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Refractory Hypoxemia in a 23-Year-Old Patient With Budd-Chiari Syndrome

Jeroen J. H. Bunge, MD; Ubbo S. Wiersema, MD; Adriaan Moelker, MD, PhD; Jasper van Bommel, MD, PhD; and Eric T. T. L. Tjwa, MD, PhD

Antiphospholipid syndrome is an autoimmune disorder characterized by a hypercoagulable state, leading to arterial and venous thrombosis. We present a 23-year-old patient, suspected of having Budd-Chiari syndrome due to antiphospholipid syndrome, who developed severe and progressive hypoxemia, requiring prolonged mechanical ventilation. After a detailed but unsuccessful workup, a contrast CT scan revealed an occluded superior vena cava and azygos vein-superior vena cava junction and massive right-to-left shunting through a network of systemic to pulmonary venous collaterals. Restoring normal blood flow from the azygos vein into the right atrium by stenting the azygos-superior vena cava junction resolved the hypoxemia immediately. Within the same procedure, the hepatic outflow obstruction was successfully treated by stenting a severe stenosis of the suprahepatic inferior vena cava caused by calcified thrombus.

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ABBREVIATIONS: BCS = Budd-Chiari syndrome; IVC = inferior vena cava; SVC = superior vena cava

Hypercoagulation may be presented by a variety of symptoms, depending on the vasculature affected. In this article, we describe a patient with dyspnea, suspected of having vascular liver disease.

Case Report

A 23-year-old woman was referred to our hospital for analysis and treatment of a suspected Budd-Chiari syndrome (BCS), with symptoms of ascites and peripheral edema. Furthermore, she complained of progressive dyspnea. Earlier CT scanning had shown a suprahepatic calcified thrombus in the inferior vena cava (IVC) as a possible cause of the hepatic outflow obstruction or BCS. Her medical history reported antiphospholipid syndrome (positive lupus anticoagulans, positive anti-β_2 glycoprotein antibodies, and positive cardiolipid IgG, and no known predisposing disorders) complicated by pulmonary emboli, and thrombosis of the superior vena cava (SVC) and brachiocephalic vein 1 year previously, for which she was still using anticoagulants, with international normalized ratio in the target range (2,5-3,5).

On physical examination, she had a respiratory rate of 20 breaths/min and peripheral capillary oxygen saturation of 85% on ambient air. Examination of the head revealed marked venous distension.

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AFFILIATIONS: From the Department of Intensive Care (Drs Bunge and van Bommel), the Department of Gastroenterology and Hepatology (Drs Wiersema and Tjwa), and the Department of Radiology (Dr Moelker), Erasmus Medical Centre University Hospital, Rotterdam, The Netherlands.

Drs Bunge and Wiersema contributed equally to this work.

CORRESPONDENCE TO: Eric T. T. L. Tjwa, MD, PhD, Department of Gastroenterology and Hepatology, Erasmus Medical Centre University Hospital, Room Hs-312, PO Box 2040, 3000 CA Rotterdam, The Netherlands; e-mail: etjwa@erasmusmc.nl

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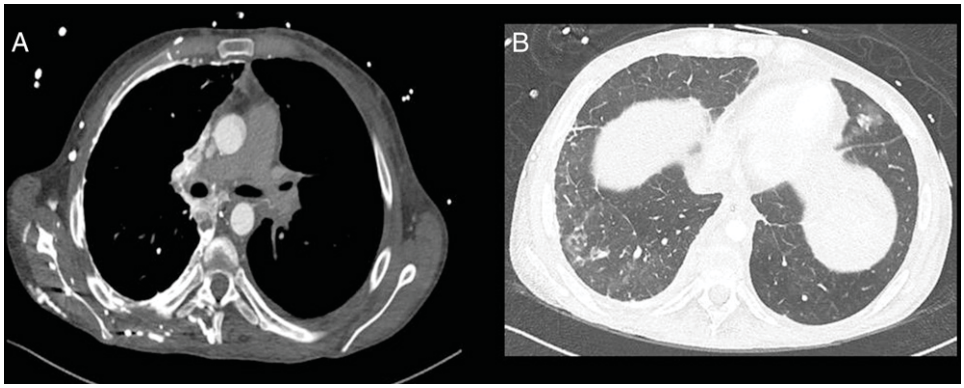

