

Unusual Right-to-Left Shunt by Single-Sided Bilateral Inferior Vena Cava

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ABSTRACT

Anomalous drainage of an inferior vena cava to the left atrium is a rare congenital cardiac anomaly that might lead to significant clinical implications. This report describes a 21-year-old male with an anomalous connection between one part of a single sided double inferior vena cava and the left atrium documented with cardiac imaging, including angiography. This rare congenital disorder should be considered in the differential diagnosis in patients with cyanosis and/or a history of paradoxical embolism. Interventional catheterization was the chosen method of intervention.

Key Words. Double Inferior Vena Cava; Amplatzer Device; Congenital Anomalies

Introduction

Developmental anomalies of the inferior vena cava are not infrequent. Congenital anomalies of the inferior vena cava (IVC) and its tributaries have become more commonly recognized in asymptomatic patients. The embryogenesis of the IVC is a complex process involving the formation of several anastomoses between three paired embryonic veins.

Anomalous drainage of the inferior vena cava to the left atrium is a rare congenital cardiac anomaly that leads to a right-to-left shunt with or without any other cardiovascular abnormalities. However, anomalies of the systemic venous return might have significant clinical implications. Only a few patients with double IVC have been reported in the literature¹ but none with a connection with an abnormal pulmonary venous drainage and the left atrium.

We report on a 21-year-old male with an anomalous connection between the double inferior vena cava, right pulmonary vein, and the left atrium documented by cardiac imaging, including angiography.

Case Report

A 21-year-old male was referred to our unit for further investigation. He had a history of severe migraine episodes, earlier diagnosed Menière syndrome, and 6 months before, a cerebellar infarction was documented and considered to be related

to a severe migraine episode. At that moment no cardiac investigation was performed. However, because of a recent syncope with prolonged loss of consciousness, the referring cardiologist who was asked to exclude a paradoxical embolism, performed a transesophageal echocardiogram and suggested an abnormal pulmonary venous return of the right lung. Although no new cerebral infarction could be documented by magnetic resonance, the event was considered by the neurologist as a transient ischemic attack. The patient recovered completely.

Physical examination revealed no abnormal clinical findings. There were no cardiac murmurs, nor signs of left or right heart failure. Transcutaneous saturation at rest was 94% and the laboratory tests were normal, including the hematocrit. A chest x-ray showed a normal cardiac silhouette and pulmonary vasculature. The electrocardiogram was unrevealing.

Computer tomography of the chest documented a posteriorly and anteriorly located IVC, an abnormal pulmonary venous return of the right lower lobe into the posteriorly located IVC, and an azygos continuation connecting to a right-sided superior vena cava (Figure 1A–C). To determine all these abnormal connections more in detail, cardiac catheterization was performed via the femoral vessels and confirmed two vessels (anterior and posterior) of nearly equal diameter at the usual site of IVC: (1) the posterior IVC received

