Radiology Case Presentation

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7/13/2018

Yale SCHOOL OF MEDICINE

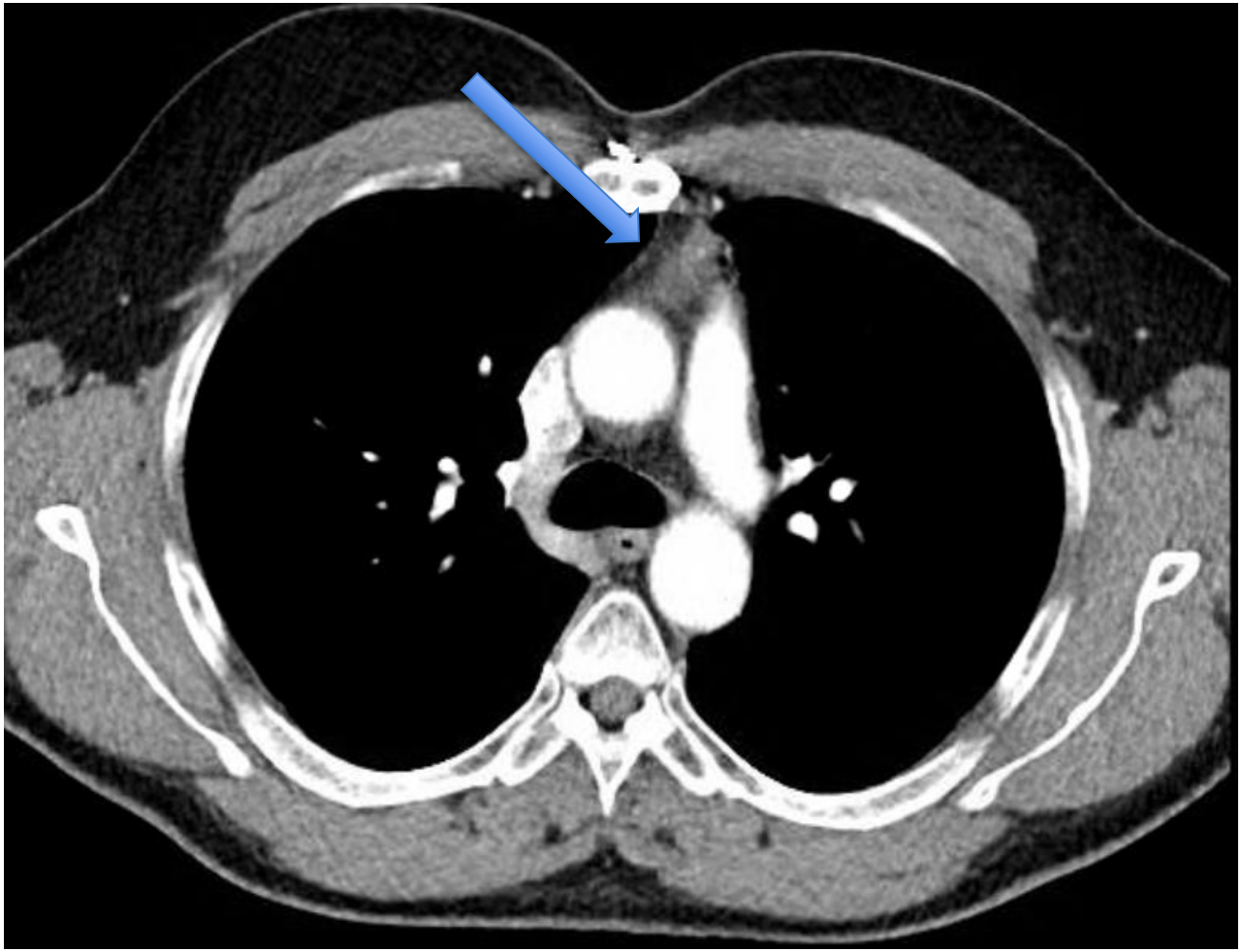
ML

- HPI
  - 54 yo male who presented with 2 months of L ptosis, bilateral upper extremity weakness, and fatigue with chewing in 9/2016
  - Subsequently diagnosed with myasthenia gravis
  - Chest CT screening showed 4cm anterior mediastinal mass consistent with thymoma
  - Underwent thymectomy, when mass was noted to be adherent to the pleura with possible invasion.
  - Surgical pathology confirmed diagnosis of Type B2 thymoma with positive margins and involving thymic capsule
  - Received postoperative radiation therapy
  - Doing well with improvement of myasthenic symptoms and no evidence of recurrent disease on follow-up imaging in 4/2017

