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CASE REPORT

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Myasthenia gravis following chicken pox infection in a Nigerian primary school girl

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Abstract We report a case of myasthenia gravis occurring post Chicken pox infection in a 9 year old Nigerian girl. The girl was growing and seeing normally until 2 weeks after a chicken pox infection when she observed progressive drooping of both upper eye lids which worsens as the day progresses and has persisted for more than one year. There was good response to Neostigmine.

Introduction

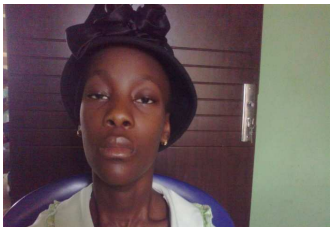
Myasthenia gravis (MG) is an autoimmune neuromuscular disorder resulting in fluctuating muscle weakness and fatigability.[1] The weakness is caused by circulating antibodies that block acetylcholine receptors at the postsynaptic neuromuscular junction inhibiting the excitatory effects of the neurotransmitter acetylcholine on nicotinic receptors in neuromuscular junctions.[2] The incidence of myasthenia gravis is 3-30 cases per million per year.[3] Myasthenia gravis occurs in both genders and affects all races. All age groups can be affected but most commonly people from 50 to 70 years. In females incidence of MG peaks in the third decade of life, whereas it peaks in the 6th and 7th decades in males.[4]

Case Presentation

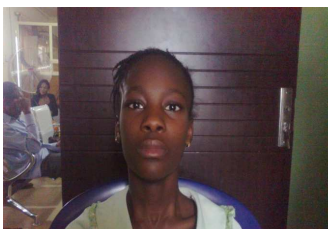
A.M., a nine year old primary 6 girl presented with 13 month history of variable drooping of both upper eye lids and double vision. She observed difficulty in lifting up both upper eye lids after engaging in some activities as the day progresses making it difficult for her to see clearly. She therefore had to lift her chin upward to allow her to see. She usually felt normal, able to see without difficulty, on waking up from sleep but becomes weak and easily gets tired as the day progresses particularly when she engages in her daily activities. There was associated double vision and difficulty seeing distant objects. She had chicken pox two weeks before onset of above symptoms. In addition she also complained of painful swallowing and chest pain.
Examination revealed a young girl who was not small for age, not pale, afebrile, anicteric . Significant findings were on ocular examination . With manual elevation of the eyelids visual acuity were normal with glasses. She had bilateral ptosis, ophthalmoplegia, pupils were 3mm equal bilaterally and reacting normally to light. Posterior ocular segments were normal bilaterally. Other systems were essentially normal.

A diagnosis of myasthenia gravis probably secondary to thymoma was made.

Investigations: Antibodies against Acetylcholine receptor was not detected; Full blood count showed relative lymphocytosis (PCV 44%, total WBC 5,300; Neutrophil 24%. Lymphocytes 72%, Monocytes 4%); Erythrocyte sedimentation rate 11mm/hr; Electrolyte and urea within normal limit; Chest X-ray, no abnormality detected, Magnetic resonance imaging of the skull and Computed tomography of the chest revealed a normal study.
She was given intravenous Neostigmine 0.05mg/kg (2.5mg) slowly with remarkable improvement: the two upper eyelids opened widely within 5 minutes of administration of Neostigmine. Thereafter she was placed on oral Neostigmine 7.5mg qds with sustained clinical improvement and patient continued to do well, able to see without lifting her chin upward. She was also reviewed by an ophthalmologist .



Before administration of Neostigmine.



After administration of Neostigmine

Discussion

Myasthenia gravis is idiopathic in most patients.[1,4] In children, three types of myasthenic syndrome can be distinguished:[1] 1. Neonatal: Pregnant mothers with myasthenia gravis in 12% of cases pass the antibodies to the infant through the placenta, causing neonatal myasthenia gravis. The symptoms will start in the first two days and disappear within a few weeks. 2. Congenital: this occurs very rarely in children of healthy mothers with symptoms beginning at birth. It is not caused by an autoimmune process but due to synaptic malformation, which in turn is caused by genetic mutations and the inheritance pattern is typically autosomal recessive. 3. Juvenile myasthenia gravis: it occurs in children but after the peripartum period. The hallmark of myasthenia gravis is fatiguability.[1] In our patient eyelid muscle weakness was the first symptom noticed. This has been reported to be the case in most cases, while in a few others difficulty in swallowing and slurred speech may be the first presentation. The muscles become progressively weaker during periods of activity and improved after periods of rest. The muscles controlling movement of the eyeballs and eye lids, facial expressions, chewing, talking, and swallowing are particularly susceptible.[1] Often, physical examination yields normal finding.[5] The degree of muscle involvement varies raging from localized form limited to the eye muscles (ocular myasthenia) to a generalized, severe form affecting many muscles, sometimes including respiratory muscles.