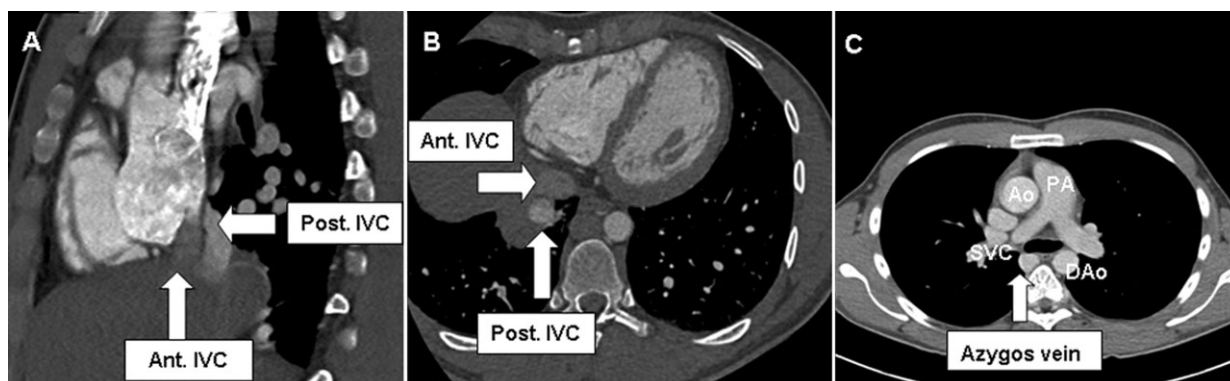


Figure 1. (A,B,C). Spiral computer tomography of the chest documented a posteriorly and anteriorly located IVC, an abnormal pulmonary venous return of the right lower lobe into the posteriorly located IVC and an azygos continuation connecting to a right sided SVC. IVC, inferior vena cava; SVC, superior vena cava; Ao, aorta; DAo, descending aorta; PA, pulmonary artery.

blood from a giant abdominal venous network and drained into the left atrium through two large collateral vessels and (2) the anterior IVC received only blood from the hepatic veins and drained correctly into the right atrium. There was a small communication between the two vessels. The heart catheterization confirmed the azygos continuation receiving blood from a paravertebral hypertrophied venous plexus and connecting to a right-sided superior vena cava. Finally, the lower right pulmonary vein was also draining into the posteriorly located IVC at the origin of the both collateral vessels that were connected with the left atrium (Figures 2 and 3). The pressures in the right heart were normal. No saturation samples were taken in the anteriorly located IVC, but were considered to be normal. Saturation samples in the posteriorly located IVC were 75%, 98% in the abnormally draining pulmonary veins, and 94% in the collaterals draining into the left atrium.

To avoid paradoxical embolism from the posterior IVC to the left atrium, it was decided first to open the small communication between the two vessels with 14-mm balloon dilatation (XXL balloon, Boston Scientific Corporation, Natick, MA, USA), and then to implant a device (Amplatzer Vascular Plug II, AGA Medical Corporation, Plymouth, MN, USA) into the posterior IVC (above the communication with the anterior IVC and beneath the drainage of the lower pulmonary vein into the collateral vessels, connected with the left atrium) (Figures 4 and 5). The closing procedure did not affect the transcutaneous saturation, which remained unchanged. The patient had a noncomplicated clinical course thereafter, was dismissed after 2 days of hospitalization with

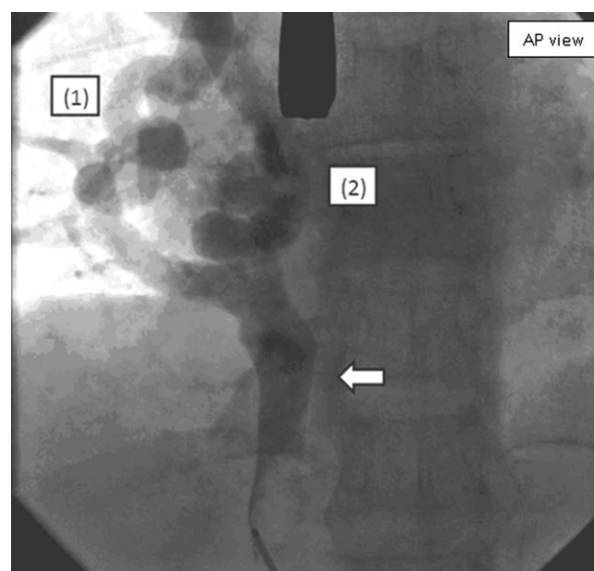


Figure 2. Angiography anteroposterior (AP) view of the posteriorly located IVC (arrow). The vena cava drains into the left atrium through two collateral veins (1) and (2), causing right-to-left shunt.

warfarin and he remains in excellent condition, free of symptoms after 1 month.

