Type A aortic dissection presenting as superior vena cava syndrome

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ABSTRACT

A 51-year-old man presented with a 5-day history of progressive facial swelling, sensation of head fullness, increasing shortness of breath and paroxysmal nocturnal dyspnea. He denied chest pain, syncope or presyncope. Past medical history included mechanical aortic valve replacement 7 years prior and atrial fibrillation treated with warfarin. A clinical diagnosis of acute superior vena cava (SVC) syndrome was made. Portable chest radiograph showed a widened superior mediastinum. Computed tomography scan of the thorax demonstrated a large type A aortic dissection almost completely effacing the SVC. Acute type A aortic dissection (AD) is an emergency requiring prompt diagnosis and treatment. Patients typically present with acute onset of chest and/or back pain, classically described as ''ripping'' or ''tearing.'' SVC syndrome is rarely, if ever, mentioned as a presentation, as it is usually due to more chronic conditions. This case illustrates a rare incidence of type A AD actually presenting as SVC syndrome.

RÉSUMÉ

Un homme de 51 ans a consulté pour une tuméfaction progressive de la face qui durait depuis 5 jours, une sensation de plénitude crânienne, un essoufflement de plus en plus marqué et de la dyspnée paroxystique nocturne. Selon le patient, il n'a pas ressenti de douleur thoracique ni fait de syncope ou de présyncope. Ses antécédents médicaux comprenaient un remplacement valvulaire aortique par une prothèse mécanique, réalisé 7 ans auparavant, et de la fibrillation auriculaire traitée par la warfarine. Un diagnostic de syndrome de compression de la veine cave supérieure (VCS) aigu a été posé. Une radiographie effectuée au moyen d'un appareil portatif a révélé un élargissement du médiastin supérieur; et la tomodensitométrie du thorax, une dissection importante de l'aorte de type A, qui effaçait presque complètement la VCS. Une dissection aortique (DA) aiguë de type A est une urgence médicale qui exige un diagnostic et un traitement rapides. Habituellement, les patients consultent pour une douleur thoracique ou dorsale d'apparition brutale, et la décrivent comme une sensation de «déchirure» ou de «rupture». Le syndrome de compression de la VCS fait rarement, voire jamais, l'objet de présentation étant donné qu'il est généralement attribuable à des affections chroniques. Il s'agit d'un rare cas de DA de type A se manifestant sous la forme du syndrome de compression de la VCS.

Keywords: aortic dissection, aortic valve replacement, cardiac surgery, computed tomography, superior vena cava syndrome

Aortic dissection is a medical emergency requiring prompt diagnosis and treatment. In type A dissection, patients typically present with the acute onset of chest and/or back pain, classically described as ''ripping'' or ''tearing.''[1,2] Associated findings, related to inadequate perfusion and end-organ damage, can include congestive heart failure, stroke, syncope, and hemodynamic changes.[1–3] Superior vena cava (SVC) syndrome is a rare finding in type A aortic dissection.[1,4] We report and discuss a case of type A aortic dissection presenting with SVC syndrome.

CASE REPORT

A 51-year-old man presented to the emergency department with a 5-day history of progressive facial swelling, a sensation of head fullness, increasing shortness of breath, and paroxysmal nocturnal dyspnea. He denied chest pain, syncope, or presyncope. His past medical history included a bioprosthetic

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aortic valve replacement and mechanical aortic valve replacement 25 and 7 years prior to presentation, respectively, and atrial fibrillation, for which he was taking warfarin.

On examination, he was nondistressed but had edema of his neck and face. He was breathing normally, and his vital signs were as follows: oxygen saturation 92% on 2 L oxygen by nasal prongs; blood pressure 114/97 mm Hg in the right arm and 119/79 mm Hg in the left arm; and heart rate 80 beats per minute and irregularly irregular. His heart sounds were normal, with no murmurs, and his air entry was normal by auscultation. Laboratory investigations showed normal electrolytes, urea, creatinine, total protein, albumin, bilirubin, liver enzymes, amylase, glucose, and cardiac biomarkers (CK-MB and troponin-I). The patient's leukocyte count was elevated at 12.7×10^9/L (normal 4.0–10.0 10^9/L), whereas his hemoglobin and platelets were normal. His international normalized ratio was increased at 2.0, and D-dimer was elevated at 294 μg/L (normal $<$ 240 μg/L). His electrocardiogram showed atrial fibrillation, and a chest radiograph showed slight widening of the superior mediastinum (Figure 1). A provisional diagnosis of acute SVC syndrome was made.