ML

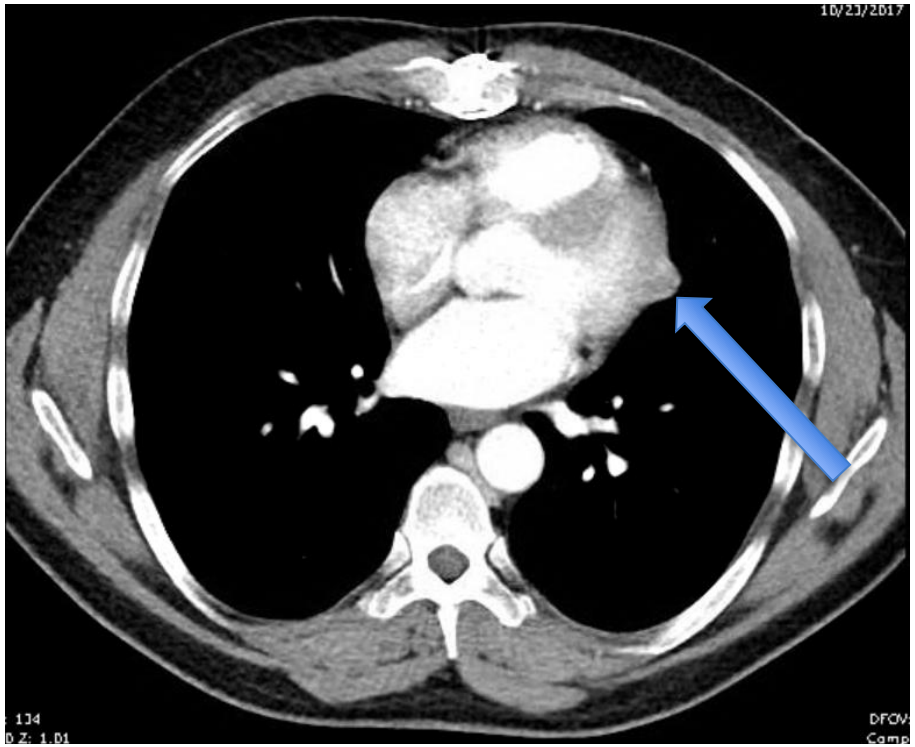
- Thymic disease recurrence
  - Follow-up chest CT in 10/2017 with evidence of recurrent disease, including multiple new soft tissue nodules along the left pleural surfaces concerning for pleural dissemination due to metastatic thymoma.
  - Repeat imaging in 1/2018 continued to show progression of soft tissue nodules and pleural disease
  - Due to recurrent thymoma, received neoadjuvant chemotherapy with cisplatin, adriamycin, cyclophosphamide
  - Planned to undergo pleurectomy in 2/2018, although aborted due to extensive number of visceral pleural nodules. Wedge resection of nodules confirmed metastatic thymoma.
  - Presented at tumor board with potential plan to undergo extrapleural pneumonectomy.

