

Malignant superior vena cava syndrome presenting after trauma

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SUMMARY: A 41-year-old man was brought to the ED after a motor vehicle crash. On presentation, he demonstrated symptoms compatible with superior vena cava (SVC) syndrome, including extreme dyspnea, face and neck cyanosis and facial swelling. A chest tube was inserted and drained large amounts of sanguineous fluid. An exploratory thoracotomy revealed an extensive tumour encasing the SVC and the hilum. Biopsy confirmed the diagnosis of T-cell lymphoma.

The most common cause of SVC syndrome is malignant disease, with bronchogenic carcinoma and lymphoma being most frequent. Review of the literature uncovered only a few anecdotal reports of traumatic SVC syndrome. There are no previous reported cases of *malignant* SVC syndrome presenting in association with trauma.

RÉSUMÉ : Un homme âgé de 41 ans est amené à l'urgence après un accident d'automobile. Ses symptômes de présentation sont compatibles avec un syndrome de la veine cave supérieure, notamment une dyspnée extrême, une cyanose du cou et du visage et une enflure faciale. Un tube thoracique est inséré et draine une grande quantité de liquide sanguinolent. Une thoracotomie exploratrice révèle une tumeur importante qui recouvre la veine cave supérieure et le hile. La biopsie confirme le diagnostic de lymphome T.

La cause la plus courante du syndrome de la veine cave supérieure est un cancer, le carcinome bronchogénique et le lymphome étant les formes les plus courantes. La revue de la littérature révèle seulement quelques rapports anecdotiques de syndrome traumatique de la veine cave supérieure. Dans le passé, on n'avait signalé aucun cas de syndrome *malin* de la veine cave supérieure associé à un traumatisme.

Key words: superior vena cava syndrome, trauma, lymphoma, dyspnea

Introduction

Superior vena cava (SVC) syndrome is a clinical entity that results from obstruction of venous return to the heart at the level of the superior vena cava. The symptoms and signs occur because of systemic venous congestion and reduced cardiac output. These include cyanosis of the head and neck, venous dilatation of the chest wall, head, neck and upper extremities, dyspnea, cough, conjunctival edema, proptosis, visual changes and a decreased level of consciousness.¹ The most common cause of SVC syndrome is malignant disease — most often bronchogenic carcinoma or lymphoma.²⁻⁶ Other nontraumatic causes include benign tumours, central venous catheters, thoracic aortic

aneurysms and mediastinal fibrosis secondary to histoplasmosis or radiation.^{3,7} Trauma is an exceedingly rare cause of SVC syndrome. Mediastinal emphysema, mediastinal hematoma, pneumothorax, hemothorax, pseudoaneurysm of the thoracic aorta and SVC thrombosis are trauma-related complications that may cause SVC syndrome.⁷⁻⁹

Case report

A 41-year-old man presented to the emergency department after a motor vehicle crash. He had been sleeping, unrestrained, in the cab of a transport truck when the driver lost control and the vehicle left the highway at 90 kph. He lost consciousness for 10 to 15 minutes but was found awake with

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stable vital signs when paramedics arrived to extricate him.

On arrival in hospital, the Glasgow Coma Scale score was 15 and vital signs were as follows: blood pressure 160/104 mm Hg, pulse 136 beats/min, and respiratory rate 30 breaths/min. The patient was extremely dyspneic, his face appeared swollen, and his head and neck were cyanotic. Auscultation of the chest revealed decreased breath sounds on the right side. There were no penetrating injuries.

Arterial blood gases demonstrated a pH of 7.32, a PCO_2 of 32 mm Hg, a PO_2 of 82 mm Hg, a bicarbonate level of 16 mmol/L and an oxygen saturation of 95% on room air. A portable chest x-ray (Fig. 1) showed a grossly widened mediastinum, measuring 12 cm, an opacity in the right middle and lower lung zones, and enlargement of the cardiac silhouette. A transesophageal echocardiogram revealed a trivial pericardial effusion and no sign of aortic injury. Computed tomography (CT) of the chest showed right pleural thickening and nodularity as well as a dense mass in the mediastinum, which encased the superior vena cava, right pulmonary artery, right main-stem bronchus and bronchus intermedius. The CT scan also showed nodular densities in the right lung, pericardial thickening, and right paratracheal and para-aortic adenopathy. Abdominal CT demonstrated a small splenic laceration, a left adrenal mass and aortocaval adenopathy.

In the ED, a right chest tube was inserted and 1.5 L of sanguineous fluid drained immediately. Because of ongoing drainage, the patient was taken to the operating room for an exploratory thoracotomy. At surgery, an extensive tan-coloured tumour involving the mediastinum was found. It encased the SVC and extended into the hilum. A biopsy was performed and pathological analysis revealed the diagnosis of high-grade, malignant T-cell lymphoma.

