Radiology Case Reports 17 (2022) 1963–1967

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journal homepage: www.elsevier.com/locate/radcr

Case Report

Coronary to pulmonary artery fistula associated with significant coronary atherosclerosis and severe aortic valve stenosis: A Case Report ☆,☆☆

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ARTICLE INFO

Article history:
Received 8 February 2022
Revised 27 February 2022
Accepted 2 March 2022

Keywords:
Coronary artery fistula
Aortic stenosis
Echocardiography
Coronarography

ABSTRACT

Coronary artery fistulas are anomalous connections between one or two coronary arteries with either a cardiac chamber or any major blood vessels (coronary sinus, superior vena cava, pulmonary veins and pulmonary artery). It is rarely reported, occurring only in 0.1%-0.2% of patients who undergo coronary angiography. We report a very rare case where myocardial ischaemia may have resulted from the presence of coronary artery fistula, significant coronary artery stenosis and severe aortic valve stenosis. Transthoracic echocardiography showed severe aortic stenosis, while coronary angiography showed a tortuous coronary artery fistula originating from the proximal left anterior descending artery, with a single opening in the main pulmonary artery. Angiography also showed significant stenosis in the middle of the left anterior descending artery. Coronary artery fistula with concomitant significant coronary atherosclerosis and severe aortic stenosis requires optimal therapeutic planning.

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Introduction

Coronary artery fistulas (CAFs) are a condition where one or two coronary arteries communicate with either a cardiac chamber or any of the great vessels, such as the coronary sinus, superior vena cava, pulmonary veins or pulmonary artery [1]. Potential complications include myocardial ischemia due to coronary steal syndrome, thrombosis, embolism, rupture, bacterial endocarditis, pulmonary hypertension, and heart failure [2]. Atherosclerosis with significant coronary artery stenosis is known to be one of the main causes of myocardial ischemia. Aortic valve stenosis is characterized by progressive narrowing of the valve, leading to impaired coronary flow reserve (CFR) and myocardial ischemia [3]. We present a very rare case where myocardial ischemia may have resulted

☆ Acknowledgments: We thank the English Editing Service Elsevier for language editing of our paper.
☆☆ Competing Interests: The authors declare that they have no competing interests.
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https://doi.org/10.1016/j.radcr.2022.03.002
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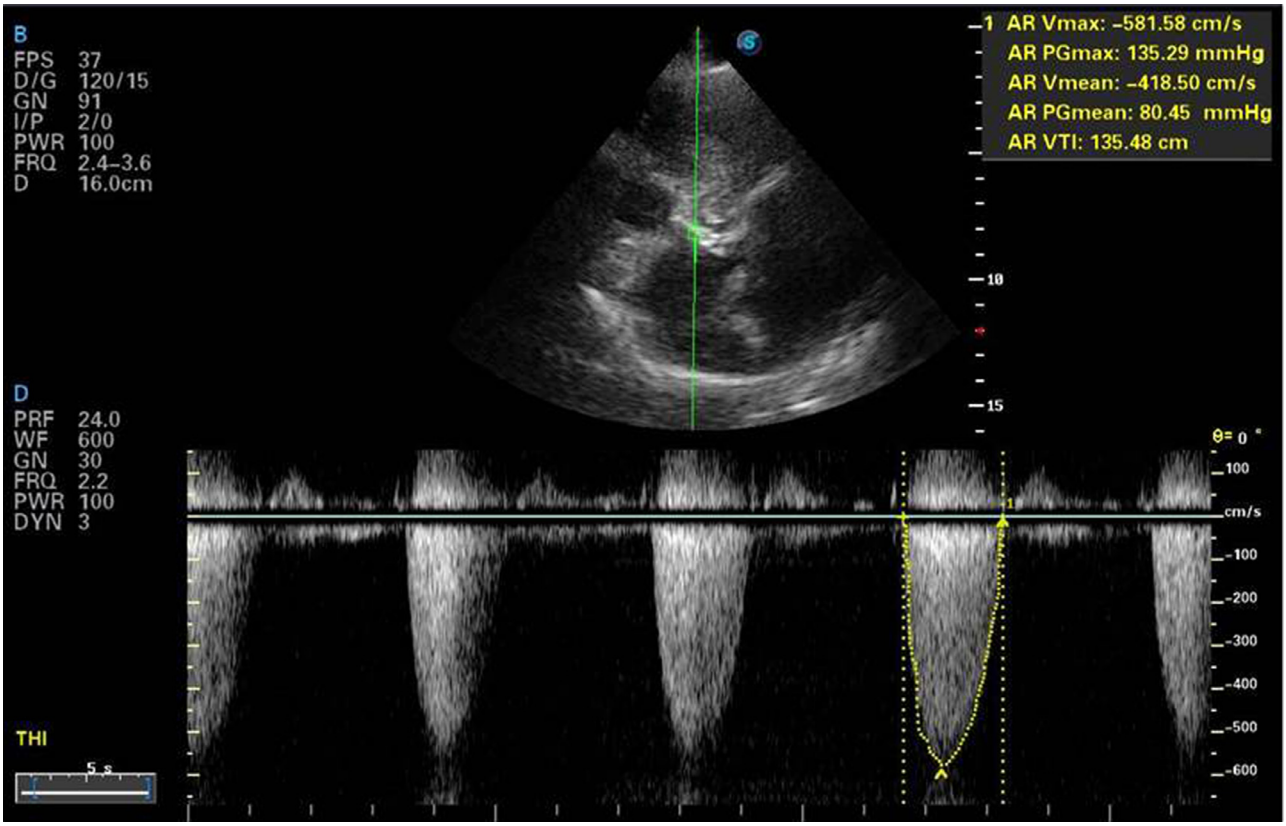

