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Persistent Left Superior Vena Cava: A Case Report and Review of the Literature

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Abstract **Case Report**

The persistence of the left superior vena cava (LSVC) is a rare and benign congenital malformation. It is often asymptomatic, and its discovery is usually incidental in the majority of cases. This venous malformation was identified incidentally in a 60-year-old woman during thoracic multi-detector computed tomography (MDCT), which was performed with the suspicion of intra-thoracic malignancy. This is associated with abnormal venous return and an atrial septal communication.

Keywords: CT scan, Vena Cava, Vascular Malformation.

INTRODUCTION

The persistence of the left superior vena cava (LSVC) is a rare and benign congenital malformation. It is often asymptomatic, and its discovery is usually incidental in the majority of cases [1]. Here, we report the case of a patient in whom this anomaly was incidentally discovered.

PATIENT AND OBSERVATION

Kh.I, aged 60 years, with no notable medical history, was admitted due to the suspicion of intra-thoracic malignancy in the presence of a general condition deterioration. Physical examination revealed a well-nourished patient for their age. There were no chest deformities, cyanosis, or digital clubbing. No signs of heart failure were present. Cardiovascular examination was normal, with no murmurs or added sounds. Neurological examination was normal. The rest of the examination showed no abnormalities. The electrocardiogram showed a sinus rhythm without conduction or repolarization abnormalities. On thoracic MDCT (Figures 1 and 2), the right superior vena cava (RSVC) was observed as a bridging vein that drained the right jugular and right subclavian veins. The persistent left superior vena cava (PLSVC) was observed as a bridging vein connecting to the left brachiocephalic vein, descending on the left side of the mediastinum, and draining into the right atrium (RA) via a dilated coronary sinus (CS), And there was evidence of abnormal pulmonary venous return with direct drainage of a right pulmonary veins into the right SVC (Figure 3). This is associated an atrial septal communication (Figure 4). The right and left pulmonary arteries were of normal caliber and course. The pulmonary veins numbered four, converging through a collecting trunk on the left side into a left atrium of normal morphology and size. Additionally, the thoracic aorta appeared normal in its various segments. The visceral organs were normally positioned.

DISCUSSION

PLSVC is the most common congenital malformation of the thoracic venous system and it affects about 0.3 to 0.5% of the general population [2]. This incidence increases 10-fold in patients with cardiac malformations. Nearly half of the patients with isolated PLSVC have other cardiac malformations, such as atrial septal defect, endocardial cushion defect or tetralogy of Fallot [2]. The PLSVC is an anomaly of organogenesis due to the persistence of the terminal part of the left anterior cardinal vein, which normally regresses around the sixth month of intrauterine life [3]. Often, both superior vena cavae are present and frequently communicate through mediastinal anastomoses or an innominate venous trunk. Absence of the right vena cava is extremely rare [4, 5]. Apart from a congenital heart defect of varying complexity, patients with this malformation are often asymptomatic [6]. The diagnosis is usually made based on the dilation of the coronary sinus on transthoracic echocardiography or during central venous catheterization [7, 8]. In some cases, this

anomaly can cause cyanosis when the return occurs at the level of the left atrium, with the risk of paradoxical embolism [7]. The diagnosis is generally incidental. Anatomical anomalies of the inferior vena cava can be classified into three main types. Normal central venous anatomy corresponds to type I. In 10-20% of individuals with persistent left superior vena cava (LSVC), there is no right superior vena cava (RSVC), which constitutes type II [9]. However, in 80-90% of cases, a RSVC is present, leading to type III, referred to as true double superior vena cava, where LSVC coexists with RSVC [9]. In approximately 35% of these cases, a left innominate brachiocephalic vein connects these two veins, known as type IIIa. In type IIIb, this innominate vein is absent. In our case, it corresponds to type IIIa [9]. This anomaly can occur in isolation or, more commonly, is associated with a congenital heart defect. Congenital cardiac anomalies commonly associated with persistent left superior vena cava include atrial septal defects and ventricular septal defects, followed by aortic coarctation, transposition of the great arteries, tetralogy of Fallot, and abnormal connections of the pulmonary veins. In our case, the investigations revealed a venous return anomaly [9]. Partial abnormal venous return most commonly affects the right side (66%), with possible drainage into the superior vena cava, right atrium, inferior vena cava, or azygos system. Less frequently, it can occur subdiaphragmatically towards the portal trunk or suprahepatic veins. Return can also occur on the left side (33%) towards the innominate trunk, left subclavian vein, or coronary sinus [9]. On chest X-ray, a crescent-shaped shadow of the PLSVC can be seen at the aortic knob or left upper mediastinum [2]. The diagnosis can be confirmed by TTE, transoesophageal echocardiography (TEE), venous angiography, computed tomography (CT) or magnetic resonance imaging (MRI) [2].

