Journal of Clinical and Biomedical Sciences

Case Report

Difficulties in Diagnosing a Case of Myasthenia Crisis

Atishaya G V[1*], Prabhakar K[2] , Anitha A[3] , Jithendra Chaitanya[4]

1. 2nd Year Postgraduate, Department of General Medicine, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy of Higher Education and Research, Kolar.
2. Professor, Department of General Medicine, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy of Higher Education and Research, Kolar.
3. Assistant Professor, Department of General Medicine, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy of Higher Education and Research, Kolar.
4. Senior Resident, General Medicine, Sri Devaraj Urs Medical College, Sri Devaraj Urs Academy of Higher Education and Research, Kolar.

Abstract

A 26 year old, female presented to EMD in severe respiratory distress. Patient had history of breathlessness since past one month with h/o intrauterine death 1month back .On further investigation it was found that anti acetylcholine receptor antibody was above normal limits and was started on cholinesterase inhibitors, initially patient showed signs of improvement but later patient condition worsened and patient was diagnosed with Myasthenia gravis in crisis. Patient was started on pulse steroid therapy but patient did not show significant response, patient attenders were advised for IVIg but they could not afford the same, later patient was transferred to a center with plasmapheresis facility where plasmapheresis was done and now patient doing well and is on oral cholinesterase inhibitors.

Keywords: Myasthenia crisis, anti-acetylcholine receptor antibodies

Introduction

Myasthenia crisis is a life-threatening condition that involves worsening of myasthenia weakness requiring intubation and mechanical ventilation or non-invasive ventilation. It usually occurs within 2 years of disease onset. It is usually associated with generalized Myasthenia and Myasthenia gravis with thymoma.

Case History

A 26 year old, female presented to EMD of R.L. Jalappa Hospital with history of breathlessness since 1month which increased since 2 days, patient was intubated and was shifted to ICU in view of respiratory distress and ABG was done which showed type 2 respiratory failure.

Patient had h/o episodes of breathlessness on and off since 1month.

H/o fever present on and off since past 1 month.

No h/o chest pain orthopnea or PND.

Patient had h/o intrauterine death 1month back.

Patient started developing above symptoms after the incident. Because of this history initially sepsis was thought off, OBG opinion was sought to look for retained products of conception as source of sepsis, USG Abdomen and pelvis was done and it was normal, since there was no improvement in clinical condition and patient kept on worsening post-partum, post-partum cardiomyopathy was thought about but echocardiogram was found to be normal.

CT pulmonary angiogram was done to rule out pulmonary embolism but was normal, it showed infective pathology in lungs,

CPK was done to rule out myopathies,

ANA levels were done to rule out vasculitic causes, GB syndrome was thought off and LP was

***Corresponding Author**
Dr. Atishaya G V
2nd Year Postgraduate, Department of General Medicine, Sri Devaraj Urs Medical College,
Sri Devaraj Urs Academy of Higher Education and Research,
Tamaka, Kolar-563101
Mobile No : +919986722473
E-mail: atishayaatishaya@gmail.com
Conflict of Interest: None
Financial Aid: Nil

done which did not show albumin cytological dissociation, since the patient had mild ptosis and the neck lift was poor and no other pathology could explain the condition of the patient Myasthenia gravis was thought off for which neostigmine test was done and turned out to be positive, there was no previous h/o diplopia or ptosis.

Anti-acetyl choline receptor antibodies were sent and was highly positive. Patient was started on pyridostigmine patient showed initial improvement and was weaned from ventilator support but patient again became breathless and was not responding to pyridostigmine, patient was diagnosed with Myasthenia gravis in crisis, pulse steroid therapy was started in view of that and patient showed only intermittent improvement and was later subjected to plasmapheresis and the patient condition improved. And now the patient is on treatment for Myasthenia and is doing well.

No family h/o neurological diseases
On examination patient was restless, anxious and labored respiration of 30/min, BP of 60 mm of Hg systolic pressure, pulse rate of 140bpmin and Spo2 of 50% at room air. Cardiovascular system was normal.

Respiratory system/L ISA, ISA crepts present, right upper zone bronchial breath sounds present

Nervous system examination:
Power 4/5 all four limbs
Reflexes diminished
Plantars b/l mute

ECG within normal limits.
ABG Showed type 2 respiratory failure
Chest x ray: Normal
CPK: 47U/L
PSEUDOCHOLINESTERASE:4753U/L
ACETYL CHOLINE RECEPTOR ANTIBODY :20.44U/L
CT THORAX: suggestive of rt lower zone infective pathology and was suggested CECT thorax to rule out thymoma as it was not visualised on plain CT thorax.
Contrast study couldn't be done because of patient condition.
CT BRAIN :WNL
2D ECHO: Normal
SOB PROFILE: Normal

Table 1: A clinical diagnosis of Myasthenia gravis in crisis was made.

Day	HB	TLC	PLT	BU	SC	Na	K
1st	12.3	23.27	399	40	0.2	129	5.8
2nd	10.6	12.29	311	42	0.2	130	3.8
3rd	11.5	13.24	341	34	0.3	131	3.3
5th	11.4	13.54	367	21	0.3	134	3.6
7th	11.3	10.46	343	20	0.4	137	4.3
10th	11	14.21	363	31	0.4	135	4.3
17th	12.4	11.38	383	15	0.3	135	4.7

Discussion

Myasthenia gravis is a neuro muscular disorder primarily characterized by muscle fatigue and weakness, usually the disorder becomes apparent by adulthood, the condition may be restricted to certain muscle groups or may be generalized.