The patient reported above did not have respiratory muscle involvement but obviously seemed to have some weakness of other muscles because of the history of easy fatiguability after engaging in her daily activities. Symptoms varies including; asymmetrical ptosis, diplopia (due to weakness of muscle that control movement of the eyeballs), unstable/ waddling gait, weakness in arms, hands, fingers, legs, and neck, a change in facial expression, dysphagia, shortness of breath and dysarthria. The patient above did not have any thymus involvement even though it has been reported that up to 75% of myasthenia gravis patients have an abnormality of the thymus and 10% have a thymoma.[1,4] In myasthenic crisis there is paralysis of the respiratory muscles necessitating assisted ventilation to sustain life. Crisis is often triggered by infection, fever, adverse drug reaction or emotional stress in patients whose respiratory muscles are already weak.[6] Cardiac muscle is generally not affected by MG since it is regulated by autonomic nervous system.[1] Myasthenia gravis is an autoimmune channelopathy featuring antibodies directed against the body's own proteins. Our patient had a history of chicken pox two weeks before onset of myasthenic symptoms. The causative role of this preceding infection cannot be proven. Although it has been proposed that sensitization to a foreign antigen which has cross reactivity with acetylcholine receptor could be a cause of MG but there is yet no known infective agent that could account for this.[4] Various drugs may also induce or exacerbate symptoms of MG which often resolve after discontinuation of the drugs.[4] Other findings associated with MG includes: female, and people with certain human leucocyte antigen types (HLA-BB, HLA-DRw3, and HLA-DQw2). We could not do an HLA typing in this patient. MG is also associated with various autoimmune diseases like Hashimoto's thyroiditis, Grave's disease, rheumatoid arthritis, diabetes mellitus type I, Lupus and demyelinating CNS diseases.[1,4]

Diagnosis of myasthenia gravis can be difficult as the symptoms can be subtle.[5] A thorough physical examination can reveal easy fatigability, with the weakness improving after rest and worsening again on repeat exercise.[1,4] There was improvement in the symptoms of the above patient after rest. Our patient possibly fell in the group of patients with negative antibodies to acetylcholine receptors much more so that her symptoms were predominantly ocular. In suspected cases serology can be performed to detect antibodies against the acetylcholine receptor. This test has a sensitivity of 80-95% in generalized severe cases but may be negative in up to 50% of ocular myasthenia gravis.[5] Patients who are negative for anti-acetylcholine receptor antibodies may be seropositive for antibodies against MuSK protein.[1,4] Electromyography considered to be the most sensitive test is not very specific for MG.[5] Endrophonium test is limited to situation where other investigations do not yield a conclusive diagnosis.[4,7] In this test intravenous endrophonium chloride or Neostigmine is administered; these drugs block the breakdown of acetylcholine by cholinesterase and temporarily increase the levels of acetylcholine at the neuromuscular junction thereby relieving weakness temporarily in ocular myasthenia gravis.[7] A chest X-ray may identify widening of mediastinum suggestive of thymoma, but CT or MRI are more sensitive ways to identify thymomas which are closely associated with MG.[4,8] MRI of the cranium and orbits is also performed to exclude compressive lesions of the cranial nerves and ocular muscles.[4] All these tests were negative in the case reported.

Treatment involves use of cholinesterase inhibitors such as Neostigmine or Pyridostigmine.[1,4] Corticosteroids are typically used in moderate or severe cases of MG that fail to respond adequately to acetylcholine inhibitors and thymectomy.[4] Thymectomy is the standard treatment for all patients with thymoma and for patients aged 10-55years without thymoma but generalized MG.[1,4,] Immunoglobulin is useful during myasthenia crisis in patients with severe weakness poorly controlled with other agents. The effect is rapid but transient and the same is true for plasmapharesis.[1,4,6] The reported case responded very well to Neostigmine tablets with relieve of symptoms.

Conclusion

we report this case of MG following chicken pox infection in a nine year old Nigerian girl to serve as a template for further investigation into the yet unknown pathogen associated with MG.

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