Myocardial infarction with non-obstructive disease and anomalous coronary origin: look for the common in the uncommon

Gindomenico Disabato1,2, Antonia Camporeale2*, Mauro Lo Rito3, Lara Tondi2, Karina Geraldina Zuniga Olaya3, Alessandro Frigiola3, Mauro Luca Agnifili4, Francesco Bedogni4, Massimo Lombardi2 and Silvia Pica2

1 University of Pavia, Pavia, Italy; 2 Multimodality Cardiac Imaging Section, IRCCS Policlinico San Donato, San Donato Milanese, Italy; 3 Congenital Cardiac Surgery, IRCCS Policlinico San Donato, San Donato Milanese, Italy; and 4 Department of Clinical and Interventional Cardiology, IRCCS Policlinico San Donato, San Donato Milanese, Italy

Abstract

Management of congenital coronary artery anomalies (CAA) is not standardized due to the variety of conditions included and their rare prevalence. Detection of CAA during myocardial infarction with non-obstructive coronary arteries (MINOCA) may induce clinicians to address the patient for surgery as CAA is not included in any algorithm1,2 for the management of MINOCA and American Association for Thoracic Surgery evidence-based guidelines suggest surgical repair for patients with anomalous aortic origin of a coronary artery and symptoms compatible with myocardial ischaemia.3 We present the case of a 35-year-old man with an anomalous origin of left coronary artery from right Valsalva sinus with pre-pulmonic course detected during urgent coronary angiography for suspected myocardial infarction. Stress cardiac magnetic resonance did not show signs of ischaemia at high-dose dobutamine but did reveal a recent myocarditis. This clinical case highlights the need for accurate risk stratification in CAA especially when confounding clinical scenarios co-exist.

Keywords MINOCA; Coronary anomaly; Cardiac magnetic resonance; Myocarditis

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*Correspondence to: Antonia Camporeale, Multimodality Imaging Section, IRCCS Policlinico San Donato, Via Morandi 30, 20097 San Donato Milanese, Italy.
Phone: +39 02 5277 4804/4376; Fax: +39 02 52774272. Email: antonia.camporeale@grupposandonato.it

Introduction

The term congenital coronary artery anomalies (CAA) include a series of rare diseases characterized by abnormalities of coronary arteries in the origin, course, size or number.1 Anomalous aortic origin of a coronary artery (AOOCA) refers to a coronary artery arising from the inappropriate sinus of Valsalva, and its prevalence varies among studies between 0.1 and 1%.1,2 The wide variability may be due to referral bias, type of imaging test used, and study population. AOOCA can manifest with sudden cardiac death or symptoms of ischaemia1; however, most patients are diagnosed incidentally, and consequently, the real prevalence on general population and cardiovascular risk remains unknown.1

Left coronary artery arising from right sinus (AOOLCA) is less prevalent than the origin of right coronary artery from the inappropriate sinus (AOORCA); nevertheless, it poses the patient at higher sudden cardiac death risk at younger age (<35 years).3

Case presentation

A 35-year-old Caucasian man was admitted to a secondary care centre for typical angina at rest. He had no previous medical records except for resting chest pain 5 years before with spontaneous resolution and no further medical assessment. At presentation, a 12-lead ECG showed infero-lateral ST elevation (Figure 1) without echocardiographic abnormal findings. Coronary angiography revealed anomalous aortic origin of left coronary artery from right Valsalva sinus (AAOLCA) with a long pre-pulmonic course and without coronary
obstruction. During the hospitalization, the patient remained asymptomatic, and the peak high-sensitivity troponin I (hsTnI) was 4366.8 ng/L (99th percentile URL 19.8 ng/L) in the first day of admission. Laboratory tests detected elevated C-reactive protein (29.5 ng/mL) with negative procalcitonin, normal blood count, and no signs of infection on urinary test and chest X-ray.

The patient was referred 3 months later to our tertiary care centre to perform a surgical evaluation of the coronary anomaly, which was suspected to be responsible of acute coronary syndrome.

At admission, the patient reported no symptoms since the previous hospitalization. Coronary computed tomography angiography (CCTA) was performed to assess the anatomy of the anomalous coronary artery (Figure 2). Subsequently, invasive coronary angiography (Figure 3) and intravascular ultrasound evaluation did not reveal any pathological finding, that is, lateral systolic compression of the coronary lumen (Video S1 and Video S2).

