

Persistent Left Superior Vena Cava, Absent Right Superior Vena Cava, Right Pectoral Muscle Agenesis: An Intriguing Association

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Abstract

Background: Persistent left superior vena cava (PLSVC) is the most common thoracic venous anomaly. PLSVC with absent right superior vena cava (RSVC) is exceedingly rare and is reported in approximately less than 0.1% of the general population and in only 0.09% to 0.13% of patients who have congenital heart defects. Combination of these anomalies with right pectoral muscle agenesis, to our knowledge, has never been reported.

Case report: A 35-year-old man with right pectoral muscle agenesis was referred for cardiac magnetic resonance (CMR) to better investigate coronary sinus dilatation incidentally detected at transthoracic echocardiography. CMR imaging showed the presence of PLSVC, draining into the coronary sinus, which appeared slightly dilated. CMR also revealed the absence of RSVC and confirmed right pectoral muscle agenesis.

Conclusion: The findings described were incidental and the patient was asymptomatic. Awareness of thoracic venous abnormalities is important, should invasive procedures be necessary.

Whether the association of thoracic venous abnormalities and pectoral muscle agenesis is casual or shares a common pathophysiological background is unknown.

Keywords

Absent right superior vena cava; Persistent left vena cava; pectoral muscle agenesis; Cardiac magnetic resonance

Introduction

Persistent left superior vena cava (PLSVC) is the most common thoracic venous anomaly occurring in 0.3% to 0.5% of individuals in the general population (1). The incidence of this anomaly is 10-fold increased in patients with congenital cardiac malformations, as reported by Danielpour and Palinkas (2-3). In most patients with PLSVC, a right superior vena cava (RSVC) is present. PLSVC with

absent RSVC is reported in approximately less than 0.1% of the general population and in only 0.09% to 0.13% of patients who have congenital heart defect (4-5). The association of PLSVC and absent RSVC with agenesis of right pectoral muscle, to the best of our knowledge, has never been reported.

Case Presentation

A 35-years-old Caucasian man with agenesis of right pectoral muscle was referred to our hospital to perform cardiac magnetic resonance (CMR) imaging to better investigate the incidental finding of dilated coronary sinus at transthoracic echocardiography. CMR confirmed the presence of PLSVC and the site of drainage into the coronary sinus, which appeared slightly dilated (1.6 cm). CMR also revealed the absence of right superior vena cava (Figure 1 Panel A, Figure 1 Panel B).

There was no evidence of valvular heart disease and the volumes of the heart chambers were within normal limits. Biventricular ejection fraction was normal; the inter-atrial and inter-ventricular septum were both intact. Pulmonary venous returns were normal. The ratio of pulmonary to systemic flow (QP/QS) was one. The thickness of pericardium was normal with no effusion. Visceral organs were normally positioned and structured. Agenesis of right pectoral muscle was confirmed (Figure 2).

Discussion

PLSVC is an embryological remnant that represents persistence of the embryonic left anterior cardinal vein (or “vein of Marshall”) and is the most common thoracic venous anomaly. PLSVC usually drains into the right atrium through a dilated coronary sinus. A recent review reveals that, in some cases, it may drain directly into the left atrium producing a right-to-left shunt and increasing the risk of paradoxical embolism with neurologic, mesenteric, and/or peripheral sequelae (6). This venous malformation is usually diagnosed incidentally, but it can pose difficulties with central venous access, pacemaker implantation and cardiothoracic surgery and it is also associated with an increased incidence of congenital heart disease, arrhythmias and conduction disturbances, as reported by Morgan (7).

PLSVC can occur in several anatomic variations. Numerous reviews reveal that PLSVC coexists with a right SVC in up to 80% to 90% of cases, but in rare cases RSVC can be absent (“isolated PLSVC”) (8-9). The incidence of PLSVC is 0.3% to 0.5% in the general population (10), while PLSVC with absent RSVC is even more uncommon, occurring in approximately less than 0.1% of the general population and in only 0.09% to 0.13% of patients who have congenital heart defects (1).

To our knowledge, the combination of these anomalies with agenesis of pectoral muscle has never been reported. Anomalies of pectoral muscle are described mainly in Poland's syndrome, a rare unilateral congenital anomaly characterized by the absence of the pectoral muscle and hand anomalies. As explained in different studies, the accepted etiological origin of this syndrome is the temporary interruption of blood supply of the subclavian artery and its branches in the early gestational period (11-12-13). It is unlikely that our patient was suffering from Poland's syndrome, as none of the other features of the syndrome, a part from pectoral muscle agenesis, were present.

The patient was asymptomatic and abnormalities of the thoracic venous system were detected occasionally. Whether pectoral muscle agenesis and venous system abnormalities share a common pathophysiological background or are casually associated in our patient is not defined.

