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Case Report

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ABSTRACT

Myasthenia gravis is an autoimmune disorder of the neuromuscular junction characterized by fluctuating weakness of certain voluntary muscles rarely involving the respiratory muscles. Presenting a case of a lady who presented with features of respiratory distress which on evaluation was found to be due myasthenia of respiratory group of muscles.

A 45yr old lady presented with progressive dyspnoea for 3 months and had orthopnoea on presentation. Preliminary evaluation including echocardiography, PFT, CTPA were normal. Electromyography studies on routine group of muscles were normal. As the symptoms persisted, EMG was done on specific respiratory group of muscle which were suggestive of myasthenia. Patient improved with steroids and Anticholinesterases thus supporting the diagnosis of isolated myasthenia of respiratory muscles.

Myasthenia gravis is a muscular weakness which in rare and mostly advanced cases include the respiratory group of muscles and even the external sphincters of bladder and bowel. Confirmation of diagnosis especially in doubtful presentations are done by electrophysiological testing, pharmacological tests and measurement of specific antibodies. Anti Ach-R and anti MUSK are sensitive and specific marker of this condition.

This particular case implies the need to consider a possibility of myasthenia of isolated respiratory group of muscles in patients presenting with respiratory distress. ENMG of the respiratory group of muscles though rarely indicated can clinch the diagnosis.

INTRODUCTION

Myasthenia gravis is an autoimmune disorder of the neuromuscular junction characterized by fluctuating weakness of certain voluntary muscles, particularly those innervated by motor nuclei of the brainstem. It manifests as weakening during continued activity, quick restoration of power with rest and dramatic improvement in strength following administration of anticholinesterase drugs.[1]

In most advanced cases all muscles are weakened, including diaphragm, abdominal, intercostal muscles and even external sphincters of bladder and bowel.[1]

This is a case report of a patient who presented with isolated respiratory distress, later diagnosed to be Myasthenia. Such a presentation was found to be extremely rare from our review of pertinent literature.

Case History

A 45year old home maker from Tamil Nadu presented with history of dyspnoea precipitated by lying down flat on bed for a certain duration. She had these symptoms since three months, however the duration required to precipitate these symptoms decreased progressively to an extent where the patient became dyspnoeic within a few minutes of lying supine.

Being a known case of Diabetes and Hypertension on treatment for 6 years, cardiac causes for this presentation was considered. Patient had no significant past history except for one episode of fever with joint pain 1 month ago, treated as Chikungunya. There was no history of chest pain, palpitation, noisy breathing with cough or seasonal variation of symptoms. Clinical examination was all within normal limits. Vital signs were normal. Preliminary cardiac, respiratory, and neuromuscular system evaluation were under normal limits.

Preliminary investigations including complete blood counts, renal function tests, liver function tests, chest X ray and ECG were normal. Following a normal Echocardiographic study including bubble contrast studies to rule out intra or extra cardiac shunts, Pulmonary function tests was considered to evaluate for restrictive/ Obstructive lung diseases which showed restrictive pattern.

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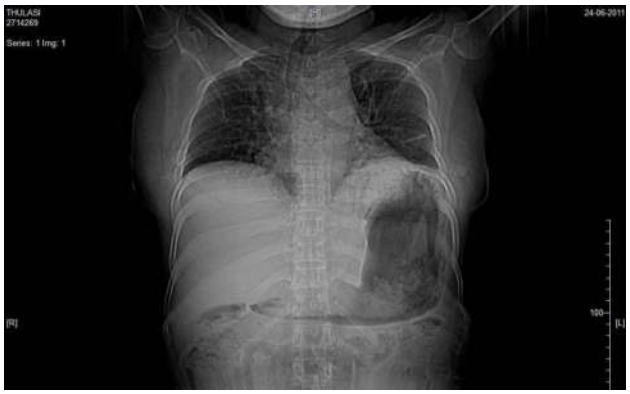
	Patient	Predicted	% predicted
FEV1	1.08	1.94	56
FVC	1.14	2.29	50
PEF	3.69	5.73	64
FEF75	0.83	1.55	54
FEF50	1.94	3.69	53
FEF25	3.62	5.27	69
FEV1% FVC	95	85	112

