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Case Report
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Myasthenia Gravis: A Differential for Foreign Body Aspiration in Adolescent

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Abstract

Foreign body aspiration is uncommon in adolescents. When faced with foreign body aspiration at an unusual age, it is important to consider potential underlying causes. We present a 14-year-old girl who presented to the emergency department twice over a 6 months period, both times aspirating whilst eating. Myasthenia Gravis was suspected and subsequently diagnosed.

Keywords: Foreign body; Aspiration; Adolescent; Myasthenia gravis

Introduction

Myasthenia Gravis (MG) in the pediatric population is rare. The disease is categorized into pathophysiological mechanisms, including neonatal MG, congenital MG, and juvenile MG.

Neonatal MG usually occurs in neonates, due to autoimmune antibodies crossing the placenta, hence leading to disruption of the neuromuscular junction. Congenital MG tends to occur in infants, due to genetically inherited diseases of the neuromuscular junction [1]. This discussion focuses on Juvenile MG (JMG), a rare autoimmune condition that occurs before the age of 19.

JMG is caused by autoimmune antibodies, like adult MG. The antibody can target different components of the neuromuscular junction. Most commonly the autoantibody binds to the nicotinic acetylcholine receptor on the postsynaptic membrane, hence impeding neuromuscular transmission. About 10% of all autoimmune MG patients are children or adolescents [2].

Compared to adult-onset MG, pre-pubertal onset MG has a higher prevalence of isolated ocular symptoms, and a higher probability of remission (both spontaneously or with treatment), and is less likely to progress to generalized MG when compared to adults. The female-to-male ratio is equal in patients with pre-puberty onset, but there is a higher female-to-male ratio from puberty onwards [1].

JMG typically presents with ocular symptoms, including ptosis, asymmetrical ophthalmoplegia, diplopia and strabismus. Bulbar involvement can lead to dysphonia and dysphagia. The generalized form of the disease causes proximal limb weakness. Rarely can it involve respiratory muscles. All symptoms are fatigable, where they are worse towards the end of the day and improve with rest [1].

The clinical presentation of MG is usually gradual with multi-system symptoms, hence not often included in the differential of dysphagia or dysphonia in an ENT setting. Here we present a case that demonstrates the possible acute and chronic ENT-related presentation of MG.

Case Presentation

A 14-year girl presented to the emergency department after choking on a pumpkin whilst eating in the supine position. She was unable to speak. She got her family's attention by pointing at her own neck. The patient received abdominal thrusts from her family member, which led to vomiting, harsh breath sounds, and drooling. Her chest sound was normal on auscultation, and her chest X-ray was normal. Her vital signs were normal in the emergency department, but due to a history of choking, she was brought to the operating room for rigid esophagoscopy and bronchoscopy. Pumpkin was found in the right main bronchus and the Posterior segmental bronchus of the left lung. Postoperatively the patient developed aspiration pneumonia. She received antibiotics and was referred to psychology for counseling.

Six months later she was brought back to the emergency department by ambulance after aspirating whilst eating broccoli. This time she was eating normally sitting up at the table. She

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collapsed and became cyanosed. Her family members called the emergency services and commenced CPR as per instructions over the phone. Paramedics arrived within minutes, and with a laryngoscope, they were able to visualize a large chunk of broccoli in her throat. The broccoli was removed with McGill forceps, and her breathing rapidly improved. Her saturation dropped temporarily to the 80s in the ambulance and was given supplemental oxygen. She rapidly improved in the emergency department, was weaned off oxygen and returned to her baseline status. She was taken back to the operating theatre for rigid esophagoscopy and bronchoscopy, which revealed no foreign objects in the upper aerodigestive tract.

On further history taking, the patient revealed that she's been having difficulty with swallowing for about two years. In particular, she noticed that she needs to turn her head to help with swallowing food, especially chunky textured food. Due to this difficulty, she has been avoiding certain food groups, such as meat. The family has also noticed that her voice has progressively gotten weaker and quieter. She developed right-sided ptosis and diplopia between her first and second presentations.

Myasthenia Gravis was suspected. Acetylcholine Receptor (AchR) antibodies were requested. This was strongly positive, at >8 nm/L, which is 20 times the upper range of normal. She was referred to pediatric neurology for further assessment and management.

She was started on pyridostigmine. MRI of the thymus showed a normal thymus. The patient was referred for thymectomy, with plans of IVIG bridging therapy and immunosuppression.

