Systemic-to-Pulmonary Venous Shunt in a Hemodialysis Patient With Extensive Thrombosis of Superior and Inferior Vena Cava

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INTRODUCTION

Hemodialysis is the most prevalent modality of renal replacement therapy in the majority of countries. Vascular access is of extreme importance for patients on dialysis and guidelines recommend specific vascular access care to guarantee the longevity of therapy, reducing complications, and providing the prescribed dose. The focus of the latest guideline was the patient life-plan before the corresponding access needs.1

In the last decades, major advances in both medical knowledge and the technology of maintenance hemodialysis have been made. It is not so rare for long-term dialysis (>10 years) patients. Therefore, complications such as systemic thrombosis are prone to happen. Deep venous thrombosis causes blood flow deviation through collateral vein formation and also shunts. Nephrologists should be aware of this phenomenon and learn to recognize it, because proper management may prevent a more serious complication and provide the best possible quality of treatment.

Here we present a case of a systemic-to-pulmonary venous shunt in a patient on hemodialysis with hypoxemia as a first clue to the diagnosis.

CASE PRESENTATION

A 43-year-old woman was admitted to the emergency department with fever, loss of appetite, and weakness for 3 days. She had been receiving hemodialysis for the last 23 years due to congenital reflux nephropathy. Ascites secondary to Budd–Chiari syndrome by thrombosis of hepatic veins had developed in the past 6 months. She had undergone parathyroidectomy due to secondary hyperparathyroidism and thyroidectomy due to papillary carcinoma 10 years prior. Her vascular access was a right internal jugular long-term tunneled catheter, chosen after an arteriovenous fistula thrombosis that had also occurred 10 years prior. Although asymptomatic on the hemodialysis sessions, the patient developed very low blood pressure in the past year, with a mean arterial pressure of 50 mm Hg.

The patient was transferred to the intensive care unit due to low oxygen saturation (84%) and low blood pressure (78/40 mm Hg). COVID-19 was excluded after 2 negative reverse transcription–polymerase chain reactions for SARS-CoV2 samples. Blood culture positive for Staphylococcus epidermidis implied that a catheter-related bloodstream infection was causing all of the symptoms. Vancomycin was started empirically and then switched to ciprofloxacin, which successfully treated the infection.

However, the patient maintained a low arterial oxygen tension of 43.9 mm Hg. Considering her previous thrombotic event and the risk of a new thromboembolic event, chest computed tomography (CT) was performed to exclude pulmonary embolism. The contrast medium was administered through a left antecubital intravenous access, and the chest CT showed thrombosis of the superior vena cava (SVC) as well as a narrowing of the supra-hepatic segment of the inferior vena cava. Multiple collateral vessels within
the mediastinum and bridging veins across the pleura apparently draining into the superior left pulmonary vein were observed, along with an early appearance of contrast in the left atrium. There was also evidence of Budd−Chiari syndrome, caused by thrombosis of the right and middle hepatic veins, and findings commonly associated with renal osteodystrophy. No signs of pulmonary embolism were found. Contrast trans-thoracic echocardiography was performed, and microbubbles appeared in the left atrium within 1 cardiac cycle, indicating a right-to-left shunt of possible cardiac origin. 99mTc-Macroaggregated albumin scanning (MAS) confirmed the presence of a right-to-left shunt, showing an important systemic shunt with extrapulmonary activity in brain and abdomen projections.

Transesophageal echocardiography was performed, and no structural abnormalities were found. In this echocardiography, microbubbles were administered through the long-term tunneled catheter, and no contrast was seen in the left cardiac chambers, implying that the cause of the left-to-right shunt was not a cardiac defect but rather a shunting pathway in the venous return superior to the right atrium. A systemic-to-pulmonary venous shunt (SPVS) generated by collateral veins secondary to inferior and SVC thrombosis was considered to be the etiology of the patient’s hypoxemia. The patient underwent endovascular treatment. Phlebography performed through the right jugular vein before the procedure showed an important filling defect in the topography of the superior vena cava to the level of the junction with the right atrium (Figure 1). When contrast was injected through the left jugular vein, phlebography showed multiple collateral vessels in the mediastinum with drainage in the left upper pulmonary field, with no contrast in the right atrium (Figure 2). Dilatation of the suprahepatic segment of the IVC, dilatation of SVC (Supplementary Figure S2A–D), and embolization of the collateral vessels were performed (Figure 3). A new MAS was performed 1 week after the procedure and showed complete resolution of the left-to-right shunt (Supplementary Figure S1A and B). The patient was completely independent of oxygen supplementation when discharged, with an arterial oxygen tension of 72.7 mm Hg, progressive reduction of ascites, and blood pressure returned to a normal range.

### DISCUSSION

In Brazil, 24.6% of patients on maintenance hemodialysis have a central venous catheter (CVC) as vascular access. Catheter-related vascular thrombosis is a common complication of hemodialysis catheters. The endothelial wall injury, the disturbance of blood flow, and the catheter surface are predisposing factors to thrombus formation. Catheter-related vascular thrombosis, although often asymptomatic, can cause vascular and catheter occlusion, contributing to increased morbidity and mortality. Loss of vascular access occurs in up to 30% to 40% of patients. Other complications related to catheter-related vascular thrombosis include infection, pulmonary embolism, formation of right heart thromboembolism, and SVC syndrome, in the case of a CVC placed on superior veins.

In this case presentation, we describe an SPVS secondary to an SVC thrombosis as a complication of a CVC placed on the right internal jugular vein. There are only a few cases reports in the literature about SPVS as

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**Figure 1.** Phlebography performed by the right internal jugular vein characterizes an important filling defect in the topography of the superior vena cava (SVC) to the level of the junction with the right atrium (white arrow). There is collateral circulation through the azygos system, with the accumulation of intravenous contrast in the azygos arch (arrowhead).

