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CASE REPORT

Levoatriocardinal Vein: An Unusual Cause of Right-to-Left Shunting

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ABSTRACT

We present a case demonstrating an anomalous vessel connecting the left brachiocephalic vein and the left superior pulmonary vein, found incidentally on computed tomography (CT) imaging. This pulmonary–systemic venous connection, known as a levoatriocardinal vein, is a rare anomaly. In previous descriptions, this vessel has typically been associated with left-to-right shunt. Here, we describe the magnetic resonance imaging (MRI) and CT findings in a case with right-to-left shunting through the anomalous vessel likely secondary to elevated right cardiac pressure.

Key words: Computed tomography, levoatriocardinal vein, magnetic resonance imaging, partial anomalous pulmonary venous connection, right-to-left shunt

INTRODUCTION

The levoatriocardinal vein (LACV) was first reported by McIntosh in 1926 and the term was used by Edwards and DuShane in 1950. This vessel is an anomalous connection between the left atrium or a pulmonary vein and any derivative of the cardinal venous system.[1,2] Most often, this anomaly has been described in the context of left-sided obstructive cardiac lesions such as mitral atresia and hypoplastic left heart.[3] It is reported that the embryological origin of this vein is an abnormal persistency of the splanchnic plexus connecting the pulmonary venous plexus and the cardinal system.[4,5] It is theorized that this connection may persist to provide alternative drainage of pulmonary venous blood in scenarios in which the left heart is malformed. However, cases of this anomalous vessel without concurrent congenital cardiac malformations have also been reported.[6] Typically, LACVs occur more commonly on the right side with blood flowing superiorly through the anomalous vessel from the pulmonary venous to the systemic venous system producing a left-to-right shunt.[7,8] In the case we present here, we describe the findings of an LACV on computed tomography (CT) and magnetic resonance imaging (MRI) with an atypical flow pattern producing a right-to-left shunt.

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CASE REPORT

A 37-year-old woman with a past medical history of bacterial endocarditis of the pulmonic, mitral, and tricuspid valves due to intravenous drug abuse, with subsequent tricuspid valve replacement and pulmonary embolism presented to our Emergency Department (ED) complaining of generalized myalgias and shortness of breath. She had no history of congenital cardiac disease. Notes in the medical record indicated that she had baseline oxygen deprivation attributed

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to a historical diagnosis of pulmonary hypertension. In the ED, she was tachypneic, tachycardic, and had persistent oxygen saturations around 88% on room air mandating the need for oxygen via nasal cannula at 2 l/min. Upon physical exam, she was in moderate respiratory distress and diaphoretic. Cardiovascular examination revealed a harsh, blowing holosystolic grade IV/VI murmur noted at the left upper sternal border. The rest of her physical exam was unremarkable. A complete metabolic panel (CMP) showed no abnormalities and a complete blood count (CBC) showed only mild anemia. No arterial blood gases were tested. An electrocardiogram (EKG) demonstrated a right bundle branch block, but no other marked abnormalities. Due to suspicion for pulmonary embolism, a CT scan of the chest was performed. The CT scan was negative for pulmonary embolism, but did reveal an anomalous vein connecting the left brachiocephalic vein to the left superior pulmonary vein [Figures 1 and 2 and Video 1]. The CT study demonstrated intravenously administered contrast passing from the left brachiocephalic vein through the anomalous vessel into the left superior pulmonary vein to the left atrium, indicating a right-to-left shunt via the anomalous vein. The anomalous vein measured 9 mm in maximum diameter. Subsequently, transthoracic and transesophageal echocardiography was performed. This demonstrated dilatation of the right atrium and right ventricle, severe tricuspid valve regurgitation, and elevated right ventricular pressure estimated at 30 mm Hg. The inferior vena cava was dilated without respiratory variation, also consistent with elevated right atrial pressure. A small fistula between the ascending aorta and the right ventricle was also seen likely secondary to endocarditis. In retrospect, this fistula was not visible on CT due to streak artifacts from the previous tricuspid valve prosthesis. Agitated saline injection showed appearance of bubbles in the left atrium from the left superior pulmonary vein, confirming an intrapulmonary right-to-left shunt. MRI of the anomalous vein was also performed to evaluate the flow characteristics [Figure 3]. As the patient was claustrophobic, only a limited evaluation of the anomalous vein could be performed. The study demonstrated monophasic, inferiorly directed flow within the vessel toward the left superior pulmonary vein, with the flow calculated as 20 ml per heartbeat.

