One week later, the patient developed increasing pain and shortness of breath. Due to his rapid deterioration, he was intubated and started on empiric chemotherapy. CT Scan after chemotherapy revealed 1800 cc of exudative, bloody fluid. Pleural biopsy was obtained, but the initial impression was equivocal, suggestive of either a synovial sarcoma or poorly-differentiated germ cell tumor. The patient was discharged to follow-up in clinic while awaiting final pathology.

One week later, the patient developed increasing pain and shortness of breath, and presented back to the ED. Repeat CT of the chest (middle image) two weeks after the initial scan showed that the tumor had grown to occupy the entire right hemithorax with further shift of the mediastinal structures to the left.

Due to his rapid deterioration, he was intubated and started on empiric chemotherapy with etoposide, ifosfamide, and cisplatin, even before a definitive diagnosis was made. His tumor responded well to chemotherapy.

Repeat biopsy was obtained (because the initial biopsy was equivocal) and was consistent with a germ cell tumor, most compatible with yolk sac origin. The patient was discharged from the hospital two weeks later after a fairly unremarkable inpatient stay.

Since discharge, he has received three additional cycles of chemotherapy. His tumor burden has decreased significantly (right image), and AFP has trended down to 7.9. Currently, discussions are taking place about possible resection of the residual tumor.

Introduction
- Extragonadal germ cell tumors (EGCT), which are defined as germ cell tumors without a primary tumor in the testes are very rare.
- Of the 8000 annual cases of GCT, only 2-5% are of extragonadal origin.
- EGCT usually occur in males between the ages of 20 and 35 years, but can occur at any age.
- The most common site is in the anterior mediastinum.

Case
- 22 year old male with no significant past medical history, presents with complaints of a non-productive cough 3 days prior to admission. The cough was associated initially with just some fatigue, but progressed to include sore throat and right-sided chest discomfort. Over the next 3 days, his fatigue progressed to the point where he could barely walk.
- Review of systems was positive for occasional night sweats as well as mild, sharp, mid-epigastric abdominal pain, and one episode of nausea and vomiting. He denied fevers, chills, shortness of breath, diarrhea, constipation, hematemesis, hematochezia, hemoptysis, and focal weakness.
- He was on no medications, had no allergies, and reported no significant medical, surgical, or family history. He was a current every-day smoker of 1 pack per day, and occasionally used marijuana. He was incarcerated four months prior to admission.
- On initial physical exam, he was afebrile, normotensive, tachypneic at 32 respirations per minute with an oxygen saturation of 98% on room air. Pulmonary exam revealed decreased breath sounds and dullness to percussion on the right. His left lung was clear to auscultation and resonant to percussion. Cardiovascular and abdominal exam was unremarkable. Testicular exam was unremarkable without swelling, mass, or tenderness.
- Labs were remarkable for a slightly elevated WBC count, mild normocytic anemia, and hypoalbuminemia. ABG revealed a metabolic alkalosis with no hypoxemia. Alpha fetoprotein (AFP) drawn on initial presentation was elevated at 860. β-hCG and CEA were within normal limits.

Case (cont’d)
- Chest X-ray revealed complete opacification of the right hemithorax. CT of the chest (left image) demonstrated a large right-sided pleural effusion with right-to-left mediastinal shift, and numerous heterogeneously-enhancing pleural-based lesions, highly concerning for neoplasm. An ultrasound of the scrotum and testicles showed a small left-sided hydrocele, but no masses.
- Thoracentesis revealed 1800 cc of exudative, bloody fluid. Pleural biopsy was obtained, but the initial impression was equivocal, suggestive of either a synovial sarcoma or poorly-differentiated germ cell tumor. The patient was discharged to follow-up in clinic while awaiting final pathology.
- One week later, the patient developed increasing pain and shortness of breath, and presented back to the ED. Repeat CT of the chest (middle image) two weeks after the initial scan showed that the tumor had grown to occupy the entire right hemithorax with further shift of the mediastinal structures to the left.
- Due to his rapid deterioration, he was intubated and started on empiric chemotherapy with etoposide, ifosfamide, and cisplatin, even before a definitive diagnosis was made. His tumor responded well to chemotherapy.
- Repeat biopsy was obtained (because the initial biopsy was equivocal) and was consistent with a germ cell tumor, most compatible with yolk sac origin. The patient was discharged from the hospital two weeks later after a fairly unremarkable inpatient stay.
- Since discharge, he has received three additional cycles of chemotherapy. His tumor burden has decreased significantly (right image), and AFP has trended down to 7.9. Currently, discussions are taking place about possible resection of the residual tumor.

Discussion
- Germ cell tumors are classified as seminomatous or non-seminomatous (NSGCT). NSGCTs are classified as yolk sac (60%) tumor, choriocarcinoma (12%), embryonal carcinoma (9%), or mixed, based on histology. These tumors are usually associated with an increased AFP, β-hCG or both. Most germ-cell tumors are located in the testes, and only a small percentage (2-5%) are found in other sites with no evidence of testicular involvement. The most common extragonadal site is in the anterior mediastinum, followed by the retroperitoneum.

Mediastinal germ cell tumors are aggressive, rapidly-growing tumors, typically found in younger men. Standard treatment consists of chemotherapy with vincristine, ifosfamide, and cisplatin, usually followed by surgical resection of residual disease. Because of the rapidity of cell growth, these tumors are typically very responsive to chemotherapy. Five year progression-free and overall survival in mediastinal NSGCT are 44 and 45%, respectively.

References