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

Gorka Bastarrika⁎, Isabel Simón-Yarza, Juan J. Gavira

⁎Department of Radiology, Clínica Universidad de Navarra, Avenida Pío XII, 36, 31008 Pamplona, Spain

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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 (ECC)-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 hepatic veins (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,
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 misses by the latter.

References