Thymoma with Paraneoplastic Myasthenia Gravis

Dallin Beus DO, Sherwin Thomas MD, Amnah Andrabi MD

Medical City Arlington

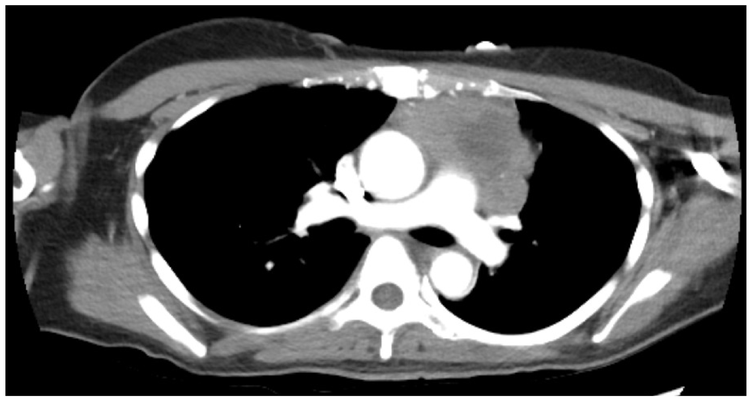
Background

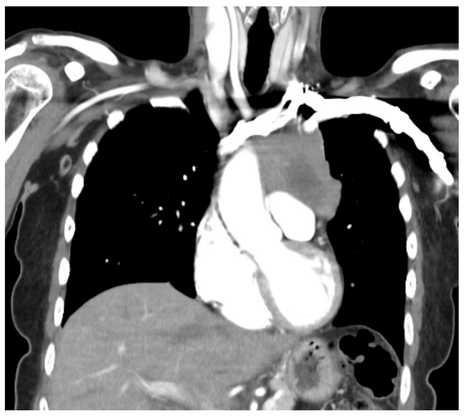
Myasthenia gravis is an autoimmune neuromuscular disorder characterized by muscle weakness and fatigue. There are several effective treatment options available for myasthenia gravis patients. About 10% of the time, myasthenia gravis is associated with thymic tumors and about 65% of the time with thymic hyperplasia. Part of the standard therapy for many of these patients involves thymectomy. However, surgery is a known risk factor for causing myasthenic crisis, a life threatening complication of myasthenia gravis. This case highlights strategies for diagnosing and treating myasthenia gravis and minimizing the risk of post-operative myasthenic crisis.

Case

A 51-year-old female with no significant past medical history presented with a one-month history of progressive, generalized muscle weakness worse in the upper extremities, weight loss and dysphagia. Portable Chest X-ray performed in the ER showed a 7.5cm lobulated density in the left chest and hilar region. CT imaging of the chest showed an 7.3 x 3.3 x 8.2cm soft tissue mass in the anterior mediastinum concerning for lymphoma vs thymoma. At this time the oncology team was consulted. Serologic testing to confirm the type of mass was performed including AFP, HCG, and flow cytometry, which were all negative. Interventional radiology was consulted for biopsy of the mass. Percutaneous CT guided biopsy of the mass was performed and histological evaluation of the tissue confirmed the diagnosis of thymoma. Further serologic testing was positive for acetylcholine receptor antibodies, which confirmed the diagnosis of myasthenia gravis. At this point neurology was consulted. The patient was initiated on therapy with an acetylcholinesterase inhibitor (pyridostigmine), corticosteroids and intravenous immunoglobulin (IVIG). The cardiothoracic surgery team was consulted for thymectomy and the patient was treated with IVIG at a dose of 0.4g/kg/day for five days prior to undergoing thymectomy. Upon completion of the surgery, she was safely extubated, but she was monitored in the ICU during the postoperative period due to requiring non-invasive positive pressure ventilation and complaints of worsening weakness and fatigue. There were concerns for possible myasthenic crisis, however none developed. The patient underwent pulmonary function testing and negative inspiratory force monitoring, which remained stable throughout her hospitalization. Over the course of the next seven days, the weakness gradually resolved and the patient was discharged with pyridostigmine and prednisone and instructions to follow up outpatient with the neurologist and oncologist.

