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Case report

Iatrogenic superior vena cava syndrome with concomitant deep vein thrombosis of azygos and hemiazygos veins ☆

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ABSTRACT

Iatrogenic superior vena cava syndrome (SVCs) represents an emergent diagnostic entity and its correlation with deep vein thrombosis is extremely rare. Recently, the increased use of indwelling lines, pacemakers and intracardiac devices has led to more cases of SVC syndrome also associated with a higher frequency of DVT. We report an unusual complication in a 74-year-old female, who has been undergoing hemodialysis via CVC for 14 years, who referred at our Emergency Department complaining of shortness of breath, headache, face and neck swelling. She underwent chest Computed Tomography Angiography (CTA), that showed a thrombus extending from the superior vena cava to the azygos and hemiazygos veins. Acute SVCs should be suspected in emergency settings in symptomatic patients with indwelling central lines, catheters and pacemakers. CTA represents an accurate and quick imaging modality for the diagnosis and the assessment of the extension of the thrombus.

Introduction

The superior vena cava (SVC) is the largest systemic vein of the mediastinum and it is formed by the confluence of the left and right innominate (brachiocephalic) veins and drains head, neck, and upper part of the body into the right atrium [1,2].

SVCS is a collection of clinical signs and symptoms resulting from either partial or complete obstruction of blood flow through the SVC; it occurs when there is direct compression or obstruction of the superior vena cava as a result of thrombus formation or tumor infiltration of the vessel wall. SVCS develops when the ability of collateral blood vessels to compensate for the SVC obstruction is exceeded [3,4]. When the SVC becomes obstructed, collateral venous return to the heart from the upper half of the body takes places through four principal pathways. The primary and most important pathway is the azygous venous system, which includes the azygos vein,

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the hemiazygos veins, and the connecting intercostal veins [2,5]. The azygos, hemiazygos and accessory hemiazygos veins originate from the last portion of the posterior cardinal veins. The azygos system is a paired paravertebral venous pathway in the posterior thorax. It passes through the aortic hiatus and ascends in the thorax to join the superior vena cava. The hemiazygos vein is the asymmetric counterpart to the azygos vein and forms part of the azygos venous system [6]. The second pathway is the internal mammary venous system plus tributaries and secondary communications to the superior and inferior epigastric veins. The long thoracic venous system, with its connections to the femoral veins and vertebral veins, provides the third and fourth collateral routes, respectively. If SVCS occurs slowly, as in the case of malignancies, adequate collateral venous return develops and minimizes the sequelae of the syndrome. In acute obstruction, as seen with thrombosis, the presentation can be much more life threatening [7,8]. SVCS has usually been associated with mediastinal malignancies, primary among which is bronchogenic carcinoma, secondarily with non-Hodgkin lymphoma and metastatic tumors. Benign and non-malignant causes and infectious disease comprise at least 40% of cases [9–11]. Non-malignant causes include benign mediastinal tumors, cardiac causes, vascular diseases and fibrosing mediastinitis, although SVCS has also been reported in infectious disease such as such as tuberculosis, syphilis, histoplasmosis, and actinomycosis [12]. The increased use of pacemaker wires and semi-permanent intravascular catheters used for hemodialysis, long term antibiotics, or chemotherapy has also led to more cases of SVC syndrome, for these reasons iatrogenic thrombus formation or SVC stenosis is a growing etiology [10,11]. Acute iatrogenic SVCS should be suspected in emergency settings in symptomatic patients with indwelling central lines, catheters, and pacemakers. The diagnosis of SVCS is clinically based on the presence of characteristic signs and symptoms with imaging being supportive to confirm and define the extension of the disease. CTA represents and accurate and quick imaging modality in ED settings [13].

Case report

A 75-year-old female on regular hemodialysis for the last 14 years via CVC was brought was to our Emergency Department complaining of shortness of breath, headache, face and neck swelling. The patient denied any history of cough, hemoptysis, chest pain or wheeze. There was no recent history of fever nor loss of weight. She denied any use of illicit drugs or alcohol. On clinical examination, the patient was afebrile but tachypneic. Dilated and tortuous veins were not observed over upper extremity and abdomen. She did not present edema in other parts of the body and blood oxygen was in the normal range. Laboratory tests showed reduction of HGB (7.5g/dl, reference range 11.5-17.5 g/dl) and RBC (3,4* 10 ^ 6/ mm^3, reference range 4* -5.8* 10 ^ 6/ mm) and net increase of fibrinogen (588 mg/dl, reference range160-380 mg/dl). Past medical history documented hypertension, chronic coronary syndrome that had required coronary artery bypass graft, antithrombotic treatment (Fondaparinux), femur fracture and CKD in end stage on dialytic treatment with CVC that was inserted from right jugular vein to the superior vena cava. During the last dialytic treatment, carried out in the morning, the CVC function was good. Upper left limb ischemia was the first suspect but Doppler-sonography showed a normal flow of the radial and ulnar arteries. The vascular surgeon required a chest-CT with intravenous contrast. The CT reported multiple blood clots around the CVC in the superior vena cava with extension to the left innominate, axillary, azygos and hemiazygos and showed also right basal pleural effusion (20 mm maximum thickness) with atelectasis of the lung parenchyma (Fig. 1 and 2). A diagnosis of superior vena cava syndrome was formulated and pharmacologic treatment was initiated (Fondaparinux and LMWH). Surgical treatment was not needed.