Blood gas values

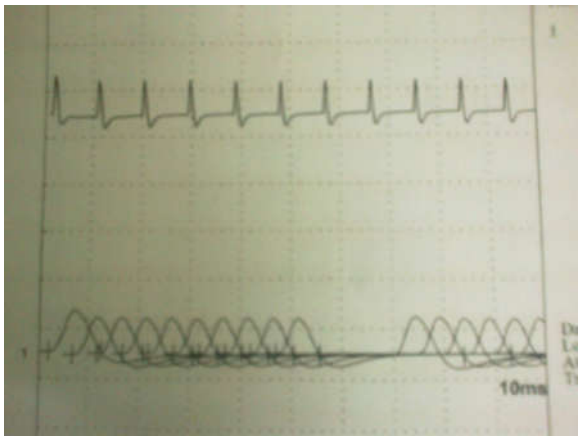
	Patient	Normal
pH	7.411	7.34 – 7.44
pCO2	42.2	35 – 45 mmHg
pO2	71.8	75 – 100 mmHg
sO2	93.2	95 – 99
pHCO3	26.4	22 – 26 mmol/ L

Patient was initially treated with low dose diuretics and bronchodilators and monitored in the ward for a period of two weeks. As no improvement was noticed further investigations including CT chest and pulmonary angiography were done. This however proved futile.

CT films



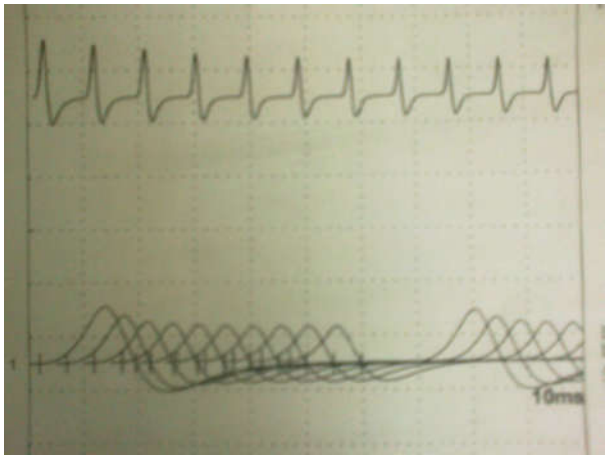
Preliminary ENMG done on right median, ulnar, peroneal, and sural nerves were normal.



RNS study of the ulnar nerve shows no signs of MG

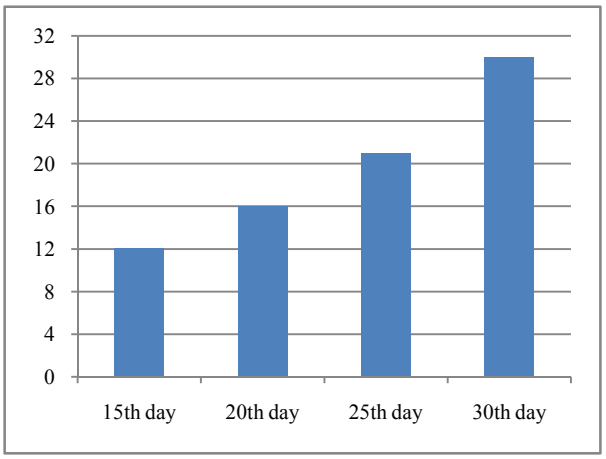
On further reassessment of the neuromuscular system by expert neurologist. Mild weakness in the neck flexors was noticed. ENMG done on respiratory muscles (Trapezius) showed significant decremental response to repeated nerve stimulation at 10 Hz and 15 Hz which is very suggestive of Neuromuscular junction disorder

ENMG Report



RNS study of Trapezius at 10 Hz shows significant decremental response s/o MG

Patient was started on Pyridostigmine and steroids and the response was monitored by assessing the single breath count and change in duration required to precipitate dyspnoea on lying supine. Both of which showed improvement as tabulated below



Thus a diagnosis of isolated respiratory muscle Myasthenia was made.