**Figure 2.** Phlebography performed through the left internal jugular vein shows multiple collateral vessels in the mediastinum with drainage in the left upper pulmonary field (arrowhead). Adjacent to the inferior vena cava (IVC) is the azygos vein (white arrow).
a complication of SVC obstruction, and the majority of patients had malignancy-associated SVC obstruction.

Our patient had experienced an episode of arteriovenous fistula thrombosis 10 years prior. Since then, she had had CVC vascular access via the right jugular vein. Chest CT performed 12 years before the admission showed SVC stenosis, most likely secondary to multiple insertions of temporary catheters. The presence of previous venous stenosis and the long-term tunneled catheter probably led to venous thrombosis, resulting in complete obstruction of the SVC. A thrombophilia screen was performed, and no additional thrombotic factors were found.

The SPVS in this context can occur through 3 different mechanisms. In the anatomic type, the high pressure against the venous return caused by an SVC obstruction leads to insufficiency of the venous valves located at the junction of the pleurohilar bronchial veins and hemiazygos veins, leading to backflow into the pleurohilar bronchial veins. In the congenital type, there are 3 possible shunting pathways: an aberrant pulmonary venous return with the reversed flow, a levocardiac embryologic remnant, and a persistent left SVC. Finally, the acquired shunt is of inflammatory origin and results in newly formed vessels bridging the subpleural pulmonary veins and the intercostal veins through pleural adhesions.

This patient had no unique veins resembling a congenital shunting pathway, but multiple collateral vessels within the mediastinum and bridging veins across the pleura draining into the superior left pulmonary vein, in a pattern consistent with an acquired shunt secondary to SVC obstruction. The systemic-to-pulmonary venous collateral pathway is an uncommon result of SVC obstruction and increases the risk of a stroke, brain abscess, and high cardiac output state. Although our patient did not develop any neurological complications, she had severe hypoxemia. The transthoracic echocardiogram showed no contrast in the right cardiac chambers after injection of microbubbles in the left antebrachial vein, whereas the left chambers were almost instantly filled with contrast in the first cardiac cycle, implying that all superior venous return was occurring through the shunting pathway.

Frequently, the management of SPVS can be conservative, mainly if the shunt does not cause relevant hypoxemia or neurological complications. However, our patient presented with serious hypoxemia, with a peripheral saturation of oxygen of 80% to 84% without

Figure 3. Embolization of the mediastinal collateral vessels. (a) Catheterization of the anomalous vessel in the topography of the left upper pulmonary field with a macrocatheter (5 Fr), followed by contrast injection. (b) Microcatheterization (2.4 Fr) of anomalous vessels demonstrating apparent drainage to the upper left pulmonary vein. After catheterization of these vessels, embolization was performed with the release of multiple Interlock fibered detachable coils (Boston Scientific, Natick, MA), in addition to the injection of n-butyl-2-cyanoacrylate (Histoacryl; B. Braun, Melsungen, Germany). (c and d) Control phlebography shows adequate contrast flow to the right atrium and closure of anomalous collateral vessels.
without intervention, the patient would need home oxygen therapy, something that would impair her quality of life. Furthermore, the untreated hypoxemia could lead to pulmonary hypertension in the long term, aggravating dyspnea and hypoxemia. Nevertheless, serious complications could occur during the cava angioplasty procedure, including embolism, stent migration, and bleeding. After a multidisciplinary discussion and with the patient’s approval, it was decided that the benefits outweighed the risks, and the patient underwent surgery.

The patient was discharged 4 days after the procedure, without oxygen supplementation. In the following months, there was a significant reduction of ascites and an increase in blood pressure due to lower hepatic vein venous pressure and greater cardiac output, secondary to a greater preload. These changes enhanced the patient’s quality of life, which was the primary goal of the procedure; therefore, it was a valid method of treatment.

**CONCLUSION**

We have presented a rare case of a patient who developed a SPVS secondary to SVC thrombosis due to multiple CVC in the right jugular vein and was successfully treated with interventional radiology. More studies are needed to establish a treatment protocol for this condition. Lessons that we have learned from this case are summarized in Table 1.

**SUPPLEMENTARY MATERIAL**

Supplementary File (PDF)

*Figure S1.* (A) MAS performed before the procedure. (B) MAS performed after the procedure.

*Figure S2.* (A–D) Images of dilations performed during the procedure and post-dilation identification and assessment with intravascular ultrasound.

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**Table 1. Teaching points**

| Teaching point | Details |
|----------------|---------|
| 1              | The central venous catheter can lead to severe complications and should not be the first option as vascular access for hemodialysis. |
| 2              | The systemic-to-pulmonary venous shunt is a possible complication of superior vena cava obstruction, by malignancy-associated causes or benign causes, such as vascular access, infection, retrosternal thyroid, sarcoidosis, radiation fibrosis, aortic aneurysm, and benign tumors. |
| 3              | Contrast transesophageal echocardiography and macroaggregated albumin scanning are useful means to confirm the presence of a left-to-right shunt. Chest computed tomography can be useful to detect which kind of shunting pathway is responsible for the left-to-right shunt. |
| 4              | Cava angioplasty and embolization of the collateral vessels can be a therapeutic alternative in systemic-to-pulmonary venous shunts in patients with superior vena cava obstruction. |

**DISCLOSURE**

All the authors declared no competing interests.

**PATIENT CONSENT**

The authors declare that they have obtained consent from the patient discussed in the report.