ESC European Society of Cardiology

European Heart Journal - Case Reports
doi:10.1093/ehjcr/ytab081

CASE REPORT
Other

A case report of a symptomatic right anomalous coronary artery with concomitant atherosclerotic disease: the benefit of a sequential comprehensive non-invasive and invasive diagnostic approach

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Received 10 November 2020; first decision 4 December 2020; accepted 4 February 2021

Background Anomalous aortic origin of a coronary artery (AAOCA) is a rare congenital disease associated with an increased risk of myocardial ischaemia, ventricular arrhythmias, and heart failure.

Case summary A 75-year-old Caucasian man was referred for invasive coronary angiography (ICA) due to atypical chest pain. Invasive coronary angiography demonstrated non-significant atherosclerotic disease of the left coronary artery and an anomalous origin of the right coronary artery (RCA); without selective intubation. Coronary computed tomography angiography (CCTA) revealed a right-AAOCA with interarterial and intramural course, and a soft plaque in the distal RCA. Subsequent physical-stress single-photon emissions computed tomography (SPECT) showed exercise-induced inferoapical myocardial ischaemia, giving a Class IC level of evidence for surgical correction of the AAOCA. Repeated ICA with selective R-AAOCA intubation confirmed an 80% distal atherosclerotic stenosis, which was treated with direct stenting. Subsequent invasive physiologic evaluation under maximal dobutamine-volume challenge (gradually increasing dose of dobutamine max. 40 µg/kg per body weight/min, 3000 mL ringer lactate and 1 mg atropine was given until the patient reached a maximum of 145 b.p.m.), revealed a haemodynamically non-relevant anomalous segment with a fractional flow reserve (FFR) of 0.91. A follow-up SPECT was normal, and the patient was completely symptom-free at 1 month.

Discussion We present the sequential diagnostic approach in a symptomatic patient with a right anomalous coronary artery and concomitant atherosclerotic disease. Using this approach, the patient could be deferred from guideline recommended open-heart surgery of the AAOCA, as direct invasive dobutamine/volume FFR revealed haemodynamic non-relevance of the anomalous segment after stenting the concomitant atherosclerotic stenosis in the distal segment within the same coronary artery.

Keywords Anomalous aortic origin of a coronary artery (AAOCA) • Multimodality cardiac imaging • Coronary computed tomography angiography • Single-photon emission computer tomography • Fractional flow reserve • Case report

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Handling Editor: Anastasia Egorova
Peer-reviewers: A. Shaheer Ahmed and Mohammed Al-Hijji
Compliance Editor: Edwina McNaughton
Supplementary Material Editor: Ross Thomson

Learning points

- Multimodality cardiac imaging is required for comprehensive assessment of haemodynamic relevance of anomalous aortic origin of a coronary artery (AAOCA), especially in the clinical setting of concomitant atherosclerotic diseases.
- In older patients, myocardial ischaemia is more likely attributable to concomitant atherosclerotic disease than to AAOCA.
- The pathophysiology in AAOCA includes a fixed and a dynamic component, which requires a specialized diagnostic workup with supramaximal stress testing.

