Unilateral superior vena cava syndrome: An uncommon complication of a commonly used procedure

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Superior vena cava (SVC) syndrome constellates symptoms of plethora and facial and upper thoracic swelling with distension of neck veins, and the consequences of this condition range from mild discomfort to upper airway obstruction. The aetiology has a strong linkage with malignancy, and is always to be excluded. SVC syndrome is commonly a bilateral presentation, and unilateral SVC syndrome is rare, with very few non-malignant causes reported. Here, we demonstrate a unilateral occurrence with an uncommon cause.

Keywords: SVC syndrome, central venous catheter, superior vena cava

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The superior vena cava (SVC), formed by the right and left brachiocephalic veins, is responsible for venous drainage of the upper head and neck, and obstruction of the venous return by various mechanisms creates a congestive syndrome.[1,2] Classically, symptoms of SVC syndrome are bilateral neck swelling, which can lead to life-threatening upper airway compromise and subclavian vein compression with upper torso oedema.[3] Current evidence attributes >70% of cases to mediastinal malignancies, although the rarer benign aetiology has a better prognosis.[4] Therefore, unilateral SVC syndromes, although uncommon, may still be due to a malignancy, and hence this should always be investigated for.[5] Device-associated SVC syndrome can be caused by pacemakers and central venous catheters, with resultant stenosis and thrombi and, as in our clinical case, may result in acute SVC syndrome with obstruction and painful unilateral symptoms.[6] In our clinical setting, with a high burden of comorbid disease, patients often present late and acutely ill, requiring interventions such as central venous catheters, and these need to be meticulously and carefully placed to avoid complications.[7] Sinister causes and even vascular anomalies need to be excluded post this rare unilateral syndrome occurrence.[8] Treatment modalities of SVC syndrome depend on the aetiology as well as the severity of the syndrome and its complications.[8] Outcomes of any treatment employed depend on the aetiology and the progression or severity of the clinical state that the patient presented with.[9] Therefore, it is important to note that treatment outcomes may range from relief of acute symptoms to resolution of the syndrome, depending solely on the underlying disease process.[10] Our patient described here depicts a rare unilateral SVC syndrome caused by a thrombus from a central venous catheter placed in a recent hospitalisation.

Case presentation

A 38-year-old female known to be HIV infected on antiretroviral therapy (ART) with suppressed viral level was diagnosed with urogenital tuberculosis (TB) and left renal abscess at a tertiary hospital. The urogenital TB was confirmed on a positive TB culture and Gene Xpert. A nephrostomy was required to manage subsequent hydroureter and hydronephrosis. During that admission she required intravenous fluids through a right internal jugular central venous catheter to resolve her haemodynamic instability after nephrostomy insertion, and was discharged when she was stable.

She presented to our district level hospital 2 weeks later for acute medical admission with a 2-week history of swelling of the right side of her upper body, which was localised to her face, neck, breast and bilateral lower limbs. She reported a 1-week history of exertional dyspnoea with reduced effort tolerance and orthopnoea.

On clinical examination, this patient appeared acutely unwell and tachypnoeic, with a respiratory rate of 30 breaths per minute (bpm), saturating at 98% in room air, while the other vitals were within normal ranges. She had unilateral and tender soft-tissue swelling of the right side of her upper body that involved her face, neck, breast, arm and the right side of her abdomen. Visibly distended right-sided external jugular vein was also present. There were no signs of airway compromise and on auscultation, no bibasal crackles. The cardiovascular findings included bipedal oedema, loud and palpable second heart sound and a raised jugular venous pressure waveform, pronounced on her right side (Fig. 1).

Special investigations performed

Chest radiography performed at the emergency department upon her arrival showed features of an enlarged cardiothoracic ratio, as well as bilateral small pleural effusions without evidence for pulmonary oedema.

An echocardiogram of the heart was performed that revealed dilated cardiomyopathy with an ejection fraction of 30%, with secondary mitral regurgitation as well as tricuspid regurgitation. No regional wall motion abnormalities were detected (Table 1).

Given the atypical right-sided presentation of oedema, further diagnostic evaluation was commenced. Contrast-enhanced computed tomography angiography scan of the neck revealed a unilateral right-sided thrombus extending from the proximal/mid-internal jugular vein to the SVC inlet (Fig. 2A and B).

The patient was started on anticoagulation. However, considering the extent of the thrombus as well as the painful unilateral SVC syndrome present, this patient was discussed with vascular surgery at a tertiary hospital. It was concluded that the likely aetiology of

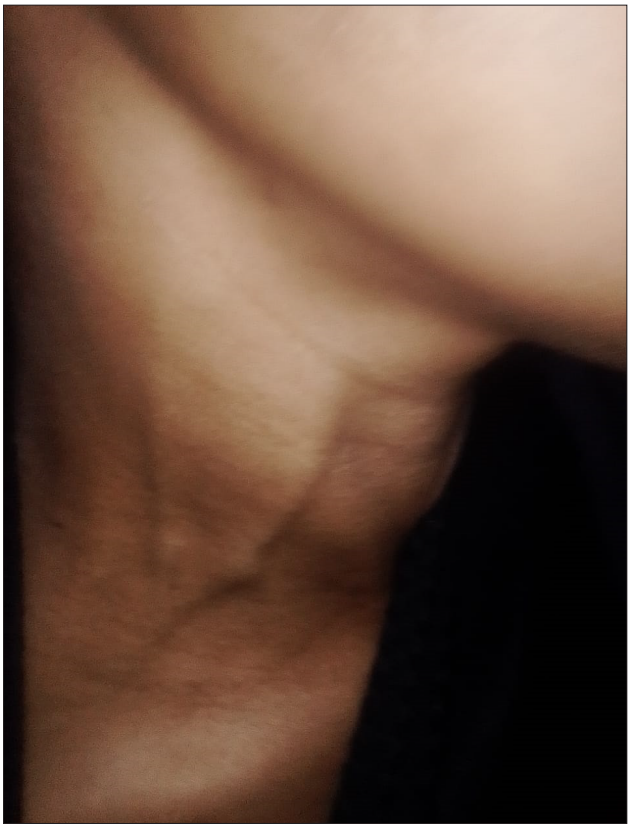

Fig. 1. Distended neck vein on the right side of the patient's body.