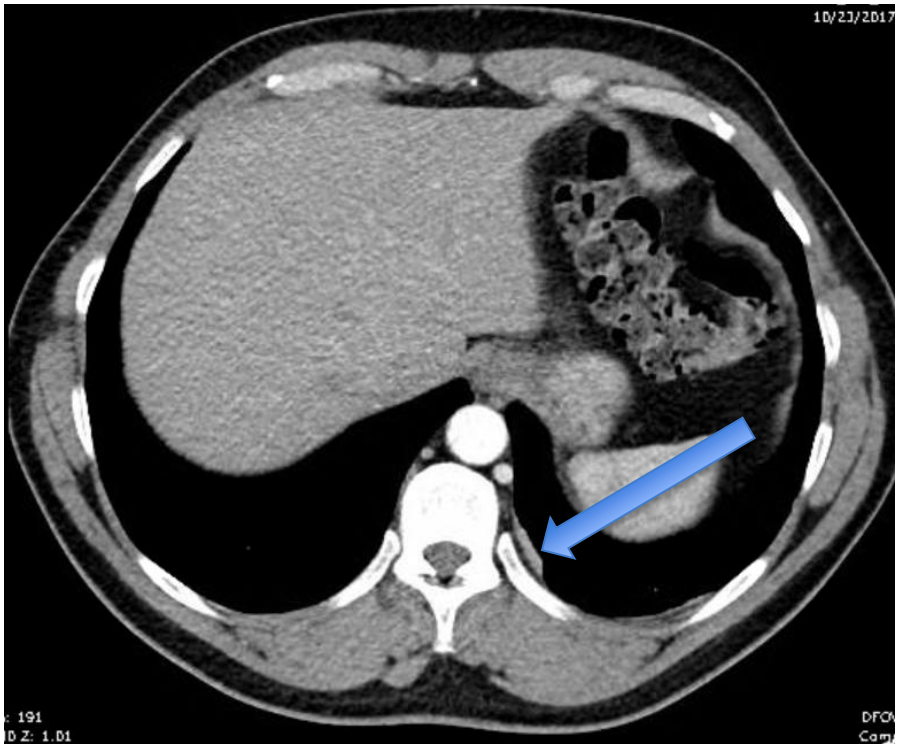
Chest CT 10/2017



- Postsurgical changes due to thymectomy and/or postradiation changes noted in anterior mediastinum
- Increased soft tissue at site of thymectomy concerning for recurrent tumor vs thymic hyperplasia