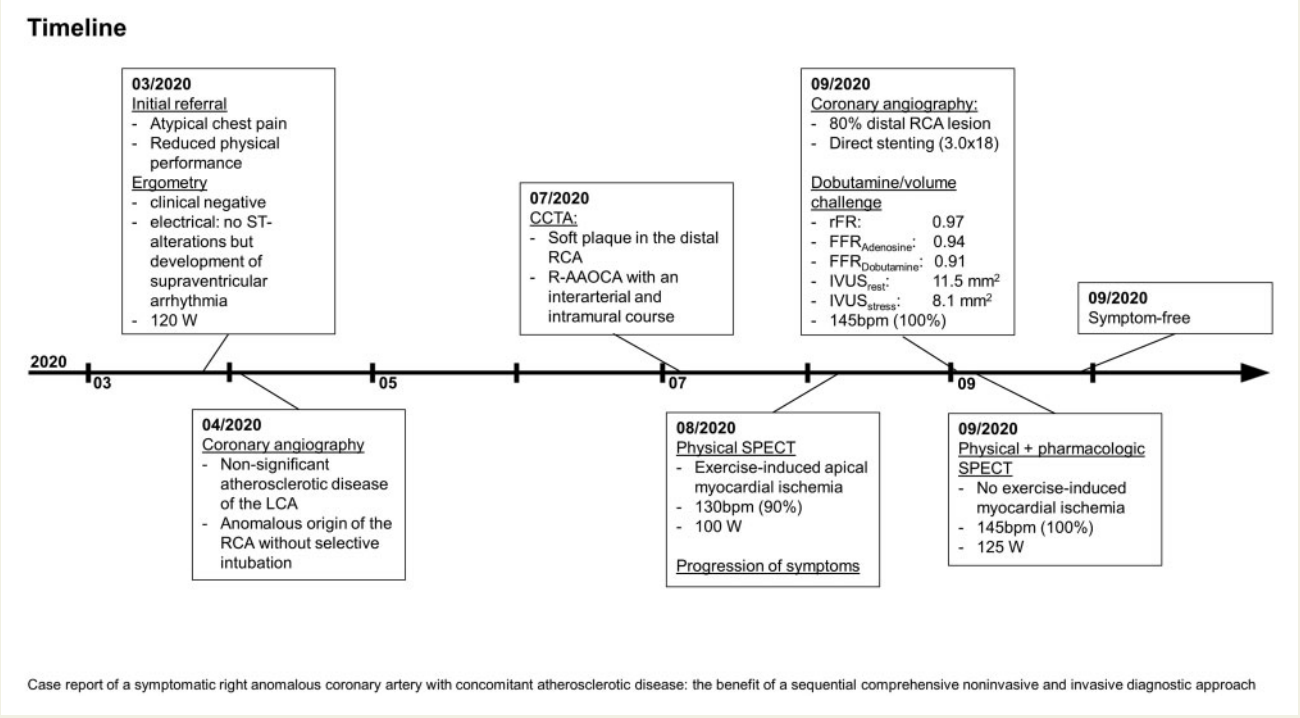
Introduction

Anomalous coronary arteries (CAA) represent a congenital disorder hallmarked by an anomalous location of the coronary ostium and/or vessel course.[1] Of particular interests are CAA with an anomalous aortic origin (AAOCA) and an interarterial course (IC) between the great arteries (i.e. aorta and pulmonary artery), a so-called 'malignant' variant which has been associated with an increased risk of myocardial ischaemia, ventricular arrhythmias, and heart failure.[1]

However, the interarterial course itself may not be the predominant cause of ischaemia, but rather represents a surrogate for other anatomical high-risk features associated with ischaemia such as slit-like ostium, acute take-off angle, intramural course (i.e. location of the proximal segment within the tunica media of the aortic wall), oval vessel shape, and proximal narrowing.[1,2] These anatomical features are considered high risk as it is hypothesized that during exercise the aortic dilation may lead to lateral compression and subsequent haemodynamically relevant stenosis of the anomalous segment.[1,2]

With increased use of non-invasive imaging in evaluation of coronary artery disease (CAD) in younger, middle-aged, and older individuals, an increase in absolute numbers of AAOCA-IC is expected. Usually, the question is whether the AAOCA-IC is a co-incidental finding or if the anomaly is causative for the patients' symptoms. Guidelines are limited and recommendations are mainly based on expert's opinions, which include strict sports abstinence in patients with interarterial courses and a low threshold for surgical coronary revascularization.[3–5] Further, in older patients with AAOCA, a higher prevalence of concomitant CAD is noted and ischaemia is more often caused by CAD than the anomaly.[6] However, the optimal management in patients with combined coronary artery anomaly and CAD remains unknown.