Fig. 1 – Continuous wave Doppler of the aortic valve showed severe aortic stenosis with a peak gradient of 135 mmHg and a mean gradient of 80 mmHg.

from the presence of coronary artery fistula, significant coronary artery stenosis, and severe aortic valve stenosis.

Case presentation

Patient information

A 68-year-old white woman came to our clinic for evaluation of chest pain and dyspnea.

The chest pain was described as pressure-like pain and effort-related, associated with numbness of left arm. In addition to pain, she also complained for dyspnea and diaphoresis with moderate entity physical activity. Patient did not report about relevant past medical history or interventions.

Clinical findings

On admission, vital signs showed a blood pressure of 130/85mmHg, heart rate of 90/minute, respiratory rate of 20/minute, and temperature of 37°C. Her cardiac examination revealed a systolic ejection murmur at the upper right sternal border, as well as along the left sternal border, while pulmonary examination demonstrated vesicular breathing, soft and low pitched, heard over both lung fields.

Diagnostic assessment

The blood analysis revealed normal values of cardiac biomarkers: cardiac troponin (cTn) level of 0.4ng/mL and creatine kinase-MB (CK-MB) level of 6.5ng/mL.

A 12-lead electrocardiogram at rest performed at hospital admission showed signs of left ventricular hypertrophy (LVH). Transthoracic echocardiography (TTE) showed severe aortic stenosis with an area of the aortic valve = 0.5cm^2 and a mean pressure gradient of 80 mmHg (Fig. 1).

Coronary angiography was performed and showed a tortuous fistula of the coronary artery originating from the proximal left anterior descending artery, with a single opening into the main pulmonary artery. The left anterior oblique (LAO) caudal view and the LAO cranial view are shown in Fig. 2 (2A and 2B), respectively. There was also significant stenosis in the mid portion of the left anterior descending artery (LAD) (Fig. 3A), while the right coronary artery was normal (Fig. 3B).

Therapeutic intervention

In a joint meeting with cardiologists and cardiac surgeons, it was agreed that due to the complexity, a surgical approach should be offered to the patient.

The surgical procedure was rejected by the patient, and thus pharmacological therapy was continued.

