

A Breathtaking Scenario: Superior Vena Cava Syndrome

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CASE REPORT

A 68-year-old woman with hypertension and asthma presented to the emergency department with three days of progressively worsening dyspnea and two weeks of left upper extremity swelling. She had a remote history of squamous cell carcinoma (SCC) of the left lung treated with total left pneumonectomy. One year before this admission, she developed cough and dyspnea, and computed tomography (CT) of the chest demonstrated a new mass encasing the right upper lobe bronchus with lymphangitic carcinomatosis. CT-guided tissue biopsy confirmed recurrence of SCC. Given her heavy disease burden and the compromised state of her airway and thoracic inlet, she was not a surgical candidate. She was treated with five cycles of systemic combined chemotherapy of carboplatin, abraxane, and pembrolizumab followed by maintenance pembrolizumab alone.

On this admission, review of systems revealed progressively worsening left upper extremity swelling, facial flushing, cyanosis of the lips, and orthopnea. She denied fever, chills, increased sputum production, and visual changes. Vital signs were significant for tachycardia to 119 beats per minute, blood pressure 162/81 mmHg, respiratory rate of 21 breaths per minute, temperature of 98.7°F, and oxygen saturation of 99% on ambient air. Physical examination revealed bilateral upper extremity edema (left greater than right), left breast edema, left periorbital edema, and extensive venous collateralization on the anterior chest wall (**Figure 1**). Bilateral arm elevation did not cause facial plethora (Pemberton's sign). Laboratory workup was unremarkable. CT of the chest with and without contrast showed marked interval disease progression from two weeks prior with increased size of numerous pulmonary nodules, a right apical mass that increased in size from 2.8 x 1.8 to 3.2 x 2.8 cm, and a mediastinal mass that increased in size from 4.8 x 4.4 to 5.4 x 5.4 cm. The mediastinal mass encased the right upper lobe bronchus, right upper lobe pulmonary artery, and superior vena cava (SVC). Numerous venous collaterals within the

Figure 1. Physical examination included extensive venous collateralization on the anterior chest wall (blue arrows).



right anterior and posterior chest wall were demonstrated, and the SVC was nearly completely occluded with only minute, threadlike flow (**Figure 2**).

These clinical and diagnostic findings were consistent with SVC syndrome. She underwent endovascular stent placement in the SVC, and her symptoms improved (**Figure 3**). Palliative radiation was then administered to reduce stress on the SVC, other vasculature, and airways. Repeat CT venacavogram demonstrated patency of the SVC stent, and she was discharged with home oxygen, a prednisone taper, and plans to continue palliative radiation.

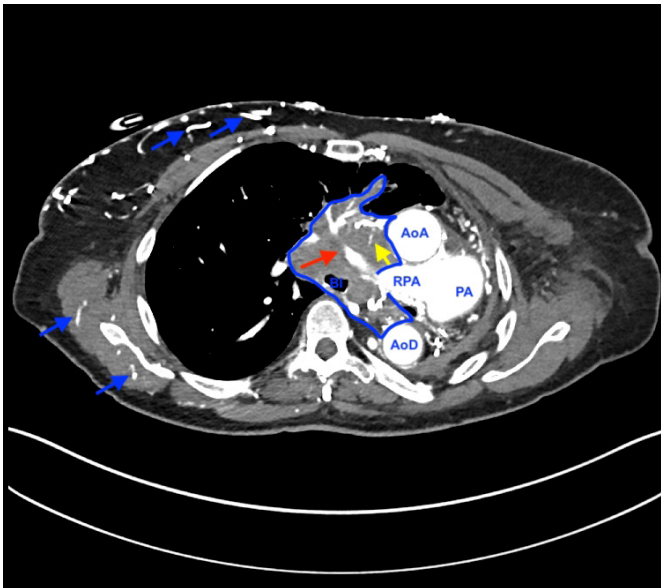
DISCUSSION

SVC syndrome was first described in 1757 by William Hunter in a patient with a large syphilitic aortic aneurysm compressing the SVC.^{1,2} The syndrome results from either partial or complete obstruction of the SVC, impeding venous return to the right atrium via external compression, vessel stenosis, or intraluminal occlusion. Malignancy is the cause

Figures 2A, 2B. Computed tomography of the chest upon presentation, prior to SVC stent placement.