A nongated contrast-enhanced computed tomographic (CT) scan of the thorax showed an extensive type A aortic dissection compressing and almost completely effacing the SVC (Figure 2, Figure 3, and Figure 4) and engorged venous collaterals of the upper right arm and chest wall (Figure 5).

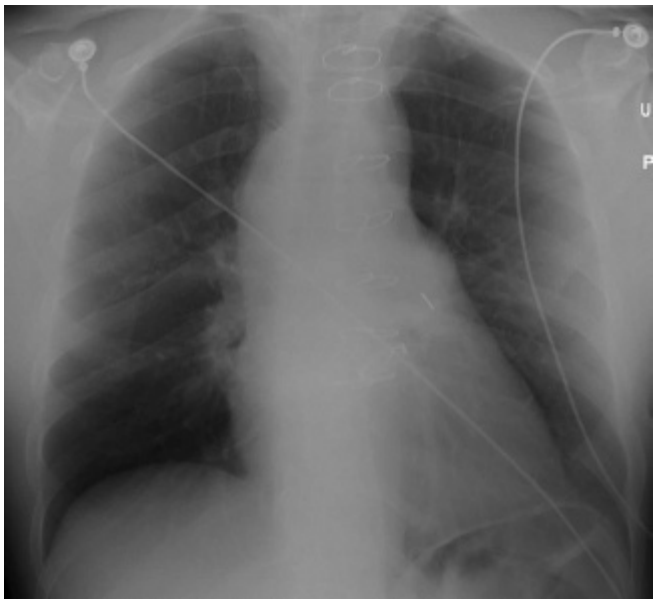

Figure 1. A portable chest radiograph showing a widened superior mediastinum.

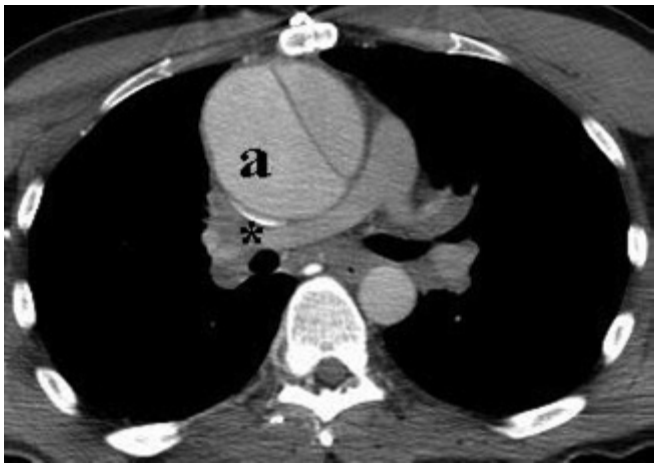

Figure 2. Nongated contrast-enhanced axial computed tomographic scan of the thorax demonstrating a large type A aortic dissection (a) almost completely effacing the superior vena cava (*).

The patient subsequently became hypertensive and was admitted to the cardiovascular intensive care unit for blood pressure and coagulation status optimization prior to emergent surgery. In the 2 years of postoperative follow-up that took place following his presentation, the patient remained well.