Discussion

A double IVC is a congenital anomaly that commonly results from persistence of the embryonal venous system, and occurs in 1–3% of the population.² Connection of the inferior caval vein to the morphologically left atrium is extremely rare. When seen, most cases are associated with atrial septal defects.³ In these patients, it is the occur-

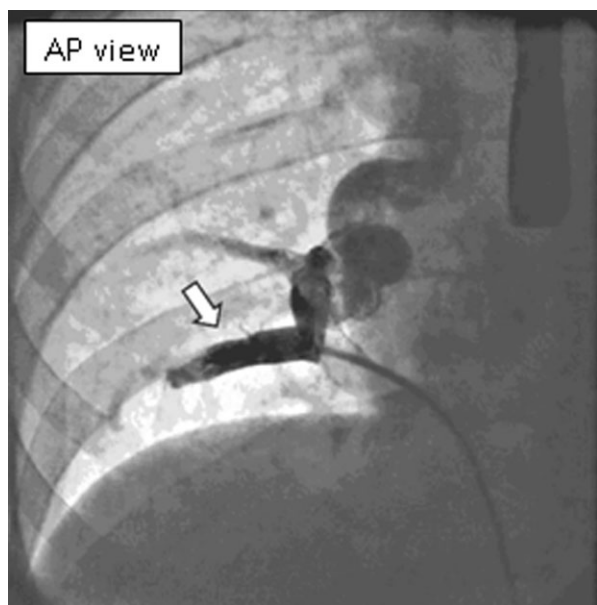


Figure 3. Angiography (AP) view of the pulmonary vein of the right lower lobe (arrow). This pulmonary vein drains into one of the collaterals that connect with the left atrium.

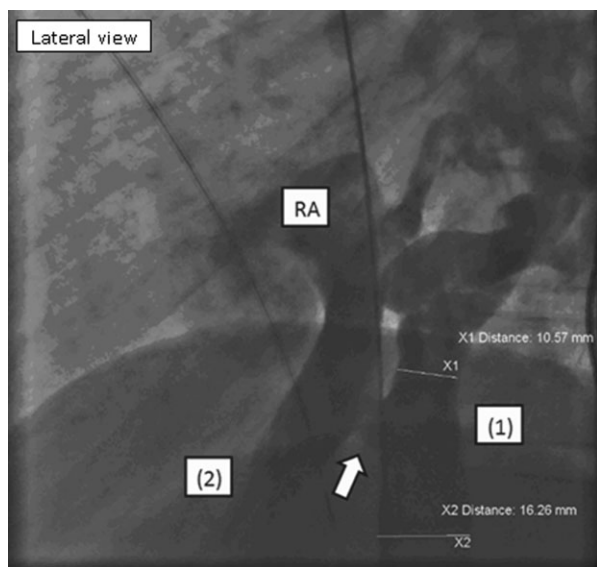


Figure 4. Angiography (lateral view) of the posteriorly located IVC (1). The posteriorly located IVC is connected (arrow) with the anteriorly located IVC (2), which drains into the right atrium (RA).

rence of severe cyanosis despite the presence of the atrial septal defect with normal right ventricular pressures that points to the correct diagnosis.

The embryology of the inferior vena cava is complex.⁴ It involves the formation, regression, and fusion among three longitudinal pairs of veins: the postcardinal, subcardinal, and supracardinal

