

Coronary-cavitary Fistula with Aneurysm in its Path: Case Report of Percutaneous Treatment

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Abstract

The authors report the case of a 29-year-old, female patient with palpitations, chest pain, and dyspnea on exertion. The patient was diagnosed with right coronary fistula and underwent percutaneous correction of the fistulous tract with an Amplatzer prosthesis.

Introduction

Coronary artery fistula is a clinical condition characterized by an abnormal connection between one or more coronary arteries and heart chambers or large vessels adjacent to the heart. It has a prevalence of around 0.002% in the population, equally in both sexes, and it accounts for 14% of all coronary artery anomalies.¹⁻³

Approximately 55% of cases of fistulas originate from the right coronary artery (or its branches) and 35% from the left coronary artery. In some cases, both arteries are involved. Most fistulas drain into the right ventricle, right atrium, and coronary sinus. Other drainage sites include the pulmonary artery, left atrium, left ventricle, and superior vena cava.⁴

The clinical picture varies according to the location and size of the fistula. Smaller fistulas usually occur without clinical repercussions, with the possibility of conservative treatment. On the other hand, large fistulas can generate a coronary steal mechanism, which causes functional myocardial ischemia and consequent angina or dyspnea on exertion. In these cases, invasive treatment with surgery or percutaneous closure of the fistula should be considered.³

Case report

A 29-year-old female patient, who was an active smoker with no previous comorbidities, came to the consultation complaining of chest pain and dyspnea on moderate

exertion for a long time. She had begun follow-up with a cardiologist 7 years prior, when she presented with palpitations, which frequently persisted until she arrived at the reference service.

Upon physical examination, she was normotensive, in regular rhythm, well perfused, without edemas or jugular swelling; lung auscultation revealed no alterations, and cardiac auscultation revealed continuous murmur (+3/+6) in the right hemithorax.

There were no significant alterations in laboratory tests. Chest X-ray showed evidence of increased cardiac area. Admission electrocardiogram revealed atrial flutter rhythm. Transthoracic echocardiogram showed preserved ejection fraction (62% by Simpson's method), paradoxical movement of the interventricular septum (diastolic dysfunction suggestive of volume overload), preserved contractility in the other segments of the left ventricle, presence of pulmonary hypertension (pulmonary artery systolic pressure = 40 mmHg, estimated by tricuspid regurgitation), and heart valves without abnormalities. Dilation of the right coronary artery was observed, beginning in the right sinus of Valsalva (Figure 1A), with a very tortuous fistulous tract and the formation of a large aneurysm in the distal portion, draining into the right atrium, close to the inferior vena cava (Figure 1B), with systo-diastolic flow and velocity of approximately 1.15 m/s (Figure 1C). The aneurysmal portion of the right coronary artery measured 73 × 85 mm in its largest diameters, with an area of 46 cm² and marked slowing of the flow inside (spontaneous contrast), exerting compression on the right atrium (Figure 1D). There was presence of reverse holodiastolic flow in the descending thoracic aorta (secondary to the right coronary fistula to the right atrium).

The transthoracic echocardiogram findings were confirmed by chest angiography, and the patient was diagnosed with coronary-cavitary fistula with an aneurysm in its tract (fistula from the right coronary artery to the right atrium).

The patient was referred for cardiac catheterization, which did not show evidence of obstructive coronary lesions; however, it showed significant slowing of the distal flow of the right coronary artery, secondary to coronary steal of proximal flow through the fistulous tract to the aneurysmal portion (Figure 2A and 2B), which drained into the right atrium.

The decision was made to occlude the fistulous tract by means of a percutaneous procedure. Closing was performed with an 18 mm Amplatzer Vascular Plug II prosthesis, implanted shortly after the emergence of the fistulous tract,

Keywords

Cardiovascular Malformation; coronary vessel anomalies; vascular fistula; coronary aneurysm

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Manuscript received November 16, 2022; revised manuscript November 30, 2022; accepted December 1, 2022.