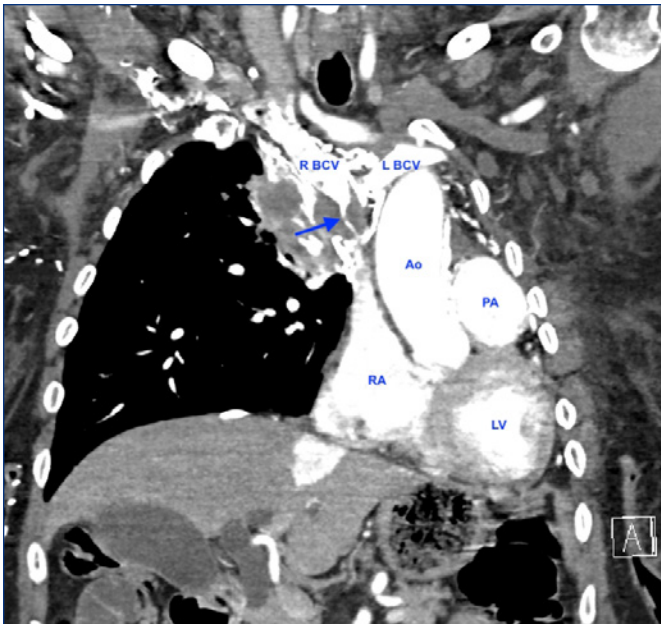
[2A] Axial image with contrast showing mediastinal mass (outlined in blue) that encases the right upper lobe pulmonary artery (red arrow) and its branches, which are diminutive (yellow arrow). No corresponding right upper lobe bronchus (implying its near-total occlusion). Numerous venous collaterals within the right anterior and posterior chest wall (blue arrows).

Abbreviations: AoA = ascending aorta, BI = bronchus intermedius, RPA = right pulmonary artery, PA = main pulmonary artery, AoD = descending aorta.



[2B] Coronal image with contrast showing minute, threadlike flow of SVC (blue arrow).

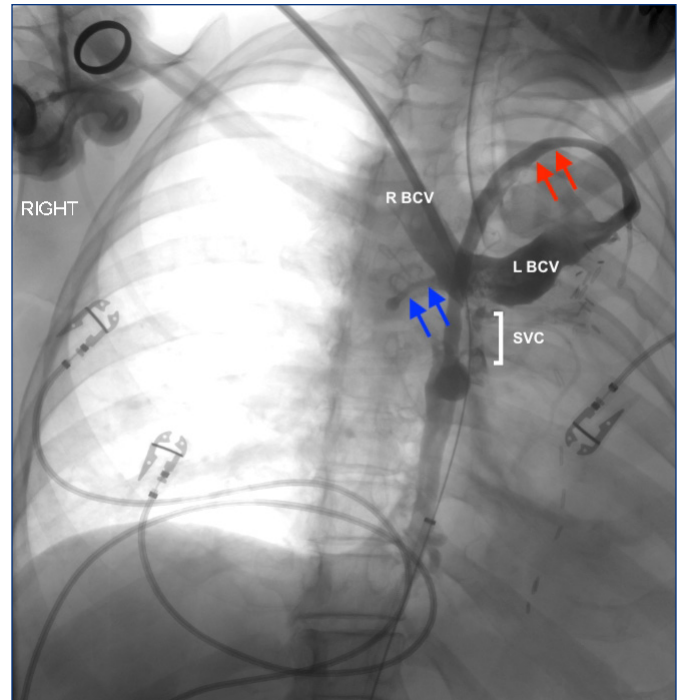
Abbreviations: R BCV = right brachiocephalic vein, L BCV = left brachiocephalic vein, Ao = aorta, PA = main pulmonary artery, RA = right atrium, LV = left ventricle.