Discussion

Superior vena cava syndrome (SVCS) is a group of symptoms caused by mechanical obstruction of the SVC that prevents the normal blood flow to the right atrium. The obstruction is associated mostly with an intrathoracic malignancy causing compression or invading the SVC, and with benign causes in up to 40% of cases [12]. Recently iatrogenic SVCS caused by intravenous catheter-associated thrombosis is more frequently reported, this may be related also to the increased use of intravascular devices [14]. Hemodialysis (HD) catheters, inserted into the internal jugular vein, may lead to catheter-related thrombosis. Moreover, as compared to other central venous catheters (CVCs), it is believed that HD catheters may be associated with an increased risk of thrombosis-related complications. The mechanism underlying HD catheter associated thrombosis is poorly understood, however factors such as recurrent vascular access, platelet dysfunction, endothelial factors, inflammation, and clotting abnormalities have been suggested [15]. Other HD thrombosis-related risk factors include catheter-related features (such as catheter caliber-to-vein), venipuncture-associated trauma (nowadays reduced with ultrasound guidance), and catheter position (i.e., higher risk if catheter tip is in the brachiocephalic vein or proximal SVC versus distal to SVC). Practice recommendations stated the catheter to vessel ratio (CVR) can assist clinicians in selecting the most appropriate-sized for device vessel, and catheter to vessel ratio CVR can increase from 33% to 45% of the vessel's diameter; this is supported by the fact that larger diameter catheters can increase thrombosis risk [16,17]. There are several risk factors associated with catheter related thrombi; catheter placement can damage the endothelial wall, the catheter can be incorrectly placed, or it can also impede blood flow within a vein, causing blood stasis. These are components of Virchow's triad, and they are factors considered to contribute to thrombosis in iatrogenic SVCS [18]. The diagnosis of SVCS is largely based on a patient's history and clinical findings which can develop acutely or over a period of days to weeks; this insidious onset is a result of a collateral vascular network. The resulting venous congestion produces a clinical scenario relating to increased upper body venous pressures. The most common signs and symptoms include face or neck swelling, upper extremity swelling, dyspnea, cough, dis-

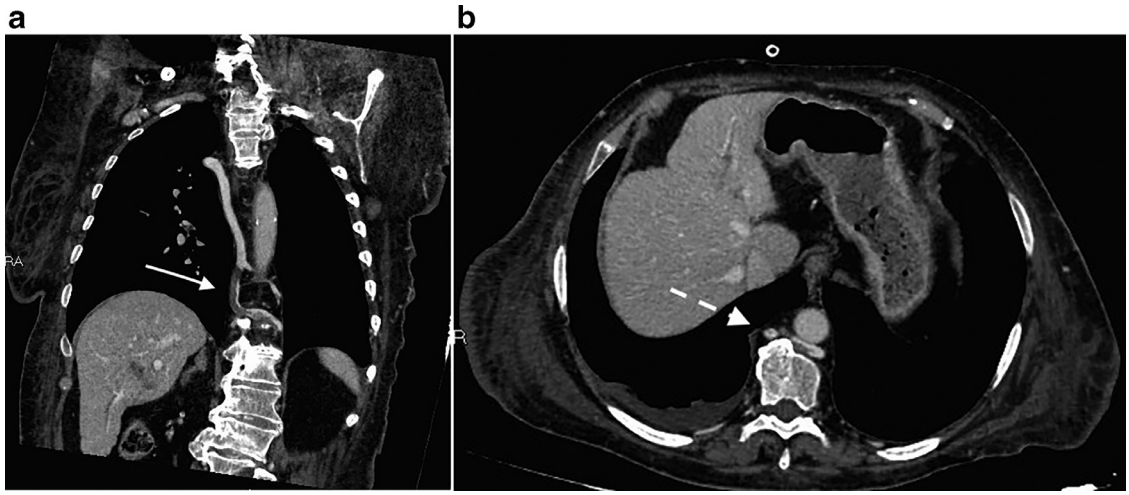

Fig. 1 – In the venous CT phase, there's the evidence of the hemiazygos vein thrombosis (arrow) leading to azygos vein on the MPR coronal plane (white arrow) (a). Evidence of the hemiazygos vein thrombosis (arrow) on the axial plane- venous CT phase (white dashed arrow) (b).