Editor responsible for the review: Simone Nascimento dos Santos

DOI: <https://doi.org/10.36660/abcimg.2022364i>

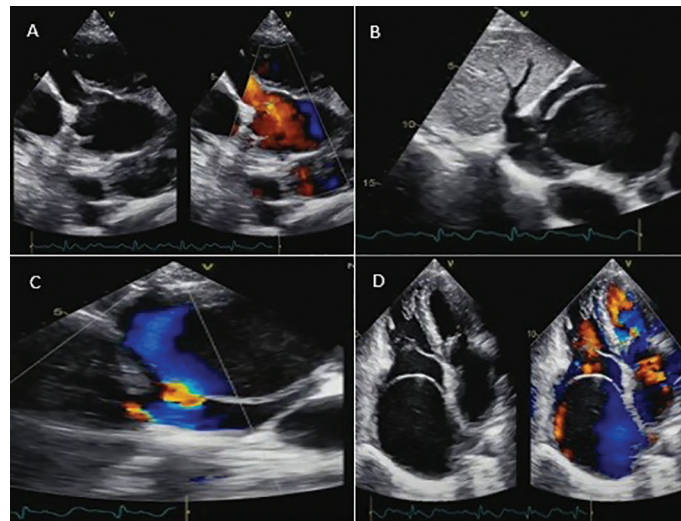


Figure 1 – Longitudinal parasternal view of the aortic root at the emergence of the right coronary artery (important dilatation) in the sinus of Valsalva (A). Subcostal view showing the draining of the aneurysmal portion in the right atrium, close to the inferior vena cava (B) and Doppler with flow between the cavities (C). Apical 4-chamber transthoracic echocardiography window, with and without color mapping, Doppler showing flow within the aneurysm cavity (D).

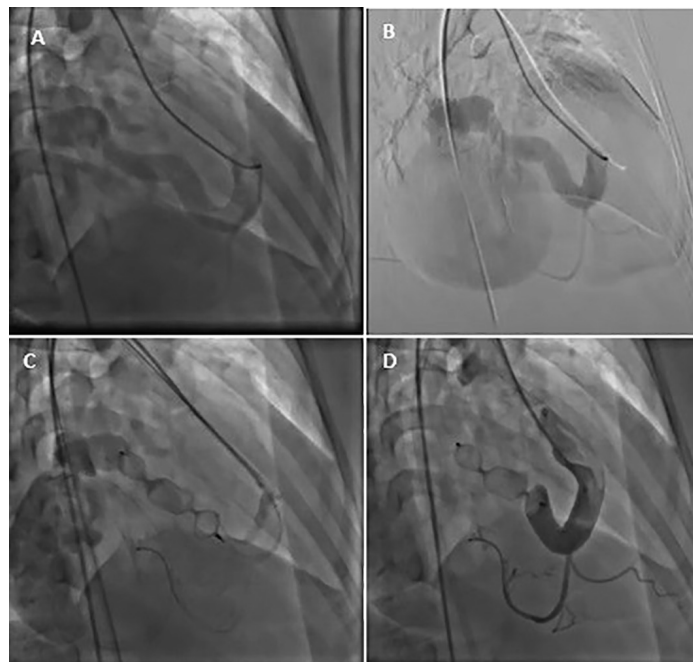


Figure 2 – Dilated right coronary artery with a distal bed with normal caliber and low flow, shortly after the emergence of the fistula toward the aneurysmal portion (A). Right coronary fistula draining into an aneurysmal sac, image with digital subtraction for better delineation of the structures (B). Amplatzer prosthesis positioned between the proximal and middle portion of the fistula (C). After release of the prosthesis and significant improvement in the distal flow of the right coronary artery (D).

blocking the flow to the aneurysmal portion (Figure 2C). During the procedure, an important improvement was already observed in the distal flow of the right coronary artery shortly after implantation of the prosthesis (Figure 2D). The procedure took place without complications.

One day after the procedure, a control transthoracic echocardiogram was performed, which showed an aneurysmal portion with thrombotic content inside (Figure 3A), already with signs of rejection by the right atrium and without blood flow between the cavities (Figure 3B), in

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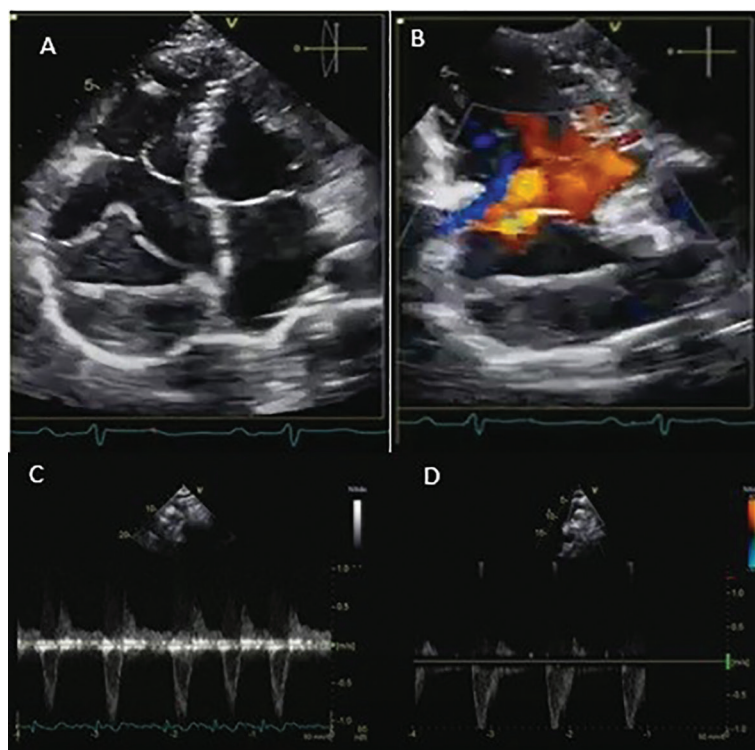


