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Double Coronary-Cameral Fistula Draining to the Right Ventricle in a Patient with Mitral Stenosis: is it Clinically Relevant?

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A 39-year-old female patient was admitted to the cardiology outpatient clinic with complaints of dyspnea on exertion and fatigue that had been present for the last 3-4 months. History assessment revealed that the patient had an episode of acute rheumatic fever at the age of 10 years, but was lost to follow-up since then. Within the last 6 months, she had been hospitalized several times for symptoms of heart failure secondary to moderate mitral valve stenosis. Transthoracic echocardiography (TTE) revealed an existing moderate rheumatic mitral valve stenosis (mitral valve area, 1.2 cm^2). However, mitral balloon valvuloplasty was not considered an appropriate option because of the relatively high Wilkins score (9 points). Accordingly, the cardiovascular team decided mitral valve replacement (MVR). Thereafter, the patient underwent coronary angiogram (CAG) as part of the preoperative examination. On CAG, coronary-cameral fistulas originating from the septal perforating branch of the left anterior descending (LAD) artery and conus branch of the right coronary artery (RCA) were detected (Figure 1a and b, Video 1). Repeat TTE demonstrated a continuous jet flow draining into the right ventricular (RV) cavity (Figure 1e, Video 1). A fistulous connection was also found to originate from both the LAD and RCA and drain into the RV cavity on computed tomography (Figure 1c and d). However, surgical closure of the fistulas was not considered because of their relatively small diameters. Following MVR, clinical improvement, manifesting as an increase in functional capacity, was observed on follow-ups 1, 3, and 6 months after surgery.

Congenital coronary artery anomalies appear to be very rarely encountered in clinical practice.[1] In particular, coronary-cameral fistula has been a very uncommon coronary anomaly.[2] More interestingly, the evolution of acquired valvular heart disease in congenital cardiac anomalies is even rarer in the literature.[3,4] In this context, our case might be regarded as an epitome of such rare cases

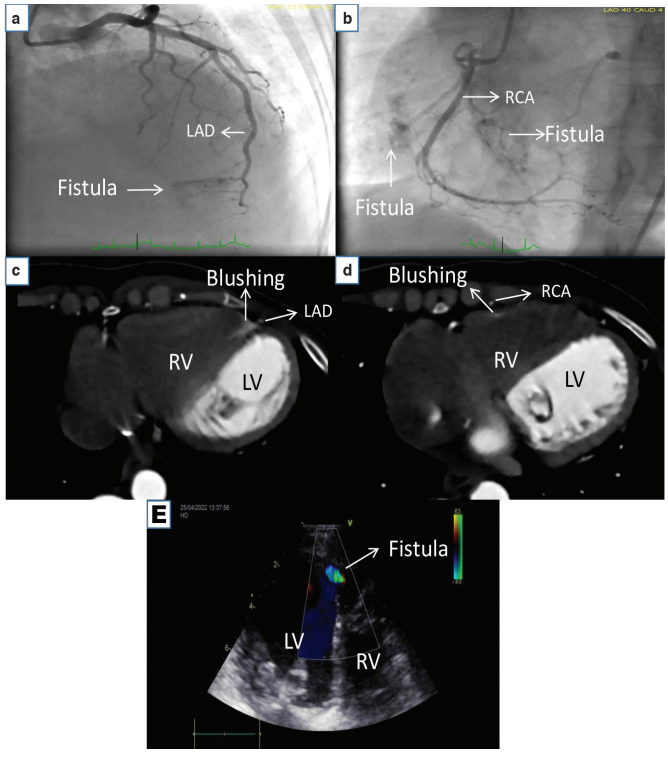

FIG. 1. (a) Coronary-cameral fistula originating from the septal perforating branch of the left anterior descending (LAD) artery on coronary angiogram. (b) Coronary-cameral fistula originating from the conus branch of the right coronary artery (RCA) on coronary angiogram. (c) Fistulous connection between the LAD and right ventricular (RV) cavity on computed tomography. (d) Fistulous connection between the RCA and RV cavity on computed tomography. (e) A jet flow toward the RV cavity on transthoracic echocardiography.

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with important clinical implications. Clinically, the presence of multiple fistulas between the coronary arteries and RV cavity might have elicited a high flow state in the pulmonary circulation, leading to early symptom onset in our patient with a moderate rheumatic mitral stenosis. Importantly, the origin, drainage site, size, number, and flow rate of coronary-cameral fistulas might be important determinants of emerging symptomatology and its severity.[5] Note that disproportionately severe symptoms in mild-moderate valvular heart disease might be attributable to the relative contribution of co-existing abnormalities including coronary-cameral fistulas. Therefore, a high index of suspicion is necessary to uncover these subtle abnormalities on imaging modalities including TTE.

Informed Consent: Written informed consent was obtained from the patient.

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Video 1. Images of coronary-cameral fistula on coronary angiogram and transthoracic echocardiography.
10.4274/balkanmedj.galenos.2023.2023-7-28.video1

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