CLINICAL VIGNETTE

Superior Vena Cava Syndrome

Adam Solis-Cohen, MD[1] and Farid Abdelmalak, MD[1]

[1]UCLA David Geffen School of Medicine, Department of Medicine

An 81-year-old male presents with shortness of breath, chest pressure, swelling of face and bilateral upper extremities (R>L) and mild bilateral foot swelling. His past medical history including COPD, CAD, and DM2. He was recently admitted to an outside hospital for shortness of breath and found to have a lung mass which was biopsied. Review of symptoms was positive for productive cough, orthopnea, and paroxysmal nocturnal dyspnea. He denied fever, chills, N/V, diarrhea. Social history positive for 50 pack years of smoking.

Home medications include aspirin, atorvastatin, insulin, nifedipine and tamsulosin.

Vital signs showed hypoxia, SpO2 98% on 5 liters nasal cannula, temperature 36.5 °C (97.7 °F), heart rate 77, respiratory rate 22, blood pressure 132/59mmHg.

Exam significant for bibasilar rales, 1+ pedal edema, 2+ bilateral upper extremity edema; R>L.

Laboratory values significant for hgb 11, wbc 11, and metabolic panel within normal limits. Troponin negative x2, BNP not elevated. EKG no acute ischemic changes, possible pericarditis.

CT chest showed: Large *right upper lobe*, *mass* 6.6 x 6.7 x 6.3cm highly suspicious for malignancy with invasion into the mediastinum and SVC. Nonvisualization of the upper and mid aspect of the *superior vena cava* also highly suspicious for invasion with prominent azygos and hemiazygos collateral vein opacification. Trace bilateral pleural effusions.

Pathology result obtained from outside lung biopsy showed *invasive adenocarcinoma.*

Bilateral upper extremity venous ultrasound positive for *thrombus* of the right *internal jugular* vein and left brachial vein.

He was admitted for acute on chronic respiratory failure, large right upper lobe mass with invasion of the mediastinum, SVC syndrome, and right IJ DVT.

The head of bed was elevated, he was started on supplemental oxygen, therapeutic anticoagulation with heparin infusion and judicious diuresis with IV lasix. Consultation was obtained from pulmonology, radiation oncology, cardiology, medical oncology and interventional radiology. Due to the large size, location and extensive venous involvement, the mass/ SVC occlusion was deemed not amenable to intervention and/or stent placement. He was started on radiation therapy.

After 4 days, despite radiation and medical management, he continued to have worsening acute respiratory failure with hypoxia requiring intubation and mechanical ventilation. He also developed atrial fibrillation with rapid ventricular response and was managed in the intensive care unit. Palliative care met with family and opted for comfort care. He was compassionately extubated and expired shortly after.

Superior Vena Cava (SVC) syndrome is a constellation of symptoms of signs caused by obstruction or occlusion of the SVC and can result in significant morbidity and mortality.[1,2]

Malignancy is the most common cause, accounting for approximately 70% of cases. Other causes include device related SVC syndrome due to central venous catheters, pacemaker or defibrillator leads.[1,3]

Historically, prior to the development of antibiotics, SVC syndrome was caused by syphilitic aortic aneurysms and mediastinal adenopathy from tuberculosis.[4]

Clinical presentation varies depending on severity, location, rapidity of onset of obstruction, and establishment of collateral veins. The most common presenting symptoms include facial and neck edema, distended neck and chest veins, watering eyes, and dizziness particularly when leaning forward.[1] Patients may also have neurological symptoms including headache, blurry vision, decreased level of consciousness, laryngopharyngeal with tongue swelling, upper extremity edema, and conjunctival/ periorbital edema.[4] Patients also typically describe worsening of their symptoms in the supine position.[1] Rarely proximal esophageal varices may be seen.[5]

Some patients with malignant SVC syndrome may present with life-threatening symptoms of cerebral, laryngeal, and pharyngeal edema due to sudden elevation in venous pressures from rapidly occluding SVC.[3,4] Median life expectancy of patients with SVC syndrome secondary to malignancy is only 6 months.[2] A scoring system grading severity of SVC syndrome

based on clinical presentation has been used to help with diagnostic approach and treatment.[6] Another rating system, the Kiski score,[7] has been used to aid in management and decision for stent therapy.

