Where Is the Bubble? A Case of Systemic–to–Pulmonary Venous Shunt in Superior Vena Cava Occlusion

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INTRODUCTION

Superior vena cava (SVC) or inferior vena cava occlusion is typically an acquired condition. Thrombosis of the SVC is a well-known complication of central venous catheter placement.[1-3]

Obstruction of the SVC causes elevated pressure in the veins draining into the SVC and increased or reversed blood flow through collateral vessels.[4] Systemic–to–pulmonary venous collateral pathway is an uncommon collateral pathway, with multiple patterns described previously. These patterns include the azygos-hemiazygos pathway, the internal and external mammary pathway, the lateral thoracic pathway, and the vertebral pathway. Regardless of the route or etiology of the pathway, the resulting right-to-left shunt leaves the patient susceptible to stroke, brain abscess, and high–cardiac output states.[5] We report a case of systemic–to–pulmonary venous shunting with inconsistent echocardiographic findings depending on the route of agitated saline administration.

CASE PRESENTATION

A 30-year-old man with a medical history of end-stage renal disease on hemodialysis through a right femoral tunneled dialysis catheter, inferior vena cava occlusion status post grating and stenting, and calciphylaxis of the left hip and right shoulder was admitted for sepsis secondary to left hip abscess, for which he underwent incision and drainage. The hospital course was further complicated by left-sided punctate stroke seen on magnetic resonance imaging of the head, for which transthoracic echocardiography with agitated normal saline was performed to exclude a cardioembolic source and an interatrial communication in relation to his symptoms of transient ischemic attack. The injection was performed via the right arm cubital fossa vein and demonstrated bubbles entering the left atrium, with later appearance of a few bubbles in the right atrium (Figure 1A and 1B, Video 1). Transesophageal echocardiography with bubble study was then performed with administration of the agitated saline via both the right and left arms. When the bubble study was performed from the right arm, bubbles first appeared in the left atrium, followed by the left ventricle (Figure 1C, Video 2). No bubbles were noted from the left upper pulmonary vein (Video 3). Bubble study via the left arm then showed initial appearance of bubbles in the right atrium, followed by the right ventricle (Figure 1D, Video 4). This inconsistency in echocardiographic findings raised suspicion of anomalous venous drainage from the right arm. Chest computed tomography with contrast (Figure 2) showed occlusion of the right brachiocephalic vein and narrowing of the distal SVC over a length of 3.5 cm, with significant reflux of contrast into the azygos vein. It also showed multiple small venous collateral vessels from the azygous vein to the pulmonary veins, explaining the aforementioned findings (Figure 3).

DISCUSSION

This patient did not have the typical symptoms present in SVC syndrome, even given the extensive central venous involvement. Central venous occlusion should be suspected in the setting of extracardiac shunting seen on contrast echocardiography.

The mechanism of this shunt in the setting of central venous system occlusion was systemic–to–pulmonary venous collateral vessels. Four general collateral pathway have been recognized in the setting of superior vena occlusion: the internal mammary pathway, azygous pathway, lateral thoracic pathway, and vertebral pathway.[6]

Venous blood return to the right atrium by any of these routes is possible, although the azygos-hemiazygos pathway is reported to be the most common.[5] It seems in this case that the development of venous anastomosis between the systemic and pulmonary system provided the mechanism whereby blood could flow from the distended central system. This could be explained by obstruction of both the inferior vena cava and the SVC, as the four major collateral systems depend on a patent inferior vena cava. This limits the systemic venous return to the right side, and instead unusual collateral vessels via the systemic-pulmonary venous anastomosis provide a return route to the left atrium. The pathway consists of mediastinal connections between the innominate veins and the superior pulmonary veins via the bronchial venous plexuses around the airways, hilar vessels, and pleura.[5] To understand this pathway, it is necessary to describe the normal bronchial venous and azygous system. The bronchial venous system includes the superficial and deep bronchial veins. The deep bronchial veins receive blood from the intrapulmonary bronchiolar plexuses and therefore drain the larger intrapulmonary bronchi. They form a common trunk that drains into the main pulmonary vein or sometimes directly into the left atrium. The superficial bronchial veins terminate by draining into the azygos vein on the right and the accessory hemiazygos or left superior intercostal vein on the left. In the setting of central venous obstruction, such as in this case, and subsequent increase in venous pressure in the azygous system, collateral vessels between the superficial (central) and deep (systemic) are formed. The presence of such collateral vessels results in a right-to-left shunt, subsequently leaving the patient susceptible to stroke, brain abscess, and a high–cardiac output state.[5]