Timeline



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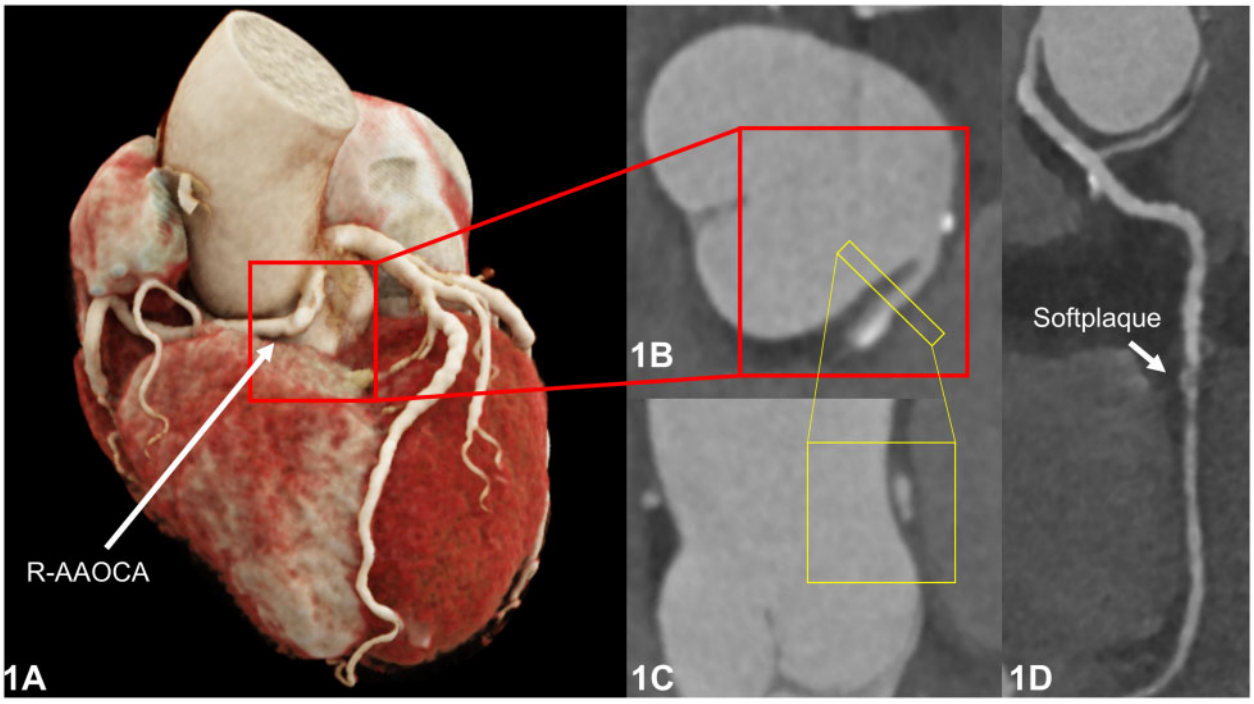

Figure 1 (*A*) Coronary computed tomography angiography of the right anomalous aortic origin of the coronary artery. (*B*) Interarterial course. (*C*) The elliptic proximal vessel shape (cross section with an increased height to width ratio) is depicted and is an indirect sign of the intramural course. (*D*) Right anomalous aortic origin of the coronary artery with softplaque in the distal part. Please note the other anatomical 'high-risk' features as acute take-off angle (*B*), proximal narrowing (*C* and *D*), and the oval vessel shape (*C*).

