

AMSER Case of the Month

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71 y/o F with myasthenia gravis found to have a
mediastinal mass

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Patient Presentation

- **HPI:** A 71-year-old female presented to primary with several month history of left eyelid drooping and fatigable weakness.
- No h/o seizures, stroke, head injury, recent illness, tick/insect bite, sick contacts, exposure to toxins, or vaccines prior to onset of symptoms.
- Denies blurred or double vision, difficulty with chewing or swallowing, dysarthria, extremity weakness, headaches, night sweats, trouble breathing.

Patient Presentation

- **PMHx:** HLD, HTN, arthritis, gallstones, chronic back pain
- **Vitals:** BP 149/86 P 69 RR 18 T 98 F SpO2 96%
- **Physical Exam:** Left eye ptosis and left facial asymmetry. Normal speech and language, no dysarthria. Negative Babinski. Normal cardiovascular exam. Normal pulmonary effort.

Pertinent Labs/Workup

- **AChR Antibodies:**

- Binding **>80 nmol/L** (Ref: 0.00-0.24)
- Blocking **36%** (Ref: 0-25%)
- Modulating **77%** (Ref: 0-45%)

- **B12:** 398 (Ref: 232-1245)

- **Tumor Markers:**

AFP 4.7 (<8.1) LDH 219 (<250) BhCG <10 (<10)

- **EMG:** Mild median sensorimotor neuropathy consistent w/ carpal tunnel syndrome
- **PFT:** No evidence of ventilatory impairment

What Imaging Should We Order?

