

Isolated Persistent Left Superior Vena Cava with Interrupted Inferior Vena Cava and Hemiazygos Continuation: A CT Angiography Case Report

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Abstract

Case Report

Congenital anomalies of systemic venous return result from complex variations in the persistence and regression of the embryonic venous system. Persistent left superior vena cava (PLSVC) is the most common congenital thoracic venous anomaly, whereas the isolated form with absence of the right superior vena cava (RSVC) is rare. Its association with interrupted inferior vena cava (IVC) and hemiazygos continuation is even less common. We report the case of a 52-year-old woman with diabetes and hypertension who was investigated for chronic exertional dyspnea. Contrast-enhanced chest CT demonstrated a single PLSVC draining into a dilated coronary sinus, absence of the RSVC, and an arteria lusoria, with suspected interruption of the IVC. Complementary thoracoabdominal CT angiography confirmed IVC interruption, persistence of a short retrohepatic IVC segment draining into the right atrium, drainage of the hepatic veins into this residual caval segment, and continuation through a dilated hemiazygos vein joining the PLSVC. This configuration creates an unusual pathway for lower systemic venous return through the hemiazygos system, the PLSVC, and the coronary sinus. This case emphasizes the value of multiplanar and maximum-intensity-projection reconstructions for accurate mapping of complex caval anomalies before endovascular, electrophysiologic, or surgical procedures.

Keywords: Persistent left superior vena cava, interrupted inferior vena cava, hemiazygos continuation, coronary sinus, CT angiography, congenital venous anomaly.

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INTRODUCTION

Congenital anomalies of systemic venous return are commonly detected incidentally on cross-sectional imaging and reflect variations in persistence or regression of the embryonic venous network [1]. PLSVC is the most frequent congenital thoracic venous anomaly; in most cases, a RSVC is also present and the PLSVC drains into the right atrium through the coronary sinus [1,2]. Concomitant absence of the RSVC defines an isolated PLSVC, a much rarer configuration [3]. Interrupted IVC with azygos or, less commonly, hemiazygos continuation results from abnormal development of the caval and supracardinal venous segments [1,4].

The combination of isolated PLSVC and interrupted IVC with hemiazygos continuation has only

been reported sporadically. We present the CT angiographic anatomy of this association, accompanied by a short retrohepatic IVC segment and an arteria lusoria.

CASE REPORT

A 52-year-old woman with type 2 diabetes and hypertension was admitted for chronic exertional dyspnea. Contrast-enhanced chest CT was performed. It demonstrated a complex systemic venous return anomaly consisting of a PLSVC with absence of the RSVC. The PLSVC descended along the left mediastinum and drained into a markedly dilated coronary sinus. An arteria lusoria was also identified. The initial examination additionally raised suspicion of interrupted IVC with a dilated hemiazygos continuation.

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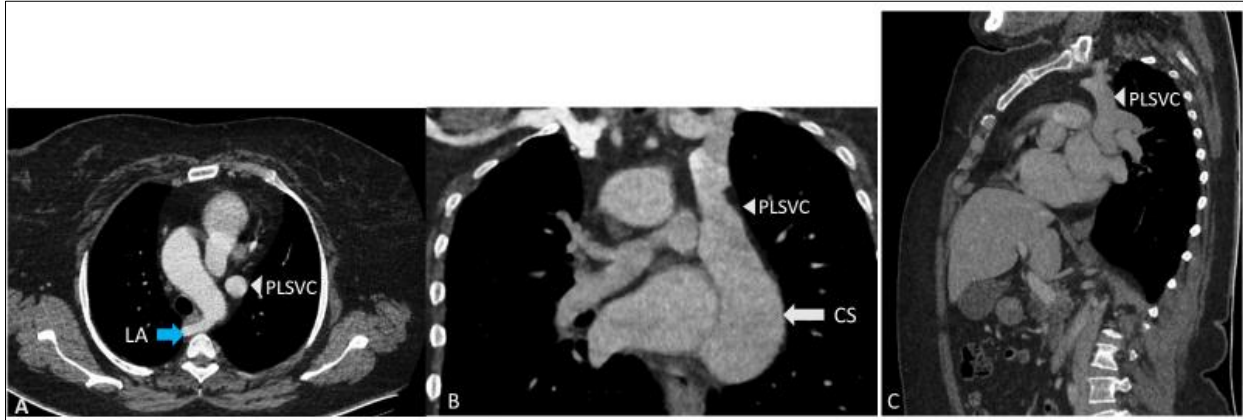