Case presentation

A 75-year-old Caucasian man was referred for invasive coronary angiography (ICA) due to atypical chest pain. Initial diagnostic workup revealed normal electrocardiographic and laboratory findings. Arterial hypertension, dyslipidaemia, and a history of smoking; cumulative five pack years, were the patient's cardiovascular risk factors. Medications were aspirin, rosuvastatin, and losartan. On physical examination, temperature was normal, blood pressure was 134/85 mmHg, and heartrate was 58 b.p.m. Cardiac examination was normal without pathological sounds or murmurs. Bicycle exercise testing (120 W, 158 b.p.m. ≙ 109% of predicted maximal heartrate) revealed an exercise-induced supraventricular arrhythmia.

Invasive coronary angiography demonstrated non-significant atherosclerotic disease of the left coronary artery and an anomalous origin of the right coronary artery (RCA), although selective intubation was not possible. Subsequently performed coronary computed tomography angiography (CCTA) revealed a right anomalous aortic origin of the coronary artery (R-AAOCA) with an intramural and interarterial course. Further, a soft plaque was seen in the distal RCA (*Figure 1*). As recommended,[3,5] a physical stress single-photon emissions computed tomography (SPECT) was performed, demonstrating exercise-induced inferoapical ischaemia (130 b.p.m. ≙ 90% of predicted maximal heartrate), within the territory of R-AAOCA, in the presence of RCA dominance (*Figure 2*). Thus, according to the 2020 ESC Guidelines, a Class IC level of evidence is given for surgical correction (i.e. open-heart surgery) of the anomalous vessel.[3]

However, in agreement with the patient, a repeat invasive angiography was performed with selective intubation of the R-AAOCA, which confirmed a significant, 80% distal atherosclerotic stenosis (*Figure 2*). This lesion was treated with direct stenting (drug-eluting stent: 3.0 mm × 18 mm). Subsequent invasive physiologic evaluation under a dobutamine/volume-challenge (maximum dobutamine dose of 40 µg/kg per body weight/min, 3000 mL ringer lactate, and 1 mg atropine),[6,7] revealed a haemodynamically non-relevant anomalous segment with a fractional flow reserve of 0.91 (145 b.p.m. ≙ 100% of predicted maximal heartrate). Intravascular ultrasound (IVUS) confirmed the presence of a non-significant anomaly with only mild lateral dynamic compression of the vessel (reduction of the minimal lumen area of 11.5 to 8.1 mm^2, i.e. 29.6% under maximal dobutamine/volume stress). Consequently, surgical revascularization was refrained. A repeated SPECT after the procedure showed absence of apical myocardial ischaemia (*Figure 2*), and the patient was completely symptom-free at 1-month follow-up.

Discussion

We present the case of a symptomatic R-AAOCA with concomitant atherosclerotic disease within the same vessel, where a maximal