In SVC obstruction, the flow of blood is diverted to the right atrium through a collateral venous network, which can take several weeks to accommodate the usual blood flow of the SVC.[3] The severity of presentation of SVC syndrome is inversely related to the development of these collateral veins and the rapidity with which SVC obstruction develops.[1,3] SVC obstruction typically causes the venous pressure to rise as high as 20 to 40 mm Hg proximal to the obstruction. The increased venous pressure produces facial, neck, and chest wall edema.[1,3] There are 4 main collateral pathways: 1) the azygous venous system, which is the largest and consists of azygos, hemiazygos, intercostal, and lumbar veins; 2) the internal mammary pathway; 3) the lateral thoracic pathway; and 4) the vertebral venous pathway.[1,8]

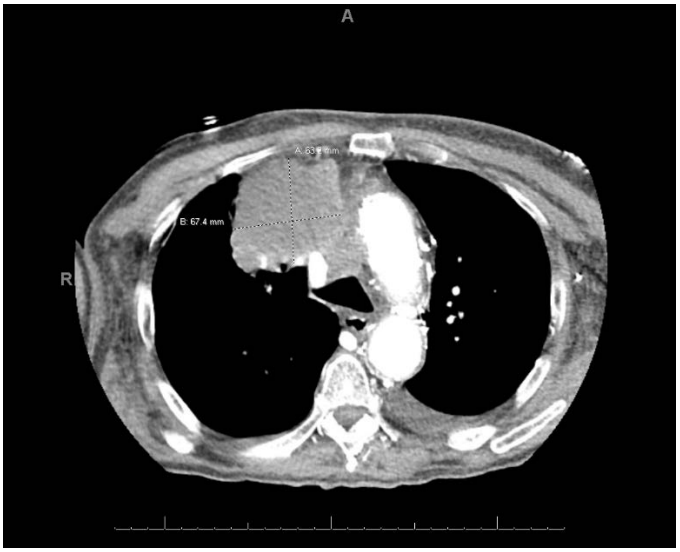
Diagnosis of SVC syndrome is based upon clinical presentation and imaging. Imaging modalities include chest radiography, contrast-enhanced computed tomography (CT) scanning, duplex ultrasound, conventional catheter-based digital subtraction venography, and magnetic resonance venography.[1] Digital subtraction venography is the gold standard for evaluation of SVC obstruction, including the presence of thrombus.[1] Magnetic resonance venography is an alternative approach in patients who demonstrate contrast dye allergy, or in cases where venous access cannot be obtained.[1]

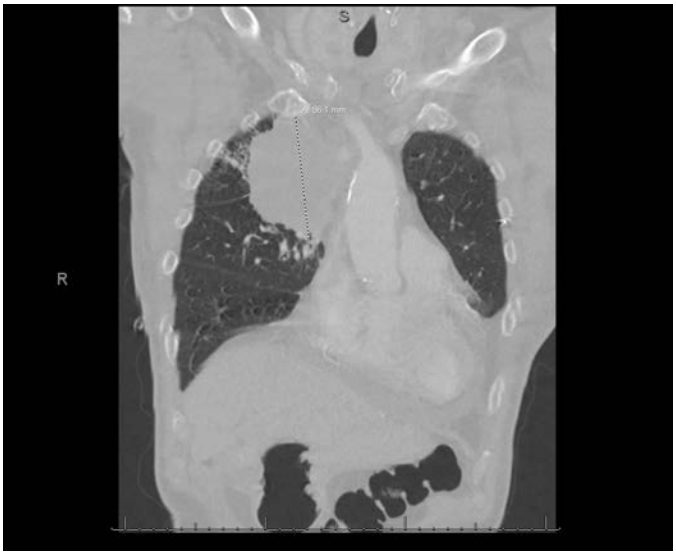
The treatment approach in patients with SVC syndrome should be multidisciplinary and may include oncology, pulmonology, radiology, surgery, and vascular and endovascular specialists. Treatment options can include chemotherapy with or without radiation therapy (RT), surgical bypass, or endovascular therapy (ET) such as angioplasty, stenting, and catheter-based thrombus removal.[1]

Initial management includes elevation of the head of bed to reduce hydrostatic pressure in the head and neck.[1] In life threatening situations initial stabilization of ABC (airway, breathing, circulation) is followed by endovascular recanalization with or without stenting.[1] Systemic anticoagulation, antiplatelet agents (if stenting is undertaken), parenteral glucocorticoids and loop diuretics are commonly used. Next, treatment options include RT and/or endovascular stenting.[1] Open surgical intervention such as bypass grafting or SVC reconstruction is reserved for cases of extensive venous thrombosis or occlusion that are not amenable to endovascular intervention.[1] In SVC syndrome with thrombosis, thrombus removal with catheter directed thrombolysis or aspiration thrombectomy is recommended prior to revascularization.[1]

Discussion

This patient presented with respiratory failure and SVC syndrome due to lung malignancy leading to invasion of the SVC. In this case, SVC occlusion was deemed not amenable to intervention or stent placement due to size, location and extensive venous involvement. Radiation therapy was initiated to reduce tumor burden. Our patient's respiratory function continued to decline leading to intubation and, ultimately, to his demise.





CT Chest with contrast axial and coronal views. Large right upper lobe, medial aspect mass highly suspicious for malignancy with invasion into the mediastinum. Nonvisualization of the upper and mid aspect of the superior vena cava also highly suspicious for invasion with prominent azygos and hemiazygos collateral vein opacification as detailed above.

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