Superior Vena Cava Syndrome

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Objectives

- Case Presentation
- SVC Syndrome
  - Pathogenesis
  - Etiology
  - Clinical Features
  - Menu of Tests
  - Treatment
  - Prognosis
Case Presentation

- 37 year old previously healthy female presenting with cough x 3 weeks, and R neck pain and swelling
- 10-15 pack year smoking history
- Physical exam:
  - Mild facial flushing
  - Trace edema in R neck and facial region
- ? SVC syndrome – Chest CT with contrast ordered
Patient’s Chest CT

SVC

Mediastinal mass
Patient’s Chest CT

R Pulmonary artery
Patient’s Chest CT

Collaterals
Hospital Course

- **Chest CT:**
  - RUL spiculated mass compatible with neoplasm
  - Mediastinal lymphadenopathy causing compression of the SVC, R main pulmonary artery and R main stem bronchus
- Transbronchial biopsy, cervical mediastinoscopy
- Lymph node biopsy: poorly differentiated carcinoma
- Dx: Unresectable stage IV non-small cell lung cancer
- Outpatient chemotherapy (cisplatin and etoposide) and radiation therapy
- Possible SVC stent for symptomatic relief
Superior Vena Cava Syndrome

Clinical manifestation resulting from partial or complete obstruction of the superior vena cava
Pathogenesis

- **SVC Obstruction**
  - Extrinsic compression
    - Benign or malignant process involving R lung, lymph nodes, mediastinal structures
  - Intrinsic or luminal obstruction
    - Neoplastic infiltration, thrombosis

- **Collateral Development**
  - Azygous, internal thoracic, lateral thoracic, paraspinous, and esophageal venous systems
Collateral Circulation

Obstruction below or at the level of the azygous vein → SVC bypass via superficial venous systems resulting in clinical evidence of SVC obstruction

Obstruction above the level of the azygous vein → direct SVC bypass and no clinical evidence of SVC obstruction
Etiology

- **Malignant - 85%**
  - Lung cancer (75-80%)
  - Lymphoma (8-10%)
  - Thymoma, mediastinal germ cell tumors, metastases (8-10%)

- **Benign - 10-15%**
  - Inflammatory – fibrinosing mediastinitis (histoplasmosis, tuberculosis), sclerosing cholangitis, sarcoidosis, postradiation fibrosis
  - Iatrogenic – thrombosis from CV line, pacemaker electrodes
Clinical Features

- **Symptoms**
  - Depend on the acuity of SVC obstruction and collateral development
    - Facial, neck, and bilateral upper extremity swelling are the most common presenting symptoms
    - Dyspnea, orthopnea, hoarseness, and cough suggest airway obstruction
    - Head fullness, syncope, and lethargy suggest cerebral edema from venous congestion

- **Clinical signs**
  - Facial plethora, tachypnea, venous distension in the neck and chest wall
  - Bending forward or lying down may worsen symptoms
SVC Syndrome

Facial and neck edema

Distended veins

www.meddean.luc.edu/.../phyabn/image15.jpg

www.UpToDate.com
Differential Diagnosis

- Pericardial tamponade and heart failure
- Nephrotic syndrome
- Mediastinal masses
- Aortic aneurysm
- Vasculitis
- Infections: Tuberculosis, Histoplasmosis, fungal
Diagnostic Tests

- **Radiologic**
  - Chest X-ray
  - Computerized axial Tomography (CT)
  - Magnetic Resonance Imaging (MRI)
  - Ultrasonography
  - Contrast-enhanced venography
  - Tc 99m scan

- **Histologic**
  - Sputum/pleural fluid cytology
  - Bone marrow biopsy
  - Lymph node biopsy

- **Procedures**
  - Bronchoscopy
  - Thoracentesis
  - Thoracotomy

- **Special Tests**
  - Increased central venous pressure (20-50 mmHg)
Mediastinal widening

Venous collaterals
- Large azygous vein
- Dilated L superior intercostal vein (aortic nipple)

Mediastinal/hilar masses

Pleural effusion

Calcifications
Computed Tomography

- Mediastinal mass
- Pulmonary lesion
- SVC obstruction
- Hilar adenopathy
- Pleural effusion
Magnetic Resonance Imaging

- Excellent anatomic visualization
- Useful if contraindication to IV contrast

Gradient echo T1-weighted MRI

Paratracheal mass

www.UpToDate.com
Ultrasonography

- **SVC**
  - cannot be directly imaged due to a lack of adequate acoustic window
  - patency can be indirectly determined with normal waveforms in the brachiocephalic and subclavian veins
- Exclusion of thrombus in the upper extremity, axillary, subclavian, and brachiocephalic veins
Ultrasonography

Patient with SVC syndrome

Patient status post SVC stent

Venous pulsatility

Respiratory phasicity
Venography

- Most conclusive diagnostic tool
- Defines SVC obstruction and collateral circulation
- Identifies thrombus

Venogram: Pt with SVC syndrome

Extrinsic compression of SVC

www.emedicine.com
Histology

- **Malignancy**
  - Small cell lung cancer
  - [www.muhealth.org/.../ thoracic/img/cellsmall.jpg](http://www.muhealth.org/.../ thoracic/img/cellsmall.jpg)

- **Histoplasmosis**
  - H. capsulatum
  - [www.med.cmu.ac.th/student/ patho/Kamthorn/](http://www.med.cmu.ac.th/student/ patho/Kamthorn/)
Treatment

- **Medical management**
  - Thrombolytics for selected cases of acute thrombosis
  - Anticoagulants to prevent clot propagation
  - Diuretics and corticosteroids for laryngeal and cerebral edema

- **Radiation and chemotherapy**
  - Non-Hodgkins lymphomas, germ cell neoplasms, limited-stage small cell lung carcinoma - responsive to chemotherapy
  - Radiation - 80-90% relieved of SVC syndrome

- **Surgical treatment**
  - Bypass of obstructed SVC
  - Mostly a palliative tool, reserved for patients with advanced intrathoracic disease

- **Endovascular treatment**
  - Minimally invasive
  - Thrombolysis, angioplasty, and stent placement
  - 80-90% procedural success rates
Endovascular Treatment

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<th>SVC syndrome</th>
<th>Stent mounted on a balloon</th>
<th>Status post SVC stent</th>
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<td>SVC occlusion</td>
<td>L superior intercostal drainage</td>
<td>Balloon deployment</td>
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<td>Patent SVC</td>
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Prognosis

- **Benign disease** – life expectancy unchanged

- **Malignant obstruction of SVC**
  - Untreated: ~30 days life expectancy
  - Treated: < 7 month life expectancy
    - 20% 1-year survival for lung cancer
      - NSCLC - poor prognosis, palliative care + radiation tx
    - 50% 2-year survival for lymphoma

- **NSCLC** - poor prognosis, palliative care + radiation tx
Summary

- SVC syndrome results from extrinsic or intrinsic obstruction of the SVC
- Clinical presentation depends on the acuity of the obstruction and adequate collateral development
- Majority of the SVC syndrome cases are caused by a malignant process
- Variety of radiologic tests are available for diagnosis
- Important to obtain a histologic diagnosis to guide treatment and determine prognosis
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References