Figures

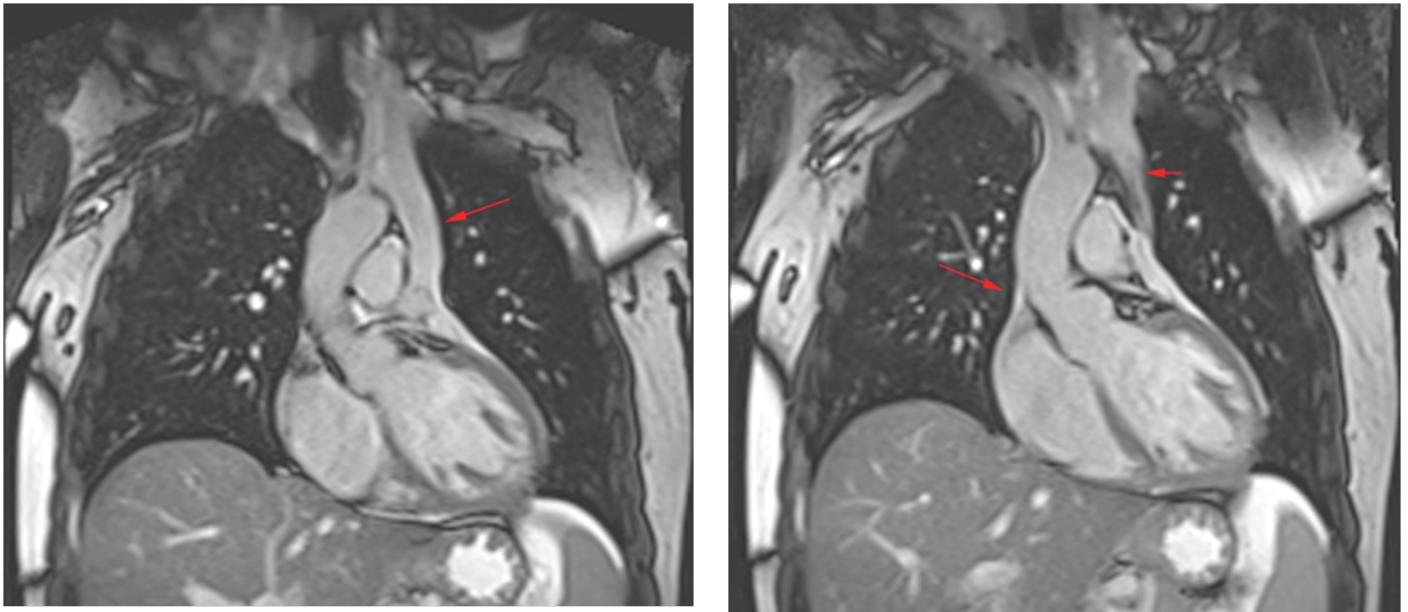


Figure 1: Panel A(left): CMR (true-FISP single-shot coronal images) showing persistent left superior vena cava (arrow); Panel B(right): CMR showing persistent left superior vena cava (short arrow) associated with right superior vena cava agenesis (long arrow).

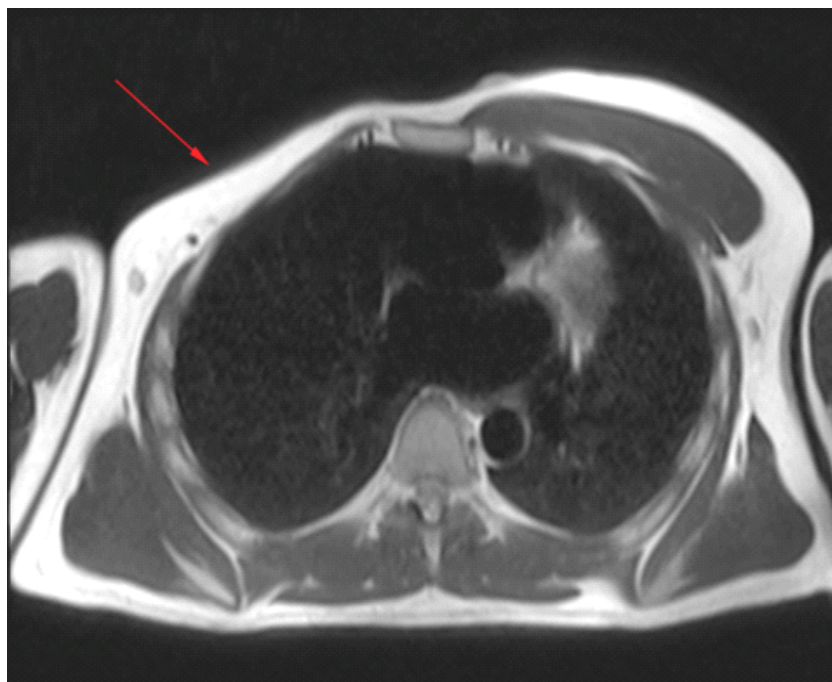


Figure 2: CMR (HASTE image, axial plane) showing right pectoral muscle agenesis (arrow).

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