

Endobronchial Ultrasound Diagnosis of a Malignant Superior Vena Cava Tumor Thrombus Extending into The Right Atrium: An Unusual Cause of Recurrent Syncope.

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Abstract

The superior vena cava (SVC) is largely responsible for the return of blood flow from the head, upper limbs and neck into the right atrium. The large vein can be subject to extrinsic tumor compression as well as invasive intraluminal tumors—metastatic and mediastinal tumors that can lead to complete or partial occlusion. SVC occlusion can also result from chronic inflammation or scarring of the vessel iatrogenically by pacemaker wires or venous access ports used for chemotherapy, long-term antibiotics or hemodialysis. Patients with SVC occlusion present with a constellation of clinical abnormalities that make up SVC syndrome. SVC syndrome includes varying degrees of facial fullness, neck and upper extremity swelling, dyspnea and classically dilated collateral veins in the upper chest. Very rarely do patients present with syncope, hoarseness, dysphagia or acute encephalopathy. The diagnosis of SVC syndrome is best established on imaging such as CT Chest with contrast, however, on rare occasions, it can be discovered by endobronchial ultrasound. We present an unusual presentation of SVC syndrome—primarily presenting as frequent syncopal episodes and diagnosed via endobronchial ultrasound (EBUS). The patient had been to the Emergency Department multiple times for recurrent syncope which resolved immediately. Interestingly a Computer Tomography with Angiography (CTA) of the Chest to rule out a Pulmonary Embolus did not initially capture the finding of an intraluminal mass within the SVC. It was during an EBUS procedure that the tumor invasion of the SVC was clearly visualized. The patient's symptoms and clinical progression correlated with the EBUS findings. Obstruction of SVC causes syncope in rare cases as it decreases venous return back to the right atrium. This presentation is not commonly seen due to the extensive preventive screening currently available to patients with risk factors for cancer. This case also presents an alternative utility for EBUS in patients who cannot tolerate contrast for diagnostic purposes.

Objective

The objective of this case report is to highlight a unique approach to solving a diagnostic dilemma that may not be easily captured on a routine CTA with contrast

Introduction

Superior vena cava (SVC) syndrome was first defined in 1757 by Scottish surgeon Dr. William Hunter in a case of syphilitic aortitis [1]. With the advent of antibiotics, syphilis no longer accounts for a majority of cases of SVC syndrome. Pathologies that can cause this condition include external tumors, tumor thrombus, pacemakers, defibrillators, large thrombus or central venous catheters. Malignancy—small cell lung cancer, non-small cell lung cancer and non-Hodgkin's lymphoma now accounts for up to 60 to 85% of SVC syndrome in the United States[1,2]. The degree of symptoms exhibited by the patient is closely correlated with not only the location of SVC invasion but also the degree of occlusion—partial versus complete occlusion. Common presenting symptoms of SVC syndrome include, but are not limited to face/neck swelling, distended neck veins, cough, dyspnea, orthopnea, upper extremity swelling, distended chest vein collaterals, and conjunctival suffusion [3, 4]. Most symptoms arise and are due to swelling at the site of blockage. There are rare symptoms such as syncope and headache which only occur in approximately 6-13% of cases of SVC syndrome [1].

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Hospital Course

The patient is a 54 year old male with a history of COPD presented to the ED one month prior with complaints of syncope and worsening dyspnea on exertion. CT Scan of the head was negative and the patient was discharged on inhalers and recommended to followup outpatient with pulmonology for a Chest CT given chronic lung history.

The patient returned to the ED a week later with concerns of non-resolving syncope. He was worked up for syncope—orthostatics were negative, carotid ultrasound was negative, head CT was repeated and negative and ECHO was normal. During the admission, the patient reported worsening shortness of breath prompting CT Angiograph of the Chest for further workup of unexplained hypoxia. The CTA Chest showed a hilar mass and at the time of the read did not mention anatomic defects in the SVC. Pulmonology was consulted for bronchoscopy and biopsy of the hilar mass. Review of prior CT scan showed suspicious tissue obscuring the SVC and The procedure revealed invasion of the bronchoscopy procedure revealed tumor invasion of the SVC. A repeat CTA further showed extension into the right atrium



Figure 1: An intraluminal tumor thrombus (red triangle) within the SVC diagnosed by endobronchial ultrasound (EBUS) with trans-needle biopsy.

Discussion

SVC Syndrome is rarely incidentally captured during routine bronchoscopy. This procedure definitely changed the course of management as the patient was placed on steroids and received 5 doses of radiation therapy targeting the mediastinal mass. After RT the patient felt better and unfortunately left against medical advice, but returned a week later with worsening facial swelling and recurrent syncope. At this time, he was no longer a candidate for radiation therapy and a stent was placed in the SVC (figure 2). This successfully resolved the symptoms of recurrent syncope.

