

CASE REPORT

Vena Caval Anomalies

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ABSTRACT

Anomalous vena cavae can have significant implications for procedures on the right side of the heart. We report a rare anatomical configuration in a 44-year-old female, which to the best of our knowledge, is the first report of such an association. She had a bicuspid aortic valve in conjunction with a persistent left superior vena cava (PLSVC) draining into the coronary sinus, and a left-sided inferior vena cava (IVC) draining into a left superior vena cava via the hemiazygos vein. Comprehensive assessment of these anomalies is crucial given the widespread use of invasive cardiac procedures.

Key words: Inferior vena cava, persistent left superior vena cava, vena caval anomalies

INTRODUCTION

Persistent left superior vena cava (PLSVC) and left-sided inferior vena cava (IVC) are both rare vascular anomalies. Amongst all inferior cava anomalies, a left-sided IVC communicating with PLSVC through the hemiazygos vein is the least common.^[1] We report a rare anatomical configuration in a 44-year-old female, which to the best of our knowledge, is the first report of such an association.

CASE REPORT

A 44-year-old fit and active female was referred by her general practitioner with palpitations, mostly occurring at rest. Electrocardiography performed in the community

demonstrated sudden onset of an atrial tachycardia of 115 beats per min. When assessed in hospital, she was asymptomatic with a blood pressure of 110/62 and a regular pulse rate of 50 beats per min. Resting electrocardiography revealed sinus rhythm with poor R wave progression with no evidence of left ventricular hypertrophy or ischemia. A 24-h electrocardiography demonstrated four beats of atrial tachycardia.

A transthoracic echocardiogram demonstrated a prominent coronary sinus, moderate mitral regurgitation, and a bicuspid aortic valve [Video 1]. Transesophageal echocardiography showed normal biventricular systolic function, left atrial dilatation, mild to moderate mitral regurgitation, and tricuspid regurgitation. The aortic valve was bicuspid with trivial aortic regurgitation but no significant stenosis. The ascending aorta was dilated for her age and body surface area, with a maximum diameter of 4.0 cm. There was no evidence of aortic coarctation or dissection.

Saline contrast echocardiography was performed via the left and right arms [Videos 2], [Videos 3], [Videos 4] and [Videos 5]. This confirmed PLSVC with no obvious shunt. Cardiac magnetic resonance imaging (MRI) demonstrated

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bicuspid aortic valve with dilated aortic root. A PLSVC was seen draining into the coronary sinus and then the right atrium. A separate right-sided non-hypoplastic superior vena cava also emptied into the right atrium. It was noted that a left-sided IVC drained into the coronary sinus and right atrium. Three hepatic veins were seen emptying into a short right-sided IVC, again draining into the right atrium.

DISCUSSION

PLSVC is the most common thoracic venous anomaly with a prevalence of 0.3-0.5% in the general population.^[2] PLSVC results from failure of regression of the left superior cardinal vein caudal to the innominate vein during embryological development and coexists with a right superior vena cava in up to 80–90% of cases, as in this case.^[2] In approximately 80-92% of cases, the PLSVC drains into the right atrium via the coronary sinus resulting in no hemodynamic consequence.^[2] Conversely, in the remainder of patients, the PLSVC can drain into the left atrium directly, or through an unroofed coronary sinus, or the left superior pulmonary vein creating a right to left shunt, predisposing the patient to paradoxical embolism.^[2] This variant confers the greatest risk in patients undergoing central venous catheter placement. In any suspected patient, venous imaging is therefore required to define the pattern of cardiac venous return at central venous catheter placement prior to use of their device. PLSVC can also complicate permanent pacemaker and cardioverter-defibrillator implantation, due to difficulties navigating through the anomalous venous anatomy and problems with lead instability and displacement.^[3] Complications of lead placement via PLSVC are relatively rare and include arrhythmia, cardiogenic shock, cardiac tamponade, and coronary sinus thrombosis.^[4]

Left-sided IVC is relatively less prevalent (0.2-0.5%) compared with PLSVC,^[5] and develops as a result of the persistence of the left supracardinal vein. A left-sided IVC usually communicates with a normally positioned infra-hepatic IVC and thus empties into the right atrium in the usual position. Occasionally, a left-sided IVC may communicate with either the azygos or hemiazygos vein.^[5] In cases of left-sided IVC, possible routes for the return of blood to the right atrium are via the azygos vein to the SVC, via the left brachiocephalic vein to right SVC or via the

hemiazygos vein to PLSVC.^[5] Our case reports a left-sided IVC communicating with PLSVC via the hemiazygos vein in association with a bicuspid aortic valve. Other reported congenital cardiac defects accompanying this combination of venous anomalies include patent ductus arteriosus and atrial and ventricular septal defects.^[6] Congenital cardiac anomalies can be associated with vena caval anomalies, which may have serious implications for procedures on the right side of the heart. Detailed investigations of these anomalies are crucial, given the increasing use of invasive cardiac and electrophysiological procedures. Diagnostic tools include saline contrast echocardiography, MRI, and venous angiography for more definitive confirmation.

CONCLUSION

Vena caval anomalies can have significant implications for procedures on the right side of the heart. Comprehensive assessment of these anomalies is crucial given the widespread use of invasive cardiac procedures.

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