Figure 3 – Apical 4-chamber view with the aneurysmal portion being rejected by the right atrium, with thrombotic content inside (A). Doppler showing that there is no more flow between the cavities (B). Tissue Doppler in descending thoracic aorta with reverse holodiastolic flow (before the procedure) (C) and subsequent control, without the initial findings (D).

addition to significant improvement in reverse diastolic flow in the descending thoracic aorta (Figure 3C and 3D).

Discussion

Coronary fistula was first described in 1886, with the first report of successful surgical correction in 1947. From an embryological point of view, it appears to represent the persistence of embryonic intertrabecular spaces and sinusoids.^{4,5} There is also an association of fistulas with other congenital anomalies, in approximately 20% of cases, such as tetralogy of Fallot, patent ductus arteriosus, and atrial and ventricular septal defects.⁶

In more than half of the cases, the fistula originates from the right coronary artery and is usually a dilated artery, with a long and tortuous tract before draining into the chamber or vessel. In approximately 19% of cases, it also presents with aneurysmal dilation. The most common drainage site is into the right heart chambers.⁵

The clinical picture is variable, and the majority of patients are asymptomatic. In the reported case, which is a high-caliber coronary-cavitary fistulous tract between the right coronary artery and the right atrium, during the entire cardiac cycle, the blood flow was directed to the right side, due to the greater pressure found in the arterioles and myocardial capillaries.⁶ As there is continuity of the distal right coronary tract after the fistula, flow competition occurs

in that territory, whose perfusion becomes compromised, promoting functional myocardial ischemia.

In asymptomatic cases, with small fistulas and without a high risk of complications, follow-up may be considered with an echocardiogram every 2 to 5 years. For symptomatic or asymptomatic patients with small fistulas and risk of complications or larger fistulas with hemodynamic repercussions, invasive treatment (surgical or percutaneous) is recommended.⁶

After the development of new hemodynamic techniques, there has been a great resurgence of indications for closing fistulas by means of the percutaneous technique. The main advantages compared to the surgical technique include lower costs of the procedure, shorter recovery time, reduced morbidity, better aesthetic results, fewer episodes of bleeding, fewer arrhythmias, fewer infections, and less myocardial ischemia.⁷⁻⁸

The first transcatheter coronary fistula closure occurred in 1983 by Reidy et al.⁶ The main techniques used are coil embolization or occlusion devices. In the reported case, an 18 mm Amplatzer Vascular Plug II prosthesis was inserted between the proximal and middle thirds of the fistula; shortly thereafter, no residual shunt was observed, and there was significant improvement in distal coronary flow. The transthoracic echocardiogram performed one day after the procedure showed no complications and showed significant improvement in hemodynamic results (reduction of the

aneurysmal portion and absence of reverse diastolic flow in the descending thoracic aorta).

Six months after the procedure, angiography was scheduled to evaluate the involution of the aneurysmal sac, recovery of right atrial volume, and intracoronary thrombus. To date, the patient has shown clinical improvement, performing usual activities without limitations.

Author Contributions

Conception and design of the research: Silva LML and Correia EB; execution of images: Silva LML, Andrade MDRM, Jesus CA; acquisition and analysis and interpretation of the data: Andrade MDRM, Jesus CA, Ribeiro MS, Alencar AL. interventional procedure and provision of images: Ribeiro MS; writing of the manuscript: Alencar AL, Silva LML.

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Potential Conflict of Interest

No potential conflict of interest relevant to this article was reported.

Sources of Funding

There were no external funding sources for this study.

Study Association

This study is not associated with any thesis or dissertation work.

Ethics Approval and Consent to Participate

This article does not contain any studies with human participants or animals performed by any of the authors.



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