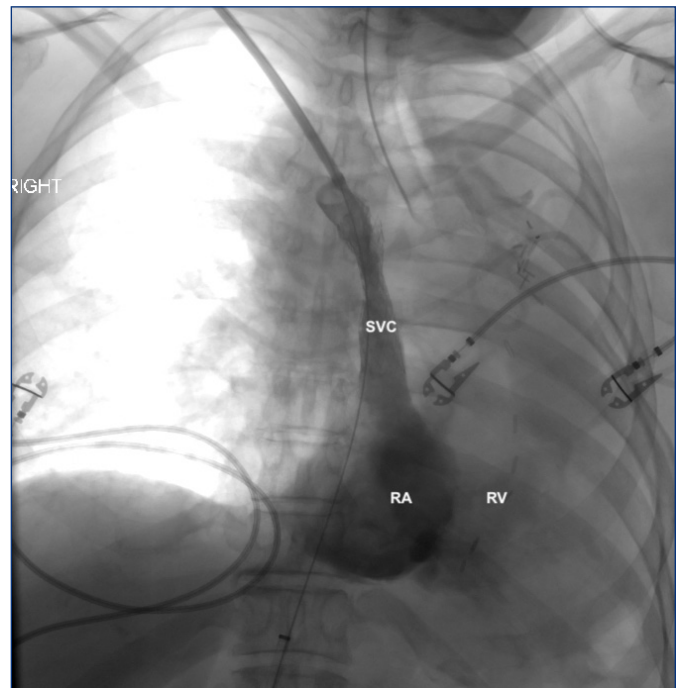
Figures 3A, 3B, 3C. Selected fluoroscopic images from endovascular stent placement in the SVC.

[3A] pre-stenting venography showing impeded flow of contrast through the SVC and resultant backflow.

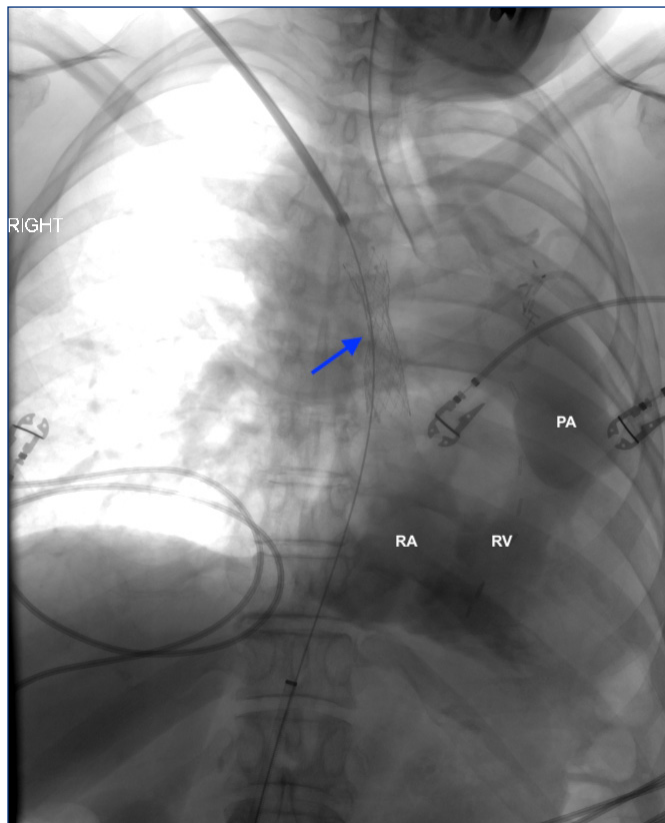
Abbreviations: R BCV = right brachiocephalic vein, L BCV = left brachiocephalic vein, red arrows = accessory hemiazygos vein, blue arrows = dilated intercostal vein draining into azygos vein.