Figure 1 – *CT scan with iodine contrast administered through the right antecubital vein. A, Extensive chest wall collaterals are visible. There is almost no contrast in the right atrium and the pulmonary arteries, but dense contrast enhancement in the right pulmonary veins, left atrium, and left ventricle. B, Lung window.*

There were no cardiac murmurs, and breathing sounds were normal. Platypnea was absent. Ascites and pedal edema were present. One day after admission, her condition deteriorated. She was intubated because of progressive hypoxic respiratory failure. With a positive end expiratory pressure of 10 cm H_2O and an F_{IO_2} of 0.7, the arterial P_{O_2} was only 60 mm Hg. Recruitment maneuvers did not improve oxygenation. A CT scan, with iodine contrast injected through the femoral vein, showed marginal pleural effusion, atelectasis, and no signs of pulmonary embolism. The liver parenchyma was homogenous, with patent flow in the portal veins. There were ascites without splenomegaly. Stenotic circular calcification in the IVC, just below the right atrium, was noted.

As a next step, in the setting of BCS, portopulmonary hypertension and hepatopulmonary syndrome were excluded as causes of the hypoxemia. Venous catheterization by right femoral access revealed a pressure gradient of 16 mm Hg (25-9 mm Hg) over the IVC stenosis. Pulmonary pressures were not elevated. Contrast echocardiography (using agitated saline injected through the femoral artery) was unremarkable and showed no signs of a right-left shunt. ^{99m}Technetium macroaggregated albumin perfusion scanning showed 22% macroaggregated albumin capture in the cerebrum/kidney, possibly indicating pulmonary vascular shunting.

Upon further respiratory deterioration, a CT scan was repeated 3 weeks later to rule out new pulmonary embolism. By chance, the iodine contrast was now administered through the antecubital vein in the right arm. This scan showed extensive thrombosis in the SVC, resulting in total occlusion above, and subtotal occlusion below, the level of the azygos ostium. The azygos vein was patent and dilated. There were extensive chest wall and neck collaterals entering the azygos and hemiazygos veins and the right pulmonary veins. There was almost no contrast in the right atrium and the pulmonary arteries, but dense contrast enhancement in the right pulmonary veins, left atrium, and left ventricle, consistent with direct shunting between systemic veins and

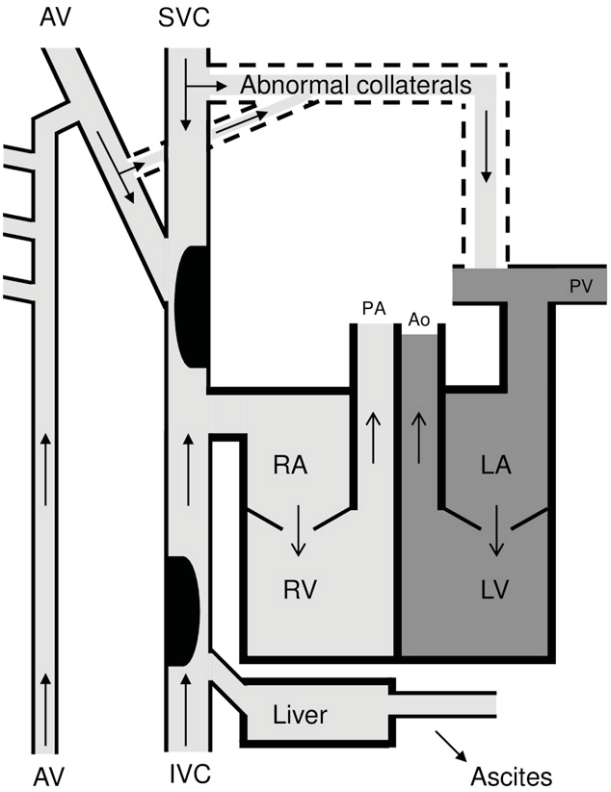

Figure 2 – *Abnormal collaterals as a result of bicaval obstruction leading to systemic hypoxemia. Ao = aorta; AV = azygos vein; IVC = inferior vena cava; LA = left atrium; LV = left ventricle; PA = pulmonary artery; PV = pulmonary vein; RA = right atrium; RV = right ventricle; SVC = superior vena cava.*