Persistent left superior vena cava with an absent right superior vena cava in a 72-year-old male with multivessel coronary artery disease

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Congenital anomalies of systemic veins are usually asymptomatic and found incidentally during ultrasonography, computed tomography (CT) or magnetic resonance examinations performed for other clinical indications. Persistent left superior vena cava (PLSVC) with absent right superior vena cava (RSVC) is the congenital aberration in the thoracic venous system which occurs in only 0.09% to 0.13% of patients who have congenital heart defects. In this paper, we present the extremely rare case of a 72-year-old male with PLSVC associated with an absence of RSVC, referred for coronary CT angiography. Multidetector CT angiography is a powerful tool for the detection of venous anomalies, which is essential before invasive procedures such as the implantation of pacemakers. (Folia Morphol 2013; 71, 3: 271–273)

Key words: systemic veins, congenital anomaly, computed tomography

INTRODUCTION

Anomalies of the superior vena cava (SVC) are the result of venous system variations occurring in the early embryonic period. The presence of persistent left superior vena cava (PLSVC) associated with the absence of a right superior vena cava (RSVC) is extremely rare. Congenital anomalies of systemic veins are usually asymptomatic and found incidentally during ultrasonography, computed tomography (CT) or magnetic resonance performed for other clinical indications. The wide availability of imaging techniques has allowed more frequent identification of vena cava anomalies, which should be correctly recognised by radiologists to enable the correct planning of surgical and interventional treatments e.g. pacemaker implantation.

CASE REPORT

A 72-year-old male with a history of arterial hypertension and chronic pulmonary obstructive disease, who was refusing to undergo cardiac catheterisation, was referred by cardiologist to the Department of Radiology for coronary CT angiography because of shortness of breath on exertion. A previously performed treadmill stress test was nondiagnostic and a transthoracic echocardiogram revealed the left ventricle to have good symmetric function with an ejection fraction of 58%. Coronary CT angiography using a 64-slice scanner (Lightspeed VCT; General Electric; Wisconsin, USA) with retrospective ECG gating was performed. A smartprep series revealed the absence of the right-sided SVC and an abnormal structure with the flow of contrast media in venous phase on the left side of the aorta corresponding to that of PLSVC. Furt-
her scans confirmed this diagnosis and also showed critical stenosis in the left anterior descending artery and significant stenoses in the circumflex branch and right coronary artery. Enlarged mediastinal lymph nodes and a tumour in the 4th segment of the right lung were additional findings. Two weeks later, venography CT of the chest was performed. For better visualisation of venous system, a contrast agent was injected intravenously in both forearms. Both right and left brachiocephalic veins drained into the PLSVC, which carried blood to an enlarged coronary sinus (Figs. 1, 2). No right sided SVC was observed. The inferior vena cava terminated normally in the right atrium. In the presented case, no other anomalies of the heart or great vessels were detected.

**DISCUSSION**

Development of the SVC starts in the 4th week of gestation, when it is derived from the right common cardinal vein and the right anterior cardinal vein. The RSVC is formed by the confluence of the left and right brachiocephalic veins and drains into the right atrium, while the left superior vena cava (LSVC) disappears during development [9]. According to Nsah et al. [8] this mechanism is due to compression of the left-sided vena cava between the left atrium and the hilum of the left lung. If the left anterior cardinal vein persists, it develops into a PLSVC and usually drains into the coronary sinus via the Marshall vein [18].

Superior vena cava anomalies are relatively uncommon [15]. PLSVC is observed in 0.3% of the general population and in 3–5% of patients with congenital heart disease [3, 5]. However, in about 90% of these cases the right homologue persists [11]. Therefore, the presence of the LSVC associated with an absent right superior vena is the rarest condition, with a prevalence of 0.09–0.13% of patients who have congenital heart defects [17].

In 1850, Marshall [7] was the first to describe a PLSVC. However, it was only in 1966 that Roszel [14] detected this phenomenon in the living, by angiocardiography and by catheterisation. Contemporary, widely available imaging techniques including echocardiography, CT, and magnetic resonance imaging have enabled the diagnosis of venous anomalies more frequently, which in the absence of congenital heart disease, usually remain asymptomatic. In the presented case, the multidetector CT venogram was crucial for the diagnosis. It should be mentioned that there are 2 protocols of performing CT venogram.

Figure 1. Computed tomography, maximum intensity projection, coronal plane; Ao — aorta; CS — coronary sinus; LA — left atrium; LBCV — left brachiocephalic vein; PLSVC — persistent left superior vena cava; PT — pulmonary trunk; RBCV — right brachiocephalic vein.

Figure 2. Computed tomography, volume rendering; Ao — aorta; AV — azygos vein; CS — coronary sinus; LV — left ventricle; LJV — left jugular vein; PLSVC — persistent left superior vena cava; RBCV — right brachiocephalic vein.

The first, called a direct venogram, is based on acquisition during the first passage of a contrast agent through the veins, which requires an intravenous line to be inserted distally to the region of interest. The main disadvantage of this method results from artifacts caused by the
CONCLUSIONS
Multidetector CT angiography has proved its worth utility in depicting vascular anatomy due to its ability to visualise not only arteries but also veins, making this technique a valuable tool in the detection of venous anomalies. Appropriate identification of vascular anomalies is crucial before invasive procedures such as implantation of pacemakers. A precise knowledge of this anatomical variation is important, especially when central catheterisation via the subclavian or internal jugular vein proves difficult.

REFERENCES