Figure 1: Axial (A), coronal (B), and oblique sagittal (C) CT images showing a persistent left superior vena cava (arrowhead) draining into a dilated coronary sinus (white arrow), with an arteria lusoria (blue arrow). PLSVC = persistent left superior vena cava; CS = coronary sinus; LA = arteria lusoria

To define the complete vascular anatomy and assess for additional developmental vascular anomalies, thoracic and abdominal CT angiography was subsequently performed. It confirmed interruption of the IVC. Venous return normally conveyed by the IVC was

diverted through a dilated hemiazygos vein, which ascended in the posterior mediastinum and joined the PLSVC. The venous pathway then continued through the dilated coronary sinus to the right atrium.

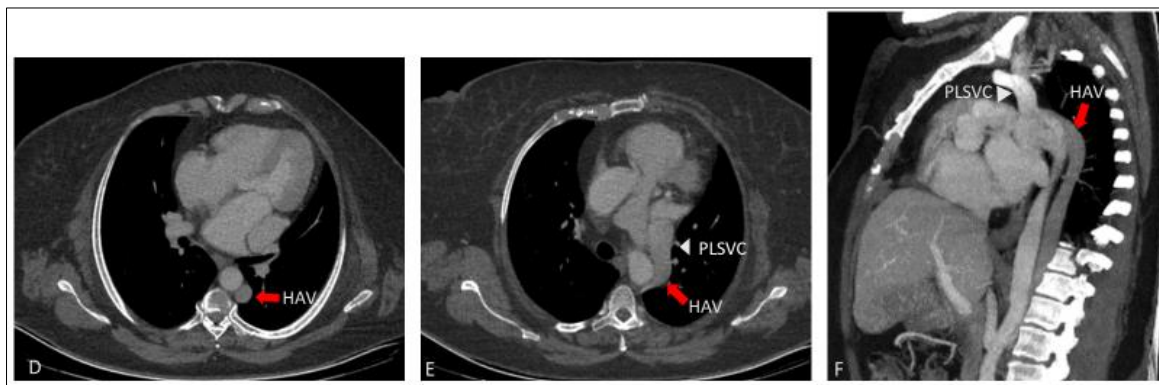


Figure 2: Axial CT images (D, E) and oblique sagittal maximum-intensity-projection image (F) showing a dilated hemiazygos vein (red arrow) draining into the persistent left superior vena cava. HAV = hemiazygos vein; PLSVC = persistent left superior vena cava

A short retro/transhepatic IVC segment persisted. The hepatic veins converged into this residual caval segment, which drained into the right atrium.

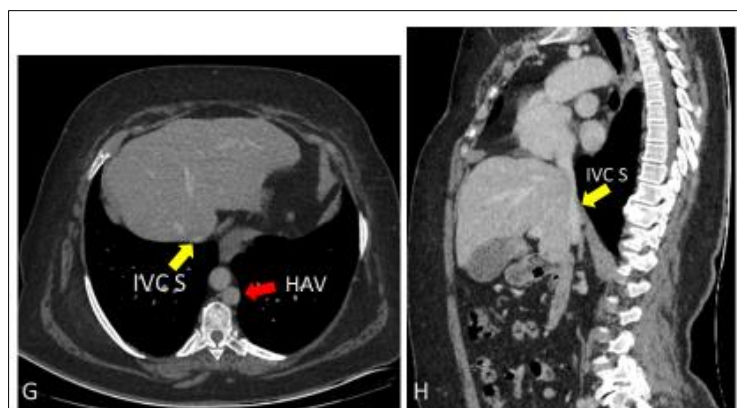


Figure 3: Axial (G) and oblique sagittal (H) CT images showing the short retro/transhepatic inferior vena cava segment (yellow arrow). The hepatic veins converge into this residual caval segment, which drains into the right atrium. IVCS = short inferior vena cava segment; HAV = hemiazygos vein