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VIDEO HIGHLIGHTS

Video 1: TTE showing bubbles from the right arm going into the left atrium.
Video 2: TEE showing bubbles from the right arm going into the left atrium.
Video 3: TEE showing the left upper pulmonary vein.
Video 4: TEE showing bubbles from the left arm going into the right atrium.

View the video content online at www.cvcasejournal.com.

The chronic states of these collateral vessels could explain the asymptomatic SVC of this patient.

However, this case leaves open the question of whether the laterality of contrast administration on echocardiography matters when describing extra- or intracardiac shunts. As mentioned previously, our case demonstrated contrast appearing in the left atrium first when using the right upper extremity for agitated saline injection. Interestingly, bubbles appeared in the right atrium when using the left upper extremity as the injection site. Although theoretical at this time, this inconsistency can be explained by collateral vessels between the distal right brachiocephalic vein and the azygos vein, possibly via the internal thoracic, as it resides on the right side of the chest cavity. Further collateral vessels between the azygous and bronchial venous system can provide the final pathway into the left system. The numerous bronchial venous complex (superficial and deep) plays a major role in this connection and providing the shunt. Although less likely, anomalous vein from the right innominate vein (brachiocephalic) to the left atrium was suspected as well but cannot be seen on existing imaging studies. Further extensive imaging, such as venography, would be required to characterize such findings. This was not performed in our patient, as he was asymptomatic, and further interventions would not have been appropriated per the vascular team.

CONCLUSION

The systemic–to–pulmonary venous collateral pathway is an uncommon collateral pathway, with multiple patterns described previously. These patterns include the azygos-hemiazygos pathway, internal and external mammary pathway, lateral thoracic pathway, and vertebral

