COMUNICACIONES BREVES

Multiple congenital coronary artery fistulae draining into the left ventricle

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Summary

Coronary artery fistula between a coronary artery and a cardiac chamber is a rare condition, especially when multiple fistulas communicate with the left ventricle. Herein we report a case of an elderly woman with multiple diffuse coronary artery-left ventricular fistulas diagnosed by angiography. Since the coronary artery-cardiac chamber communications were multiple and diffuse neither surgery nor transcatheter coil occlusion was considered in this case.

Resumen

FÍSTULAS CORONARIAS CONGÉNITAS MÚLTIPLES DRENANDO EL VENTRÍCULO IZQUIERDO

Las fístulas de las arterias coronarias que drenan a las cavidades cardíacas son una anomalía infrecuente, especialmente cuando son múltiples y drenan hacia el ventrículo izquierdo. Presentamos el caso de una mujer octogenaria con múltiples fístulas difusas que se originan de la coronaria izquierda y que drenan al ventrículo izquierdo. El hecho de que fuesen múltiples y difusas imposibilitó una intervención quirúrgica o percutánea como se recomienda en estos casos.
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Key words: Multiple coronary artery fistulae. Congenital heart disease. Diagnose by angiography.
Palabras clave: Fístulas múltiples de arterias coronarias. Cardiopatía congénita. Diagnóstico angiocardiográfico.

Introduction

Coronary artery fistulas between a coronary artery and a cardiac chamber constitute a rare finding in about 0.2% of patients undergoing cardiac catheterization.[1-3] In more than 90% of cases, a single fistula drains into the right heart chambers or into the pulmonary artery resulting in a left to right shunt.[2] Single or multiple fistulas draining to left heart chambers is a rather rare condition.[2-6]

Herein, we report a patient with angina in whom cardiac catheterization showed multiple diffuse fistulas from left anterior descending artery and left circumflex artery draining into left ventricle chamber. Eventually, a striking pattern of complete filling of this chamber was visualized.

Case report

A 78 year-old woman was admitted to our institution due to 2-months history of typical an-

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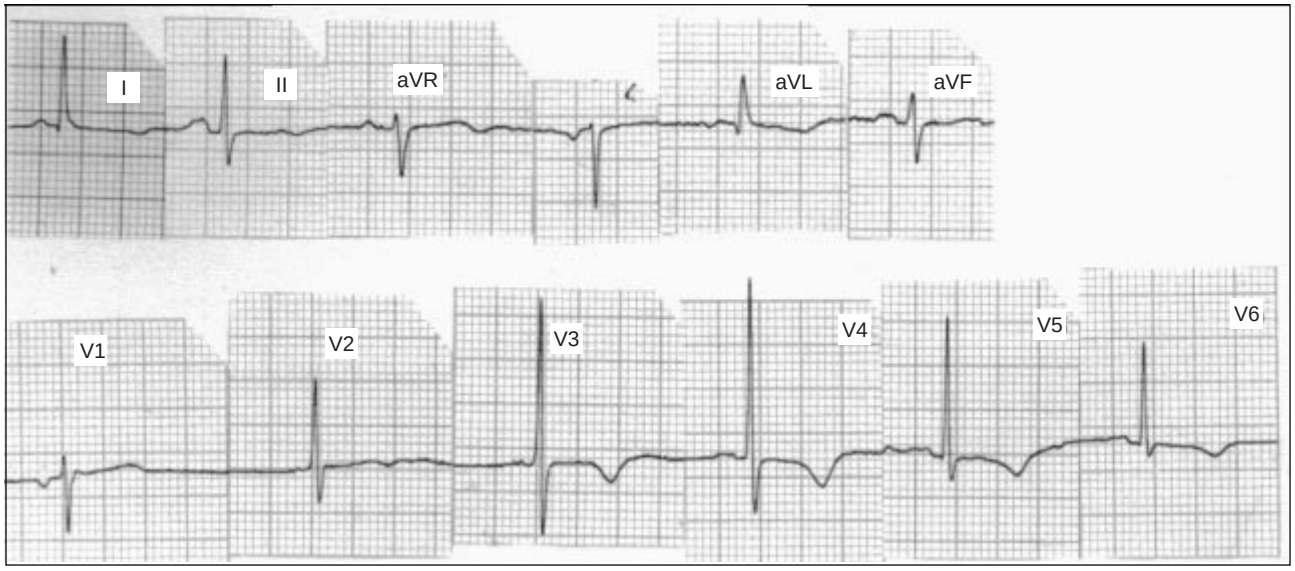

Fig. 1. Surface 12-leads electrocardiogram showing sinusal rhythm, $AQRS_F$ +4°, PR 165 msec, T-axis 125°, PR 157 msec, QRS 82 msec, QTc 394 msec, intrinsecoid deflection time inscription in V6 38 msec. Negative T-waves on leads I, aVL, V2-V6 are evident.