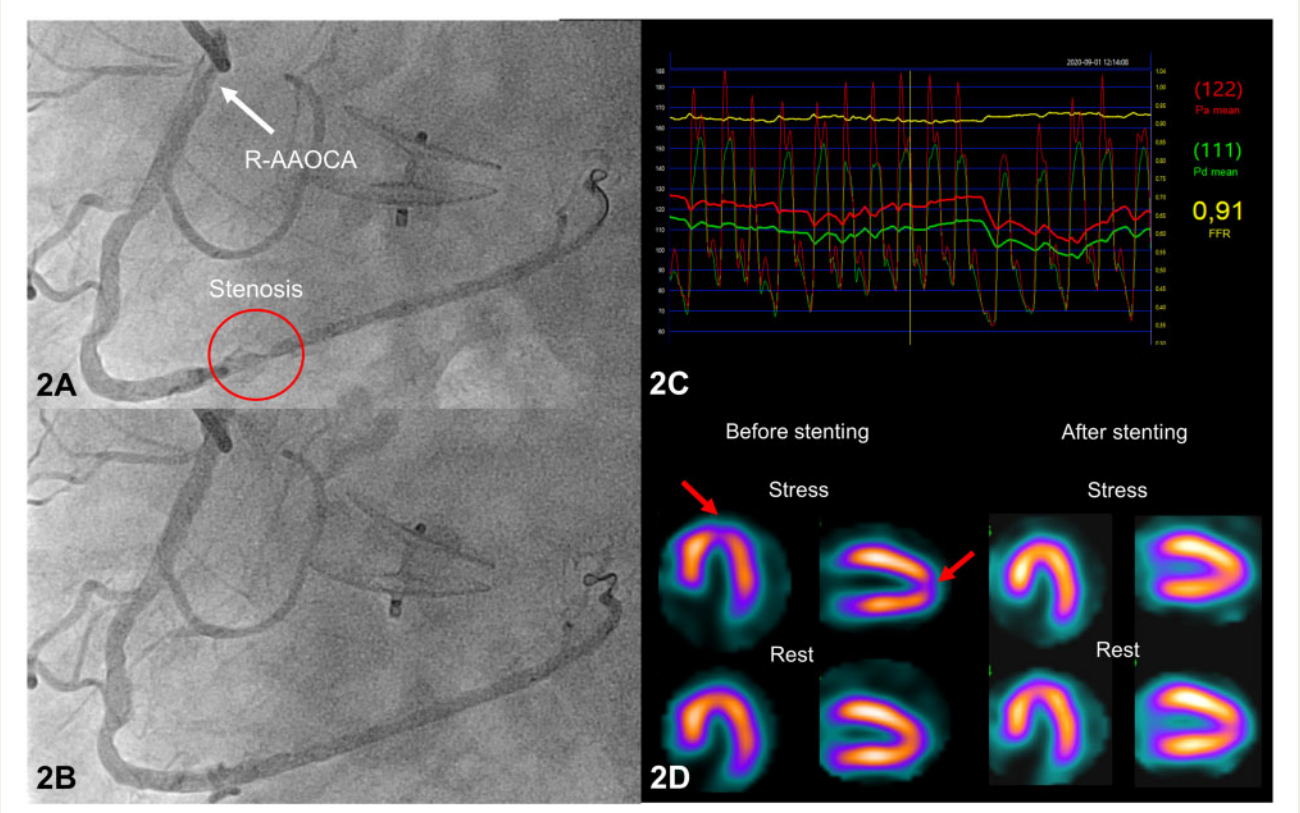

Figure 2 (*A*) Invasive coronary angiography with significant distal stenosis. Please note the 'Amplatzer PFO Occluder Cribriform 25 mm'-device, which was implanted after a major stroke in the year 2009. (*B*) Post-stenting invasive angiography. (*C*) Invasive assessment with non-significant dobutamine/volume fractional flow reserve. (*D*) Single-photon emissions computer tomography before stenting with inferoapical ischaemia and follow-up SPECT without ischaemia.

physical exercise non-invasive imaging testing revealed ischaemia in the supplied myocardial territory. While surgical *unroofing* is the guideline recommended therapy for symptomatic R-AAOCA, percutaneous coronary treatment with drug-eluting stents is preferred treatment in single RCA stenosis. As it was unclear, whether the AAOCA or the atherosclerotic stenosis was responsible for the patient's symptoms and ischaemia, a sequential treatment and diagnostic approach was chosen. Direct stenting of CAD and succeeding invasive diagnostic approach with haemodynamic evaluation of the anomalous course under pharmacologic inotropic stress revealed that the AAOCA was an innocent bystander. The patient was symptom free and could be prevented from unnecessary open-heart surgery using this approach.