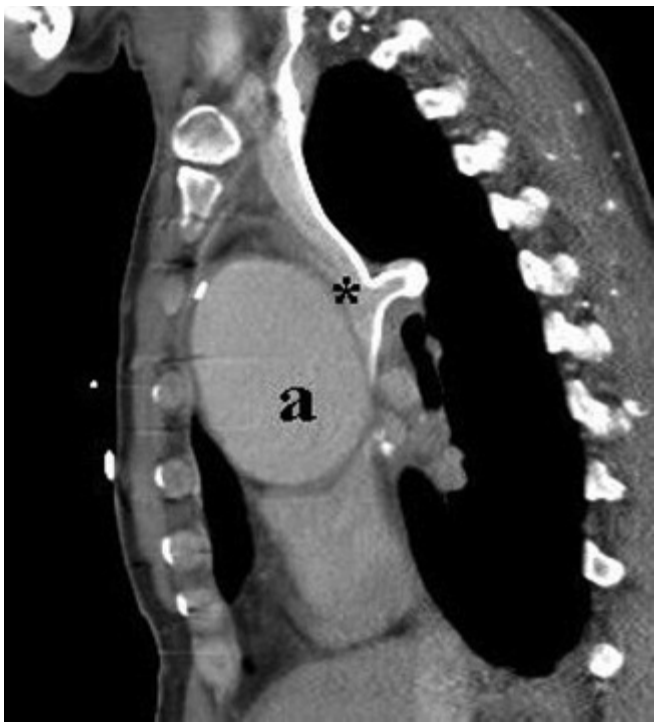

Figure 3. Sagittal reformatting of a computed tomographic scan showing aortic dissection (a) compressing the superior vena cava (*).

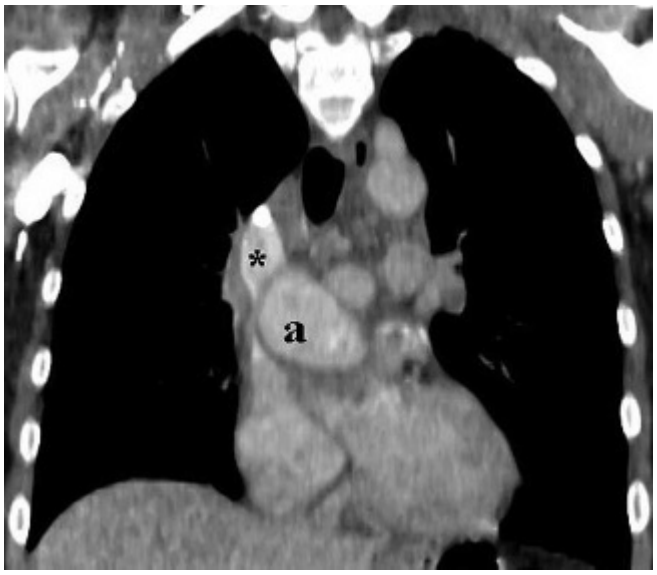

Figure 4. Coronal reformatting of a computed tomographic scan showing aortic dissection (a) compressing the superior vena cava (*).

DISCUSSION

SVC syndrome is the term used to describe a constellation of signs and symptoms resulting from obstruction of venous return at the level of the SVC, the superior atrial-caval junction, or the great veins as they empty into the SVC.[4] Historically, infectious etiologies were common; however, now most cases of SVC syndrome arise from a thrombogenic intravascular device or malignancy, most commonly lymphoma or primary lung cancer.[4,5] Common signs and symptoms include head and neck edema and plethora, upper extremity edema, dyspnea, cough, head pressure, hoarseness, dysphagia, jugular and thoracic venous distention, and syncope.[6,7] It is rare for SVC syndrome to require emergent intervention on presentation.[4]

