctuating weakness and diplopia, who was diagnosed as seropositive myasthenia gravis with B2 variant of thymoma. 9 months after the disease, he developed anasarca with nephrotic range proteinuria and deranged renal function tests and was found to have minimal change disease.

Key words: Myasthenia gravis, thymoma, minimal change disease.

Introduction

Myasthenia gravis is a common autoimmune neuromuscular junction disorder but its association with proteinuria is a rare entity. Minimal change disease is a rare paraneoplastic manifestation of thymoma. Both myasthenia gravis and minimal change disease are related to the dysfunction of T-lymphocytes. We report here a case of myasthenia gravis who developed minimal change disease after thymectomy.

Case report

A 27-year-old male with no comorbidities, presented with complaints of fluctuating weakness in bilateral upper and lower limbs for 3 months, and diplopia for 1 month – with aggravation during the night; and gradually progressed to a level when he had difficulty chewing hard food, and even getting up from bed during evening time. On examination, the patient was conscious, oriented, and vitally stable. There was no pallor, icterus, pedal oedema, skin rash. Nervous system examination was suggestive of ptosis in both eyes with bilateral medial rectus palsy which improved on ice pack test. There was weakness in all 4 limbs on repeated activity, with demonstrable improvement on rest. Higher mental functions, other cranial nerves, and sensory system examination was normal. Other systemic examination showed no abnormality.

On investigation, patient had a haemoglobin of 15.7 gm/dl, TLC of 13,000 cells/mm^3, platelet count of 2,00,000 cells/mm^3; muscle enzyme CPK was 68 U/L, CK-MB was 2 U/L and LDH was 667 U/L; thyroid profile was normal; ANA was negative and myositis profile was also negative. In the background of fluctuating weakness, Acetylcholine Receptor antibody levels were checked which turned out to be 22.14 nmol/l (normal: < 0.40). Diagnosis of myasthenia was confirmed. CECT chest was planned, which was suggestive of 3.9 x 10.0 x 5.4 cm, horizontally placed cystic density with heterogeneous contents and peripheral enhancement, likely a Cystic thymoma (Fig. 1). PET scan done to rule-out metastasis, turned out to be normal. The patient was started on IV steroids and pyridostigmine. Thymectomy was done and biopsy was suggestive of B2 variant of thymoma (Fig. 2).

After 9 months of treatment, he noticed swelling over his face and gradually all over his body over the course of 15 days (Fig. 3, 4). It was associated with frothing in urine and gradually a reduction in urine output to around 400 ml/24 hrs. Blood pressure was in the normal range.

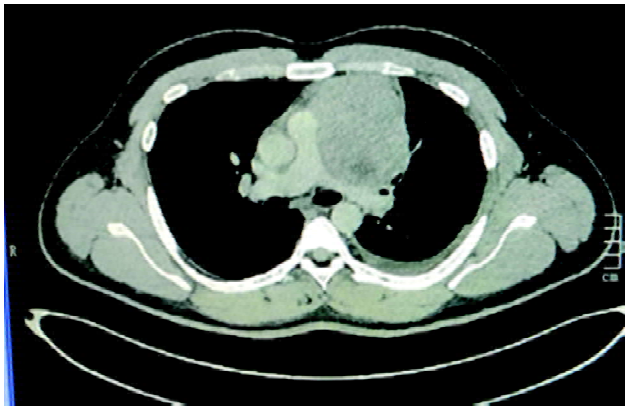

Fig. 1: *CECT chest demonstrating a mass in superior mediastinum with heterogenous content and peripheral enhancement suggestive of cystic thymoma.*

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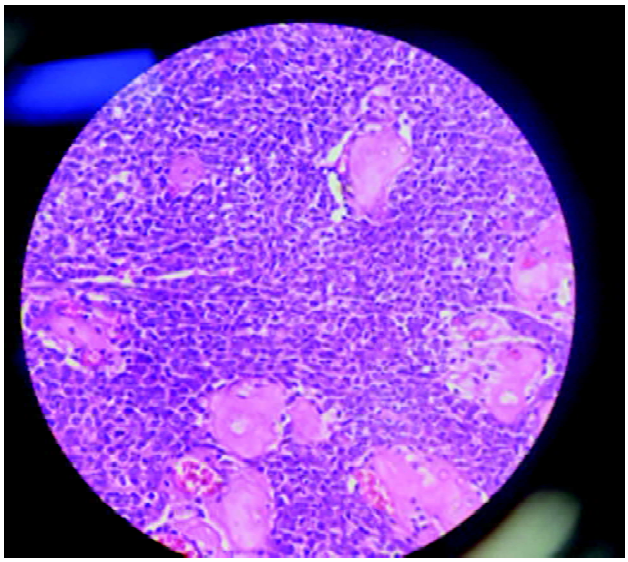

