A multidisciplinary Myasthenia and Thymoma Program at Penn Medicine has been established to coordinate and expedite the management of patients with myasthenia gravis (MG), thymoma and related diseases. Staffed by neurologists, thoracic surgeons, radiologists, ophthalmologists and many other specialists, the program has the objective of providing accurate diagnoses and medical management for MG and surgical intervention for thymoma and other thymic disorders.

Myasthenia gravis is an autoimmune disorder caused in most patients by antibodies that destroy acetylcholine receptors (AChR) at the neuromuscular junctions of striated muscles. The resulting loss of AChR manifests in patients with MG as progressive muscle weakness. The course of MG is variable and symptoms are generally nonspecific, leading frequently to missed or mistaken diagnoses.

Treatment for MG can substantially improve the symptoms of the disease. Thus, a missed diagnosis means, at the least, a continued deterioration in quality of life. However, for myasthenia gravis patients with thymoma (an associated disorder that affects up to a third of patients with the disease), or any of a host of comorbidities treated with drugs that inhibit neuromuscular transmission, a missed diagnosis can have profoundly deleterious effects.

At Penn, specialists across the therapeutic spectrum are trained to recognize the early signs of MG so that a confirmatory diagnosis can be made early in the course of the disease. This is typically achieved by blood tests for the presence of antibodies to acetylcholine receptors, electromyography (EMG) and if needed, single-fiber electromyography. Once confirmed, patients have radiographic scans to check for thymoma.

Medical treatment for MG at Penn may include cholinesterase inhibitors (e.g., Mestinon®), steroids and immunosuppressants, which increase the relative amount of acetylcholine by reducing the immune-mediated attack against the AChR. Two procedures are also commonly used to treat MG at Penn. The first, plasmapheresis, removes AChR antibodies from the blood through a process resembling dialysis; the second, intravenous immune globin (IVIg) therapy, involves infusing pooled gamma globulin to restructure the immune response to AChR. Expanding upon these options, Penn is also a thriving source of clinical trials for patients who are refractory to therapy.

With improvement in MG symptoms, patients with early stage thymoma and selected individuals without thymoma may be candidates for thymectomy. When possible, robotic surgery is preferred as an alternative to sternotomy and transternal procedures, which are associated with longer hospital stays, increased operative duration and greater blood loss.

CASE STUDY

Mr. D, a 68-year-old man, was referred to a specialist at Penn Otorhinolaryngology-Head and Neck Surgery after a 15-month history of episodic choking and recurrent aspiration pneumonia. Noting mild ptosis that increased in severity upon sustained upward gaze, Mr. D was referred to Penn Neurology, where an anti–acetylcholine receptor (AChR) antibody (Ab) test and electromyography confirmed myasthenia gravis. A chest CT subsequently identified a 4 cm mass in the anterior mediastinum consistent with a thymoma.

Following plasmapheresis and IVIg with concomitant Mestinon® (pyridostigmine bromide) therapy, Mr. D’s symptoms improved sufficiently to permit surgery, and he was scheduled for a robotic thymectomy.

Prior to surgery, an epidural was placed to optimize postoperative pain management; a right chest approach was utilized along with a 3-port technique. Robotic dissection began with the right pericardial fat pad and progressed both cranially and to the left. Both phrenic nerves could be well visualized using the 3-dimensional camera system, and both were preserved during skeletonization, which included all the surrounding fat and tissue (Figure 1). There was no evidence of pericardial involvement as the specimen was dissected free. The brachiocephalic vein was skeletonized and dissection proceeded into the neck to include both upper poles.

On postoperative day 1, Mr. D’s single chest tube was removed and he was discharged home that afternoon on oral narcotics. He required the narcotics for only the first week, resumed his dose of Mestinon, and was able to return to his normal activities after the second week. He continues on Mestinon therapy but at a declining dose two years later.
FACULTY TEAM
The Penn Medicine Myasthenia and Thymoma Program is comprised of a multidisciplinary team of thoracic surgeons, neurologists and other specialists dedicated to the comprehensive management of patients with myasthenia gravis and its associated effects and conditions, including thymoma.

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Figure 2: Enhanced chest CT scan demonstrating a 3cm solid mass (arrow) in the anterior mediastinum consistent with a thymoma.

Computed Tomography for Thymoma
At Penn Medicine, patients with myasthenia gravis typically undergo computed tomography (CT) imaging to rule out concomitant thymoma. When a thymoma is identified (as in Figure 2), CT is used to characterize the tumor and to investigate the presence and extent of local invasion. CT scans are also an important source of incidental findings of thymoma in patients assessed for diseases other than myasthenia gravis.