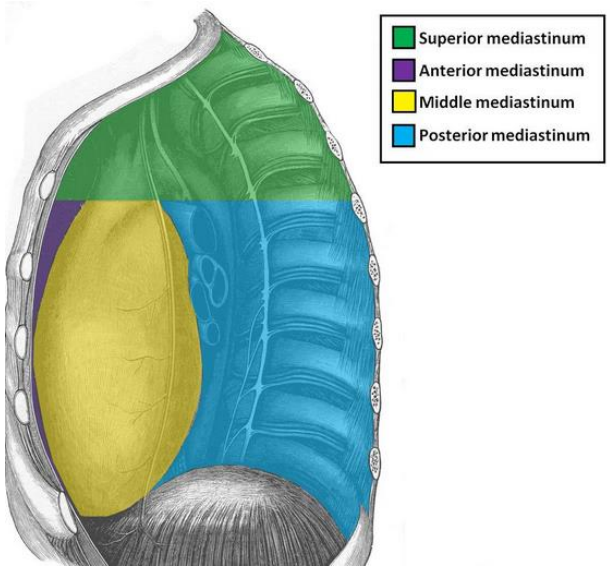
CT Findings



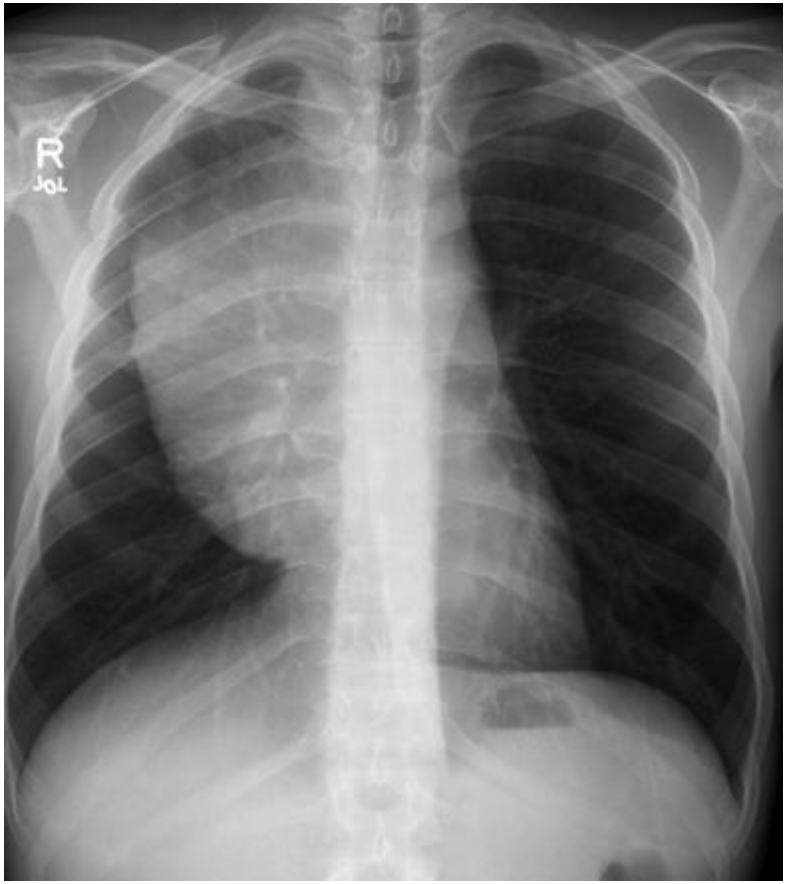


- New soft tissue nodules noted along the left pleural surface and left pericardium, compatible with pleural implants.

Anterior Mediastinal Masses

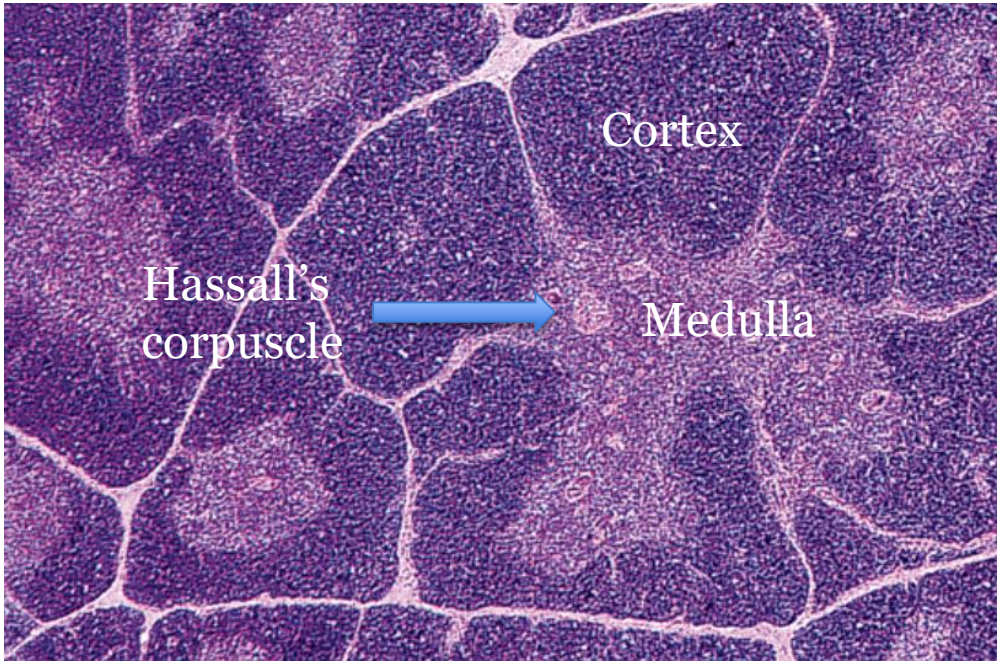


- "5 T's"
  - Thymoma
  - Thyroid mass
  - Teratoma
  - Thoracic aortic aneurysm
  - "Terrible" lymphoma