Shortly after admission, treatment was started for bacterial endocarditis and subsequent blood cultures were positive for *Escherichia coli*. Her symptoms were attributed to bacterial endocarditis and right-heart overload secondary to the severe tricuspid valve regurgitation and the small fistula between the aortic root and the right ventricle. Right-to-left shunting through the anomalous pulmonary vessel was also believed to further exacerbate her hypoxia. No interventions were undertaken with regard to the

Video 1: 37–year year-old female who presented with dyspnea and hypoxia with a diagnosis of levoatriocardinal vein. Contiguous axial images from contrast–enhanced CT scan of the chest demonstrate the abnormal vessel. Beginning at the level of the left brachiocephalic vein, bright undiluted contrast drains inferiorly as it flows through the anomalous vessel, to the left superior pulmonary vein, and finally into the left atrium. Therefore, this flow demonstrates an extra-cardiac right–to–left shunt.

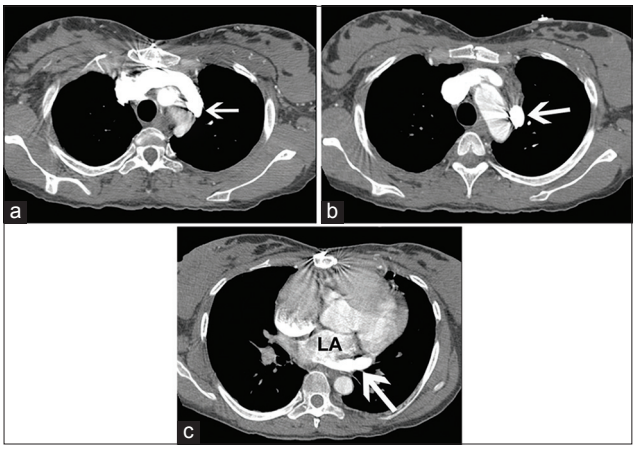

Figure 1: 37-year old female who presented with dyspnea and hypoxia with a diagnosis of levoatriocardinal vein. Axial images from contrast-enhanced CT scan of the chest demonstrate the abnormal vessel. (a) Image at the level of the left brachiocephalic vein shows connection with the anomalous vein (arrow). (b) Image at the level of the aortic arch shows bright undiluted contrast within the anomalous vein (arrow). (c) Image at the level of the left atrium shows bright undiluted contrast in the left superior pulmonary vein (arrow) draining into the left atrium (LA). Thus, the inferiorly directed flow through the anomalous vein produces an extracardiac right-to-left shunt.

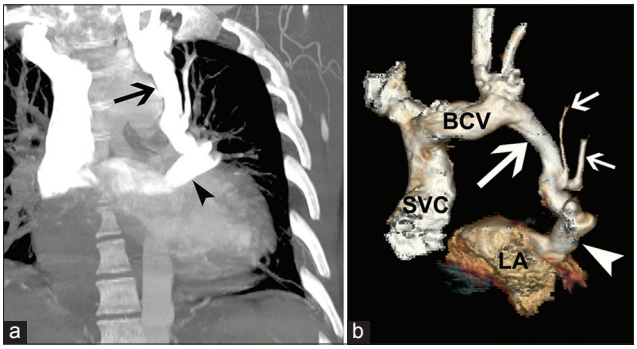

Figure 2: 37-year-old female who presented with dyspnea and hypoxia with a diagnosis of levoatriocardinal vein. Reformatted images from chest CT scan demonstrate the abnormal vessel. (a) Reformatted coronal image shows flow through the levoatriocardinal cardinal vein (arrow) to the left superior pulmonary vein (arrowhead). (b) Volume-rendered image from an anterior perspective shows the levoatriocardinal vein (large arrow) connecting the left brachiocephalic vein (BCV) to the left superior pulmonary vein (arrowhead). Note small pulmonary venous branches (small arrows) connecting to the anomalous vessel. SVC = Superior vena cava; LA = Left atrium.