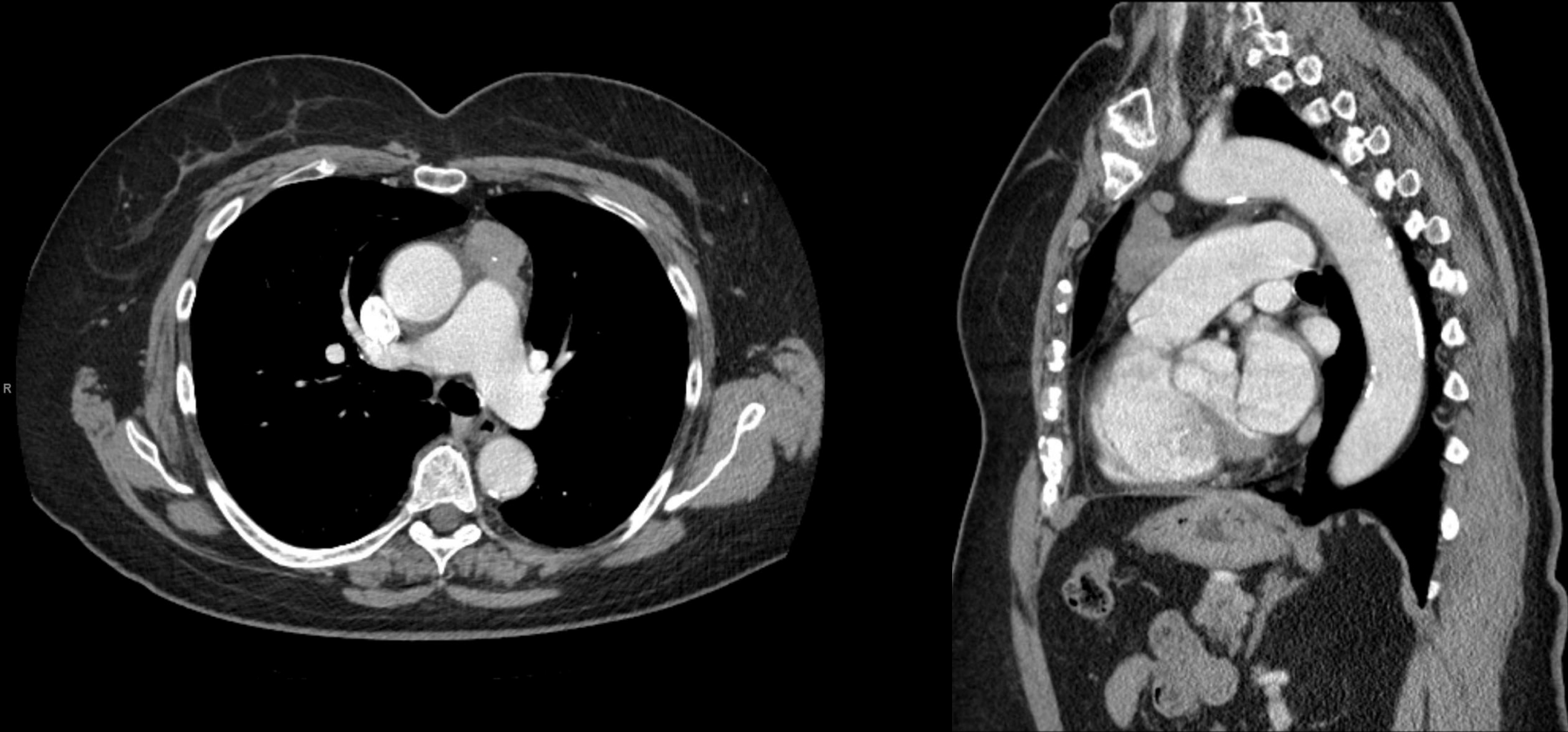
ACR Appropriateness Criteria

Scenario	Scenario ID	Procedure	Adult RRL	Peds RRL	Appropriateness Category
Mediastinal mass suspected, initial imaging	3194009	● Radiography chest	<0.1 mSv ⊕	<0.03 mSv [ped] ⊕	Usually appropriate
		● MRI chest without and with IV contrast	0 mSv ○	0 mSv [ped] ○	Usually appropriate
		● MRI chest without IV contrast	0 mSv ○	0 mSv [ped] ○	Usually appropriate
		● CT chest with IV contrast	1-10 mSv ⊕⊕⊕	3-10 mSv [ped] ⊕⊕⊕⊕	Usually appropriate