Fig. 2: *Thymic biopsy (Haematixylin-eosin, 10 x magnification): Cyst separated by fibrous septae in background of normal thymic tissue.*

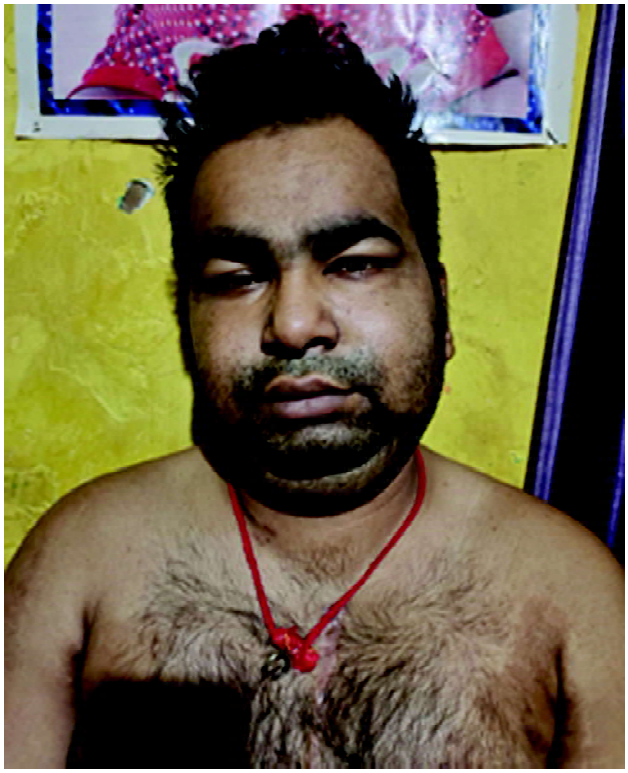

Fig. 3: *Facial puffiness.*

Investigations were suggestive of proteinuria with a 24 hrs urine protein of 7g/24 hrs (nephrotic Range). There were no casts or haematuria. Urea was 40 mg/dl, creatinine was 2.2 mg/dl, Na^+/K^+ was 133/4.2 mEq/l, C3, C4 levels were normal, ANA was negative, and ultrasonography revealed normal kidneys. Renal biopsy was done which was