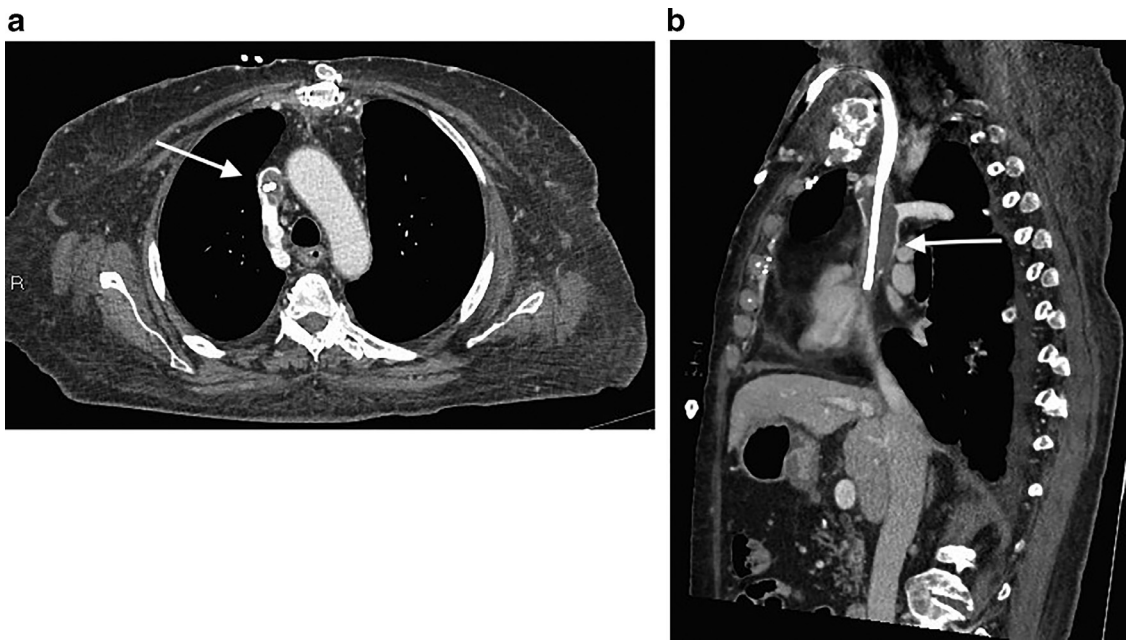

Fig. 2 – Venous CT phase-confluency of the azygos vein into superior cava vein with the evidence of thrombosis (white arrow) enveloping the CVC (star) in a muff, on the axial (a) and MPR sagittal (b) planes.

tended neck and chest vein collaterals, orthopnea, and conjunctival suffusion. Other less common symptoms of SVC syndrome include stridor, hoarseness, dysphagia, headache, nausea, lightheadedness, syncope, change in vision, altered mental status, upper body edema, cyanosis, papilledema, stupor, and coma. Some rare but serious clinical consequences reported in SVC syndrome include cerebral edema and upper respiratory tract obstruction secondary to edema of the larynx and pharynx. Thrombosis can also remain clinically silent and be discovered only later during examinations performed for different reasons [3,4,19]. The survival in patients with SVCS depends mainly on the course of the underlying disease. No mortality, per se, results directly from mild venous congestion. In patients with benign SVCS, life expectancy is unchanged [3]. A careful clinical examination is often sufficient to rule out a cardiogenic origin of the patient's symptoms. Patients with high clinical suspicion of SVCS syndrome should undergo imaging of the upper body and its vascular system. A traditional radiological exam such as chest X-ray can't show direct signs of SVC thrombosis, but only indirect signs as lymphadenopathies or pathologic opacities in lung parenchyma or mediastinum in case of malignant SVCS [4]. Ultrasonography can provide information about the presence and extension of a thrombus, sonographic findings of superior vena cava

obstruction are represented by the venous waveform in the subclavian and brachiocephalic veins that may show dampening, loss of pulsatility and minimal respiratory variation [20], although SVC cannot be directly visualized with ultrasound. Chest-CT Angiography is crucial to define the urgency of medical intervention and to establish a definitive diagnosis. In our case, CT angiography showed the thrombus around the tip of the catheter inserted in the SVC extending to the azygos vein. The thrombus determined an eccentric tubular and elongated filling defect and presented a small attachment to the roof of the vessel, forming an acute angle with the vessel wall. The rest of the azygos vein was fully opacified, perhaps and another tubular filling defect was recognized into the hemyazygos vein and in an intercostal vein (Fig. 1 and 2). The case we presented is very rare because of the extension of multiple thrombi in the SVC with associated deep venous thrombosis of the azygos and hemiazygos system, which involved the right and the left thoracic venous system and both the upper and lower body venous system. CT angiography represents the imaging gold standard and should be part of the diagnostic algorithm for the diagnosis of SVCs and to determine the possible extension into the deep veins.

Patient consent

Patient's consent not required as patients identity is not disclosed or compromised.

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