

When the Tumor Presses Back: Recognizing Malignant Superior Vena Cava Syndrome

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INTRODUCTION

Superior vena cava (SVC) syndrome results from obstruction of blood flow through the SVC, producing a recognizable clinical syndrome.¹ It is most often caused by malignancy, particularly lung cancer and lymphoma.² We report a case of malignant SVC syndrome due to small cell lung cancer presenting to the emergency department.

CASE PRESENTATION

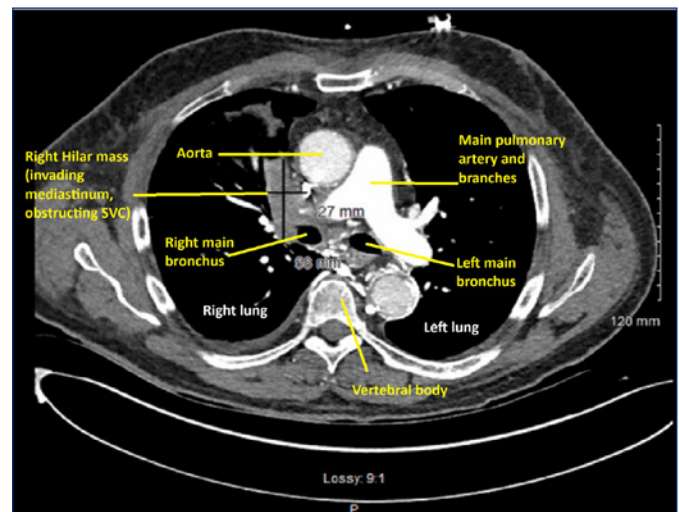
A 78-year-old man with hypertension, hyperlipidemia, and a 64-pack-year smoking history presented with several weeks of progressive facial and bilateral upper extremity swelling. His symptoms were initially attributed to pneumonia and treated empirically with antibiotics by his primary care physician but continued to worsen, and the day before presentation he noted new dilated veins across his chest wall. He reported mild exertional dyspnea and difficulty clearing sputum but denied hemoptysis, chest pain, fevers, or other constitutional symptoms; a recent colonoscopy was normal, and he had no family history of malignancy.

On arrival, vital signs were stable, and he was afebrile with normal oxygen saturation. An electrocardiogram showed new-onset atrial fibrillation with a heart rate of 98 beats per minute. Examination revealed facial plethora, marked venous distension of the neck and chest wall, bilateral upper extremity edema, hoarseness, and a positive Pemberton sign, with facial plethora and cyanosis developing within one minute of arm elevation. Laboratory studies showed mild hypercalcemia (10.4 mg/dL) and leukocytosis ($14.9 \times 10^9/L$), with normal coagulation studies. Computed tomography angiography of the chest revealed a $5.6 \times 2.7 \times 2.8$ cm right hilar mass invading the mediastinum and obstructing the SVC, with extensive chest wall venous collaterals consistent with chronic obstruction [Figures 1,2]. He underwent endobronchial ultrasound-guided fine needle aspiration of the hilar mass and mediastinal lymph nodes, along with endovascular SVC stenting and radiation therapy.

Figure 1. A portable AP Chest Radiograph on admission showing elevation of right hemidiaphragm, and mild right paratracheal soft tissue fullness.



Figure 2. High Resolution Chest CT showing a right hilar mass suspicious for malignancy that invades the mediastinum and obstructs the superior vena cava.



DISCUSSION

This case illustrates how malignant SVC syndrome can mimic more common, benign conditions, often delaying diagnosis; facial and upper extremity edema are frequently attributed to allergic reactions, venous thrombosis, or infection before malignancy is considered.⁹ SVC syndrome most often results from external compression by an enlarging tumor in the right hilum or anterior mediastinum, though direct invasion or thrombus formation in a hypercoagulable malignant state can also contribute.^{3,4} Classic findings—facial plethora, neck and upper extremity edema, dilated chest wall veins, and a positive Pemberton sign—should prompt urgent evaluation, particularly when accompanied by red flags such as stridor, hemodynamic instability, or altered mental status, which signal impending airway or cerebral compromise.^{5,6} A five-grade classification system can help stratify severity and guide management [Table 1].⁷

Histologic diagnosis should be obtained before starting therapy whenever feasible, since treatment is directed by tumor type.⁸ In this case, tissue sampling confirmed small cell lung cancer, one of the most common causes of malignant SVC syndrome given its central, mediastinal growth pattern.¹¹ Management is guided by symptom severity. Patients with mild to moderate symptoms, as in this case, can often be stabilized with supportive measures—head-of-bed elevation, supplemental oxygen, and avoidance of upper-extremity procedures—while diagnostic workup proceeds. Glucocorticoids are sometimes used empirically, though supporting evidence is limited.⁴ For patients with severe or refractory symptoms, endovascular stenting offers the most rapid and durable relief and was effective in achieving symptom control in this patient.^{10,12}

Table 1. Grading System for Superior Vena Cava Syndrome

Grade	Findings	Estimated incidence (%)
0	Asymptomatic – Radiographic superior vena cava obstruction in the absence of symptoms	10
1	Mild – Edema in head or neck (vascular distention), cyanosis, plethora	25
2	Moderate – Edema in head or neck with functional impairment (mild dysphagia, cough, mild or moderate impairment of head, jaw, or eyelid movements, visual disturbances caused by ocular edema)	50
3	Severe – Mild or moderate cerebral edema (headache, dizziness), mild/moderate laryngeal edema, or diminished cardiac reserve (syncope after bending)	10
4	Life-threatening – Significant cerebral edema (confusion, obtundation), significant laryngeal edema (stridor), or significant hemodynamic compromise (syncope without precipitating factors, hypotension, renal insufficiency)	5
5	Fatal – Death	<1

Superior vena cava syndrome is classified using a five-grade severity scale developed by Yu and colleagues (2008), which guides clinical decision-making and treatment urgency

CONCLUSION

Malignant superior vena cava syndrome is a serious oncologic emergency that requires prompt recognition, particularly in the emergency department where patients may first present. As demonstrated in this case, delays in diagnosis can occur when symptoms are attributed to more common etiologies. Increased awareness of SVC syndrome's clinical presentation can result in earlier diagnosis for this high-risk population.

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Disclosures

None

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