Imaging



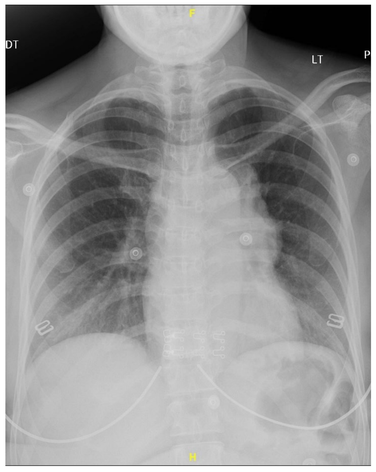


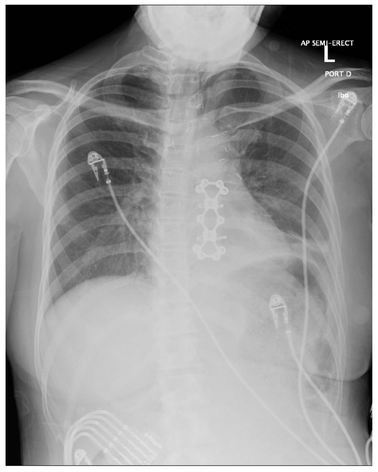
Top: transverse view of thymoma on CT chest w/ IV contrast.

Left: Coronal view of thymoma on CT chest w/ IV contrast.

Bottom Left: Pre-operative chest x-ray.

Bottom Right: Post-operative chest x-ray. Note absence of mediastinal irregularity and post-op clips.





Conclusion

This case illustrates the complex diagnostic approach to an anterior mediastinal mass and the importance for a high clinical suspicion for thymoma and myasthenia gravis. It also highlights the importance of quickly initiating appropriate therapies for preventing life threatening complications such as myasthenic crisis and respiratory failure once myasthenia gravis has been confirmed. While these steps are vital, there are well established guidelines to diagnosing and treating myasthenia gravis. Here we explored a solution to a known problem; myasthenic crisis related to surgery and anesthesia. This is especially relevant for patients with myasthenia gravis related to a thymoma as thymectomy is a standard therapy for all patient able to safely undergo surgical intervention. This has been an area of ongoing research, with suggestions of different doses and durations of IVIG as well as plasma exchange therapies. In concert with neurology, oncology and cardiothoracic surgery teams we attempted a strategy to help minimize the risk of postoperative complications and the patient received IVIG for 5 days prior to the surgery. This strategy seemed beneficial as no life threatening postoperative complications occurred in this patient and there was no evidence of myasthenic crisis. Further investigation may be needed to determine which patients will benefit from preoperative IVIG and the most effective dose and duration of this therapy.

Lessons Learned

In patients with myasthenia gravis associated w/ thymoma, the addition of preoperative IVIG therapy appears to be beneficial in reducing the risk of postoperative myasthenic crises in patients undergoing thymectomy.

References

1. Alipour-Faz A, Shojaei M, Peyvandi H, et al. A comparison between IVIG and plasma exchange as preparations before thymectomy in myasthenia gravis patients. Acta Neurol Belg. 2017;117(1):245-249.
2. Sanders DB, Wolfe GI, Benatar M, et al. International consensus guidance for management of myasthenia gravis: Executive summary. Neurology. 2016;87(4):419-425.
3. Álvarez-Velasco R, Gutiérrez-Gutiérrez G, Trujillo JC, et al. Clinical characteristics and outcomes of thymoma-associated myasthenia gravis. Eur J Neurol. 2021;28(6):2083-2091.

This research was supported (in whole or in part) by HCA Healthcare and/or an HCA Healthcare affiliated entity. The views expressed in this publication represent those of the author(s) and do not necessarily represent the official views of HCA Healthcare or any of its affiliated entities.

HCA Healthcare®