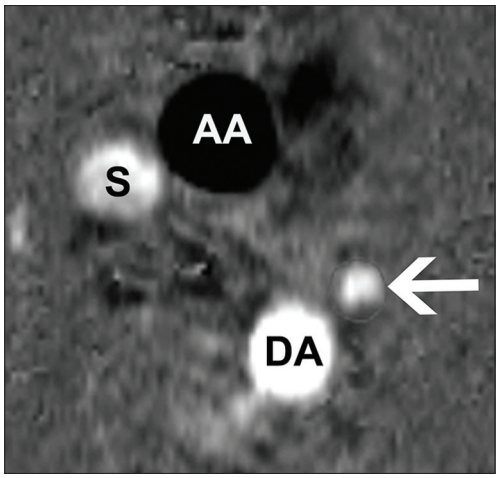

Figure 3: 37-year-old female who presented with dyspnea and hypoxia with a diagnosis of levoatriocardinal vein. Axial image from velocity-encoded phase-contrast MRI sequence shows the flow direction in anomalous vessel. The levoatriocardinal vein (arrow) is seen in cross section. In this sequence, superiorly directed blood flow is encoded as black while inferiorly directed flow is encoded as white. Flow direction in the anomalous vein is the same as that in the descending thoracic aorta (DA) and superior vena cava (S), confirming inferiorly directed flow. From this sequence, absolute volume of flow per heartbeat can also be calculated. AA = Ascending thoracic aorta.

anomalous vessel during the patient's hospital course. The patient left the hospital against medical advice before completion of therapy and was lost to follow-up.

DISCUSSION

As stated previously, the LACV has classically been described as a vessel producing a left-to-right shunt with the flow directed superiorly. These characteristics have typically been demonstrated using contrast-enhanced imaging studies as well as phase-contrast MRI.[8,9] As demonstrated in our case, predominantly right-to-left shunting through the anomalous vessel can occur with elevated right heart pressure. In a case report by Cullen et al., the authors describe a similar phenomenon with bidirectional flow in an LACV demonstrated on color Doppler and MR cine phase-contrast imaging. The authors report superiorly directed flow (left-to-right shunt) in an LACV at rest, but inferiorly directed flow toward the left atrium (right-to-left shunt) during the Valsalva maneuver when the right cardiac pressure is transiently elevated. We postulate that in our patient, the combination of severe tricuspid regurgitation, pulmonary hypertension, and the fistulous tract from the ascending aorta to the right ventricle all contributed to elevated right heart pressure producing elevated pressure in the left brachiocephalic vein promoting flow in the anomalous vessel toward the left atrium. We suspect that the continuous nature of this right-to-left shunt contributed to the hypoxia in our patient.

Typically, LACVs have been surgically managed with ligation with or without intra- or extracardiac rerouting. Following temporary occlusion of the distal portion of the LACV (portion closest to the systemic vein), simple ligation can be performed as long as the mean pressure in the LACV does not exceed 30 mm Hg. If the venous pressure exceeds 30 mm Hg, then rerouting of the vein to the left atrium is indicated to allow adequate decompression of pulmonary tributaries draining into the LACV.[10] However, the effects of ligation of this vessel have not been studied in cases such as ours with right-to-left shunting.

CONCLUSION

In summary, our case shows that flow direction in an LACV can be ascertained on contrast-enhanced CT by noting the location of undiluted contrast on sequential images as demonstrated in Figure 1. In our case, the demonstration of a column of undiluted contrast extending into the left atrium along the course of the LACV indicates inferiorly directed flow along the anomalous vessel producing a right-to-left shunt. Phase-contrast MRI is valuable for both determining the flow direction and quantifying the amount of flow in an LACV. While flow in an LACV usually produces a left-to right shunt, a right-to-left shunt may occur when the right cardiac pressure is elevated.

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