Management of patients with anomalous aortic origin of a coronary artery and coronary artery disease

AAOCA is a coronary anomaly with a prevalence of 0.26% in the general population.[8,9] Hence, the level of evidence is limited and current guidelines are often vague with regard to management recommendation. Mostly, surgical revascularization is recommended with a low threshold based on the increased frequency of sports-related sudden cardiac deaths in younger individuals, observed in autopsy series.[10,11] However, the significance of newly diagnosed AAOCA in older patients remains unclear, especially since the haemodynamic relevance of AAOCA may decrease with age while concomitant CAD become increasingly significant.[12] However, also in advanced age, first time appearance of AAOCA-related myocardial ischaemia is possible, as recently described by our group.[6]

For the optimal diagnostic and therapeutic management of the AAOCA, one has to consider the underlying pathophysiology. Our group supported a two-tier concept for the pathomechanisms of ischaemia in AAOCA. In detail, this concept states 'the occurrence of ischaemia is based on the extent of a fixed (i.e. anatomic high-risk features as slit-like ostium and proximal narrowing) and a dynamic (i.e. acute take-off angle, intramural course with lateral compression) component'.[13] The latter, which occurs during strenuous physical exercise promoted by aortic dilation and reduced diastolic perfusion time. Further, the mass of viable myocardium supplied by the anomalous coronary artery as well as the distensibility of the aortic wall directly affects the haemodynamic relevance. Taken into account the various components, it is obvious that a thorough diagnostic evaluation requires a multimodality approach.

CCTA is the preferred technique for the qualitative and quantitative assessment of the anatomical high-risk features with

simultaneous evaluation of the presence of concomitant atherosclerotic disease.[3] Further, a maximal physical (non-pharmacological) functional imaging is recommended according to the most recent ESC guidelines (Class IC).[3] If a physical stress test is not possible, dobutamine/atropine imaging might be used as an alternative to mimic physical conditions, whereas vasodilatation stress tests are not ideal modalities. In older patients, a hybrid imaging technique with simultaneous CCTA and functional imaging should be preferred to discriminate between AAOCA and atherosclerotic diseases-related perfusion defects.[12,14]

However, this diagnostic approach fails when CAD occurs within the same vessel as the anomalous coronary artery. A non-invasive functional imaging test may not adequately discriminate whether possible ischaemia is due to AAOCA- or CAD-related stenosis within the same vessel. This situation is not covered by recent guidelines, nor exist expert consensus on how to proceed with the decision-making in this particular setting. Direct surgical treatment with unroofing may be inadequate, as with advanced aging haemodynamic relevance of AAOCA might be less relevant.[15] Alternative aorta-coronary bypass grafting to the distal RCA, by treating CAD and AAOCA at the same time, would not be the primary preferred approach, taking into account the uncertain haemodynamic relevance of the AAOCA. Further, direct percutaneous coronary intervention of a focal, non-calcified lesion is expected to be unproblematic. Hence, we recommend the presented sequential approach with initial non-invasive evaluation (anatomical and functional imaging) followed by interventional treatment of the atherosclerotic lesion, if an independent indication for CAD-related stenosis treatment is given. Within the same intervention after treatment of the atherosclerotic stenosis, invasive assessment of the anomalous coronary artery with combined haemodynamic (pressure indices) and morphologic (IVUS) assessment during a supramaximal dobutamine/volume/atropine stress test should be performed to identify whether surgical correction of AAOCA is required.

Lead author biography

Marius Reto Bigler (1991) is currently a research fellow and cardiologist in training at the Department of Cardiology, Bern University Hospital in Switzerland. His PhD project involves the functional assessment of myocardial ischaemia using the intracoronary electrocardiogram as well as structural and haemodynamic parameters. He has specific interests in the (patho)-physiologic comprehension of myocardial perfusion and in various cardiac imaging modalities. Within such a multidisciplinary collaboration, the comprehensive diagnostic workup described in this case report was performed.

Supplementary material

Supplementary material is available at *European Heart Journal - Case Reports* online.

Acknowledgements

We would like to thank Yasushi Ueki, MD and Tatsu Otsuka, MD from the Department of Cardiology for their experience and help with the invasive assessment.

Slide sets: A fully edited slide set detailing this case and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case report including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: None declared.

Funding: This work was supported by the general research fund of the Department of Cardiology, Inselspital, Bern University Hospital, University of Bern, Switzerland.

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