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Case Report

A case of Myasthenia Gravis masquerading as GBS

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Abstract

Myasthenia gravis (MG) is an autoimmune neurologic disease that affects the post synaptic portion of the neuromuscular junction. Usually MG occur in young adults or in the elderly characterized by weakness of fatigue of skeletal muscles due to repetitive use.[1-2] It represents a challenge for clinicians due to the diversity of disease manifestations. It is therefore important to monitor the neuromuscular blockade (NMB) due to the multiple presentations of MG. In our case the patient presented with clinical features suggestive of Guillen Barre Syndrome (GBS) which is an autoimmune disease against myelin nerve sheath. Initially he was planned to be treated as GBS because of it's unusual descending paralysis like presentation. Nerve conduction studies were done a few times and were found to be normal. Anti acetylcholine receptor antibody test was found positive. Plane X Ray of chest and CT scan of Chest showed a mediastinal mass suggestive of Thymoma. Patient recovered dramatically after receiving treatment for Myasthenia Gravis.

Key word: *Myasthenia gravis (MG), Guillen Barre Syndrome (GBS), Nerve conduction study (NCS), Anti acetylcholine receptor anti body.*

Case Report:

A 12 years old boy was admitted in neurology in patient unit of National Institute of Neurosciences and Hospital (NINSH) on Dec 20,2017 with the complaints of drooping of upper eye lid of right eye, double vision followed by pain & weakness of neck muscle, nasal regurgitation of food with nasal intonation of voice for 10 days. He also developed weakness of all limbs during this period for which he was unable to perform his daily activities and had to quit going to his school. Two days after admission he developed respiratory distress on exertion which needed him to be admitted in NINSH ICU for better management. He had no H/O fever, cough, diarrhea, trauma or taking canned food prior to this illness. On examination, he was found conscious, oriented, with right sided incomplete ptosis with bilateral facial muscle weakness and broken neck sign. His cough and gag reflexes were absent and his neurological examination of four limbs revealed quadriparesis with reduced muscle power 3/5 of both proximal as well as distal muscle group, generalized arreflexia with flexor plantar response but intact sensory function. His fundus was normal.

On ICU admission he was immediately intubated for airway protection and was kept on mechanical ventilation for type 2 respiratory failure evidenced on ABG. His Glasgow coma scale (GCS) was 15/15 and he was febrile with rectal temperature 101^0 F and was haemodynamically unstable with Spo_2 – 94% in room air, Pulse was 120bpm, BP-80/60mm of Hg, Breathing- Laboured. Lungs – B/L diminished breath sound in lung base & crackles in right lung more than left.Investigations were done to find out the cause.

Case Report:

NCS of cross limb and repeated nerve stimulation (RNS) of right median nerve was done and showed normal study. Lab investigations showed Hb 13%, ESR 33, Leukocytosis(WBC 31 x 10^9/ L), RBC 06 x 10^{12}/ L, Platelet 229x 10^9/ L, S. Creatinine 0.61 mg/dl, serum CPK 211u/ L S.Total calcium 2.6 mmol / L, serum Magnesium 0.84 mmol/L, CSF study showed Glucose 5.43 mmol /L, protein 72.1 mg /dl. MRI of brain was negative for hemorrhage, infarction or SOL. Blood sugar, renal function& liver function tests were normal.

X Ray Chest showed small mass on left hilum. (Fig 1). CT scan of Chest revealed a hilar mas on left medistinum (Fig 2)

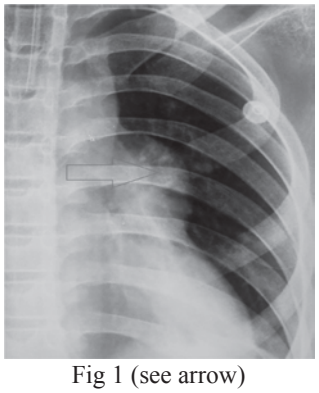

Fig 1 (see arrow)

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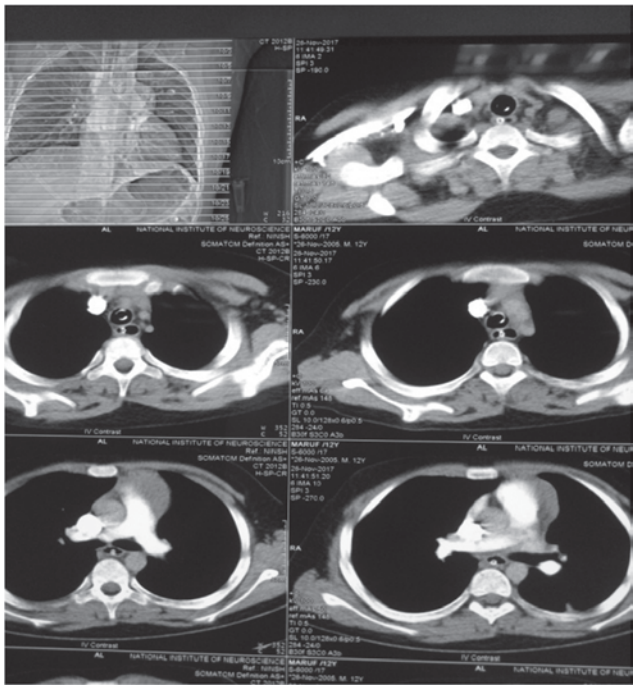

Fig 2 (see arrow)

Cultures of blood, urine & tracheal aspirate showed no growth. Acetyl choline receptor antibody test was sent and meanwhile injectable Neostigmine along with methyl prednisolone was started for prior to test report which showed significant improvement of his sign and symptoms within next 5 days and later on his tests results showed highly positive Acetylcholine Receptor Auto-antibodies to postsynaptic Ach receptor diagnostic of MG.

