Journal of BMANA

Case Report

Myasthenic crisis: atypical presentation

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Abstract

Myasthenia gravis (MG) is a chronic, autoimmune, neuromuscular disease that causes weakness in the skeletal muscles. These muscles connect to the bones, allowing body movement in the arms and legs and breathing through contraction. Muscle weakness worsens after periods of activity and improves after rest. Some patients with MG can present with a myasthenic crisis, which is life-threatening and can result in respiratory failure. It is necessary to take a detailed and thorough history when patients have atypical presentations in order to make timely and accurate diagnoses.

Introduction

MG is a disease of neuromuscular disorder. The most common presenting symptoms include visual disturbances, impaired speech, and limb weakness. The following antibodies are involved in the pathogenesis of myasthenia symptoms:

• **Acetylcholine receptor (AChR) antibodies:** 80% of MG patients have detectable levels in the serum of autoantibodies against AChR. These antibodies are believed to play a central role in disease pathomechanism.

• **Muscle-specific receptor tyrosine kinase (MuSK) antibodies:**

Some patients are seronegative for AChR but positive for MuSK antibodies. Patients with both antibodies are rare, although the frequency may be as high as 10% in some regions in Asia.

Myasthenic crisis is a life-threatening exacerbation of myasthenia gravis that is defined as the worsening of myasthenic weakness requiring intubation or noninvasive ventilation (1). While respiratory failure is due to the weakness of respiratory muscles, severe bulbar (oropharyngeal) muscle weakness often accompanies respiratory muscle weakness or maybe the predominant feature in some patients. Intubation and mechanical ventilation are necessary when this results in upper airway obstruction or severe dysphagia with aspiration.

Although data is limited, the portion of patients with myasthenia gravis who experience at least one myasthenic crisis may be as high as 10 to 20% (2), and the annual risk of myasthenic crisis among the patients with myasthenia gravis is approximately 2 to 3% (3). Myasthenic crisis is the first manifestation of MG for 13-20 % of patients with this disorder (3-5). Most myasthenic crises occur in the first few years after the diagnosis of MG. This case is about a patient who presented with myasthenia crisis and had no previous history of MG.

Case presentation

Chief complaint:

Trouble speaking and swallowing for two days.

HPI:

A 63-year-old man with a history of HTN, HLD was fine until two days ago. He worked in the yard two days ago and developed chest pain. He then started to have slurred speech and dysphagia. His condition was worsening, so he came to the ER.

Past medical history: HTN and HLD

Past surgical history: Gall bladder removed.

Personal history: No smoking, no alcohol or illicit drug use.

Allergy: NKDA

Family history: HTN.

Medication history: Lotensin, HCT

Review of the system: Normal except slurred speech, difficulties in swallowing, dizziness, and chest pain.

Neurological examination:

- **Mental status:** AxOx3
- **Speech:** slurred speech, no aphasia.
- **CN:** II-XII, normal.
- **Sensory:** Normal
- **Coordination:** Normal.
- **DTR:** Normal.
- **Gait:** Normal.

Differential Dx:

- Rule out acute CVA
- Cardiac event
- Physiological
- Non-neurological.

Battery of tests:

- CBC-Normal with MCV 96.2(H), PT/INR -Normal.
- CMP: -Normal. LFT: -normal
- CPK: -52, Troponin: <0.04

Radiology:

- CT head: No stroke.
- MRI of the brain: No acute stroke on DW image.
- CTA of brain: No blood clot, mild distal atherosclerotic disease.
- Carotid u/s: mild plaque
- Xray chest: OK

Cardiac w/u:

- Echo (N) with EF 55%, EKG: sinus @72

Disposition:

- D/C home with advice and f/u with Neuro, Cardio and PCP.
- Meds: ASA, Benazepril and Niaspan

Readmission: He came to ER after two days: with **C/C**

- "I can not breath"
- "I cannot swallow, and I cannot lie flat."

ER evaluation:

- Patient is sitting upright, breathing fast, hard, shallow and gasping for air.

Lying down on bed is causing respiratory stridor. Speech-slurred and dysarthric. No double vision. Motor and Sensory examination: Normal.

ABG: 7.4/43.5/75/27.1/95%

DDX in ER

- PE. vs acute oropharyngeal dysmotility vs CVA
- Consult: Pulmonary, GI and Neurology.

Tests done at this point:

- Xray chest. ultrasound of chest, U/S for DVT, came out - Normal.

Pharyngogram: could not do because he could not lie flat.

Condition deteriorated and the patient was moved to MICU. Pulmonary, GI and Neuro on the case hunting for diagnosis.

Neurology:

- Patient gasping, hardly speaking, no double vision on exam, DTR (N),but diplegia of facial muscles.
- Ddx: Atypical GBS vs brainstem stroke
- Plan: MRI of the brain, spinal tap. The patient could not do MRI; he could not lie flat.

Pulmonary:

- Bilateral diaphragmatic failure. VQ scan –(N),

Bronchoscopy: (N)

Gastro:

- EGD –normal.

Medical Boards:

- Neurology, Pulmonary, and Gastroenterology sat together and discussed the case:

Neuro got a history from the daughter that his problem was getting worse over two months. Neuro suspected neuromuscular disease like Myasthenia Crisis, new onset without a history of Myasthenia Gravis before.

Plan:

- Immediate Bedside Tensilon Test. While waiting for Tensilon test, a spinal tap was done.

- Tensilon Test was done by author. The patient improved instantly. Speech got better and respiration improved.

- **Myasthenia Graves was confirmed .**
- Plasmapheresis and mestinon were ordered.
- Discharged home after 10 days with full strength, normal speech and respiration.

Discussion:

A rapid and accurate diagnosis is very important for the well-being of a patient in myasthenic crisis and to decrease the mortality and morbidity from the crisis. Obtaining a thorough history of the present illness, including collateral information from the family, can increase the likelihood of making the proper diagnosis. It is important to remember that a patient's presentation can be unique or different from those symptoms most commonly found. Collaboration between specialties and the early involvement of consultants can assist in ruling out diagnoses and confirming a suspected one.

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