Clinical vignette

Multimodality imaging of circumflex artery fistula to coronary sinus with persistent left-sided superior vena cava

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A 26-year-old man presented with chest pain. Examination and electrocardiogram were normal but cardiac enzymes were elevated (CK 530 U/L, Troponin I 5.3 μg/L). Cardiac catheterization revealed an aneurysmal left circumflex artery (LCX) fistula draining into coronary sinus with a persistent left superior vena cava (PLSVC), but no significant coronary stenoses (Panel A). Oxygen saturation increased from mixed venous blood (87%) to right ventricle (92%) with a calculated shunt of 2 : 1. Cardiac MRI confirmed bilateral SVC with left brachiocephalic vein communication (Panel B). Delayed contrast enhancement and T2-weighted spin echo hyperintensity was present in the basal infero-lateral segment, consistent with myocardial injury. Echocardiography with saline contrast differentiated coronary sinus from LCX (Panel C). Colour Doppler documented the site of fistulous drainage into coronary sinus (Panel C/D). The patient was referred for fistula closure.

PLSVC occurs in <0.5% of the population. Coronary artery fistulas (CAF) are rare vascular abnormalities that usually arise in the right coronary or left anterior descending artery; the LCX is rarely involved. Most CAF manifest as a continuous murmur or complications including heart failure, myocardial ischaemia, arrhythmia, or endocarditis. Associated cardiac abnormalities include ventricular septal defects. To the authors' knowledge, this combination of findings has not been described. Multimodality imaging is paramount in the evaluation of coronary anatomy, documentation of atherosclerosis, haemodynamic assessment, assessment of fistula origin, course and drainage, ventricular function and shunt quantification, thereby enabling informed management decisions with regards to percutaneous versus open closure techniques.

See online supplementary material for a colour version of this figure available at European Heart Journal online.

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