Finally, a wide spectrum of clinicians (radiologists, sonographers, intervenists, intensivists, anaesthesiologists, cardiothoracic surgeons) should be aware of PLSVC and its variations in order to avoid possible complications.

CONCLUSION

LSVC is a rare anomaly of systemic venous return. Its diagnosis should be suspected in cases of visualization of a dilated coronary sinus on transthoracic echocardiography (ETT) and confirmed by cardiac magnetic resonance angiography (MRA).

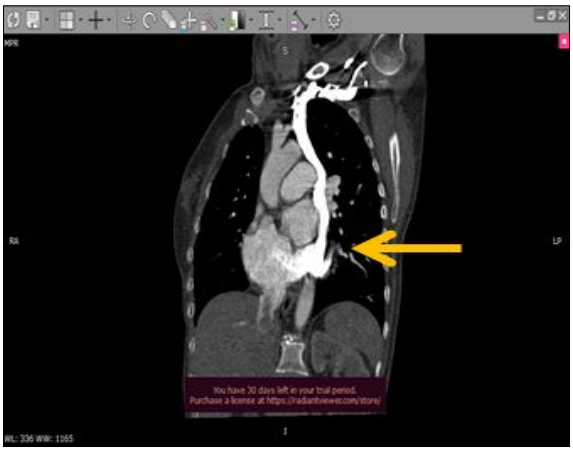

Figure 1: Thoracic CT angiography in coronal reconstruction showing the left superior vena cava with drainage into the coronary sinus

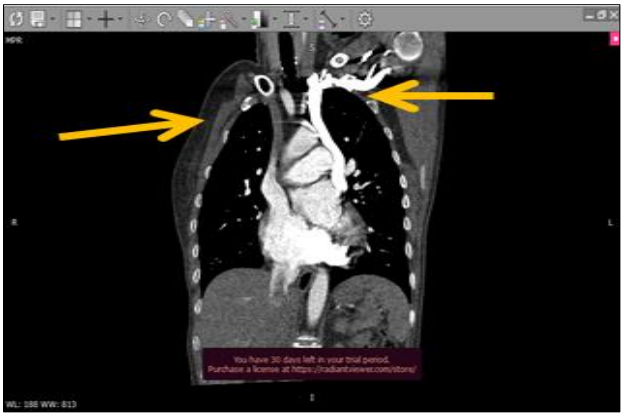

Figure 2: Thoracic CT angiography in coronal reconstruction showing the left superior vena cava and the right superior vena cava

Figure 3: Thoracic CT angiography in coronal reconstruction showing an abnormal pulmonary venous return with direct drainage of a right pulmonary vein into the right SVC

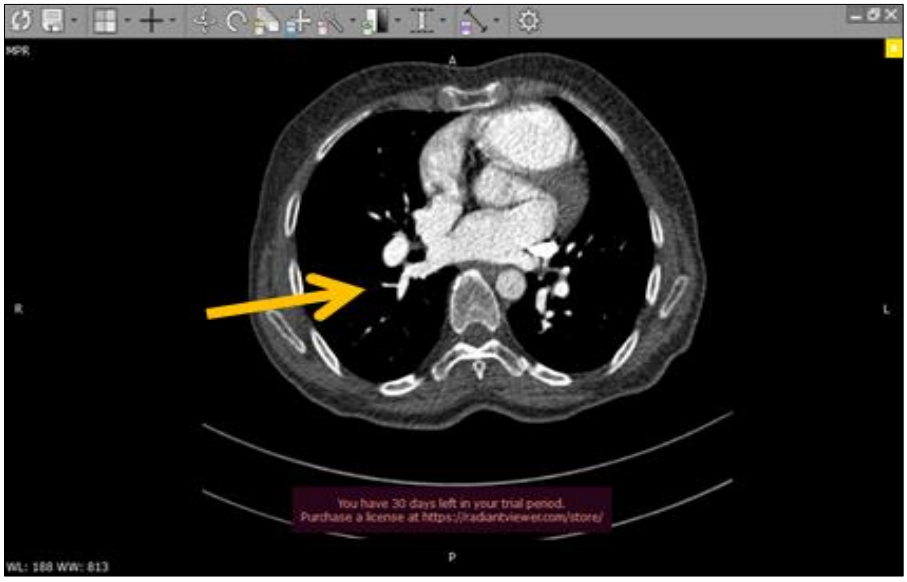

Figure 4: Thoracic CT angiography in coronal reconstruction showing the CIA.

Conflicts of Interest: The authors declare no conflicts of interest.

Author Contributions

All authors contributed to the development of this work. All authors have read and approved the final version of the manuscript.

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