

Case report - Congenital

Incidental computed tomography diagnosis of a rare triad consisting of absence of coronary sinus, persistent left superior vena cava, and scimitar syndrome

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Abstract

We report a case of an unusual congenital triad consisting of absence of coronary sinus, persistent left superior vena cava and scimitar syndrome incidentally found in a CT-scan performed on a female complaining of exertional dyspnea.

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1. Case report

A 73-year-old female presented with a five-years' history of exertional dyspnea. She had a past history of arterial hypertension and hypercholesterolemia. She did not have cyanosis. A chest radiograph showed cardiomegaly and dextroposition of the heart, right lung hypoplasia, and signs of pulmonary hypertension. Transthoracic echocardiogram showed normal left and right ventricular morphology and systolic function, mild-to-moderate tricuspid regurgitation [estimated systolic pulmonary artery pressure (sPAP) 80 mmHg], biatrial dilation, and marked dilation of the inferior vena cava (IVC) and hepatic veins (HV) with inversion of the systolic flow. An echocardiography (ECG)-gated dual-source computed tomography (DSCT) (Somatom Definition, Siemens Healthcare, Forchheim, Germany) angiography was performed to evaluate the status of pulmonary vasculature. The chest CT angiogram revealed hypoplasia of the right lung and a large vertical scimitar-like vein formed by the three right pulmonary veins draining into the IVC near the confluence of the HV, just above the diaphragm (Fig. 1a). The two left pulmonary veins drained normally into the left atrium (LA). On the left side, a left persistent superior vena cava (SVC) formed by the left subclavian vein and the left internal jugular vein was observed. This vein descended vertically into the left side of the mediastinum and drained directly into the LA [1] (Fig. 1b). Incidentally the study revealed an absence of the coronary sinus with the great cardiac vein draining in the anterolateral aspect of the LA (Fig. 2a), and the middle cardiac vein, the posterior vein and the posterolateral

marginal vein draining independently into the posterior region of the LA (Fig. 2b). The atrial situs was solitus, and both left and right appendages showed normal morphology. No interatrial septal defect was found.

Prior case reports have documented sporadic cases of scimitar syndrome with persistent left SVC [2] or total coronary vein–left atrial drainage with an associated persistent left SVC [3, 4]. This case report is unique as it gathers an unusual triad consisting of absence of coronary sinus with drainage of all of the coronary veins into the LA,

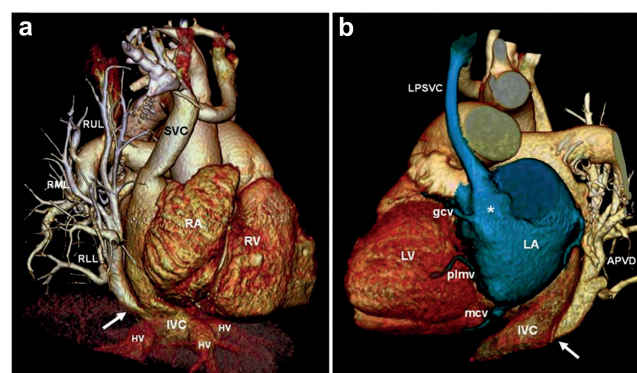


Fig. 1. Volume rendered ECG-gated DSCT angiography. (a) Right oblique view. A large vertical scimitar-like vein formed by the three right pulmonary veins draining into the inferior vena cava near the confluence of the hepatic veins is shown (arrow). (b) Left oblique view. Left persistent superior vena cava (LPSVC) draining directly (*) into the left atrium (LA) is demonstrated. Independent drainage of all the coronary veins (GCV, great cardiac vein; PLMV, posterolateral marginal vein; MCV, middle cardiac vein) into the left atrium is clearly seen. Note confluence (arrow) of the right anomalous pulmonary venous drainage (APVD) and inferior vena cava (IVC) on the right side. RUL, right upper lobe pulmonary vein; RML, right middle lobe pulmonary vein; RLL, right lower lobe pulmonary vein; SVC, superior vena cava; HV, hepatic veins; RA, right atrium; RV, right ventricle; LV, left ventricle.

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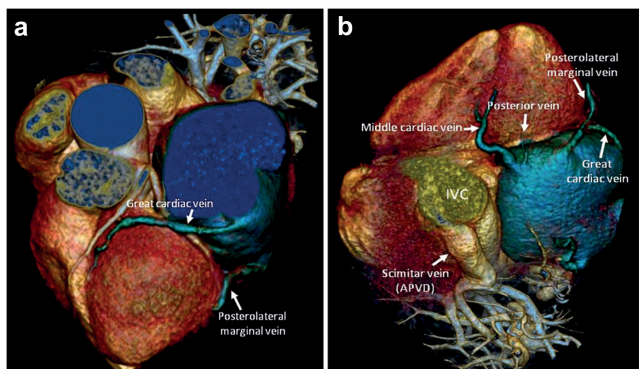
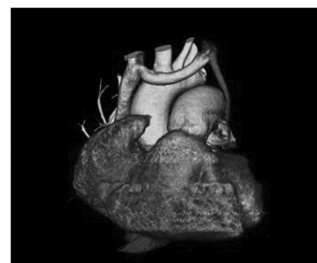


Fig. 2. Volume rendered ECG-gated DSCT angiography. (a) Anterior-cranial view. The cardiac study revealed absence of the coronary sinus with the great cardiac vein and the posterolateral marginal vein draining independently in the anterolateral and posterolateral aspect of the left atrium, respectively. (b) Inferior view. Note direct drainage of the middle cardiac vein, the posterior vein and the posterolateral marginal vein into the posterior region of the left atrium. This view showed nicely the convergence of the scimitar vein (right anomalous pulmonary venous drainage, APVD) and inferior vena cava (IVC).

persistent left SVC and scimitar syndrome (Video 1). Moreover, the case presented here emphasizes the usefulness of CT to assess complex congenital heart diseases, often confirming and completing information available from echocardiography and not infrequently demonstrating anatomical malformations missed by the latter.



Video 1. Volume rendered ECG-gated DSCT angiography. The absence of coronary sinus with drainage of all of the coronary veins into the left atrium, persistent left superior vena cava (in blue) and right anomalous pulmonary venous drainage is shown.

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