		● CT chest without IV contrast	1-10 mSv ⊕⊕⊕	3-10 mSv [ped] ⊕⊕⊕⊕	Usually appropriate
		● US chest	0 mSv ○	0 mSv [ped] ○	Usually not appropriate
		● Image-guided transthoracic needle biopsy	Varies	Varies	Usually not appropriate
		● CT chest without and with IV contrast	1-10 mSv ⊕⊕⊕	3-10 mSv [ped] ⊕⊕⊕⊕	Usually not appropriate
		● FDG-PET/CT skull base to mid-thigh	10-30 mSv ⊕⊕⊕⊕	3-10 mSv [ped] ⊕⊕⊕⊕	Usually not appropriate



This imaging modality was ordered by the physician.

CT Chest W/ Contrast; Findings

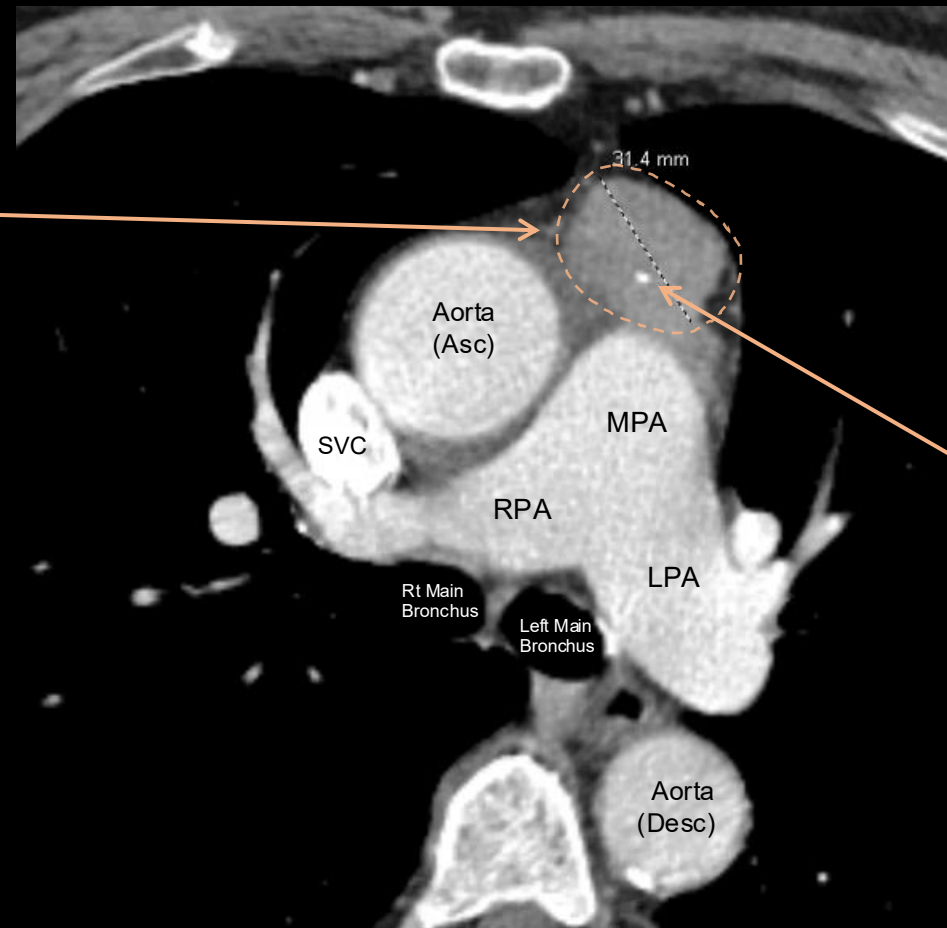
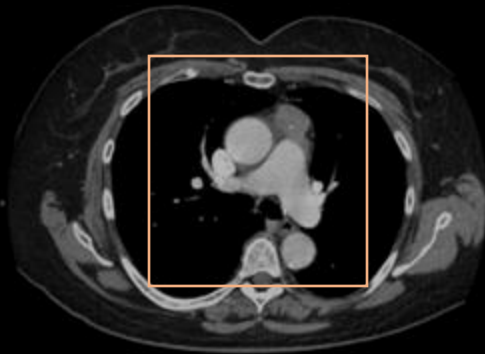


