Right superior vena cava draining into the left atrium, associated with abnormal pulmonary vein drainage

Drainage de la veine cave supérieure droite dans l’oreillette gauche associée à une anomalie du retour veineux pulmonaire

Alexis Théron, Philippe Paule, Dominique Métras

Service de chirurgie cardiaque adulte, hôpital La Timone, 264, rue Saint-Pierre, 13385 Marseille cedex 5, France
Service de cardiologie, hôpital d’instruction des armées Laveran, 13013 Marseille, France

A 20-year-old woman was hospitalized for recent onset of dyspnoea with marked oxygen desaturation (80%). Her medical history included minor thalassaemia and unexplored digital cyanosis. The transthoracic echocardiogram at admission was considered normal (no atrial septal defect). A computed tomography scan of the chest was performed to rule out pulmonary embolism; unexpectedly, the scan revealed abnormal direct drainage of the superior vena cava (SVC) into the left atrium and of the right superior pulmonary vein (RSPV) into the SVC, resulting in a right to left shunt. Magnetic resonance imaging (MRI) confirmed the anatomy with dynamic imaging of the blood flow (Appendix A, Fig. 1). The complications of this very rarely described congenital anomaly (only nine cases in the literature) — usually reported in infants and children — are related to a direct connection between systemic venous return and the left cardiac chambers; besides hypoxaemia, complications include systemic emboli and brain abscesses (3 cases: 2 in childhood and 1 in adulthood). These serious potential complications justify prompt surgical treatment, even when the functional consequences are limited.

In this patient, surgical correction revealed a fibrotic structure (Fig. 2) on the anterior wall of the SVC corresponding to the right SVC orifice involution. Repair comprised transection of the right SVC proximal to its connection with the right superior pulmonary vein.
Anomalie du retour veineux pulmonaire ;
Imagerie par résonance magnétique cardiaque ;
Abcès cérébral ;
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Figure 1. MRI cine in gradient echoes acquisition (SSFP). MRI 1,5 Tesla, echospeed Gems. Abnormal drainage of the right superior vena cava into the left atrium (LA) with abnormal drainage of right superior pulmonary venous return. LV, left ventricle; white arrow, right superior pulmonary vein; dashed arrow, superior vena cava.

Figure 2. Photograph taken after right posterolateral thoracotomy and before repair; note the fibrotic structure on the anterior aspect of the SVC, which corresponds to the involution of the right SVC orifice (arrow) (Ao: aorta; RA: right atrium).

and in connection with the right atrial appendage. The patient’s postoperative course was uneventful, with immediate resolution of dyspnoea and cyanosis.

This case is a typical clinical presentation of systemic venous anomalies that induce the physician to look for this disease before complications supervene. Cardiac MRI allows accurate diagnosis, and avoids the use of radiation and iodine perfusion.

Conflicts of Interest

None.

Appendix A. Supplementary data

Supplementary data associated with this article can be found in the online version at doi:10.1016/j.acvd.2008.11.004.