Stress cardiac magnetic resonance (CMR) was performed to assess induced myocardial ischaemia: Neither perfusion defect nor regional kinetic abnormality was detected at peak dobutamine infusion (i.e. 40 mcg/kg/min). However, sub-epicardial late gadolinium enhancement involving infero-lateral segments was detected, raising the suspicion of myocarditis (Figure 4). The day after, CMR was integrated by acquiring T2-weighted images, showing signal hyperintensity compatible with residual myocardial oedema in lateral segments (Figure 4). According to the new diagnosis of recent myocarditis, laboratory tests were repeated, revealing mild elevation in high-sensitivity troponin I (75 ng/L, 99th percentile URL 45 ng/L), normal CK-MB (1 ng/mL, normal value 0.18–5 ng/mL), and negative inflammatory markers. A full panel for myocarditis, as recommended by recent European Society of Cardiology guidelines,4 tested negative for infections or autoimmune diseases.

Discussion

We present a clinical case of a 35-year-old patient with no cardiovascular risk factors initially admitted with signs and symptoms compatible with myocardial infarction with ST segment elevation. Coronary angiography revealed the presence of pre-pulmonic AAOLCA and no coronary obstructions.

Considering the diagnosis of MINOCA, further testing would have been needed for diagnosis as suggested by several diagnostic algorithms of recent guidelines.5,6 None of these algorithms, however, contemplates the presence of CAA, and the finding of anomalous origin of the left coronary artery was suspected to be related to the clinical presentation. The American Association for Thoracic Surgery evidence-based

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Figure 1  ECG at first admission. ECG showing ST elevation in leads V4–V5–V6 and in DII-DIII-aVF with specular ST down-sloping in anterior leads (V1–V2–V3) and aVR.

Figure 2  Coronary computed tomography angiography (CCTA) showing the anatomy of the anomalous coronary artery (AAOLCA) arising from the right sinus of Valsalva.

Figure 3  Invasive coronary angiography revealing the presence of anomalous coronary origin.

Figure 4  Stress cardiac magnetic resonance (CMR) with late gadolinium enhancement showing residual myocardial oedema in lateral segments.
guidelines suggest surgical repair for patients with AAOCA and symptoms compatible with myocardial ischaemia\(^2\); therefore, the patient was referred to our centre. The usual benign prognosis of AAOLCA without inter-arterial course\(^1,2\) suggested a cautious approach despite the recent acute episode. We routinely perform dobutamine stress CMR to search for signs of inducible myocardial ischaemia, as suggested by ESC guidelines on congenital heart disease\(^3\), and to exclude the presence of ischaemic scar. We are aware that non-invasive stress imaging has a high positive predictive value but lacks enough sensibility to rule out reduced coronary flow reserve in this setting of patients\(^2\); however, no sufficient data are available for stress CMR, which has proven to have higher diagnostic accuracy compared with other non-invasive functional stress imaging in general population\(^7,8\).

In our case, the possibility to exclude reduced coronary flow reserve and detect myocardial tissue alterations by CMR allowed the diagnosis of myocarditis as the most likely cause of the acute presentation. Myocarditis can also explain the high level of hsTnI and C-reactive protein detected during first hospitalization. Intracoronary ultrasound was performed as suggested by guidelines\(^1,9\) to detect potential mechanisms of flow reduction, but no pathological finding was revealed. In our case report, negative intravascular ultrasound and stress CMR prevented the patient from undergoing unnecessary surgical repair.
Conclusion

CA enlist a series of uncommon conditions whose management is not well standardized. After appropriate study of coronary anatomy, accurate risk stratification should go beyond history of symptoms suggestive of ischaemic nature. Our clinical case highlights the importance of CMR both to rule out myocardial ischaemia and to check for tissue abnormalities and other possible causes of chest pain in a patient firstly admitted for suspected acute myocardial infarction. In this regard, CMR plays a pivotal role and should be suggested as part of the diagnostic algorithm of MINOCA despite the evidence of CAA.

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Conflict of interest

The authors have nothing to disclose.

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Supporting information

Additional supporting information may be found online in the Supporting Information section at the end of the article.

Video S1. Supporting Information
Video S2. Supporting Information

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