Treatment For Myasthenia Gravis: Neostigmine And Pyridostigmine Are Commonly Used Drugs
Other drugs which can be used are corticosteroids, non-steroidal immunosuppressant.

Myasthenia Crisis: Myasthenia crisis is a condition characterized by worsening of muscle weakness, resulting in respiratory failure that requires intubation and mechanical ventilation in a patient of myasthenia gravis.

1/5th-1/3rd of patients get affected by myasthenic crisis minimum once in their lives once they are diagnosed with generalized myasthenia gravis.[1]

Patient may present in crisis at the time of initial presentation in 1/5th of patients Women are twice more affected than men.

The median time duration from onset of myasthenia gravis to onset of crisis is 8-12 months.

Usually myasthenia crisis occurs following a precipitant like infection, pregnancy aggravates MG and carries high risk of perinatal mortality in these

patients, surgery, sleep deprivation, stress, medical treatment for bulbar myasthenia including steroids, anticholinesterases can precipitate crisis, other commonly used drugs like aminoglycosides, macrolides, anticonvulsants, antimalarial etc. can also precipitate crisis.

Crisis can be early or late onset myasthenia, that is <50 years or after 50 years.

In early onset incidence of crisis is 15-20% but in late onset the incidence may go up to 50% and relapse rate is also higher.

Currently, mortality is 4% and is primarily the result of comorbid medical conditions.

Treatment Of Myasthenia Crisis: They require invasive or non-invasive ventilation in most of the cases.

About 2/3rd to 90% of the cases require intubation and mechanical ventilation

BIPAP is effective for the treatment of acute respiratory failure in patients with myasthenia gravis.[2]

The two important pharmacological therapies for myasthenia crisis are intravenous immunoglobulin or plasma exchange, corticosteroids can be used in conjugation with these two.

Cholinesterase inhibitors usually discontinued during crisis to avoid excessive secretions.[3]

Thymectomy is the surgical option for patients with myasthenia that has the possibilities of complete remission.[4]

Thymic tumors, found in 15% of patients with MG and in 32% of patients with myasthenic crisis, should be treated with thymectomy[5,6.] Patients with non-thymomatous MG can consider thymectomy to improve the likelihood of improvement or remission of the disease.[7]

Differential diagnosis for myasthenic crisis : cholinergic crisis , GB syndrome, Botulism , MND, spinal cord diseases.

Investigations

ANTI -Acetylcholine Receptor Antibody Levels (Highly Specific)

ANTI-Musk Antibody

ANTI LRP4 ANTIBODY

CT AND MRI THORAX: Thymoma an anterior mediastinal mass can be seen.

ELECTRODIAGNOSTIC STUDIES: Repetitive nerve stimulation, SFEMG.

PHARMACOLOGICAL TESTING: Edrophonium test, Neostigmine test.

ICE PACK TEST: Lightly placing ice that is in a surgical glove or that is wrapped in a towel over the eyelid will cool it within 2 minutes.[8] A positive test is clear resolution of the ptosis. [9,10]

Conclusion

Myasthenia gravis is an important neuromuscular disorder and the diagnosis requires high degree of suspicion and hence early diagnosis and treatment reduces complications associated with it and hence mortality and morbidity of patient.

References

1. Jaretzki A 3rd, Barohn RJ, Ernstoff RM, Kaminski HJ, Keesey JC, Penn AS et al. Myasthenia gravis: recommendations for clinical research standards. Task Force of the Medical Scientific Advisory Board of the Myasthenia Gravis Foundation of America. Neurology 2000; 55(1):16-23. doi: 10.1212/wnl.55.1.16.
2. Zhang B, Tzartos JS, Belimezi M, Ragheb S, Bealmear B, Lewis RA et al. Autoantibodies to lipoprotein-related protein 4 in patients with double-seronegative myasthenia gravis. Arch Neurol 2012;69(4):445-51
3. Padua L, Stalberg E, LoMonaco M, Evoli A, Batocchi A, Tonali P. SFEMG in ocular myasthenia gravis diagnosis. Clin Neurophysiol 2000;111 (7):1203-7
4. Li Y, Arora Y, Levin K. Myasthenia gravis: newer therapies offer sustained improvement. Cleve Clin J Med 2013; 80(11):711-21. doi: 10.3949/ ccjm.80a.13044.
5. Meriggioli MN, Sanders DB. Advances in the diagnosis of neuromuscular junction disorders. Am J Phys Med Rehabil 2005; 84(8):627-38.
6. Spillane J, Higham E, Kullmann DM. Myasthenia gravis. BMJ 2012. 345:e8497. doi: 10.1136/ bmj.e8497
7. Juel VC. Myasthenia gravis: management of myasthenic crisis and perioperative care. Semin Neurol 2004; 24(1):75–81. doi: 10.1055/s-2004-829595.
8. Chaudhuri A, Behan PO. Myasthenic crisis. QJM 2009; 102(2):97–107. doi: 10.1093/qjmed/ hcn152.
9. Thomas CE, Mayer SA, Gungor Y, Swarup R, Webster EA, Chang I et al. Myasthenic crisis: clinical features, mortality, complications, and risk factors for prolonged intubation. Neurology 1997;48(5):1253–60
10. Gronseth GS, Barohn RJ. Practice parameter: thymectomy for autoimmune myasthenia gravis (an evidence-based review): report of the Quality Standards Subcommittee of the American Academy of Neurology. Neurology 2000; 55(1):7–15.