Rev Port Cardiol. 2013;32(1):63-66

Revista Portuguesa de Cardiologia
Portuguese Journal of Cardiology
www.revportcardiol.org

CASE REPORT

Right-sided scimitar syndrome in a patient with a single aortic trunk and coronary-cameral venous fistula

Mehmet Cilingiroglu[a], **Nuri Ilker Akkus**[b,*]

[a] *University of Pittsburgh Medical Center, Heart and Vascular Institute, Pittsburgh, United States*
[b] *LSU Shreveport Health Sciences Center, Shreveport, LA, United States*

Received 2 April 2012; accepted 9 May 2012

KEYWORDS
Scimitar syndrome;
Abnormal pulmonary venous return;
Coronary venous fistula

Abstract Scimitar syndrome (SS) is a rare congenital anomaly characterized by partial or complete anomalous pulmonary venous drainage of the right or left lung into the inferior vena cava. The syndrome is commonly associated with hypoplasia of the right lung, pulmonary sequestration, persistent left superior vena cava, and dextroposition of the heart. We report a rare variant of SS in a 44-year-old man together with a single aortic trunk, as well as a coronary-cameral venous fistula.

PALAVRAS-CHAVE
Síndrome de Cimitarra;
Drenagem pulmonar anómala;
Fístula coronária

Síndrome de Cimitarra direita num doente com *truncus arteriosus* e fístula venosa coronário-ventricular

Resumo A síndrome de Cimitarra (SC) é uma malformação congénita rara, caracterizada por uma drenagem pulmonar anómala parcial ou total do pulmão direito ou esquerdo, na veia cava inferior. Esta síndrome é frequentemente associada a hipoplasia do pulmão direito, sequestração pulmonar, persistência de veia cava superior esquerda e dextroposição do coração. Reportamos uma variante rara de SC num homem de 44 anos com tronco supra-aórtico único e fístula coronário-auricular.

Introduction

Scimitar syndrome (SS) is a rare and complex congenital anomaly, characterized by partial or complete anomalous pulmonary venous drainage from the right or left lung into the inferior vena cava (IVC), though drainage into the hepatic vein, right atrium (RA) or left atrium (LA), or the

* Corresponding author.
E-mail address: iakkus@hotmail.com (N.I. Akkus).

portal vein can also occur. According to the original definition, the syndrome is commonly associated with hypoplasia of the right lung, pulmonary sequestration, persistent left superior vena cava, and dextroposition of the heart.[1,2] In two-thirds of cases, the scimitar vein provides drainage for the entire right lung, but in one third, this vein drains only the lower portion of the right lung.[3] Partial abnormalities of pulmonary venous return (PAPVR) are seen in 0.4–0.7% of adult autopsies, while patients with SS account for 3–5% of these cases.[4,5] SS constitutes 0.5–1% of all congenital heart defects. This rare anomaly has an incidence of approximately 1-3 per 100 000 live births; however, the true incidence may be higher because many patients remain asymptomatic.[6] The etiology is not completely understood. In several patients with total anomalous pulmonary venous return, the gene locus has been mapped to chromosome 4q12,[7] and females are more frequently affected than males. The scimitar sign refers to the crescent (resembling the Turkish sword) described by the descent of the anomalous pulmonary vein (the tip of the crescent points inferiorly and medially to the diaphragm/right heart border junction). The concavity of the crescent is adjacent to the junction of the diaphragm and right heart border.