Discussion

Foreign body aspiration occurs predominantly in children under 3 years of age. This is caused by an immature neuromuscular system for airway protection whilst swallowing, and immature dentition, in a phase of life where exploration is done by inserting objects into the mouth. Foreign bodies in older children are much less common. A retrospective study including 148 children showed that the mean age of aspiration in 1- to 18-year-olds was 4.23 years [3]. This finding was again confirmed by larger studies. Chapin et al. found that in over 111,000 children 14 years old or younger who presented to the emergency department for non-fatal food-related choking in the United States between 2001-2009, the mean age was 4.5 years and 37.8% of the children treated were 1 year old or younger [4].

As our patient had 2 episodes of foreign body aspiration within 6 months, at an age when aspiration is uncommon, with long-term issues with swallowing, voicing, and ptosis, the suspicion of underlying causes was raised. MG was included in the differential diagnoses.

Diagnosis of MG composes of symptoms suggestive of MG, autoantibodies against AchR, and electrophysiological testing which demonstrates gradually reduced action potential with repeated nerve stimulation. Our patient's AchR antibodies were strongly positive. 85% of the time AchR antibodies are detected. Negative autoantibodies do not exclude the diagnosis of MG, as they can develop against other components of the neuromuscular junction [2].

The ENT service can be the first point of contact for patients with MG. Liu et al. presented a case series, based on 7 patients who presented with dysphonia as a primary complaint, out of 1,520 patients who were diagnosed with myasthenia gravis over a 15-year period. Half of these patients also had concomitant dysphagia. Just under half of these patients developed subsequent ocular symptoms [5]. Yang et al. presented a case series on 30 MG patients with primary laryngeal manifestation. The ENT department was the first point of specialist contact in 63.3% of these patients. Dysarthria was the most common primary symptom (46.7%), followed by dysphagia (36.7%), slurred speech (13.3%) and dysphonia (3.3%). Subsequent symptoms included ptosis, cough, nasal regurgitation, loss of chewing power and limb weakness [6].

MG can cause acute vocal cord paresis. Khan reported a 71-year-old man who had repeated presentation of rapid progressive shortness of breath and dysphagia. The patient was found to have bilateral vocal cord palsy with cords fixed in the paramedian position. He required 2 emergency tracheostomies on 2 separate occasions. The patient was initially diagnosed with stroke due to MRI showing bilateral ischemic changes. MG was later diagnosed with positive anticholinesterase antibodies [7]. Vocal cord paresis is rare, with a study in 1972 documenting 6 out of 147 MG patients over a 12-year period having abductor paresis, of which 4 required tracheostomies [8].

The thymus must be imaged, usually with either CT or MRI. The thymus is believed to be central in the pathogenesis of MG, due to the lack of self-tolerance of T cells, or *via* the paraneoplastic process of a thymus neoplasm. MG is associated with thymus hyperplasia and thymoma. 10% to 15% of MG patients have thymoma [4].

The treatment of MG consists of symptomatic treatment and immunosuppressive treatment. Symptomatic treatment includes acetyl cholinesterase inhibitors such as pyridostigmine. Immunosuppressive treatment includes glucocorticoids, azathioprine, cyclosporine, methotrexate, mycophenolate mofetil and Tacrolimus [4].

In addition to the above, IVIG and plasmapheresis are used in myasthenic crisis, or special circumstances such as pregnancy [4]. In this case, IVIG was planned for bridging to thymectomy.

Thymectomy is always performed unless there are contraindications [4]. The MGTX trial has shown that in non-thymomatous MG, a patient receiving thymectomy and prednisone, compared to prednisone alone have less symptom burden, and was on lower doses of prednisone [9]. This is the reason that despite MRI showing a normal thymus in our patient, she was regardless, referred for thymectomy.

A PubMed search reveals only one other similar case. Patel and Murray reported an 8-year-old boy who was diagnosed with MG after aspiration of popcorn. The child was found to be cyanotic on paramedic arrival. Heimlich Maneuver was performed which improvement in ventilation, but the child had ongoing tachypnea, stridor and coarse breath sounds. The patient was noted to have poor spontaneous ventilation. Undigested popcorn was seen in the pharynx and the trachea. His parents later revealed that he had 6 months history of deterioration in speech, difficulty writing, and inability to keep up with sports. JMG was diagnosed with an electrophysiological study. He was treated with 5 sessions of plasmapheresis and pyridostigmine. He underwent a thymectomy 4 days later. At 6 months the patient was in remission, taking no medications [10,11].

Conclusion

The ENT services can be the first point of contact for MG; hence it is necessary to keep in mind MG as a differential when facing patients with symptoms such as dysphagia or dysphonia, in both acute and

chronic settings. This is especially important to consider in cases with atypical demographic or clinical features.

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