The patient is a 56-year-old woman admitted to the Intensive Care Unit for an acute traumatic subdural hematoma. Her medical history was significant for progressive, severe hypoxia reported to be secondary to chronic obstructive pulmonary disease, despite absence of supporting pathology on chest radiograph (Figure 1). An electrocardiogram on admission revealed sinus rhythm with left atrial enlargement and a prolonged QT (Figure 2). She underwent emergent craniectomy and evacuation, thereafter experiencing a prolonged Intensive Care Unit course with difficulty weaning off mechanical ventilation. During the hospitalization, intravenous access was obtained with a subclavian venous line (Figure 1). Intravenous agitated saline was injected into this subclavian line under echocardiographic guidance to confirm correct line placement. Within seconds, saline microbubbles were visualized in the left atrium (LA), without transit into the right atrium (Figure 3 and Movie 1 in the online-only Data Supplement), consistent with venous shunt into the LA. Echocardiography demonstrated mild left atrial dilation and otherwise normal cardiac structure and function (Figure 3). Three-dimensional reconstruction of contrast computed tomography of the chest obtained on admission revealed an isolated right superior vena cava draining into the LA (Figure 4A and 4B). Right superior vena cava drainage into the LA represents an exceedingly rare congenital anomaly of the systemic thoracic veins, with only 20 cases reported, predominantly in children. Our case demonstrates the importance of considering abnormal shunt physiology in a patient with progressive hypoxia.

The most frequently seen anomaly of the thoracic veins is a persistent left superior vena cava draining into the right atrium via a dilated coronary sinus. This anomaly is often detected as an incidental echocardiographic finding, usually without significant clinical consequence. However, an isolated right superior vena cava draining into the LA is characterized by right-to-left shunt physiology, which may lead to progressive dyspnea, hypoxia, and cyanosis. Furthermore, as a direct conduit between the venous and systemic circulations, there is the attendant risk of paradoxical embolization; several case reports describe such patients with shunt complicated by cerebral abscesses. An intravenous line placed inadvertently along the route of this anomalous venous system would serve as a direct nidus for paradoxical emboli. Fortunately, our patient did not experience any embolic or infectious complications from her anomaly, and the left subclavian line was carefully removed without adverse event.

There is no consensus as to the embryological mechanism of this anomaly, but echocardiographic and postmortem anatomic findings suggest a deficiency in the wall that is shared by the right superior vena cava and the right upper pulmonary veins. Contrast echocardiography, cardiac magnetic resonance imaging, and contrast-enhanced computed tomography have been used as noninvasive modalities for the diagnosis. Regardless of the initial clinical presentation, surgical correction of the systemic venous connection to the LA is indicated to prevent complications of paradoxical embolization.

Disclosures

None.

References

Figure 1. Chest radiogram showing no evidence of chronic obstructive pulmonary disease. There is correct placement of left subclavian line (arrowhead).

Figure 2. Resting electrocardiogram shows normal sinus rhythm, left atrial enlargement, and prolonged QT, the latter likely a manifestation of the patient’s neurological injury.

Figure 3. Transthoracic echocardiogram after injection of agitated saline solution into the right subclavian venous line revealed opacification of the left atrium without microbubbles in the right atrium or ventricle. There is mild left ventricular hypertrophy (1.2 cm ventricular septal thickness) and normal systolic function (71% ejection fraction), with mild lateral atrium dilation (27.9 cm² area). Right ventricular size and systolic function were also normal (57% right ventricle ejection fraction, 12.4 cm/s lateral tissue Doppler systolic velocity, 2.3 cm tricuspid annular plane systolic excursion).

Figure 4. A, Two-dimensional coronal view demonstrating the anomalous connection of the right superior vena cava (RSVC) to the left atrium (LA). B, Contrast chest computed tomography with three-dimensional reconstruction showing the anomalous connection.
Isolated Right Superior Vena Cava Into the Left Atrium Detected by Contrast Echocardiography
David Cohen, Jonathan Reiner, Jalil Ahari and Jannet Lewis

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Movie Legend

**Movie 1.** Transthoracic echocardiogram following injection of agitated saline solution into the right subclavian venous line revealed opacification of the left atrium (LA) without microbubbles in the right atrium or ventricle. Movie file best viewed with Quicktime application.