CT Chest W/ Contrast; Findings (Labeled)

Well-defined, oval-shaped mass with smooth contours and homogenous soft tissue attenuation measuring up to 3.14 cm in AP dimension. No fluid or cystic areas noted.

Mass is located left of midline. Although not definitive, midline location favors germ cell tumor or lymphoma. Thymoma/thymic epithelial tumors are characteristically off midline.

Punctate calcification helps to narrow differential (NOT present in thymic hyperplasia, pericardial cysts, abscess. May be present in thymoma, lymphoma, teratoma, seminoma, NSGCT).

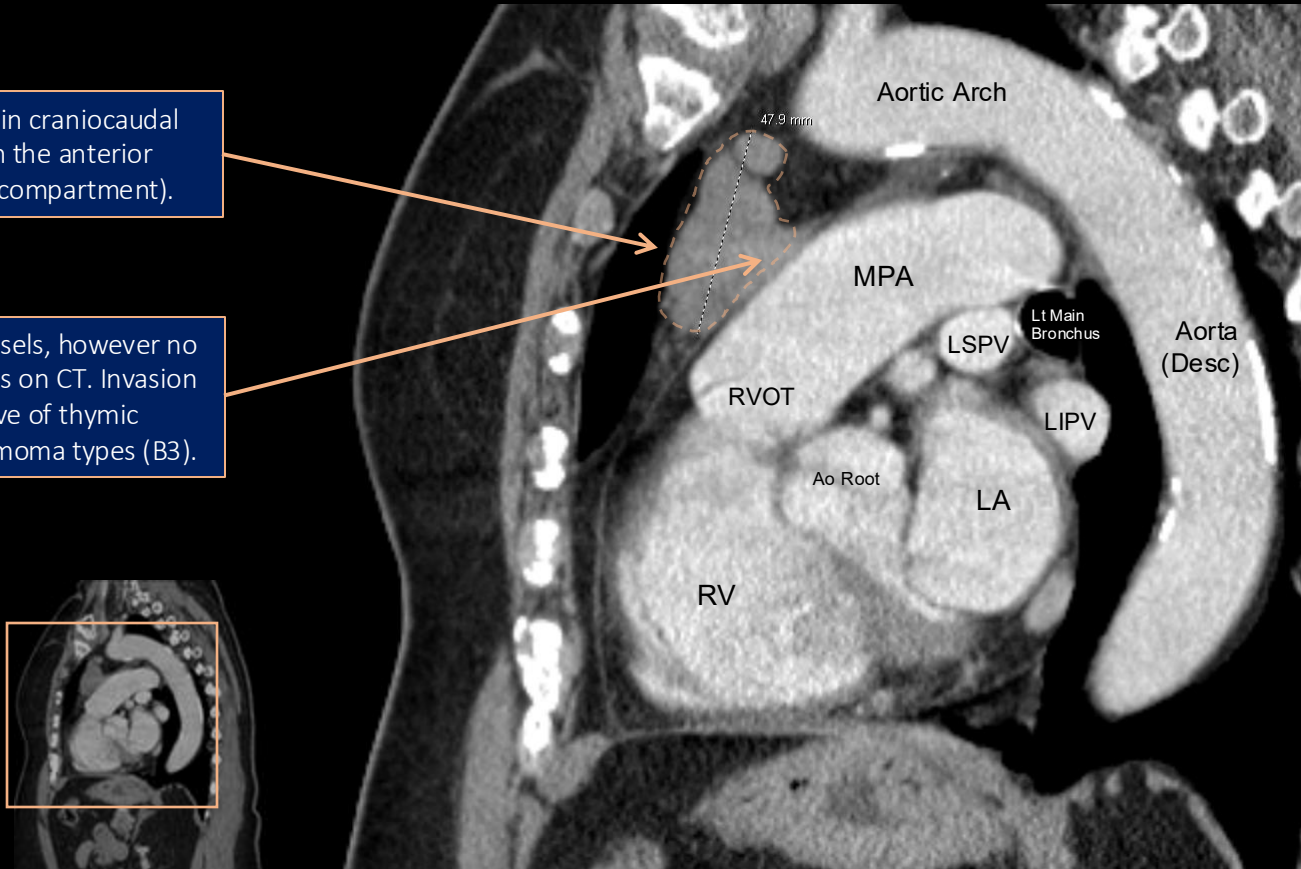


MPA: Main Pulmonary Artery. RPA/LPA: Right/Left Pulmonary Artery

CT Chest W/ Contrast; Findings (Labeled)

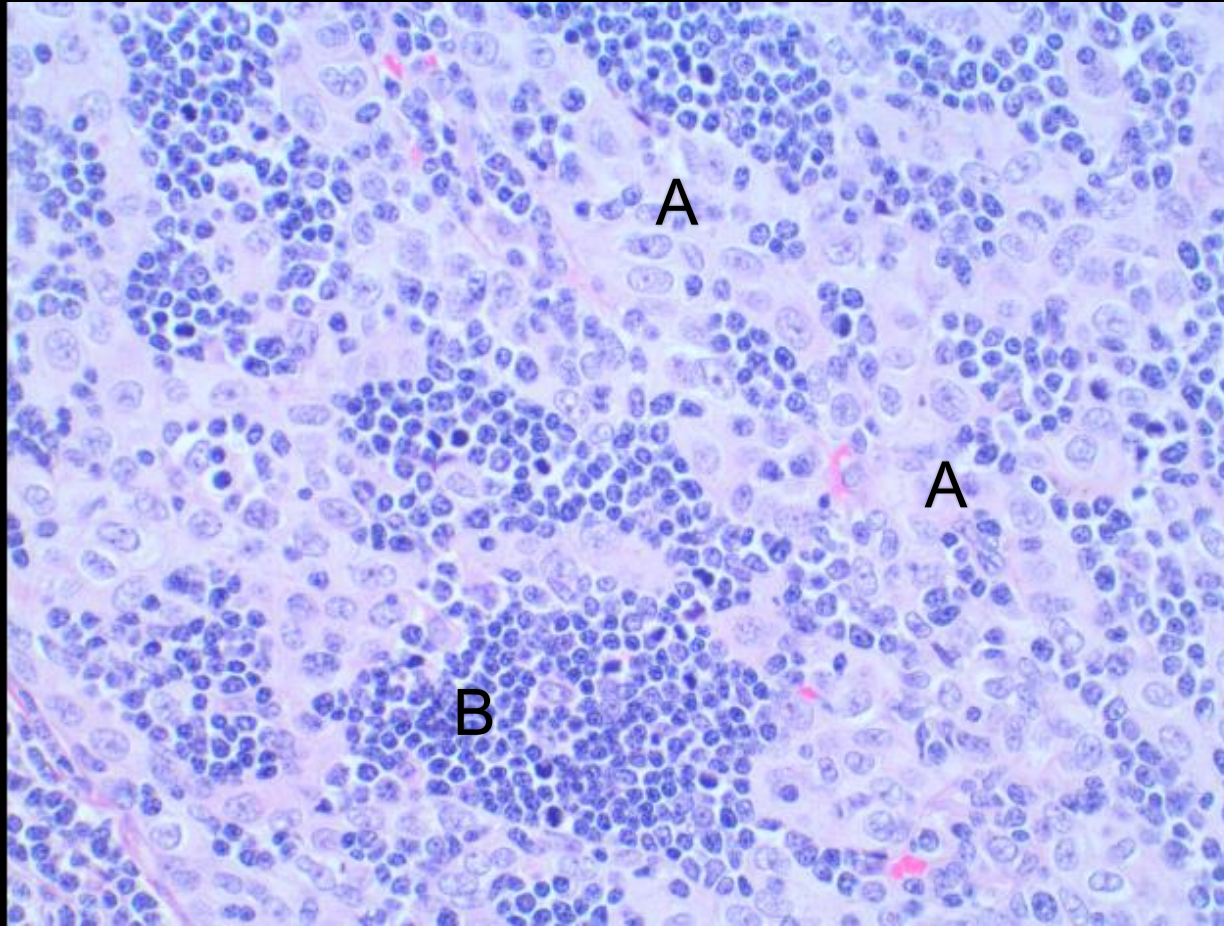
Mass measures up to 4.79 in craniocaudal dimension and is located in the anterior mediastinum (prevascular compartment).