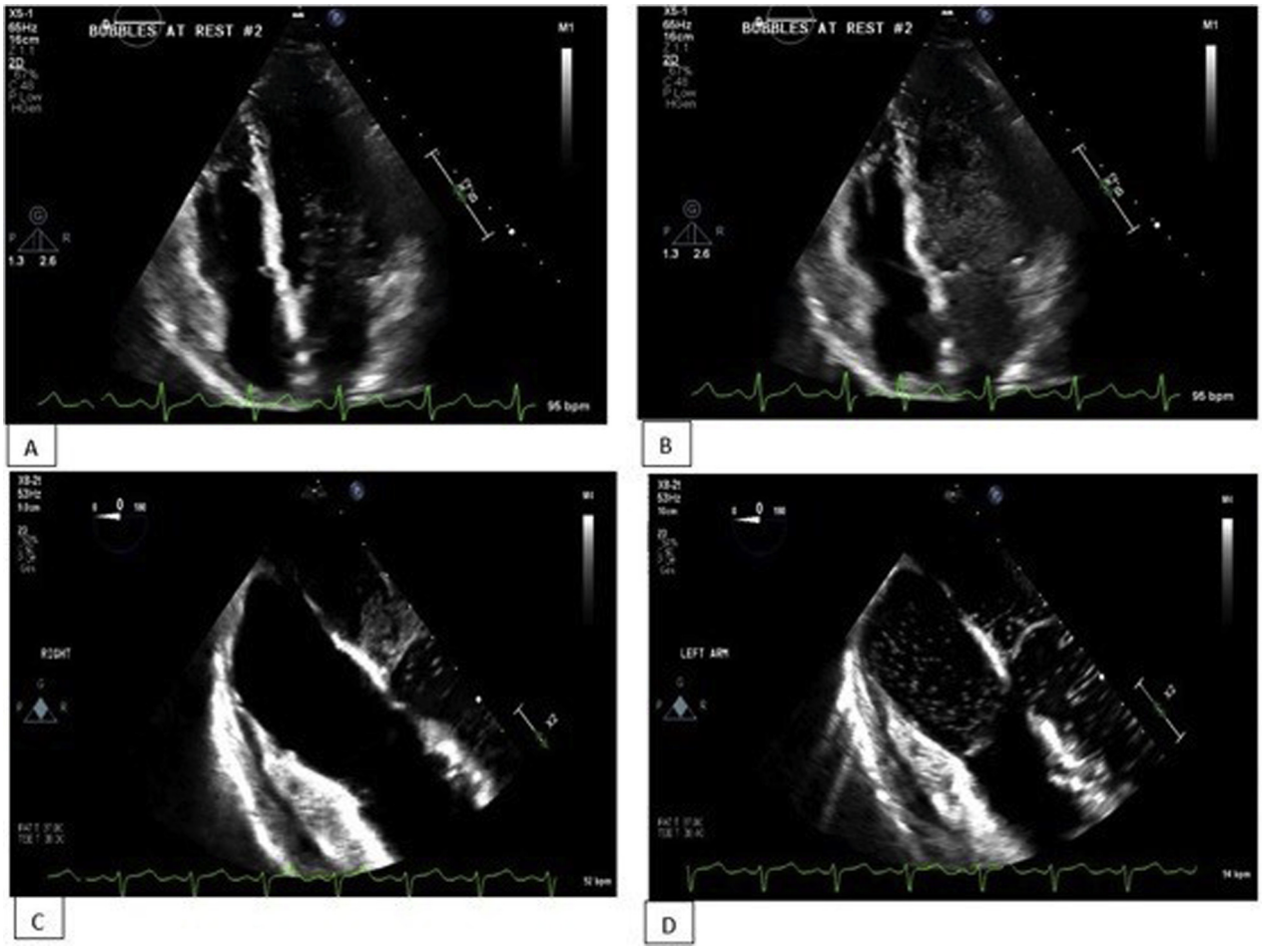

Figure 1 Transthoracic echocardiography, four-chamber view, showing bubbles appearing in the left atrium and then the left ventricle, with none on the right side of the heart, when agitated saline was injected from the right arm **(A, B)**. Transesophageal echocardiography similarly showed bubbles in the left atrium when contrast was injected from the right arm **(C)**. The same view demonstrates bubbles in the right atrium when contrast was injected in the left arm **(D)**.