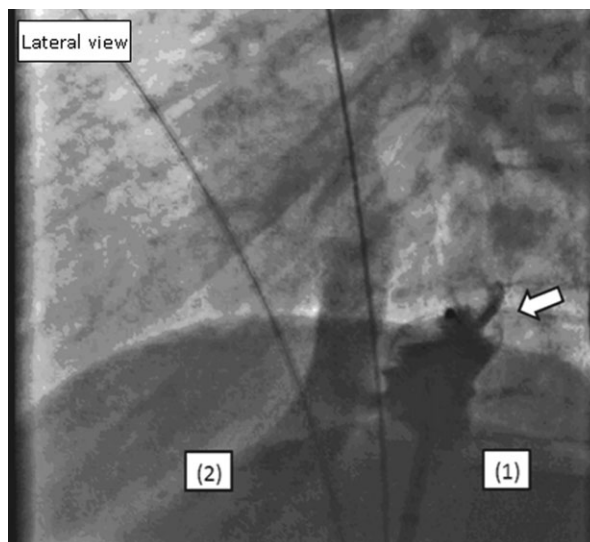


Figure 5. Angiography (lateral view) of the posteriorly located IVC (1) after closing the connection with the two fistula (arrow). The blood drains now preferentially to the anteriorly located IVC (2).

veins. Briefly, the normal IVC is composed of four segments: hepatic, suprarenal, renal, and infrarenal. The hepatic segment is derived from the vitelline vein. The right subcardinal vein develops into the suprarenal segment by subcardinal-hepatic anastomoses. The renal segment develops from right suprasubcardinal and postsubcardinal anastomoses. In the thoracic region, the supracardinal veins give rise to the azygos and hemiazygos veins. Progressive changes in the abdomen lead to replacement of the postcardinal veins by the subcardinal and supracardinal veins. In the pelvic region, the postcardinal veins persist as the common iliac veins.⁵ This complex process of IVC formation results in several variations in systemic venous return.^{6,7} Because of the many transformations that occur during the formation of the inferior vena cava, anomalies in its final form may occur. Such anomalies occur in 0.3% of otherwise healthy individuals and in 0.6–2% of patients with other cardiovascular defects.⁸

In our case report, there might be a “semantic discussion” concerning the name of the veins that were documented, also possible would be “interrupted right sided IVC”—or—nonfusion of the hepatic IVC segment with the suprarenal, renal, and infrarenal part.

Double IVC occurred due to the persistence of the left supracardinal vein.⁶ The most major anomalies that the IVC encountered are; duplication of the IVC, transposition of IVC (left IVC),

circumaortic (left) renal vein, retroaortic (left) renal vein and the absence of the hepatic portion of the IVC.⁹ Duplications of the IVC constitutes the major portion having a prevalence of 2–3%.^{10,11}

The double IVC in the presented case is considered as a single sided, whereas from the most cases in the literature, the IVC duplication is double sided. We believe that our patient is the one of the first to have been diagnosed with IVC duplication single sided and we agreed with previous reports.⁷

Previous study described long-term survival of a patient with left atrial connecting inferior vena cava with no anatomic intracardiac shunt.¹² Nevertheless, surgical or interventional correction of this cyanotic cardiac disease is indicated for prevention of paradoxical embolism, pronounced effort intolerance, and low peripheral oxygen saturation, with moderate polycythemia and an increase in left ventricular output as a compensatory mechanism.

Isolated drainage of the inferior vena cava into the left atrium is a rare congenital disorder, which should be considered in the differential diagnosis in patients with cyanosis without cardiac murmurs.

The majority of cases are clinically silent and diagnosed incidentally on imaging for other reasons. In our patient the saturations were normal, probably because the pulmonary vein drained into the collaterals from the posterior located IVC to the left atrium. We hypothesise that this oxygen jump masked the potential desaturation coming from the posteriorly located IVC. Probably, the flow in the abnormally draining pulmonary veins was competitive against the flow in the posteriorly located IVC, so that the saturations were relatively high in the collaterals draining into the left atrium.

Venography is considered to be the gold standard for the diagnosis of anomalies of the IVC. Computed tomography and magnetic resonance imaging may occasionally mistake the duplicated IVC for lymph nodes or other retroperitoneal structures.⁶ Operation or interventional procedures are the only methods for correction.

In conclusion, correct preoperative or precatheterization identification of such anomalies allows the interventionalist to avoid catastrophe during procedures with the intention to correct or to optimize abnormal systemic or pulmonary venous return.

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