Hilum overlay sign

Thymoma



- WHO classification
  - Type A thymoma – spindle cell or medullary thymoma
  - Type B thymoma – cortical thymoma
  - Type AB thymoma – mixed thymoma
  - Type C thymoma – thymic carcinoma

Thymoma in myasthenia gravis

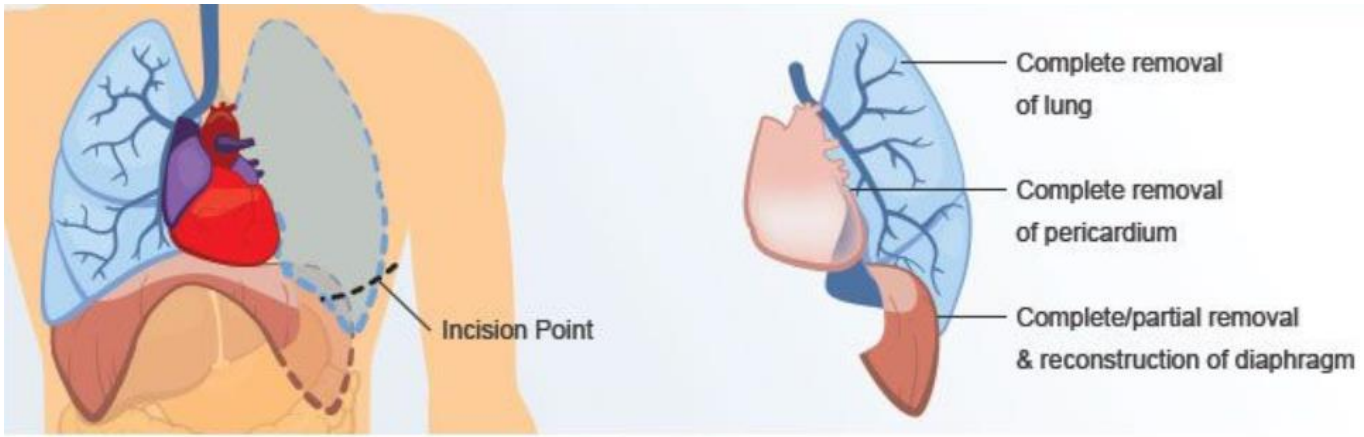
- Myasthenia gravis
  - NMJ disorder characterized by muscular weakness and fatigability
  - Classic symptoms include ptosis, diplopia, dysphagia, fatigable chewing, proximal limb weakness, respiratory muscle weakness
  - Most commonly caused by ACh receptor antibodies
- 15% of patients with myasthenia gravis have thymoma, and up to 50% of patients with thymoma have myasthenia gravis
- MG associated with Type B thymomas as a paraneoplastic syndrome
- Neoplastic cells in thymomas express epitopes cross-reactive with skeletal muscle proteins, including ACh receptor, titin, and ryanodine receptor, which are presented to maturing autoreactive T cells and lead to symptoms of myasthenia.

Surgical treatment of thymoma

- Thymectomy for definitive treatment of thymoma
- Evidence for preoperative treatment with plasmapheresis and/or IVIG to reduce circulating antibody load and decrease risk of subsequent myasthenic crisis with respiratory failure
- If complete resection not possible, can also treat with radiation therapy and/or chemotherapy.
- With phrenic nerve involvement, often can sacrifice the nerve to resect the tumor vs dissection of tumor away from nerve.
- Recurrent thymic disease –
  - Ruffini et al and Okumura et al suggest surgical resection of recurrent thymic tumor when complete resection is feasible is recommended and can lead to improved long-term survival with outcome directly related to WHO classification of tumor.
  - In disseminated disease with incomplete resection, surgical resection likely still contributes to improved long-term survival.

Invasive thymoma with pleural dissemination

- Extrapleural pneumonectomy (EPP) has demonstrated safety and efficacy in pleural malignancies like mesothelioma, now being studied in other cancers including thymoma and NSCLC.



