Metastatic Anterior Mediastinal Mass Causing Simultaneous Chylothorax and SVC Syndrome

Kelly Schulte DO, MA[1]; Kavanya Feustel MD[1]; Jeffrey Ramsey DO[1]; Elizabeth Hicks MD[2]

- [1]Sky Ridge Medial Center Department of Internal Medicine, [2]Critical Care & Pulmonary Consultants

Background

- Chylothorax occurs from accumulated chyle in the pleural cavity as a result of a disruption in the thoracic duct.[1]
- Right sided chylothorax is more common due to location of thoracic duct and diagnosed by triglyceride levels greater than 110 mg/dL in the pleural fluid.[1,2]
- Superior Vena Cava (SVC) syndrome generally occurs from a disruption in the venous blood flow either intrinsically or extrinsically.[3]
- Malignancy is a common cause for chylothorax and SVC syndrome individually from extrinsic compression.[1,3]
- There has only been a few cases documented in literature with both chylothorax and SVC syndrome occurring at the same time and only one other case in the setting of breast cancer.[4-7]
- We present a case of a metastatic anterior mediastinal mass obstructing the thoracic duct and compressing the bilateral brachiocephalic veins.

Case Presentation

- A 41 year old female with stage IV right sided invasive ductal breast carcinoma with metastasis to bone, lung, and mediastinum status post bilateral mastectomy and chemo-radiation presented with several days of dyspnea and progressive swelling of bilateral arms, face, and neck.
- She was recently diagnosed with acute deep vein thrombosis (DVT) of internal jugular and innominate veins and was on Apixaban prior to hospitalization.
- Imaging showed bilateral brachiocephalic vein occlusions from her mediastinal mass with collaterals and bilateral pleural effusions.
- Diagnostic and therapeutic thoracentesis of the right pleural effusion removed approximately 1.6L of milky fluid. Triglyceride levels were 1666 ng/dL, and cytology was negative for malignancy.
- Patient tolerated the procedure well and reported improvement in her dyspnea.
- However, patient had to undergo a pleural catheter placement for her re-accumulating pleural effusion.
- She was also placed on an octreotide infusion and a low fat diet to reduce the production of chyle.[1,2]
- Labs were significant for hypoalbuminemia (2.5 g/dl) and worsened leukopenia (2 10^9/L) while being on Letrozole from the chyle leak.
- Bilateral stents were placed in the occluded brachiocephalic veins as well as started on anticoagulation therapy (therapeutic Enoxaparin) for 3 months durations. This provided immediate improvement of her upper extremity and facial swelling.
- External beam radiation therapy was initiated for her anterior mediastinal mass which also provided symptomatic relief and tolerated well.
- Patient's history of chest cavity radiation precluded the consideration of thoracic duct ligation/embolization.

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Images

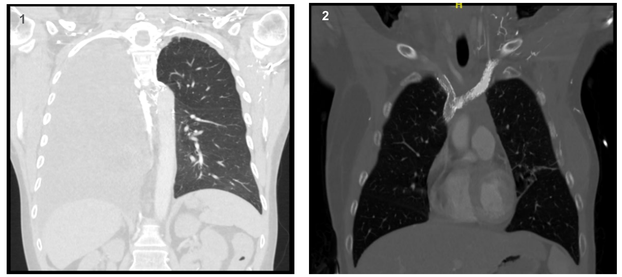

Figure 1: CT scan with right sided chylothorax. **Figure 2**: CT scan with bilateral brachiocephalic vein stents placed and resolution of chylothorax after pleural catheter.

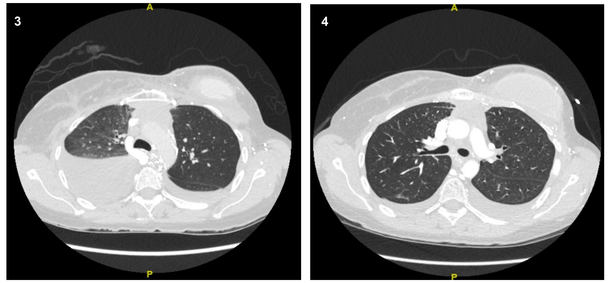

Figure 3: CT scan with right side chylothorax and small left side pleural effusion.
Figure 4: CT scan with resolution of chylothorax after pleural catheter.

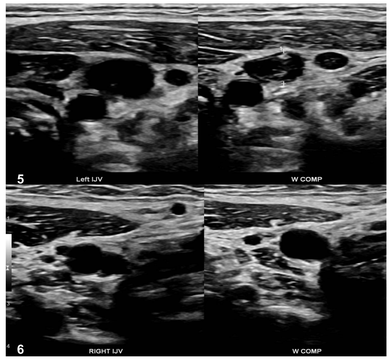

Figure 5: Doppler ultrasound with acute thrombus in left internal jugular vein (IJV).
Figure 6: Doppler ultrasound with chronic thrombus in right internal jugular vein (IJV).

Discussion

- Patients with large chylothorax present with progressive breathlessness and pressure from the mass effect on lung expansion.[1]
- Obstruction of the SVC presents with upper extremity/head swelling, cyanosis, facial plethora, thrombosis. Significant facial and neck edema can cause hoarseness, dyspnea, and dysphagia.[8]
- Chylothorax was not diagnosed in this patient until her pleural fluid was noted to be milky and triglyceride levels confirmed the diagnosis.
- Continued chyle leak can lead to leukopenia and malnutrition as seen in this case.[1]
- Management of chylothorax includes diet modification (high protein low fat diet with medium chain triglycerides), octreotide, thoracentesis, indwelling pleural catheter placement, thoracic duct ligation/embolization.[1,2]
- Dietary modifications and octreotide (somatostatin analogue) both reduce lymphatic flow via decreasing gastric blood flow and subsequently fat absorption.[1,9]
- CT imaging (or venography which is the gold standard) is essential in diagnosing SVC syndrome while identifying its etiology and type of compression.[3]
- Management of SVC syndrome includes elevating head, endovascular therapy, anticoagulation, and chemotherapy/radiation for the underlying malignancy.[8,10]
- In this case, the patient's history of radiation and active malignancy (metastatic breast cancer) increased her risk of developing both SVC syndrome and chylothorax.[7]

Conclusion

- Suspect SVC syndrome in patients with dyspnea, upper extremity/head swelling, facial plethora, thrombosis.
- While dyspnea can occur with SVC syndrome, consider chylothorax in the differential diagnosis as an accumulating pleural effusion can present with dyspnea.
- Disruption of both venous blood flow to the SVC and thoracic duct can occur from malignancy.[1]
- Management of both conditions should provide symptom relief while ultimately addressing the underlying cause.

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