Case report

A 44-year-old man presented with several weeks of palpitations. He also described symptoms of dyspnea with mild exertion and substernal chest discomfort at rest. He had a history of long-standing systemic hypertension. The 12-lead ECG showed atrial fibrillation with rapid ventricular rate. His 2D Doppler echocardiogram revealed a mildly enlarged RA and right ventricle (RV). He subsequently underwent right and left heart catheterization with selective coronary angiography. Right and left heart catheterization with a full oximetry run to calculate shunts revealed Qp/Qs of 1.3. Selective coronary angiography was performed using right radial artery access, and showed no significant coronary disease. However, angiography of the aortic arch showed a single trunk takeoff for the large vessels from the aortic arch (Figure 1). Using right femoral vein access, a 5-Fr multipurpose diagnostic catheter was advanced into the upper (Figure 2) and middle (Figure 3) right pulmonary veins as they opened into the superior vena cava (SVC). To exclude any possible associated atrial septal defect, an MP-1 catheter was engaged into what proved to be a coronary-cameral fistula (CCF) opening into the RA separately from the coronary sinus (Figure 4), the CCF went from the coronary vein to the RA and the coronary sinus was also filled with contrast retrogradely from the vein (Figure 4). Pulmonary angiography using a 5-Fr pigtail catheter showed a moderately dilated pulmonary trunk (Figure 5). Following consultations with the cardiothoracic surgery and pediatric cardiology teams, it was felt that the best course of management would be to follow the patient clinically with serial echocardiography, as there was no significant right-to-left shunt.

Discussion

SS is a complex form of PAPVR, which is a connection failure between the right pulmonary veins and the LA during fetal development. Variations in PAPVR include the right pulmonary veins draining into the SVC-LA junction, RA, or IVC, or as in our case, separately to a high SVC. In SS, an anomalous right pulmonary vein generally draining the entire right lung but occasionally the middle and superior lobes, may descend in a cephalad-to-caudal direction toward the diaphragm with a crescent (scimitar) shape. This vein then curves sharply to the left just above or below the IVC-RA junction.[8] The anomalous right pulmonary venous trunk usually courses anterior to the hilum of the right lung and connects to the IVC just superior to the orifices of the

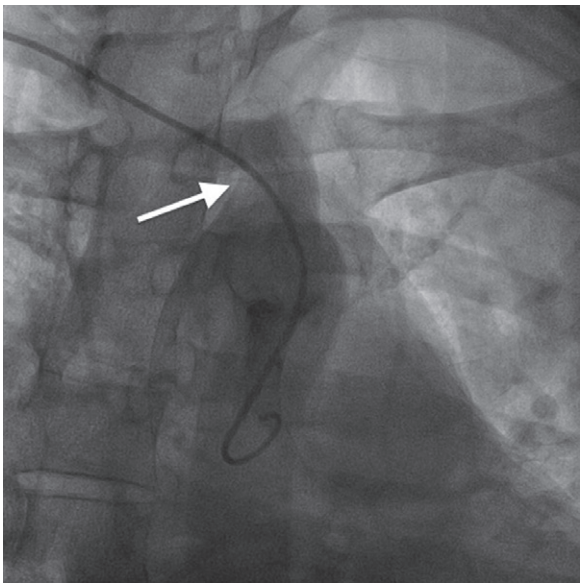

Figure 1 Aortic arch angiography. Takeoff of large vessels from the aortic arch via a single trunk (white arrow).

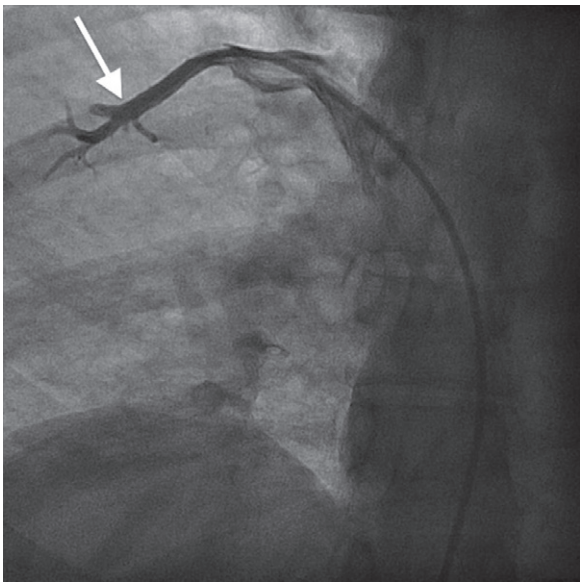

Figure 2 Right upper pulmonary venous angiography. Abnormal pulmonary venous drainage from the upper lobe of the right lung draining into the high SVC via the right upper pulmonary vein (white arrow).