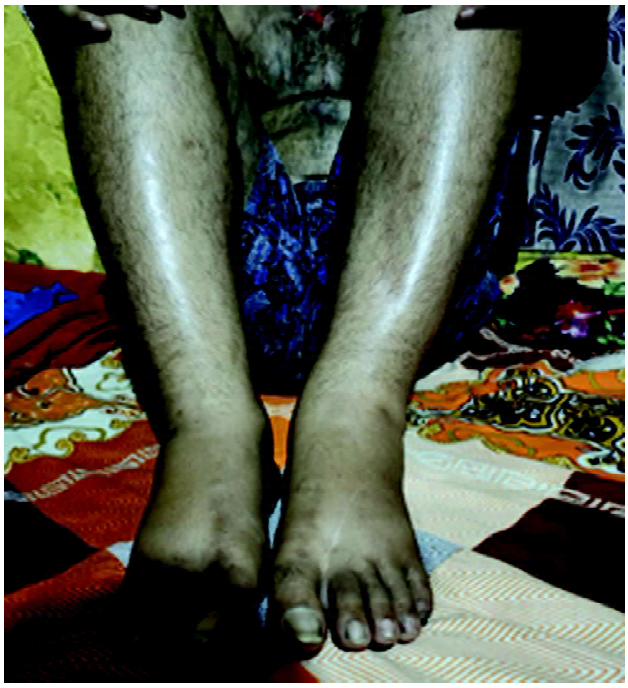

Fig. 4: *Swelling in both legs extending up to thigh.*

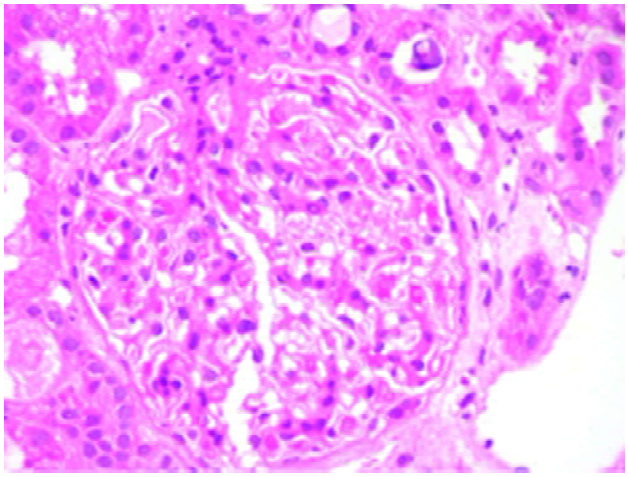

Fig. 5: *Kidney biopsy (Haematoxylin-eosin 10 x magnification): Normal glomeruli.*

suggestive of minimal change disease (Fig. 5, 6).

The patient was started on pulse steroids and ACE-inhibitors, after which there was decrease in anasarca and proteinuria. Patient was discharged on oral steroids. However, the patient returned 1 month later with generalised weakness, therefore the patient was switched to tacrolimus and steroid was gradually tapered after which there was some gain in function.

Discussion

Myasthenia gravis is a neuromuscular disorder

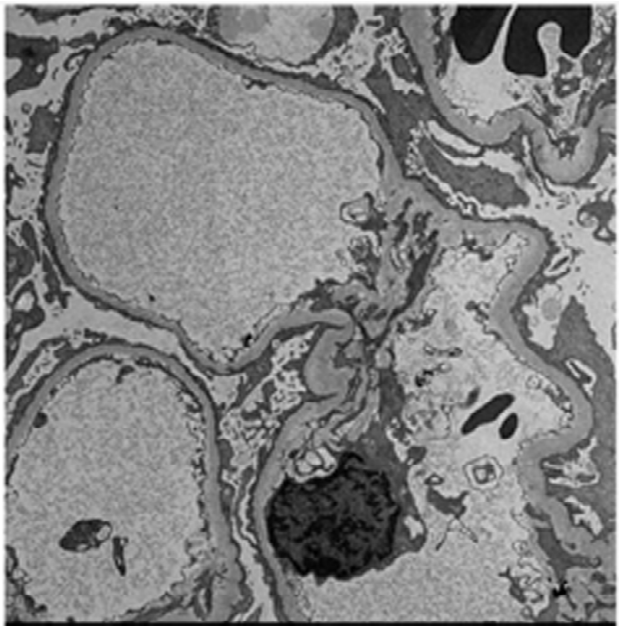

Fig. 6: *Kidney biopsy (electron microscopy): Extensive effacement of foot processes and absence of electron dense deposits.*

characterised by fluctuating weakness of ocular, bulbar, limbs, and rarely respiratory muscles[1]. These symptoms occur as a result of autoantibodies against acetylcholine receptors at neuromuscular junction causing synaptic rundown[2]. Association between minimal change disease and myasthenia gravis is a rare entity. Minimal change disease is a podocytopathy due to T-cell dysregulation and presents with anasarca and nephrotic-range proteinuria. It has a predilection in children and its occurrence in adults is rare.

There may be association between minimal change disease and thymoma. Thymectomy can act as a precipitating factor for minimal change disease as it is followed by changes in lymphocyte function[3].

Our case was a myasthenia gravis patient who underwent thymectomy following which he developed nephrotic range proteinuria and turned out to be minimal change disease on renal biopsy within a year of surgery, whereas the occurrence of minimal change disease in myasthenia gravis is generally a late presentation[4]. The treatment of the same is challenging as high dose steroids are the mainstay of management for minimal change disease; however, the same dose may cause worsening of myasthenia gravis[5] which was quite apparent in our patient and he had to be switched to tacrolimus.

Conclusion

In a patient with myasthenia gravis and thymoma, minimal change disease may occur in the course of illness secondary to T-cell dysfunction post-thymectomy. Therefore, clinician may keep this association in mind. Steroids must be titrated such that it does not worsen the myasthenia.

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