PRIMARY MEDIASTINAL T-CELL ALL PRESENTING WITH SUPERIOR VENA CAVA SYNDROME: A CASE REPORT

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Background

- Superior vena cava syndrome (SVCS) is a medical condition characterized by obstruction of the superior vena cava (SVC), often resulting from compression, invasion, or thrombosis. The typical presentation includes facial edema, venous congestion of the neck, and tachypnea, though the specific symptoms may vary depending on the underlying cause. [1] Intrathoracic malignancy, particularly small-cell bronchogenic carcinoma and non-Hodgkins's lymphoma, is the most common cause of SVCS. [2] This article highlights a rare and unusual case of SVCS caused by T-cell acute lymphoblastic leukemia (T-Cell ALL).

Case Presentation

- A previously healthy 30-year-old female presents to the Emergency Department (ED) with 2 weeks of dry cough, fevers maxing out at 102 F, and 5 lbs of unintentional weight loss. For the past week, she has also had bilateral chest pain that radiates to her back and left shoulder. Patient noticed the dyspnea worsens while supine. She noticed intermittent redness and warmth in her face and neck. The past 4 days, she noted post-tussive non-bloody emesis. As the symptoms worsened, she presented to the ED for further evaluation. Initial vital signs were notable for tachypnea with a respiratory rate of 24 breathes per minute but were otherwise within normal limits. Her exam was remarkable for diminished breath sounds on posterior right lower lobe and frequent cough. When patient was laid supine, she became dyspneic with audible rhonchi. The rest of her exam was unremarkable.

- Differentials included:
  - 80% of SVC obstructions relate to Malignant SVC syndrome
    - Small cell lung cancer, non small cell lung cancer (75-80%)
    - Non-Hodgkin's lymphoma (10-15%)
  - 15-40% of SVC obstructions occur due to indwelling intravascular devices, which were absent in this patient, making it less likely.
  - Given patient's lack of smoking or tobacco use, lung cancer was less likely. Large B cell lymphoma remained the top differential diagnosis until more tests were performed.

Intervention/Timeline

- At admission: laboratory studies displayed normal WBC count, normocytic anemia, mild thrombocytosis, elevated ESR
- Chest X-Ray: widened mediastinum (Figure 1)
- CT Angiogram of chest with and without IV contrast (Figure 2):
  - Large mediastinal mass (10.0 x 7.9 x 11 cm) extending from thoracic inlet
  - Bilateral lower lobe ground glass opacities and interlobular septal thickening
- Flexible laryngoscopy/nasopharyngoscopy: no lesions or airway obstructions
- CT neck with and without IV contrast: demonstrated bilateral enlarged subclavian nodules
- Transthoracic Echocardiogram: confirmed moderate pericardial effusion
- US-guided Core Biopsy of mass: showed non-Hodgkin's lymphoma on one core, and Hodgkin's lymphoma on second core
- Staging PET: consistent with Lugano Stage 1
- Final biopsy demonstrated T-cell ALL
- Intrathecal chemotherapy: CALG10403 Protocol
  - Vincristine
  - Daunorubicin
  - Oral Dexamethasone
  - Pegasparase
- Within a few days of Vincristine administration, patient began experiencing bilateral jaw pain (resolved without intervention)
- Induction Therapy (1-8 Days):
  - Intrathecal Methotrexate
  - IV Hydrocortisone
- Discharged after course of medications to continue treatment in the inpatient/outpatient (IPOP) program

Images

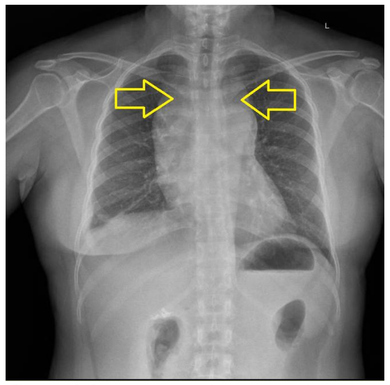

Figure 1. CXR showing widened mediastinum (yellow arrows)

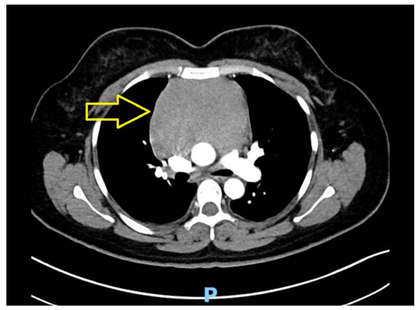

Figure 2. CTA of chest, abdomen, pelvis with and without IV contrast revealing a large mediastinal mass (yellow arrow) approximately measuring 10.0 x 7.9 x 11 cm extending from the thoracic inlet just inferior to the thyroid extending inferiorly to the right pericardial border.

Outcomes

- Patient is currently undergoing chemotherapy in IPOP program, which she is tolerating well

Discussion

Primary mediastinal T-cell ALL is a rare and aggressive form of ALL that originates in the thymus due to an abnormal development of immature T-cell precursors [6] and commonly manifests as an anterior mediastinal mass. Due to its close anatomical position to many essential midline structures, tumor expansion can lead to variety of mass effects including compression of SVC, essentially leading to the development of SVCS.

In this case, the large anterior mediastinal mass produced the classic symptoms of SVCS, including facial swelling, dyspnea, redness and warmth. To the best of the authors' knowledge, the current literature reports similar findings in only 6 cases [9-11].

This case highlights the importance of utilizing appropriate diagnostic methods, such as chest X-Ray, CT scan and most importantly tissue biopsy, which is an essential tool used to guide the decision-making process when developing the most optimal treatment course and deciding whether chemotherapy is a necessary addition to other common interventions, such as radiotherapy and endovascular procedures. [13]

Initial management targets reduction of hydrostatic pressure in the head and neck and can be achieved by elevating the head. Studies are ongoing on the most effective treatment regimen. In the past, radiation therapy was considered first-line, however, chemotherapy both alleviates the symptoms of SVCS and addresses hematologic malignancy. Most commonly seen treatment includes vincristine, an anthracycline and a steroid medication. [15]

Even though T-cell ALL prognosis has improved because of treatment advancement, its presentation in conjunction with SVCS symptoms at diagnosis may indicate a more aggressive disease and may require a more intense treatment due to increased risk of relapse in a few years.

Conclusion

The presentation of primary mediastinal T-cell ALL with SVCS is a medical emergency requiring prompt evaluation and management. This case adds to the growing body of literature on the various causes of SVCS and underscores the critical importance of tissue diagnosis and timely intervention in improving patient outcomes.