DISCUSSION

Myasthenia gravis is a muscular weakness formerly of grave prognosis. Usually the eyelids and the muscles of the eyes and somewhat less often of the face, jaws, throat and neck are the first to be affected. In rare and mostly advanced cases all muscles are weak including the respiratory group of muscles and even the external sphincters of bladder and bowel. A later age of onset was also associated with a higher incidence of fatal respiratory crises.[5]

Although classical clinical features of effort dependent fatiguability and reversal with rest or neostigmine can hardly be overlooked in diagnosis of myasthenia, confirmation of diagnosis especially in doubtful presentations are done by a) Electrophysiological testing, b) pharmacological tests and c) measurement of specific antibodies.

Characteristic of myasthenia is a rapid reduction in the amplitude of compound muscle action potentials evoked during a series of repetitive stimulations of a peripheral nerve at a rate of 3 per second. *Single-fiber electromyography (EMG)* represents an even more sensitive method of detecting the defect in neuromuscular transmission.

Although pharmacological testing including Edrophonium and Neostigmine tests are equally valuable to electrophysiological tests, measurement of antibodies to acetylcholine receptors [Anti-AchR] provides a sensitive and highly specific test for the diagnosis of myasthenia .Anti Ach-R antibodies found in 80-90% of patients with generalized and 60% of those with symptoms restricted to the ocular muscles.[6]

Recently, the majority of such cases which are negative for Ach-R antibodies havebeen ascribed to Ig G antibodies directed against a muscle-specific kinase (MuSK). Patients with MuSK antibody, mostly women, have a special clinical syndrome of prominent oculobulbar weakness, often with persistently severe disease and respiratory crises.[7,8]

Importance of ENMG studies on respiratory group of muscles.
Interestingly, in our case, the ENMG done on routine nerve muscle groupsi. eradial, ulnar the ocular were negative. Only on further evaluation of specific respiratory muscle groups (egTrapezius) could the diagnosis of myasthenia be made. This implies the importance to consider myasthenia as a possibility in patient with respiratory failure when all other evaluation for the same turn futile. ENMG of the respiratory group of muscles though rarely indicated can clinch the diagnosis.

References

1. Rooper AH, Brown RH. Adams and Victor's principles of neurology. 8th ed. New York (USA): McGraw-Hill 2005. P.1251-64.
2. Siu AYC, Wong TW, Lau CC. An undiagnosed myasthenia gravis presented with acute respiratory failure. Hong Kong J Emerg med 2001; 8:106-107.
3. Kim WH, Kim JH, Kim EK, Yun SP, Kim KK. Myasthenia gravis presenting as isolated respiratory failure a case report. Korean J Intern MED 2010; 25:101-104.
4. Dushay KM, Zibrak JD, Jenson WA, Myasthenia gravis presenting as isolated respiratory failure. Chest 1990; 97:232-235.
5. Bever CTJ R, Aquino AV, Penn AS, *et al*: Prognosis of ocular myasthenia. *Ann Neurol 1983*14:516.
6. Vincent A, Newaom, Davis J, Martin V: Antiacetylcholine receptor antibodies in *d*-penicillamine associated myasthenia gravis. *Lancet 1978* 1: 1254.
7. Scuderi F, Marino M, Colonna L, *et al*: Anti-p110 autoantibodies identify a subtype of 'seronegative' myasthenia gravis with prominent oculobulbar involvement. *Lab Invest 2002*82:1139.
8. Evoli A, Tonali P, Padua L, *et al*: Clinical correlates with anti-MuSK antibodies in generalized seronegative myasthenia gravis. *Brain 2003*126: 2304.

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