Injectable Neostigmine and methyl-prednisolone was then switched to oral pyridostigmine and prednisolone respectively. Along with other supportive management single session plasmapharesis was given. Steroid sparing immune-modulatory drug Mycophenolate Mofetil was started also. His condition was gradually improving and he was kept on weaning trial from ventilator in next 8 days and 3 days after extubation was done. Patient was observed for 2 days then discharged from ICU to general neurology ward of the hospital with proper medication & follow-up advice.

Discussion:

Myasthenia gravis is a chronic neuromuscular disorder affecting the post synaptic acetylcholine (Ach) receptor or its MSK enzyme of NMJ. Myasthenia gravis is caused by an error in the transmission of nerve impulses to muscles. It occurs when normal communication between the nerve and muscle is interrupted at the neuromuscular junction—the place where nerve cells connect with the muscles they control.Neurotransmitters are chemicals that neurons, or brain cells, use to communicate information. Normally when electrical signals or impulses travel down a motor nerve, the nerve endings release a neurotransmitter called acetylcholine. Acetylcholine travels from the nerve ending and binds to acetylcholine receptors on the muscle. The binding of acetylcholine to its receptor activates the muscle and causes a muscle contraction.

In myasthenia gravis, antibodies (immune proteins) block, alter, or destroy the receptors for acetylcholine at the neuromuscular junction, which prevents the muscle from contracting. In most individuals with myasthenia gravis, this is caused by antibodies to the acetylcholine receptor itself. However, antibodies to other proteins, such as MuSK (Muscle-Specific Kinase) protein, can also lead to impaired transmission at the neuromuscular junction.

These antibodies are produced by the body's own immune system. Myasthenia gravis is an autoimmune disease because the immune system which normally protects the body from foreign organisms mistakenly attacks itself.

The thymus is a gland that controls immune function and maybe associated with myasthenia gravis. Located in the chest behind the breast bone, the gland is largest in children. It grows gradually until puberty, and then gets smaller and is replaced by fat. Throughout childhood, the thymus plays an important role in the development of the immune system because it is responsible for producing T-lymphocytes or T cells, a specific type of white blood cell that protects the body from viruses and infections[3]. In our case X ray chest and CT scan of chest showed a small mediastinal mass which could be indicative of thymoma. Thymoma may be associated with MG according to standard text books.

Myasthenia gravis geo epidemiology shows that it is a rare disorder with and prevalence in the world, except for infantile MG, which is more similar incidence common in Asia.The incidence has increased in the last decades, going from 2-5/1,000,000 to 9-21/1,000,000, but without proportional increase in mortality[4].

Where GBS is another very important autoimmune disease with similar molecular immune-pathology in our subcontinent in which auto-antibodies develop against myelin nerve sheath of peripheral nerve. GBS can unusually presents acutely with descending paralysis started from eyelid to facial muscle, neck muscle, respiratory muscle to limb muscle along with arreflexia and opthalmoplegia, bulbar palsy as observed in our case. For this reason our patient was initially suspected to have GBS and was planned to be treated for GBS. But suspicion of another diagnosis grew after repeated normal NCS study prior to commencing treatment for GBS.

Neck pain & dysphagia is a rare clinical manifestation in MG as the initial presentation. Considering the extensive diagnostic hypotheses involving the swallowing pathology, the neuromuscular pathology must be suspected after a medical history and physical examination,valuing the accompanying symptoms, daytime evolution of the pathology and aggravation factors.According to electrophysiological assessments, the most affected muscles when the disease presents with dysphagia are the muscles of the tongue,the larynx elevators, pharyngeal constrictors, cricopharyngeal muscle[5]and the muscles of the distal esophagus[6].

Misra et al [7] reported a similar case of MG presenting with areflexia, ascending paralysis and bilateral ptosis suggestive of GBS and diagnosis of MG was confirmed with positive anticholinesterase antibodies and tensilon test. MG and GBS can occur concurrently but it is very rare. Kung et al [8] reported a patient with concurrent MG and GBS as a first case in Taiwan. The author hypothesized that such concurrence can be associated with molecular mimicry between infectious agents and self antigens, such antibodies may show cross reactions against both myelin proteins of peripheral nerves and acetylcholine receptors of neuromuscular junctions. Thymoma associated multi-organ autoimmunity may also play role in initiating autoimmune process.

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