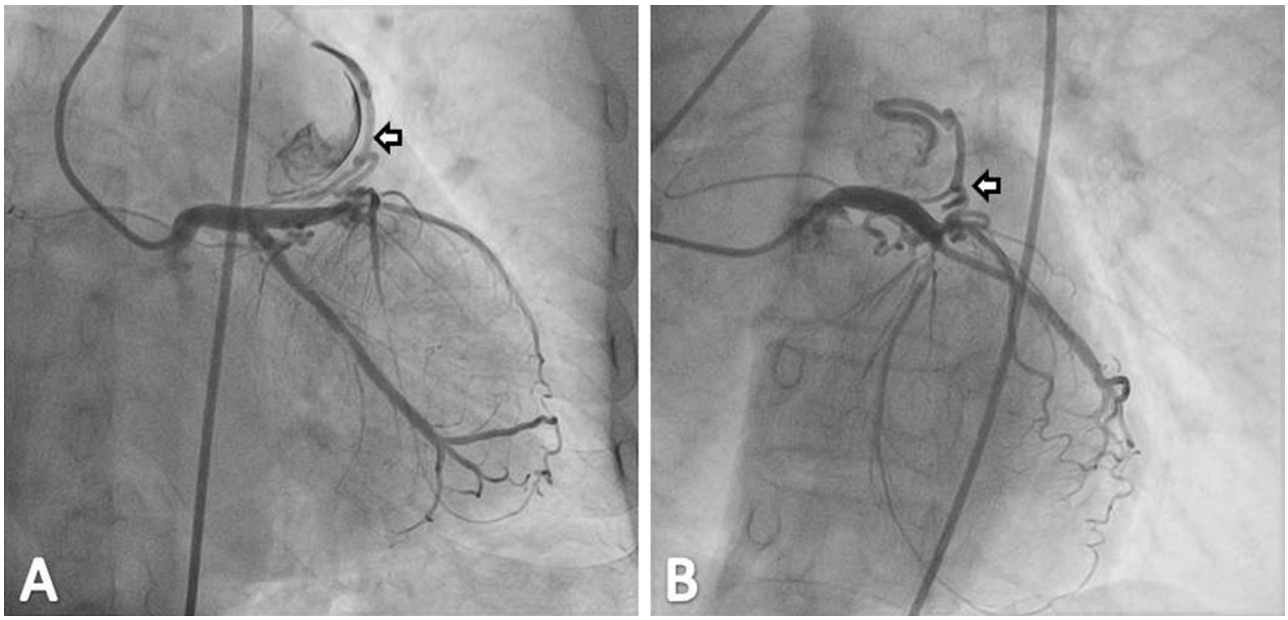

Fig. 2 – A tortuous coronary artery fistula originating from the proximal left anterior descending artery, with a single opening into the main pulmonary artery. Left anterior oblique caudal view (A) and left anterior oblique cranial view (B).