the thrombus waslinked to a prolonged right internal jugular central venous catheterisation. The discussion regarding a thrombectomy was considered. However, it was deferred on the basis of limited data available on prognosis and outcomes post surgery. During the patient's hospital stay, the bipedal oedema cleared up within 3 days on antifailure treatment, yet the right-sided oedema of the face, neck and upper torso persisted, and ultimately resolved with anticoagulation therapy within 3 weeks.

Discussion

Our patient's oedematous state was clearly a combination of unilateral SVC syndrome and dilated cardiomyopathy. The overwhelming right-sided upper-body painful oedema was atypical for dilated cardiomyopathy, which led us to investigate further. The aetiology here was not common nor of malignant origin; however, the syndrome can still manifest with severe consequences. Adequate history taking and seeking common causes through imaging remain important. Unilateral SVC syndrome has a variable outcome that is dependent on the cause of the syndrome. In our case, the cause of internal jugular vein thrombosis was benign, and the proposed mechanisms for thrombus formation around the catheter site may be attributed to venous stasis and possibly inflammatory cytokine adhesion molecules. Early surgical involvement remains essential to prevent life-threatening consequences. Our literature search did not find much information on the duration of anticoagulation for internal jugular vein thrombosis, and it was decided by both the medical and surgical teams to treat with life-long anticoagulation, and to continue antifailure therapy for the dilated cardiomyopathy. Central venous catheterisation is a much-needed lifesaving intervention, and care

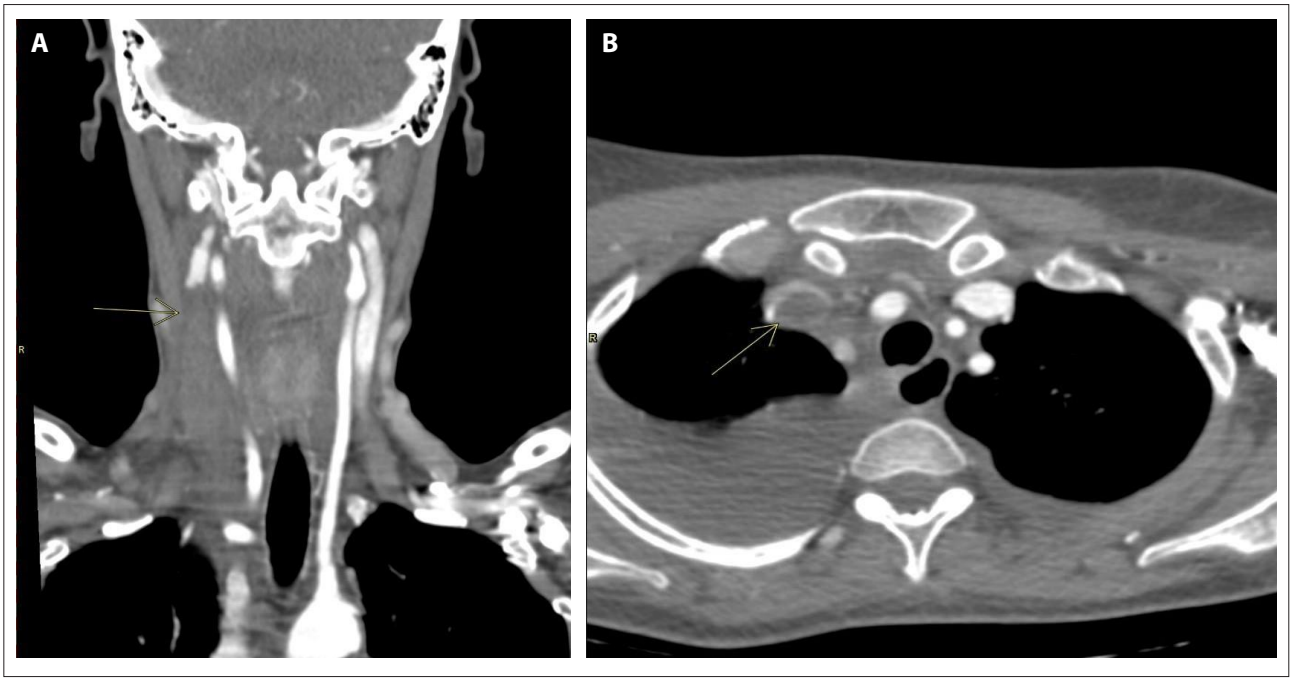

Fig. 2A and B. Long segment non-opacification of the right internal jugular vein (IJV), extending from the proximal IJV to the superior vena cava inlet.

Table 1. Laboratory findings

Parameter	Finding	Reference range
CD4+	1 125 cells/μL	332 - 1 652 cells/μL
Creatinine	90 μmol/L	49 - 90 μmol/L
White cell count	12.59 × 10^9/L	3.90 - 12.60 × 10^9/L
C-reactive protein	39 mg/L	<10 mg/L

should always be taken to ensure correct insertion and frequent monitoring of complications post insertion.

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Conflicts of interest. None.

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