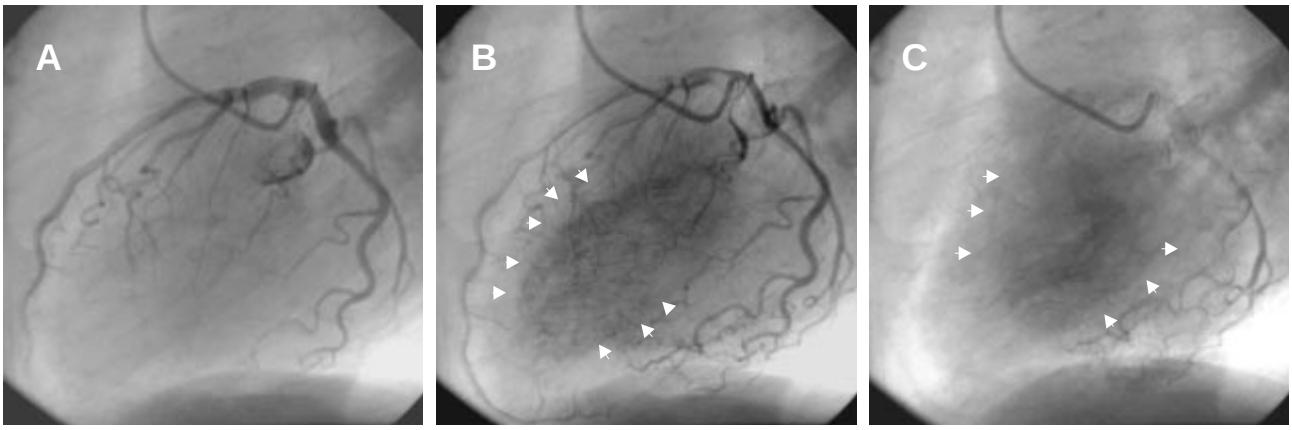

Fig. 2. Coronary angiogram of the left coronary artery in left lateral view showing normal epicardial vessel (panel A), the sequence demonstrates multiple fine fistulas draining to the cavity of the left ventricle, as result, a striking pattern of complete filling of this chamber is visualized (arrow heads in panel B and C). Fistulas emerge from the proximal, mild and distal segments of the left anterior descending and left circumflex artery.

gina both at rest and on exertion. She had a 10 years history of arterial hypertension and hypercholesterolemia. Physical examination revealed normal first and second heart sound without murmurs. Her blood pressure was 130/80 mm Hg. No signs of heart failure were observed. The electrocardiogram showed sinus rhythm, 65 beats per minute, with $AQRS_F$ +4º, PR 165 msec and negative T-waves in leads I, aVL and V2-V6 *(Fig. 1)*. Chest *x*-ray appeared normal. A transthoracic echocardiogram showed normal left ventricular wall motion with normal wall thickness.

A perfusion Thallium-201 stress scintigraphy with adenosine was also normal and no pain developed during maximal hyperaemia. Because of persistent angina despite treatment with aspirin, nitrates, calcium antagonist (amlodipine, 5 mg, PO qd), and beta blocker (propranolol, 20 mg, pO tid), heart catheterization and coronary angiogram was performed. Left ventricular angiography revealed a normal sized left ventricle without regional wall motion abnormalities, with an estimated ejection fraction of 70% with and end diastolic pressure of 8 mm Hg. Selective arteriography of the left coronary showed normal epicardial vessels, but multi-

ple fine fistulas were revealed in the left ventricle. These fistulas were emerging from the proximal, mid and distal segments of the left anterior descending and left circumflex artery *(Fig. 2)*. The dominant right coronary artery was normal without coronary fistulas. Since the coronary-cardiac chamber communications were multiple and diffuse neither surgery nor transcatheter coil occlusion were considered in this patient as it has been recommend in some cases.[6,7] The patient was discharged under beta-blocker (metoprolol, 95 mg PO bid) but nitrates were discontinued obtaining adequate control of symptoms and heart rate at rest of 56 beats per minute; a new electrocardiogram performed 7 days later showed negative T waves only in leads I, avL and V4-V6. A new stress test was not performed.

Discussion

Multiple coronary artery-left ventricle fistulas are uncommon anomalies, although it represents one of the most common major congenital malformations of the coronary circulation compatible with adult survival.[3] Other different causes of fistulas include chest trauma, myocardial infarction, surgery and endomyocardial biopsy.

The embryogenic origin of the malformation is obscure. With regard to the development and maturation of the normal coronary arterial system, it begins with migration of the epicardium from the proepicardial organ with an important intervention of the adhesion molecules. The migrating epithelial sheet interacts with the underlying myocardium to cooperatively create a subepicardial matrix, which is subsequently invaded by at least three eventual subpopulations of cells from the advancing epicardium. Morphological investigations suggest a partial or abnormal persistence of embryonic myocardial sinusoids that arise from endothelial protrusions into the intertrabecular spaces.[5-8] The development of abnormal connections between the coronary arterial network and its neighbouring structures results in abnormal communications that may resemble fistulas. Fetal regression of these structures results in the formation of the Thebesian vessels of the adult heart.[4,9] On the other hand, an early phenomenon during heart and blood vessels development has been recently described in which the epicardium and endocardium are in close relationship through the myocardium.[10] Under abnormal circumstances these counterpoints could persist originating fistulas between heart chambers and coronary vessels.[10,11]

Only half of the patients with large fistulas remain asymptomatic. The remaining 50% develop complications, such as congestive heart failure, infective endocarditis, rupture of an aneurysmatic fistula, or myocardial ischemia.

Some patients with this anomaly present typical or atypical angina during their adult life.[2] The clinical syndrome has been attributed in part to a coronary steal phenomenon,[2,5,12] although, ischemic responses during exercise stress testing and thallium scintigraphy have not been unanimously detected in previous cases, including the present one.[2,13] A unique finding in our patient was the striking pattern of complete filling of the left ventricle from the left coronary artery.

Finally, in some patients, a large fistula can cause a considerable shunt volume draining into the left ventricle that might result in diastolic overload mimicking that of aortic regurgitation.[14,15]

Conclusions

Coronary artery-cardiac chamber fistula is unusual. Physicians should be aware of this entity in the differential diagnosis of patients with angina pectoris and consider that in some cases stress test are negative.

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