Mass adjacent to great vessels, however no clear invasion of the vessels on CT. Invasion of great vessels is suggestive of thymic carcinoma or high risk thymoma types (B3).



RVOT: RV Outflow Tract. Ao Root: Aortic Root. LSPV/LIPV: Left Superior/Inferior Pulmonary Vein.

Pathology



Type AB thymoma. Paler areas of predominantly bland spindle/oval cells and lymphocyte-poor component (type A) admixed with lymphocyte-rich component (Type B) ⁶

Final Dx

Type AB Thymoma

Myasthenia Gravis Workup

- Myasthenia gravis is characterized by autoantibodies against acetylcholine receptors (AcR) or muscle-specific kinase (MuSK)
- Presents with fatigable weakness often affecting eye, bulbar, respiratory, or limb muscles
- Antibody production T cell dependent and mainly occurs in intrathymic environment; explains 50% of seropositive MG co-occurring with thymic hyperplasia and 10-15% with thymic tumor. In addition, 30-50% of patients with a thymoma have myasthenia gravis ^{2, 3}
- EMG can confirm diagnosis (RNS or jitter analysis in SFEMG) ²
- CT should be performed in patients with MG to exclude thymoma ²
- Tumor markers (AFP, LDH, BhCG) to differentiate mediastinal masses (4 T's)

Mediastinal Mass Differential

- Approach considers anatomical compartment (prevascular, visceral, and paravertebral), density pattern, and patient age/gender
- Prevascular compartment differential: ⁷
 - Thymic Hyperplasia – Diffuse enlargement or soft tissue lesion similar in appearance to normal thymus
 - Thymic Cyst – Soft tissue attenuation on pre-con CT, mean HU 31. Higher T2 signal than solid masses on MR
 - Pericardial Cyst – Cystic lesion in cardiophrenic angle, high MR fluid signal
 - Mediastinal Hemangioma – Lo/med T1, Hi T2 signal. Uniform vs peripheral nodular enhancement +/- centripetal enhancement
 - Lymphoma – Lobular shape, mild enhancement, conglomerate lymph nodes. Younger patient age. B symptoms
 - Teratoma/GCT – Various internal constituents (fat, soft tissue, calcifications, fluid). Younger patient age
 - Thymoma (next slide)

Thymoma Characteristics

- Thymoma
 - Most common primary neoplasm in anterior mediastinum (20%) ^{4,5}
 - Typically spherical or ovoid, encapsulated, off-midline soft tissue lesion with homogeneous attenuation and contrast enhancement. May be round or lobulated due to septa ^{4,5,7}
 - May display heterogeneity due to hemorrhage, necrosis, or cystic changes ^{4,5}
 - May contain punctate, curvilinear, or coarse calcifications ^{4,5}
 - Heterogeneity/calcifications/lobulated margins are more likely in invasive tumors ⁴
 - Slow growing with local extension or spread along serosal surfaces (pleura/pericardium) ⁴
 - Local invasion correlates with prognosis; Masaoka-Koga or TNM staging systems commonly used ^{4,1}
 - Thymoma histotype affects prognosis with Type AB associated with a 2% recurrence rate (vs up to 7% with B3). Type AB tends to occur at higher patient age and presents in a lower stage as compared to Types B1, B2, B3 ⁸

Thymoma Staging

Stage	Definition
I	Grossly and microscopically completely encapsulated tumor
IIa	Microscopic transcapsular invasion
b	Macroscopic invasion into thymic or surrounding fatty tissue, or grossly adherent to but not breaking through mediastinal pleura or pericardium
III	Macroscopic invasion into neighboring organ (i.e., pericardium, great vessel, or lung)
IVa	Pleural or pericardial metastases
b	Lymphogenous or hematogenous metastasis

Masaoka-Kaga Staging System ¹

Treatment

- Pharmacological treatment is focused on controlling symptoms ²
 - First line therapy is cholinesterase inhibitors (pyridostigmine) titrated to symptomatic improvement
 - Immunosuppressants (Glucocorticoids/Azathioprine)
 - AZA can be used alone or in conjunction with prednisone
- For refractory cases may consider plasmapheresis, cyclophosphamide, rituximab, eculizumab ²
- If thymoma present, must be excised with only rare exceptions. Further treatment of thymoma based on histology and staging ²
- **Our Case:** Type AB (mixed) thymoma based on histology. Patient was referred for surgical resection

References

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