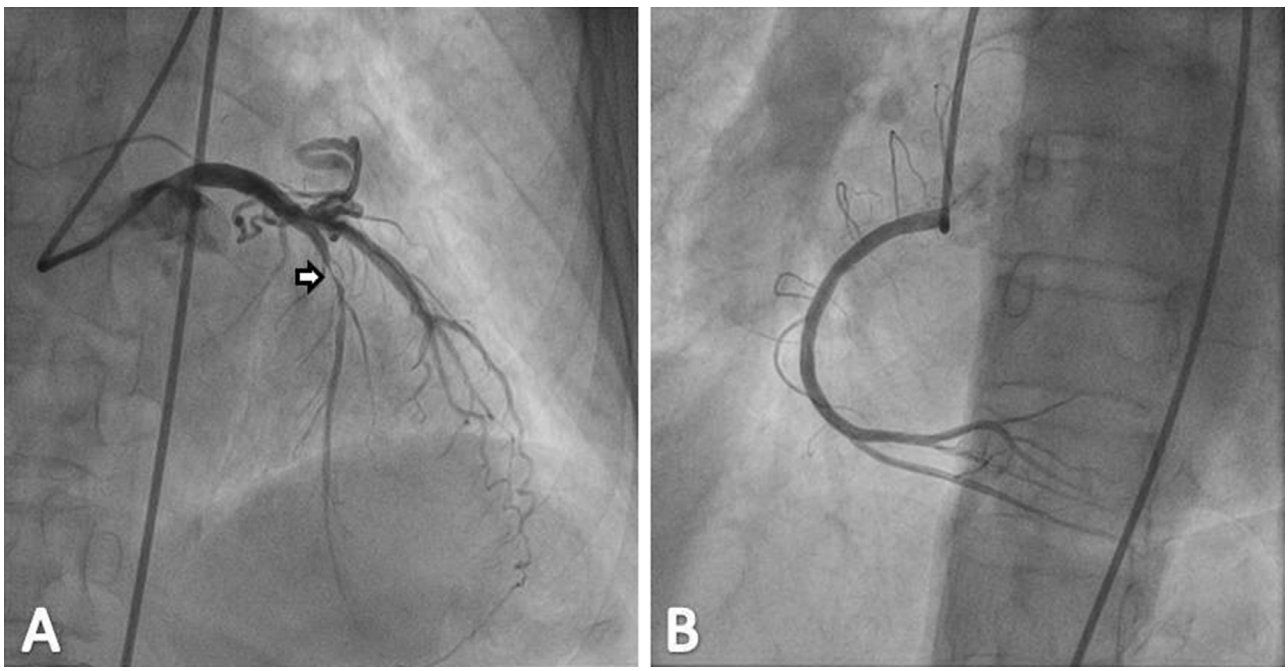

Fig. 3 – (A) A significant mid- left anterior descending artery stenosis; (B) Normal right coronary artery.

Discussion

Our patient presents the only known case in the medical literature, to the best of our knowledge, that presents with a combination of coronary artery fistula, coronary artery atherosclerosis, and significant aortic stenosis, all these pathologies related to angina.

CAFs are a rare finding, reported only in 0.1%-0.2% of patients who undergo coronary angiography [1]. CAFs drain most often into the right ventricle, and less commonly into the right atrium and pulmonary artery. CAFs draining into the pulmonary trunk are observed in most coronary-pulmonary arterial fistulas (CPAF) [4]. Depending on resistance and drainage site, the consequences of CAF on venous structures or the right cardiac chamber can be a diastolic 'coronary steal'

with signs of myocardial ischemia [5]. The onset of symptoms (angina pectoris and dyspnea) usually occurs during the fourth through sixth decades of life, with the tendency of fistulas to grow with age [6].

In patients with obstructive coronary artery disease, myocardial ischemia is the result of an imbalance between myocardial oxygen supply and demand, usually presenting as angina. Aortic stenosis (AS) is a progressive disease characterized by valve narrowing that clinically manifests itself as dyspnea, syncope, and angina. Coronary flow reserve (CFR) is defined as the maximal increase in myocardial blood flow at maximal hyperemia above its resting level. Impairment of CFR is the main cause of angina pectoris in patients with AS [7].

Furthermore, myocardial ischemia due to the fistula steal phenomenon cannot be clinically distinguished from that of coronary atherosclerosis and aortic stenosis.

The diagnosis of CAF is often incidental during a routine clinical examination with the presence of atypical systolic, diastolic, or continuous murmur, regardless of symptoms. TTE and transoesophageal echocardiography (TEE) provide a view of the anatomy of the coronary artery, chamber dilation and fistula (origin, course, and termination) [8]. Multidetector computed tomography (CT) allows anatomical description and can provide useful information regarding the presence or absence of coronary obstruction [9].

However, coronary angiography remains the main diagnostic technique for the detection of CAF in the presence of concomitant atherosclerosis [10].

Treatment options include medical therapy (for asymptomatic patients), surgical ligation and interventional closure with occlusion coils, umbrellas, vascular plugs and covered stents [2]. Taking into account the complex pathology of our patient, the final recommendation was to performed a combined surgical procedure that included fistula ligation in association with CA bypass grafting and replacement of the aortic valve.

Conclusions

Coronary artery fistula with concomitant significant coronary atherosclerosis and severe aortic stenosis is an exceptional finding in the medical literature with three pathologies that cause angina. A complicated case such as this requires optimal therapeutic planning, guided by the patient's symptoms, the size of the fistula and the severity of the associated pathologies.

Patient Consent

Written informed consent was obtained from the patient for the publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Declarations

This manuscript or essential parts of it have not been previously published or are under consideration by another journal, in English or another language.

Cover letter

We are pleased to submit a case report "Coronary to pulmonary artery fistula associated with significant coronary atherosclerosis and severe aortic valve stenosis" for publication in Radiology Case Reports. In this manuscript, we describe a very rare case of coronary to pulmonary artery fistula associated with significant coronary atherosclerosis and severe aortic valve stenosis so cardiologist should think also in multiple causes of myocardial ischemia in the same patient. We confirmed that none of the authors have any conflicts of interest associated to this manuscript. In addition, we confirm that this manuscript has been approved by all authors to be published in the Radiology Case Reports.

Availability of data and materials

All data from this study are included.

Authors' contribution

XK was the first author. XK, AB and DK prepared the final manuscript. All authors contributed to data collection and read and approved the final manuscript.

Ethics approval and consent to participate

The Approval of our local ethics committee for publication was obtained.

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