

## Case Report

# MYESHENIA GRAVIS PRESENTED WITH RESPIRATORY FAILURE

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## ABSTRACT

A case of 41 year old female known case of depression since 10 years, developed dry cough, low grade fever, breathlessness and drowsiness since 4 days was admitted in ICU and initially diagnosed as type 2 respiratory failure due to pneumonia but on further investigating for altered sensorium patient was found to be NCV positive and was diagnosed as seronegative myasthenia gravis.

**Key Words:** Myasthenia gravis, Acetylcholine receptor, Muscle specific tyrosin kinase

## INTRODUCTION

Myasthenia gravis (MG) is a neuromuscular disorder characterized by weakness and fatigability of skeletal muscles. The underlying defect is a decrease in the number of available acetylcholine receptors (AChRs) at neuromuscular junctions due to an antibody-mediated autoimmune attack.<sup>[1]</sup>

However, some patients (about 15%) with generalised MG do not have detectable AChR antibodies. There is some evidence, however, that this "seronegative" MG is an antibody-mediated disorder. Plasma from patients with the disorder seems to contain various distinct humoral factors: IgG antibodies that reversibly inhibit AChR function; a non-IgG (possibly IgM) factor that indirectly inhibits AChR function; and an IgG antibody against the muscle-specific kinase (MuSK). The presence of antibodies against MuSK appears to define a subgroup of patients with seronegative MG who have predominantly localised, in many cases bulbar, muscle weaknesses (face, tongue, pharynx, etc) and reduced response to conventional immunosuppressive treatments. Moreover, muscle wasting may be present, which prevents complete response to these therapies.<sup>[2]</sup>

## CASE

**Clinical Presentation:** A case of 41 year old female known case of depression since 10 years, developed dry cough, low grade fever, breathlessness and drowsiness since 4 days. She went to the psychiatrist and was found hemodynamically unstable and severely dyspnoeic. Patient was advised for hospitalisation and was admitted in ICU. Patient was initially diagnosed as right lower zone pneumonitis with Type-II respiratory failure. Even after treatment of

same, patient remain drowsy and disoriented. On further evaluation we found absent gag reflex, bulbar muscle weakness and ptosis. Neurophysician opinion was taken and advised CSF examination, MRI brain with spine and NCV study.

**Investigation and Diagnosis:** The CSF and MRI were normal but NCV study showed decreased amplitude on repeated stimulation. So myesthenia gravis was suspected. Anticholinesterase antibody was negative. So we diagnosed it as seronegative myesthenia gravis.

**Treatment:** Patients was admitted in ICU, Initially bipap and then invasive ventilatory support was given. Antibiotics and other supportive treatment was given. After NCV study , injection neostigmine was given along with plasma pheresis.

**Outcome:** After treatment there was improvement in patient's condition, but she developed sudden onset of breathlessness and cardiac arrest due to pulmonary embolism(Portable ECHO suggested clot in cardiac chamber) and patient died because of this.

## DISCUSSION

Muscular weakness in 90% of patients with myasthenia gravis is caused by an antibody mediated autoimmune response to muscle nicotinic acetylcholine receptors (AChRs). Immunization of animals with purified AChRs includes experimental autoimmune MG, in which autoantibodies to AChRs cause muscle weakness by impairing neuromuscular transmission through the same mechanism found to occur in MG: loss of AChRs caused by increased internization of antibody cross-linked AChRs and destruction of AChRs by complement-mediated focal lysis, disrupted synaptic morphology caused by postsynaptic morphology caused by postsynaptic membrane dam-

age and (to limited extent) direct impairment of AChR function by bound autoantibodies. [3]

An explanation for muscular weakness in 10 to 15% of patients with the seronegative MG who lack autoantibodies to AChRs had appeared to be autoantibodies to muscle-specific receptor tyrosin kinase (MuSK). It was found that in 30 to 70% of patients with MG seronegative for autoantibodies to AChRs but not in patients with MG with autoantibodies to AChRs. MuSK mediates agrin induced clustering of AChRs during synapse formation. These autoantibodies to extracellular domain of MuSK inhibited its function in tissue culture. [3]

There have not been reports of inducing muscular weakness in animal by immunization with purified MuSK. Thus, It has not been demonstrated that autoantibodies to MuSK can actually cause muscle weakness through impairing neuromuscular transmission. The possibility remains that autoantibodies to other presynaptic proteins could account for some or all seronegative MG. [4]

Patients with seronegative MG respond to plasma exchange and immunosuppressive therapies.

## CONCLUSION

Many of the diseases like hypothyroidism, myasthenia, collagen vascular diseases etc can represent with non specific symptoms, if not given proper attention particularly in female patients, can turn out to be life threatening when diagnosed late and remain untreated.

## REFERENCES

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