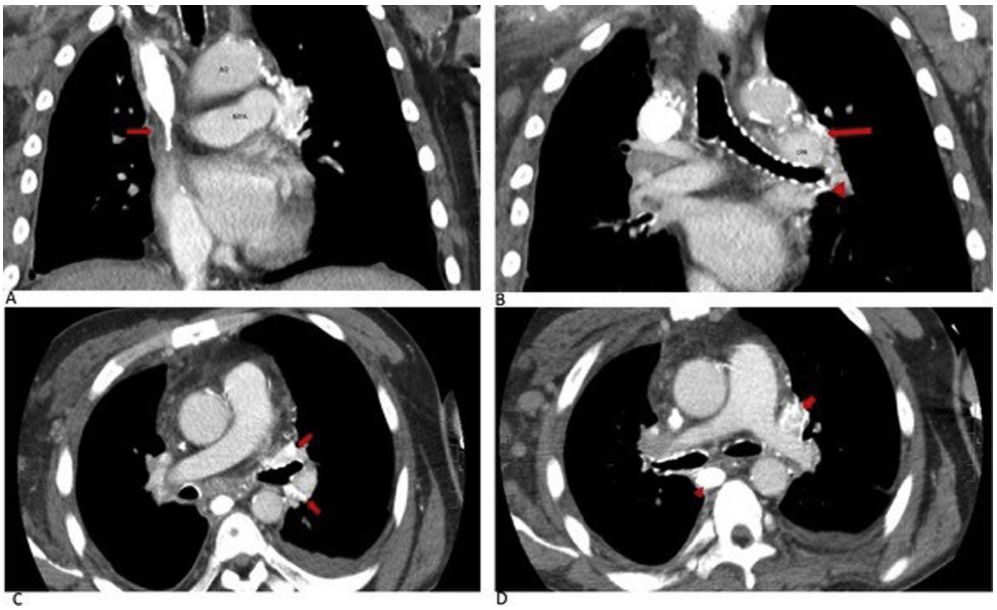

Figure 2 Systemic-to-pulmonary collateral vessels in a 30-year-old man with a history of end-stage renal disease and multiple dialysis catheter placement. He underwent chest computed tomography with contrast to evaluate the venous system given inconsistent findings on contrast echocardiography. **(A)** Coronal image showing narrowing of the distal SVC. **(B)** Coronal computed tomographic image shows systemic-to-pulmonary collateral vessels (*arrow*) surrounding the left pulmonary artery, with contrast visible in the left superior pulmonary vein (*arrowhead*). **(C)** Axial computed tomographic image showing collateral vessels around the left main stem bronchus (*arrow*). **(D)** This is also seen around the left pulmonary artery (*arrow*), along with dilation of the azygos vein (*arrowhead*). Dilation of azygous system along with nexus of collateral blood vessels throughout the bronchopulmonary trunk suggests involvement of the bronchial venous system in this right-to-left shunt.

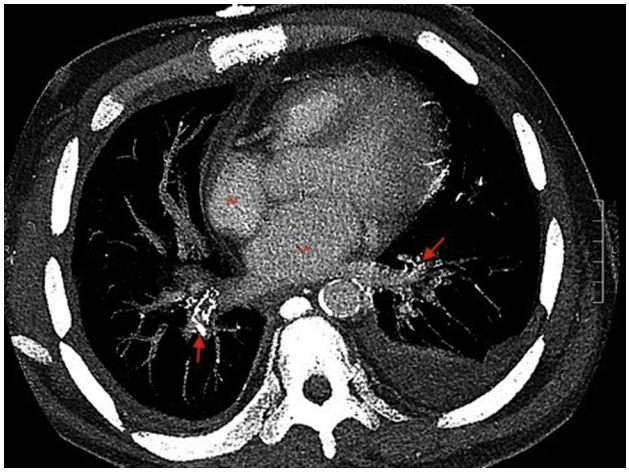

Figure 3 Computed tomography with contrast of pulmonary vein, left atrium, and right atrium in one view. Collateral vessels are seen in the pulmonary vein (*arrows*) that are draining into the left atrium. Collateral vessels seen by contrast enhancement in this image are better appreciated on the right pulmonary vein. Given proximity to the bronchus, this suggests bronchial venous complex involvement in collateral vessels and the final pathway into the left system. Normally, superficial bronchial veins drain to the azygous system, whereas deep bronchial veins drain into the pulmonary vein and sometime the left atrium. Anastomosis between these two venous systems in the setting of raised central venous pressure can explain the present findings.

pathway. In this case, collateral vessels were initially suggested after findings on bubble echocardiography that prompted further imaging, highlighting the importance of agitated saline studies as a valuable screening tool. It is important to recognize such unusual collateral vessels because the acquired right-to-left shunt, regardless of route, leaves patients susceptible to stroke, brain abscess, and a high–cardiac output state.

SUPPLEMENTARY DATA

Supplementary data related to this article can be found at https://doi.org/10.1016/j.case.2020.08.003.

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