

## Case Report

# Cardiac Dysfunction in Myasthenic Crisis

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### Abstract

**34 year old female presented with complains of abdominal pain and slowly progressing weakness of bilateral upper limbs along with respiratory muscular weakness leading to CO<sub>2</sub> retention and intubation. On evaluation, patient was found to have 25% ejection fraction which was unexplained even after full evaluation. Repetitive nerve stimulation-Nerve conduction velocity-Electromyography test showed neuromuscular disorder. Patient was given pyridostigmine along with steroid pulse for myasthenia gravis which resulted in dramatic improvement of ejection fraction to normal within 4 days.**

**Keywords:** Myasthenia gravis, Cardiac dysfunction, Striational antibodies, Anticholinesterase drug, Steroid pulse.

## INTRODUCTION

Myasthenia gravis is an autoimmune neuromuscular disorder characterized by skeletal muscle weakness and fatigue. The prevalence of the disease is approximately 1:7500 with a maximum during second to third decade in women and fifth to sixth decade in men with female predominance (3:2). The disease may involve a group of muscles (extracurricular muscles) or may occur in generalized form (84% of patients), (Călin et al., 2009). Cardiac involvement in myasthenia gravis has been described over 100 years but literature is limited to only case reports and case series. An article by Gibson 1975, concluded that the involvement of myocardium in myasthenia gravis is indicated clinically or by ECG, vector cardiography and autopsy. A study done by Hakon et al. 1984 studied 108 cases of myasthenia gravis and found 17 patients had cardiac involvement that could be due to myasthenia gravis.

## Case Report

A 34 year old housewife had complaints of acute pain abdomen (sudden onset, moderate intensity, radiating to

back, not relieved with medication) 7 days prior to admission. Patient was evaluated and was found to have cholelithiasis with cholecystitis. Patient underwent laparoscopic cholecystectomy at a private hospital. On postoperative period day 2, patient started having difficulty in lifting arms above shoulders along with respiratory distress and fall in oxygen saturation for which she required intubation and mechanical ventilatory support. Patient was then referred to our hospital which is tertiary care centre in north India.

On examination patient was calm, conscious and cooperative with blood pressure 110/70 mmHg, pulse rate 76/-, temperature- 98.4 °F. Neurologic examination showed normal higher mental functions, cranial nerves were normal while power was 4-/5 in all limbs and rest of systemic examination was normal. On evaluation, leukocyte counts were 16900/ul, C – reactive protein (CRP) was 8 while her thyroid profile, lipid profile, renal function tests (RFTs) and liver function tests (LFTs), cardiac markers and ECG were within normal range. Echocardiography showed an ejection fraction (EF) of 25% and global hypokinesia (at rest). Patient was initially managed with broad spectrum intravenous antibiotics, proton pump inhibitors and ventilatory support. In view of persistence of weakness of limbs and ventilatory weaning failure, Nerve conduction velocity-Repetitive nerve stimulation-Electromyography (NCV-RNS-EMG) was planned which showed post-synaptic

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neuromuscular disorder. In view of above presentation, patient was diagnosed to have myasthenic gravis presently in myasthenic crisis precipitated by surgery and sepsis. Patient was given intravenous steroid pulse (methylprednisolone) 500mg for 4 days along with pyridostigmine 60 mg twice daily. Patient showed marked improvement in weakness and started maintaining oxygen saturation without any support. After completion of 4 days of steroid pulse, echocardiography was repeated which showed improvement of EF to 60%. AchR (acetylcholine receptor) antibodies were positive (raised) in the patient whereas CT chest showed no evidence of thymoma.

## DISCUSSION

In myasthenia gravis, the pathophysiology involves decrease of the number of acetylcholine receptors and flattening of post-synaptic membrane. It is characterized by autoimmune antibodies against the acetylcholine receptors at the level of the neuromuscular junction. Thymus plays an important role in the initiation of the autoimmune response in 75% of myasthenic patients [hyperplastic (65%) or tumor (thymoma-10%)]. Anti-AchR antibodies are present in 85% of patients with myasthenia gravis and MuSK (muscle-specific tyrosine kinase) antibodies in 50% with absent anti AchR antibodies. (Călin et al., 2009).

Cardiac involvement of the heart in myasthenia gravis was described by Weigert in 1901 while Leopold Laquer termed it as Herzmyasthenie in the same year (Johan, 2009). Cardiac involvement in myasthenia gravis ranges from asymptomatic ECG changes to ventricular tachycardia, myocarditis, conduction disorders, heart failure and sudden death. Cardiac involvement occurs upto 50% of patients with myasthenia gravis with thymoma while 12% in patients without thymoma. It was suggested that myocarditis could be a paraneoplastic phenomenon (Bonow et al., 1981).

In myasthenia gravis the characteristic AchR autoantibodies bind to nicotinic AchR in skeletal muscle but do not bind to cardiac muscle (Bonow et al., 1981). The myocardial involvement in myasthenia gravis is immunologic response to non-AchR antigens present in heart muscle i.e. titin and ryanodine receptors (RyR) thereby leading to altered cardiac function (Suzuki et al., 2009). In 2005, anti-KV1.4 has also been described. These three antibodies have been described as striational antibodies because they bind to cross-striational pattern in heart muscles. Anti-titin antibody is detected in 20–40% of all myasthenia gravis patients, anti-RyR in 13–38%, and anti-Kv1.4 in 12–15% of all myasthenia gravis patients (Aarli et al., 1990). The presence of titin antibodies in patients with MG

correlates with the electromyographic evidence of myopathy (Somnier et al., 1999). Anti-RyR antibodies cause allosteric inhibition of RyR function in vitro, inhibiting  $Ca^{2+}$  release from the sarcoplasmic reticulum (Skeie et al., 2003).

Myasthenia gravis patient having thymoma is at increased risk for myocardial involvement. Heart symptoms in these patients should always be suspected as being caused by myocarditis or pericarditis. It has been shown that early diastolic atrioventricular plane velocity and tissue Doppler peak systolic strain was lower in myasthenia gravis patients than in controls. The differences disappeared following administration of pyridostigmine (Owe et al., 2008). Johannessen et al. 1992 found myasthenia gravis patients have decreased peak diastolic filling rate which improved after pyridostigmine medication. Our case is non-thymoma myasthenia gravis in crisis with cardiac dysfunction which improved after anticholinesterase drug and steroid pulse.

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