

A Rare Coronary Anomaly in a Patient with Pulmoner Hypertension Secondary to Scleroderma: Multiple Coronary Cameral Fistula



Sklerodermaya Bağlı Pulmoner Hipertansiyonlu Bir Hastada Nadir Koroner Bir Anomali: Multipl Koroner Kameral Fistül

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A 63-year-old female with a history of scleroderma was referred to our institution for the investigation of dyspnea and atypical chest pain. On physical examination, her blood pressure was 132/88 mmHg and pulse rate was 116 beats/min. She was cyanotic and had an arterial oxygen saturation level of 85%. On cardiac examination, a left parasternal heave was noted; there were soft S1 and hard S2 with a grade III/VI holosystolic murmur heard over the left sternal border, which increased on inspiration. A 12-lead electrocardiogram demonstrated sinus tachycardia and right bundle branch block. Echocardiography showed a normally functioning left ventricle and enlarged right atrium, right ventricle and D-shaped septum (Figure 1). There was severe tricuspid regurgitation, and the estimated pulmonary artery systolic pressure was 106 mmHg. The patient was hospitalised for medical stabilisation. Furthermore, cardiac catheterisation revealed no atherosclerotic coronary artery disease; however, it incidentally revealed diffuse coronary cameral fistulae (CCF) involving the circumflex-obtuse marginal system and diagonal branches emptying into the left ventricle (Figure 2), delineating the endocardial border very well (Figures 3, 4). Because of the fistulae were diffuse and many smalls, an intervention was not considered. CCF are rare congenital or acquired anomalous with abnormal vascular communications between the coronary arteries and cardiac chamber. A vast majority (approximately 90%) of CCF communicates with the right-sided chambers of the heart, and in the remaining cases, they drain to the left side of the heart or to both sides⁽¹⁾. The pathophysiology of CCF is unclear. Rudraiah et al. suggested that angiogenesis secondary to hypoxia may play a role in the development of CCF in the absence of acquired causes (e.g. exogenous trauma, previous invasive cardiac procedures or atherosclerosis)⁽²⁾.

In this respect, hypoxia occurring in the setting of scleroderma may contribute to the development of CCF in the present case.

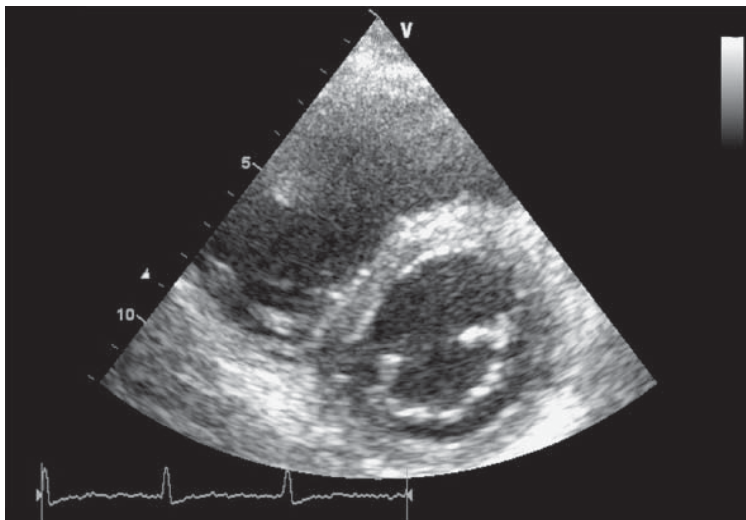


Figure 1. Parasternal short-axis view showing a D-shaped inter-ventricular septum and an enlarged right ventricle.

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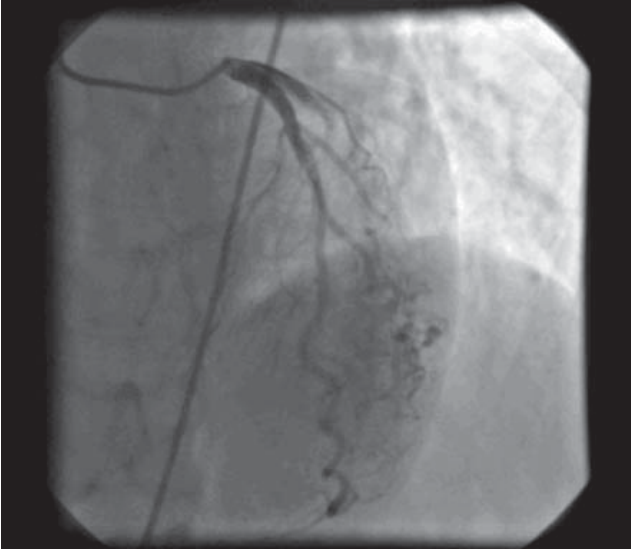


Figure 2. Coronary angiography shows the fistulae from the diagonal branches of the left anterior descending artery to the left ventricle.

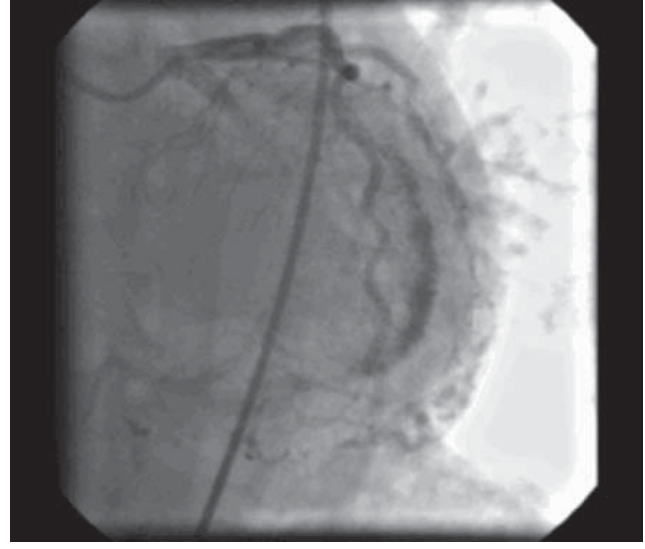


Figure 4. Coronary angiogram demonstrates the opacification of the left ventricle endocardial border.

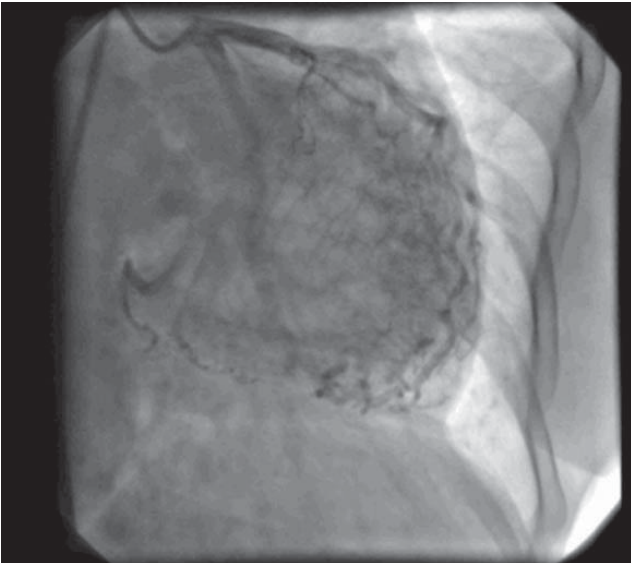


Figure 3. Coronary angiography reveals the diffuse fistulae from the left coronary system to the left ventricle.

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