Voluminous coronary fistula between the left coronary artery and the right atrium

Une fistule coronaire volumineuse entre l’artère coronaire gauche et l’oreillette droite

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We report the case of a 4-year-old girl for whom a CAF between the left coronary artery and the right atrium (Fig. 1A, Video 1) was diagnosed at the age of 7 months after echocardiographic exploration of a continuous cardiac murmur. As the patient was asymptomatic, the CAF was not initially closed. Because both the left coronary artery (Fig. 1B) and the left ventricle were dilated in the last echocardiograms (10 mm for left coronary artery diameter; 42 mm for left ventricular end-diastolic diameter), percutaneous closure was decided upon. Electrocardiography was normal. Computed tomography revealed a huge CAF (12 mm maximal diameter) originating from the left coronary artery, passing along the left atrial roof (Fig. 1C) and draining into the roof of the right atrium (Fig. 1D). Transcatheter closure was performed using an anterograde approach. Angiographies accurately showed the horizontal course of this voluminous CAF (Fig. 2A and 2B, Video 2). A vascular plug (diameter, 4 mm) was then deployed in the narrowest portion (diameter, 2 mm) to close the CAF (Fig. 2C). Complete occlusion was confirmed by postinterventional echocardiography (Fig. 2D). To prevent extensive coronary artery thrombus formation, antiplatelet therapy was prescribed for 6 months. The left coronary artery and left ventricle remained dilated a few weeks after the closure before recovering normal size.

Abbreviations: CAF, Coronary Artery Fistula.
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Figure 1. Echocardiographic views showing (A) a markedly dilated left coronary artery (white arrow) and (B) a huge fistula draining into the right atrium. (C) Computed tomography scan showing the fistula (hollow arrow) passing along the left atrial roof and (D) the fistula (hollow arrow) communicating with the right atrium. Ao: aorta; LA: left atrium; LAA: left atrial appendage; LV: left ventricle; RA: right atrium; RV: right ventricle.

Figure 2. Selective angiographies showing (A) the voluminous tortuous coronary fistula originating from the left coronary artery and draining into the right atrium, (B) its horizontal course along the left atrial roof and (C) the successful deployment of the device (white arrow), allowing complete occlusion. (D) Postinterventional echocardiographic view showing the absence of any residual shunt. Ao: aorta; F: fistula; LAD: left anterior descending coronary artery; RA: right atrium; RCA: right coronary artery; SVC: superior vena cava.
CAF is a rare congenital anomaly defined as a fistulous communication between a major coronary artery and a cardiac chamber or thoracic vessel. CAF is usually fortuitously diagnosed in adult patients. When CAF is responsible for a high-flow shunt, children may present congestive heart failure. Spontaneous regression of CAF is possible during childhood but rarely occurs after 2 years of age. Closure of CAF, either by surgery or a transcatheter procedure, is currently indicated for symptomatic patients or children with a high-flow shunt.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

Appendix A. Supplementary material