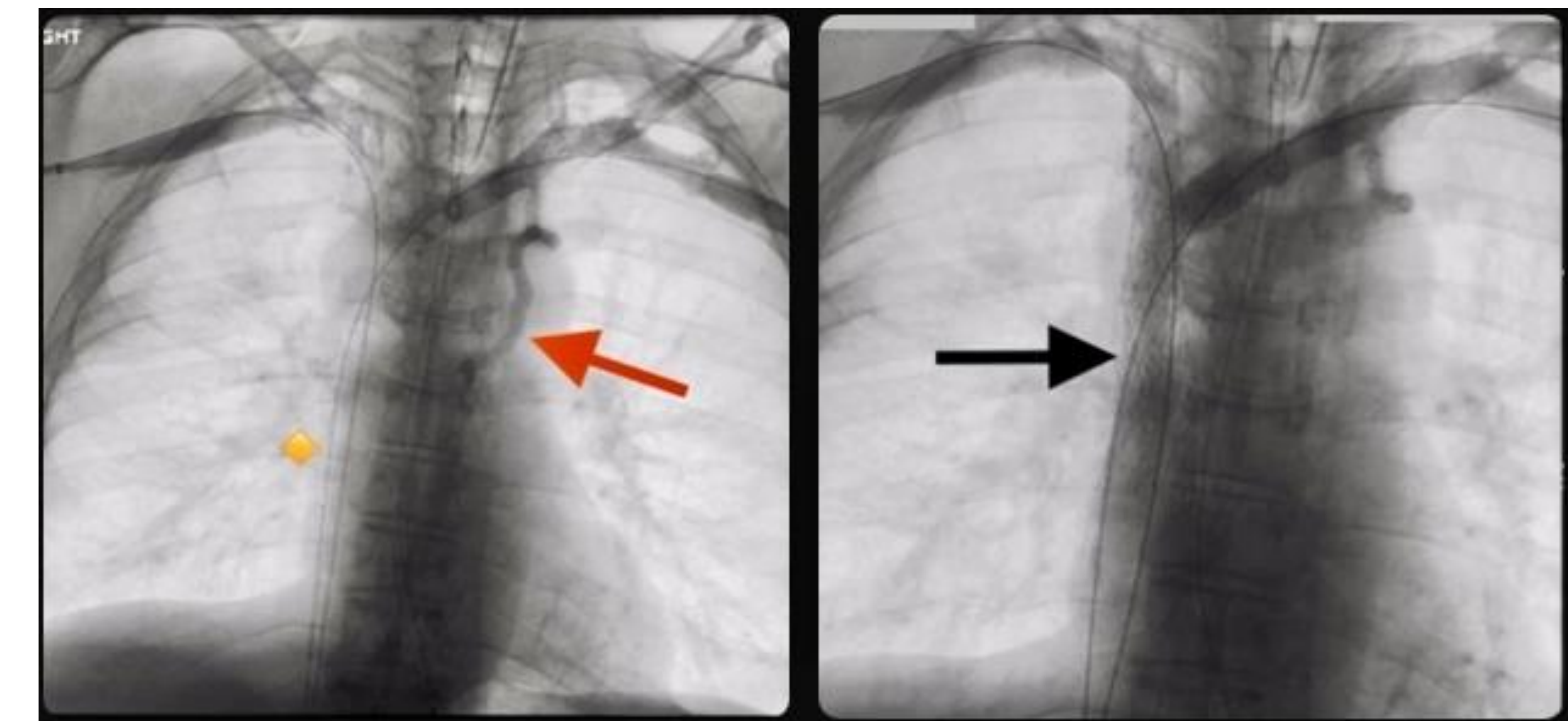


Figure 2: Central venography from bilateral axillary vein injections demonstrating severe SVC stenosis due to known tumor (yellow diamond shape) and engorgement of azygos collateral vein (red arrow). After percutaneous balloon angioplasty and stent, there was significant flow through the SVC without significant residual stenosis (black arrow).

Conclusion

SVC syndrome is an established as an oncologic emergency that can result in significant facial swelling, airway obstruction, coma, and death. Diagnosis is achieved with a high degree of clinical suspicion and advanced imaging such as a CT Chest with contrast. However Endobronchial Ultrasound with Trans-bronchial Needle Aspiration (EBUS-TBNA) provides high diagnostic yield and alternatives for patients who cannot obtain CT with contrast. EBUS is also an invasive procedure requiring anesthesia and thus must be discussed with the patient as part of a shared decision-making process.

References

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