Clinical Presentation

11 year old male presents in April with chief complaint:

- Tiredness and weakness after physical activity
- Difficult to raise arms over head.
- Speech dysarthria
- Left eyelid ptosis compared to right
- 3-4/5 strength in proximal muscles, 4-5/5 in distal muscles.

Background Info on Myasthenia Gravis

Pathophysiology: Myasthenia Gravis (MG) is an autoimmune disorder mediated by autoantibodies directed against components of the neuromuscular junction, usually the Acetylcholine receptor (see above). Compare to adults, children are more likely to have no detectable autoantibodies. Most MG-positive children have thymoma. MG-positive children are most common. One case report in 2012 stated that there were 1.6 per million are affected per year. ~80% of cases are female. Thymomas in children are even less common. One case report in 2012 stated that there were only 10 cases of MG associated with thymoma in children.

Symptoms:
- Occular symptoms are most common presenting symptoms. ~70% of cases present with ptosis, along with unilateral ophthalmoplegia, strabismus and blepharospasm.
- Symptoms are made worse by sustained upward gaze. May also experience painless fatigueability of bulbar and limb muscles which manifests as dysphagia, dysphonia and proximal limb muscle weakness. Myasthenia crisis is a life threatening crisis and failure is imminent and often mechanical ventilation is required.

Diagnositc Tests:
- Autoantibodies: Acetylcholine receptor antibodies (ex. myasthenoidis) are first line therapy, but only provide several hours of symptomatic relief. Thymectomy increases the chance of remission in AChR seropositive prepubertal patients. Steroids are often an adjunctive therapy to improve symptoms and are used for crisis. Improvement after IVIG is temporary, usually lasting 4-6 weeks.

Special Considerations:
- Autoantibodies to muscle-specific kinase (MuSK) is a subset who present with more severe disease. There are no reports of thymoma in MuSK-positive children. Patients with MG also have very unpredictable reactions to neuromuscular blocking agents, so special consideration must be taken if they are to undergo anesthesia.

Treatment Options:
- Acetylcholinesterase inhibitors (ex. pyridostigmine) are first line therapy, but only provide several hours of symptomatic relief. Thymectomy increases the chance of remission in AChR seropositive prepubertal patients. Steroids are often an adjunctive therapy to improve symptoms and are used for crisis. Improvement after IVIG is temporary, usually lasting 4-6 weeks.

Treatment and Outcome for Our Patient

He was initiated on pyridostigmine and underwent x3 IVIG treatments in order to manage his weakness. Thymectomy was performed on 5/18/17 (pathology above). He did not experience exacerbation of weakness or respiratory deterioration post-OP, and has been recovering as expected.

References
5. Gilbreath, Peter, MD, PGY3; Greer, Chelsee, DO, PGY1; Kahlden, Kyle, MD, PGY2; Cossey, Melissa, MD. (2018). Autoimmune Myasthenia Gravis: A Clinical Experience. Pediatric Myasthenia Gravis: A Clinical Experience

Chest X Ray

![Chest X Ray Image]

Chest MRI

![Chest MRI Image]

Thymus must be imaged to evaluate for thymoma. The first image is an H&E of the resected lesion. The upper right shows thymic tissue with intact cortical-medullary architectural pattern that would be seen in normal thymus. The lower left is effaced thymic cortex with thymic hyperplasia. This feature is more in keeping with thymoma (WHO Type B1), rather than thymic hyperplasia, which is most common in children. The lower right is a thymus with thymic hyperplasia and a normal cortical pattern in the preserved thymus. The second image is an MRI of the resected lesion. The upper right shows thymic tissue with intact cortical-medullary architectural pattern that would be seen in normal thymus. The lower left is effaced thymic cortex with thymic hyperplasia. This feature is more in keeping with thymoma (WHO Type B1), rather than thymic hyperplasia, which is most common in children. The lower right is a thymus with thymic hyperplasia and a normal cortical pattern in the preserved thymus.