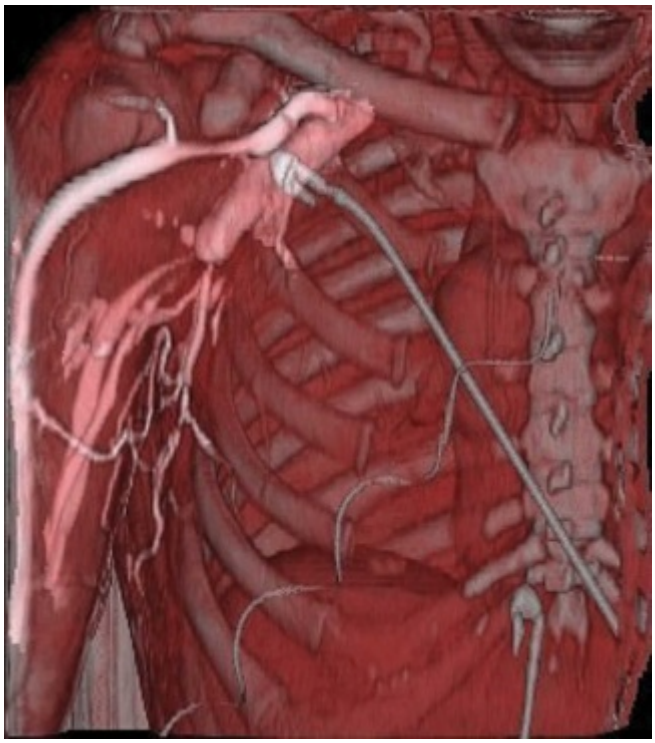

Figure 5. Three-dimensional volume rendering exhibiting engorged venous collaterals of the upper arm and chest wall.

Aortic dissection results from an intimal tear that allows blood to divide the medial layer, resulting in a true and a false lumen.[8] In 70% of cases, these two lumens are separated by an intimal flap.[9] The Stanford classification, which is most commonly used, is based on involvement of the ascending aorta. It defines a type A dissection as involving the ascending aorta, regardless of distal extent, and a type B dissection as originating from anywhere distal to the left subclavian artery. Hypertension is the most common predisposing factor.[8,10] Other factors include connective tissue diseases such as Marfan or Ehler-Danlos syndrome, congenital or acquired aortic valvular defects, aortic coarctation, aortic aneurysm, infection and other causes of aortitis, pregnancy, and cocaine use.[8] In otherwise healthy patients younger than 40 years of age, Marfan syndrome is the most common predisposing condition.[10]

Aortic dissection is deemed "acute" when the diagnosis is made within 2 weeks of symptom onset and "chronic" when symptoms are greater than 2 weeks in duration.[10] Type A dissection is the most common and accounts for 75% of cases.[8] As the great majority of deaths arise from acute dissections, immediate repair of acute type A dissection is indicated to avoid complications such as extension into the pericardium, pleural space, coronary arteries, or aortic valve ring and their resulting increase in mobidity and mortality.[8] Type B dissection is usually treated medically unless end-organ ischemia is present.

The International Registry of Acute Aortic Dissection (IRAD) published in 2000 stated that the most common presenting symptom of type A aortic dissection is pain (93.8%) in either the chest (78.9%), back (46.6%), or abdomen (21.6%).[11] Other signs and symptoms include aortic insufficiency murmur (44.0%), hypertension (systolic blood pressure [SBP] > 149 mm Hg; 35.7%), pulse deficit (18.7%), shock/tamponade (13.0%), hypotension (SBP < 100 mm Hg;

11.6%), congestive heart failure (8.8%), and stroke (6.1%).[11]

CT is the most common imaging modality employed to investigate suspected acute aortic dissection owing to its speed and wide availability.[8–10] Helical CT has a specificity and a sensitivity that approach 100%.[8,9] Furthermore, CT aids in distinguishing acute aortic dissection from other similarly presenting conditions, such as acute intramural hematoma and penetrating atherosclerotic ulcer.[8,9] Most institutions use a protocol that begins with an unenhanced scan and is followed by a contrast-enhanced arterial phase scan.

Acute type A aortic dissection presenting as SVC syndrome is extremely uncommon. We identified only nine English-language case reports of patients presenting with SVC syndrome resulting from compression by a type A aortic dissection. Of these, four described patients with no chest pain who presented, as in our case, because of symptoms related to SVC obstruction.[1,12–14] The remaining patients reported presented with chest pain or other complaints not related to SVC syndrome. In 1994, Link and Pietrzak reviewed 17 such cases from the English and non-English literature.[1] Interestingly, six of the patients had previously undergone cardiac surgery, as was the case with our patient. Many of the characteristics of these 17 patients contrasted with those of our case; however, in two-thirds of the cases, patients were found to have a murmur, for example.[1]

CONCLUSION

This case highlights how, on occasion, an emergent, life-threatening condition can present in an atypical and seemingly nonemergent fashion. Although a handful of cases have been published previously, it is likely that most emergency physicians consider type A aortic dissection as a possible etiology when evaluating patients with SVC syndrome. Aortic dissection is a condition to consider when evaluating a patient with acute-onset SVC syndrome, particularly if they have a history of cardiac surgery.

Competing interests: None declared.

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