[3B] Post-stenting venography showing flow of contrast through SVC stent into right atrium (RA) and right ventricle (RV).



[3C] Post-stenting venography showing flow of contrast past SVC stent (blue arrow) and into right atrium (RA), right ventricle (RV), main pulmonary artery (PA), and downstream pulmonary vasculature.



in up to 90% of cases, due to tumor invasion or external compression of the SVC (most commonly a bronchogenic squamous cell carcinoma). Other common etiologies of SVC obstruction include thrombus formation, infection, intravascular device malfunction, and fibrosing mediastinitis.³

Anatomically, the right and left brachiocephalic veins join to drain into the SVC. A major tributary to the SVC is the azygos vein, which can potentially connect the body's venous supply in the event of SVC obstruction.⁴ The resultant backflow and increased pressure to upstream vessels can lead to edema of the head, neck, brain, chest wall, and upper extremities. Symptoms include cough, dyspnea, stridor, hoarseness, and dysphagia, while cerebral edema can cause headaches, confusion, and visual disturbances.⁵ Plethora and cyanosis might be observed, but the diagnosis should not be precluded in darker skinned individuals. Symptom severity is correlated with the tempo of disease progression and the degree of SVC narrowing. Rarely, the condition can be fatal via one or multiple mechanisms: brainstem herniation leading to obtundation and coma, laryngeal edema leading to respiratory failure, or interruption of cardiac preload leading to hemodynamic collapse⁶ (though the latter is more likely to result from compression of the right atrium itself).

Regardless of symptom severity, any patient with a known malignancy that has the possibility of causing SVC syndrome deserves cross-sectional imaging; venous phase CT with contrast is often the study of choice, but magnetic resonance venography can be used as well. Point-of-care ultrasound (POCUS) is also helpful when making the diagnosis more urgently or in outpatient settings.⁷⁻⁹ Once SVC obstruction is confirmed, symptom severity scoring systems can be used to guide management.¹⁰ The Kishi score can aid in assessing the need for stent placement.¹¹ The median life expectancy among patients with SVC obstruction is approximately six months, but this duration can vary depending on the underlying malignancy. However, survival among patients presenting with malignant SVC syndrome does not significantly differ from survival among patients with the same tumor type and disease stage who present without SVC obstruction.⁵

For patients with malignant SVC syndrome, treatment should focus on reducing tumor burden and promptly relieving symptoms. Supportive care for patients often includes head elevation to relieve the effects of cerebral edema, though data supporting this practice is lacking.⁵ An SVC stent should be considered in these patients if they cannot tolerate optimal treatment of their malignancies or if symptoms have persisted or recurred after treatment.¹⁰ This patient presented with moderate symptoms secondary to a known radiosensitive malignancy, which would at first indicate radiation therapy as appropriate treatment. Her tumor progression on imaging paired with her acute decline in respiratory status, however, prompted the care team to pursue stenting. Post-stenting radiation was protective against recurrent SVC obstruction as well as progression of right bronchus obstruction.

The role and duration of anticoagulation and antiplatelet therapy after intravascular stenting remains an area of uncertainty in the management of SVC syndrome. Some studies suggest the use of aspirin and a P2Y12 inhibitor, while others advocate for therapeutic anticoagulation for a period of one to nine months.³ A 2013 retrospective review of patients with malignant SVC syndrome treated by endovascular stenting showed that long-term anticoagulant therapy did not influence the risk of recurrence or complications,¹² while a 2018 retrospective review of benign SVC syndrome showed the same results.¹³ However, further studies are required to develop a consensus on management.

The management of superior vena cava syndrome is focused mostly on alleviation of symptoms and treatment of underlying causes. There are no universally accepted guidelines on staging or management, but the proposed approaches might be helpful to clinicians. This patient's case highlighted pertinent physical exam and imaging findings, with notable improvement in her symptoms after endovascular stent placement, which is now the preferred treatment.

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Disclaimer

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