Extrapleural Pneumonectomy

Extrapleural pneumonectomy

Thymoma Patients With Pleural Dissemination: Nationwide Retrospective Study of 136 Cases in Japan

Katsuhiro Okuda, MD, PhD, Motoki Yano, MD, PhD, Ichiro Yoshino, MD, PhD, Meinoshin Okumura, MD, PhD, Masahiko Higashiyama, MD, PhD, Kenji Suzuki, MD, PhD, Masanori Tsuchida, MD, PhD, Jitsuo Usuda, MD, PhD, and Hisashi Tateyama, MD, PhD

- Okuda et al retrospectively reviewed 136 cases of thymoma with pleural dissemination
  - 128 patients underwent tumor resection, 8 patients underwent EPP
  - Overall 5-year survival of thymomas with pleural dissemination after surgical resection or EPP = 83.5%
  - Overall 5-year survival of pts undergoing EPP = 70%
  - Better prognosis with fewer disseminated pleural nodules (10 or fewer) and achievement of macroscopically complete resection.
  - No significant improvement in long-term survival for patients treated with adjuvant chemoradiotherapy as compared to those without.
- Due to high mortality and low quality of life associated with pneumonectomy, further studies need to be done in patients with disseminated thymic disease to confirm overall favorable postoperative outcome.

RD

- HPI
  - 18 yo male with PMH of simple bone cyst involving left proximal humerus c/b multiple pathologic fractures presenting with left upper extremity shortening.
  - After progressing through growth spurt, noticed progressive shortening of the left humerus due to growth arrest.
  - Has been experiencing musculoskeletal strain with heavy lifting attributed to overcompensation of right arm for left arm, as well as difficulty with activities of daily living due to unequal arm lengths and mild shoulder pain.
  - Due to functional limitations, bone lengthening procedure desired.
  - On exam, left humerus noted to be approximately 11-12cm shorter than the right humerus.

Bilateral humerus Xray 11/2015

- Right humerus length = 35cm and left humerus length = 24cm
- Postsurgical changes with bony deformity and irregularity of proximal diaphyseal cortex of the left humerus noted.

Bone lengthening surgery

- External fixation and lengthening (Ilizarov apparatus)
  - 1951: Soviet surgeon Dr. G.A. Ilizarov created a circular external fixator providing appropriate stabilization following osteotomy and distraction to allow formation of new osseus tissue.
  - Classic procedure for limb lengthening procedures
- Intramedullary rod magnetic lengthening
  - Magnetically driven, activated by applying magnetic field generator by using an external remote controller (consists of 2 rotating magnets) to the skin to control rate of distraction.
  - Magnet inside intramedullary nail rotates in response to remote, leading to lengthening
  - Maximum lengthening of 5cm at one time
  - Newer technology to minimize disadvantages of external fixation.

Left humeral Xray

8/23/16 – 24cm

9/1/16 – 25cm

9/20/16 – 27 cm

Left humeral Xray

11/15/16 – 29cm

Complete lengthening of nail achieved

1/17/17 – 29cm

5/16/17 – 29cm

Left humeral Xray

- Second intramedullary nail placed 11/29/17 for further lengthening

- Most recent film 7/2/18
- Left humeral length = 33cm
- Bony remodeling and bridging seen across the osteotomy site
- Increased lucency around distal screw suggesting component of subsidence of hardware

External fixation vs. intramedullary nail lengthening

Femoral lengthening with a motorized intramedullary nail

Joachim Horn, Øyvind Grimsrud, Anita Hoddevik Dagsgard, Stefan Huhnstock & Harald Steen

- Horn et al performed matched-pair case-control study of 30 femoral lengthening procedures
  - 15 patients undergoing motorized intramedullary nail and 15 patients undergoing external fixator
  - Primary outcomes measured were lengthening and alignment achieved, consolidation index (time from osteotomy to radiographic consolidation divided by lengthening distance achieved), knee ROM, and complications.
  - Planned lengthening and alignment achieved in all patients.
  - Intramedullary nail group had statistically significant superiority in consolidation index (1.5 months/cm vs 1.9 months/cm), fewer complications, and, most significantly, improved knee ROM.

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- The Anterior Mediastinum – Teach Me Anatomy