Postoperatively, the patient stated that he had been experi-

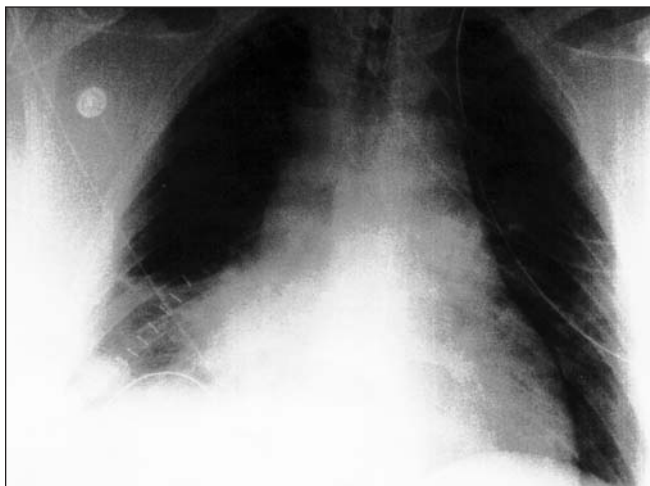


Fig. 1. Portable chest x-ray post-thoracotomy

encing cough and dyspnea prior to the accident. He improved in hospital, was discharged in satisfactory condition and was referred for definitive outpatient management of his lymphoma.

Discussion

This case of SVC syndrome was interesting because the initial presentation suggested it was traumatic in origin. Trauma is, however, a rare cause of SVC syndrome. The differential diagnosis of traumatic SVC syndrome includes mediastinal emphysema, mediastinal hematoma, pneumothorax, hemothorax, pseudoaneurysm of the thoracic aorta and SVC thrombosis.⁷⁻⁹ A MEDLINE search using the terms "trauma" (text word and subject heading) and "superior vena cava syndrome" located only 3 articles. A hand search uncovered 3 more. Three of these were articles excluded because they did not refer to cases of traumatic SVC syndrome, and the remainder are cited here as references.

One study⁸ detailed 17 patients with pseudoaneurysm of the thoracic aorta: 13 cases occurred after aortic or cardiac surgery and 4 after blunt chest trauma. A few of these patients demonstrated frank SVC syndrome; the others experienced symptoms compatible with SVC syndrome but not exclusive to it (e.g., dyspnea). Interestingly, none of these presentations were acute, and the minimum time from "injury" to presentation was 3 months.

Anecdotal reports of traumatic SVC syndrome date back to 1883.¹⁰ In 1936, Ochsner and Dixon reported an unusual case of SVC syndrome that developed gradually in a 24-year-old woman after forceful anterior chest trauma while opening a soft drink bottle.¹⁰ In 1979, Matthews and coworkers¹⁰ reported the case of a 44-year-old man who had face and neck swelling, plethora, headache and dilated chest wall veins 12 hours after falling forcefully on his chest during a softball game. A superior vena cavagram in this case revealed complete occlusion of the SVC, yet chest x-ray bronchoscopy, mediastinal lymph-node biopsy, and skin tests for tuberculosis, histoplasmosis and coccidioidomycosis were negative. Although no treatment was provided, the patient's symptoms gradually subsided, presumably due to the development of collateral circulation.¹⁰

Another report involved a young labourer who fell 2 m from a box car, landing on his back on a hard surface. Progressive SVC syndrome developed and, upon his arrival at the hospital, an upper mediastinal mass was discovered on chest radiography. At thoracotomy, the mass was found to be an organized thrombus in the SVC. As in the previous case, the patient's symptoms gradually resolved as collateral circulation developed.⁹

Existing evidence²⁻⁵ suggests that bronchogenic carcinoma, lymphoma and breast carcinoma are, respectively, the most common causes of SVC syndrome. Benign causes are rare, but mediastinal fibrosis, SVC thrombosis and retrosternal goitre have been reported.

As our case report shows, SVC syndrome presenting after trauma may be due an incidental acute presentation of a chronic condition. The patient discussed in this report suffered SVC syndrome after blunt chest trauma. The nature of the presentation (trauma), not the underlying etiology (non-Hodgkin's lymphoma) was unique.

When SVC syndrome occurs in the setting of trauma, emergency physicians should look for known causes such as SVC thrombosis, pneumothorax, hemothorax, mediastinal hematoma, mediastinal emphysema and pseudoaneurysm of the thoracic aorta. All patients should have chest radiography, and other investigations, including CT, Doppler imaging, aortography and endoscopic studies, should be ordered depending on the clinical suspicion of the physician and the stability of the patient.

In the absence of trauma, the most likely cause is malignant disease, and a tissue diagnosis should be obtained before radiotherapy or chemotherapy is begun. A discussion of the specific methods of tissue acquisition is beyond the scope of this article.

Patients with SVC syndrome rarely die as a result of the syndrome, and many cases of the syndrome will resolve spontaneously as collateral circulation develops. The long-term prognosis depends on the underlying cause.^{2,4}

Conclusions

SVC syndrome is a complex clinical entity that is rarely seen as an emergency. Emergency physicians can, however, play a key role in diagnosis and have direct impact on the patient's outcome.

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