Rare disease

Anomalous venous circuit: a detour to the heart

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Summary

We report a case of a complicated vascular access secondary to systemic venous defects, which have not been previously reported. Evaluation revealed an atypical coursing of a subclavian vein-persistent superior vena caval system into the hemizygos-azygos system and ultimately into the right (proper) superior vena cava. The clinical and surgical considerations of such an anomaly are discussed therein. We look at embryological origins of such a merger and share our experience in troubleshooting through the anomaly.

BACKGROUND

The most commonly encountered systemic thoracic venous anomaly is a persistent left superior vena cava (PLSVC).¹ We share an interesting case of a patient with a constellation of systemic venous anomalies, which have not been previously reported. An understanding of these anomalies is of paramount importance because of the association with various congenital defects.² Anomalies present potential problems with cardiovascular access^{3–6} for therapeutic or diagnostic studies pertaining to the fields of cardiology, oncology, critical care, anaesthesiology and general/thoracic surgery.

CASE PRESENTATION

A 77-year-old woman with a history of dilated cardiomyopathy and non-sustained ventricular tachycardia presented for the evaluation of recurrent episodes of syncope. Her vitals and examination were unremarkable. An admission EKG revealed for evidence of right bundle-branch block with QRS duration of 160 ms. Left anterior fascicular block was also noted. A prior echocardiogram revealed for an ejection fraction of 35%, with moderate aortic regurgitation and left atrial enlargement with a global hypokinesis. Her recurrent syncope was thought to be due to recurrent ventricular tachycardia. Implant a cardioverter-defibrillator (ICD) placement was considered for (A) protection against recurrent syncope and/or death (B) cardiac resynchronisation therapy for congestive heart failure and (C) implementation as a pacing unit for developed conduction system disease. Following appropriate consent and sterile preparation, a peripheral venography was performed to guide venous access. A micropuncture needle was then used to gain access to the extrathoracic left axillary vein. An introduced glide wire could not be advanced, taking a mid-loop position in the thorax.

INVESTIGATIONS

A repeat venogram (figure 1A,B) revealed a left subclavian vein draining into what appeared to be PLSVC. This drained into an accessory hemizygos vein, azygos vein, its arch, the superior vena cava proper and the right atrium. Given technical difficulties, the approach was deemed

unfavourable for device implantation. Therefore, haemostasis was achieved and the patient was prepared for an alternate attempt from the opposing right side. Venous access was then obtained in the extrathoracic component of the right axillary vein. Initially, advancement of the pacing electrode catheter into the coronary sinus was met with resistance. Multiple attempts were made without success. Next, a straight sheath was utilised with successful placement into the coronary os. Atrial and ventricular leads were then placed and hooked up to the device with appropriate capture.

DIFFERENTIAL DIAGNOSIS

The utilisation of the contrast venogram during our approach towards ICD placements made our relative differential limited. We had encountered a venous anomaly comprising of the subclavian vein, PLSVC, accessory hemiazygos and azygos vein draining into the superior vena cava proper.

OUTCOME AND FOLLOW-UP

No immediate complications were encountered during this dual-chamber ICD implantation.

DISCUSSION

Marshall (in 1850) was the first to account for the development of the great anterior veins. He went on to describe the fetal remnants as found in adults.⁷ A century later, Edwards and DuShane described that the PLSVC was the commonest venous anomaly of the thorax, accounting for up to 2% in the general population and higher (up to 4.3%^{8–10}) in patients with other congenital cardiac abnormalities.¹ This PLSVC may occur in the absence of a right superior vena cava, in up to 10% of subjects.^{11–12} Other thoracic venous anomalies reported include azygos and hemiazygos continuation of an interrupted inferior vena cava, and anomalous pulmonary venous return.¹³

Venous anomalies were often reported for academic interest. This was because with the exception of an anomalous left-to-right shunt, the great majority of cases were without haemodynamic consequences. Clinical interest

Figure 1 Venogram revealed a left subclavian vein (SV) draining into a left persistent superior vena cava (LPSVC), left accessory hemiazygos vein (HAV), azygos vein (AV) the azygos arch (AA), the superior vena cava (SVC) and right atrium (RA).

has increased with the reporting of association with other congenital heart abnormalities, the most common being atrial septal defects. The true association is under reported, given that subjects without associated lesions are not the focus of investigation. Our patient did not have associated congenital anomalies.

Surgical considerations are equally important. As with our patient, technical difficulties imposed by convoluted coursing may be encountered when attempting to ICD.^{2 5 14 15} Similar issues may arise when other transvenous procedures (via left subclavian approach) are attempted. These include central line placement,^{6 16} right heart catheterisation, electrophysiological studies, cardiopulmonary bypass⁶ and biventricular pacing.¹⁶ In addition to incompleteness of the intended procedure, there may be derangement of ICD and/or pacemaker leads. More significant consequences include angina, cardiogenic shock and production of arrhythmia leading to sudden cardiac death.¹⁷

Our patient had a left subclavian vein draining into a PLSVC with continuation into the hemiazygos-azygos vein system. This atypical course ultimately concluded into the right atrium via the superior vena cava. (figure 2A,B)

Embryologically,^{18–20} the PLSVC is a result of a failure of natural obliteration of the left anterior cardinal vein. In the great majority of cases (greater than 90%), the persistent vessel enters the coronary sinus through its oblique branch. In the presence of associated coronary sinus atresia, drainage is retrograde through a bridging left innominate vein. This arrangement is without clinical consequence, as blood drainage of the head and neck is not compromised. During fetal venous system development, the hemiazygos vein develops from the upper left supracardinal vein; the azygos vein is a derivative of the right posterior cardinal vein and the azygos arch from an upper segment of the right posterior cardinal vein. In our case, it is postulated, the thoracic venous anomaly

Figure 2 Comparison of normal and anomalous venous systems. (A) Demonstrates the development of the normal venous system. (B) Shows the anomalous venous circuit with the left subclavian vein draining into the left persistent superior vena cava, left accessory hemiazygos vein, azygos vein, the azygos arch and the superior vena cava.

developed from the communication of the left subclavian vein and the cardinal vein systems. This union, ultimately derived the anterior cardinal vein, which in the adult forms the superior vena cava.

In a simple PLSVC-coronary sinus anomaly, a left-sided approach may be trialled and is often successful. In the case above, a left-sided approach was met with technical limitations. A right-sided approach proved successful on utilisation of straight sheaths. Although it has largely been replaced with trans-venous lead placement, epicardial lead implantation by thoracotomy can be utilised in situations where thoracic venous anomalies pose technical difficulties.

Learning points

- We encountered a venous anomaly involving a left subclavian vein draining into a persistent left-sided superior vena cava. This, in turn, drained into the accessory hemizygous vein, the azygos vein and ultimately into the superior vena cava.
- It is postulated that during embryological development, the thoracic venous anomaly developed from the communication of the left subclavian vein and the cardinal vein systems. This union, ultimately derived the anterior cardinal vein, which in the adult forms the superior vena cava.
- The discovery of a venous anomaly is of academic, clinical and surgical importance. Clinical interest has increased with the reporting of association with other congenital heart abnormalities, the most common being atrial septal defects. This could pose a potential problem with cardiovascular access for therapeutic or diagnostic studies pertaining to the fields of cardiology, oncology, critical care, anaesthesiology and general/thoracic surgery.
- Technical difficulties imposed by convoluted coursing may be encountered when attempting to implant a cardioverter-defibrillator.

Competing interests None.

Patient consent Obtained.

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