DISCUSSION

Development of the SVC primarily depends on the anterior cardinal venous system. Persistence of the left anterior cardinal vein results in a PLSVC; when the right cardinal system regresses simultaneously, the RSVC is absent and the PLSVC becomes the single superior caval pathway [1–3]. In the usual configuration, the PLSVC enters the right atrium through the coronary sinus. Coronary sinus dilatation is therefore an important imaging clue that should prompt a search for PLSVC [2,5]. In the present case, the PLSVC-coronary sinus continuity was clearly depicted on axial, coronal, and oblique sagittal reconstructions.

IVC embryogenesis is more complex and involves the vitelline, subcardinal, and supracardinal venous systems. Interruption of the hepatic IVC segment is classically related to failure of the right vitelline-subcardinal anastomosis, with diversion of lower systemic venous return into the supracardinal system [1,4]. Drainage most commonly occurs through the azygos vein; hemiazygos continuation is considerably less frequent. The hepatic veins then retain a direct route to the right atrium, often through a residual hepatic or suprahepatic caval segment [1,4]. The anatomy in our patient is particularly illustrative: the interrupted IVC is functionally replaced by a dilated hemiazygos vein that joins the PLSVC, whereas the hepatic veins converge into a short residual retro/transhepatic IVC segment that drains directly into the right atrium.

Several previously published cases help place this configuration in context. Kim *et al.*, described interrupted IVC with hemiazygos continuation to a PLSVC and hepatic venous drainage to the right atrium, but with a preserved RSVC [6]. Kabakus *et al.*, reported a left-sided IVC continuing through the hemiazygos vein to a PLSVC in the absence of the RSVC, with direct hepatic venous drainage to the right atrium [7]. Mondal *et al.*, described absent RSVC, PLSVC, and interrupted IVC continuing as the hemiazygos vein to the PLSVC and then the coronary sinus [8]. More recently, Lin and Sun illustrated a PLSVC associated with hemiazygos continuation of a left-sided IVC on CT [9]. The present case therefore belongs to a very uncommon spectrum of combined caval connections, with the additional findings of a short retrohepatic IVC segment and an arteria lusoria.

The radiologic relevance is twofold. First, each venous trunk should be followed along its entire course rather than simply identifying a PLSVC or a dilated paravertebral vein. Multiplanar, maximum-intensity-projection, and, when available, volume-rendered reconstructions allow complete depiction of the venous pathway and help avoid mistaking a dilated vascular structure for a mediastinal mass or lymphadenopathy [1,4,5]. Second, accurate venous mapping directly affects planning of central venous catheterization,

femoral venous access, pacemaker lead implantation, electrophysiology procedures, and selected cardiothoracic operations [3,5,8]. In this patient, a femoral venous route would ascend through the hemiazygos system and the PLSVC before reaching the right atrium via the coronary sinus, a markedly different course from normal anatomy.

These venous anomalies are often well tolerated when systemic venous blood ultimately reaches the right atrium without a significant shunt [1,2]. However, the available data did not include detailed echocardiographic or hemodynamic assessment; consequently, a causal relationship between the chronic exertional dyspnea and the venous anomaly cannot be established. The arteria lusoria represents an associated arterial variant, but no compressive symptoms or additional data were available to determine its clinical significance.

CONCLUSION

The association of isolated PLSVC and interrupted IVC with hemiazygos continuation is a rare configuration of systemic venous return. CT angiography provides comprehensive anatomic mapping by demonstrating PLSVC drainage into the coronary sinus, the compensatory hemiazygos pathway, and direct hepatic venous return to the right atrium. Recognition of this anatomy is essential to avoid diagnostic pitfalls and to anticipate difficulties during venous, electrophysiologic, or surgical procedures. The concomitant arteria lusoria further supports the value of a complete thoracoabdominal vascular assessment.

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