

Malignant Superior Vena Cava Syndrome

Obstruction of the superior vena cava (SVC) in the setting of cancer leads to decreased venous return to the right heart leading to a variety of potentially life-threatening effects. It commonly presents late in the course of the malignancy and prognosis is typically poor. Lung cancer is the most common underlying cause, but it can be seen in any cancer with mediastinal involvement including lung cancer, non-Hodgkin lymphoma, thymoma, germ cell tumors, or any solid tumor with mediastinal metastases. For lung cancer patients, SVC syndrome may be the initial presentation.

Pathophysiology

SVC obstruction in patients with cancer can occur a variety of ways:

- A mass in the lung or mediastinum causing external compression
- Direct tumor invasion into the SVC
- Thrombosis

When blood return from the head and upper extremities is impaired more proximal vessels dilate and collaterals form with some symptoms of venous congestion. However, if the SVC obstruction occurs quickly, there may not be enough time for collaterals to form to avoid a dangerous rise in venous pressure.

Clinical Presentation

Patients may present with a range of symptoms based on the rate of SVC occlusion and collaterals.

Common presenting symptoms:

- Facial swelling
- Arm swelling
- Visible collateral vessels in the upper body
- Shortness of breath
- Cough

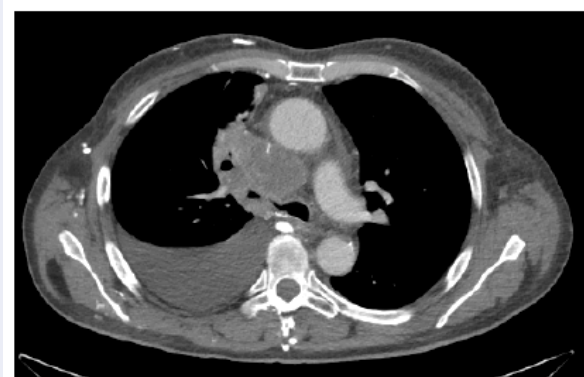
More concerning symptoms include those associated with **airway edema** (hoarseness, stridor, dysphagia) or **cerebral edema** (headaches, visual changes, altered level of consciousness). Rarely, patients may also present with shock due to decreased cardiac output from compression of the great vessels.

Investigations

For patients with suspected SVC syndrome urgent imaging should be arranged. It is important to trial having the patient lay flat prior to transporting them to radiology to ensure they can tolerate it. An experienced clinician should also accompany the patient for the test.

- **CT venogram is best imaging modality** to identify the cause and extent of obstruction.
- MRI is also an option → May provide more information about collaterals and location of the obstruction. However, given the amount of time required for an MRI, this may not be a safe alternative.

The patient's chest x-ray may have clues to the diagnosis including a widened mediastinum suggesting presence of a mass or evidence a pleural effusion.



Andersen. Imaging Med. (2015)

Management

1

Look for Life-Threatening Symptoms:

- Altered mental status
- Stridor
- Hemodynamic instability

2

Stabilize the Patient:

- Ensure adequate pre-load with IV fluids
- Avoid upper extremity IV access
- Sitting the patient up may help relieve symptoms
- Call for advanced airway expertise if the patient needs to be intubated → Airway management is very high risk in these patients

3

Recognize Potential Airway Complications:

- Avoid hypotension with potential cerebral edema
- Airway compression or edema preventing passage of the endotracheal tube
- Worsening hypotension and/or tracheal collapse with sedation and loss of spontaneous respiration

Preferred approach = Awake fiberoptic intubation with an armored endotracheal tube and elevated head of the bed.

Airway management should be performed by the most experienced provider following a multidisciplinary discussion.

4

Definitive Management:

To temporize the patient and improve their symptoms while a definitive treatment plan can be arranged, endovascular stenting is the mainstay of treatment.

Catheter-directed thrombolysis can also be considered if thrombosis is contributing to the SVC occlusion.

Although there is insufficient evidence, additional treatments including steroids and systemic anticoagulation may be recommended based on expert opinion.

If the symptoms are not life-threatening, it is important to have a multidisciplinary discussion prior to arranging endovascular therapy.

- In some circumstances, chemotherapy or radiation may be preferred if clinical stability allows.