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Multimodality imaging of an incidental anomalous coronary artery

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Short title: Multimodal imaging of an anomalous coronary artery

Anomalous Right Coronary Artery arising from the Main Pulmonary Artery (ARCAPA) is commonly identified on an incidental basis in asymptomatic individuals.

This case report of an adult male admitted with troponin-positive chest pain highlights the incidental nature via which ARCAPA may present. Invasive coronary angiography suggested a potential anomalous right coronary origin, before CT coronary angiography confirmed ARCAPA whilst also identifying extensive, acute bilateral pulmonary emboli, in itself sufficient to explain the presentation. Stress perfusion cardiac MRI accordingly excluded both myocardial infarction and inducible hypoperfusion. We discuss multimodality investigation in the management of incidental ARCAPA.

A 50-year-old male ex-smoker presented with a one-day history of central chest tightness and dyspnoea. The patient did not report any fever, haemoptysis, or orthopnoea. Obstructive sleep apnoea was his only comorbidity. He took no medications. Clinical examination was unremarkable with normal JVP, heart sounds and clear lung fields. Clinical observations were 135/82 mmHg, pulse 90/min, SpO2 97%/air, respiratory rate 18/min.

Electrocardiography (ECG) (**Supplementary Figure S1**) demonstrated sinus tachycardia rate 120 bpm, T-wave inversion in V1 to V3 and minor ST depression in V3. High sensitivity troponin I rose from 305 to 563 ng/L over three hours. Full blood count and inflammatory markers were normal. PCR for SARS-CoV-2 was negative. Plain chest radiography demonstrated cardiomegaly, engorged hila and moderate upper lobe venous blood diversion but no overt pulmonary oedema or pleural effusion (**Supplementary Figure S2**). Transthoracic echocardiography demonstrated mild concentric left ventricular hypertrophy with good global left ventricular systolic function; there was only trace tricuspid regurgitation. Mr A was given a working diagnosis of non-ST elevation myocardial infarction by the on-call team and loaded with aspirin and clopidogrel.

Diagnosis and Management

On invasive coronary angiography, direct cannulation of the right coronary system failed. Angiography (**Video 1**) of the left coronary artery demonstrates epicardial connection of the right coronary artery to the pulmonary artery. A prospectively gated CT coronary angiogram (CTCA) confirmed the anatomy of the aberrant coronary system with a normal left main stem origin, but the right coronary artery arising from the main pulmonary artery (ARCAPA; **Supplementary Figure S3**). The coronary system was aneurysmal in all territories, with no focal stenosis, thrombi or plaques. A retrospective review of the transthoracic echocardiogram highlighted some findings that provided the first clue towards the presence of an ARCAPA, including prominent transeptal collateral vessels on colour Doppler imaging (**Video 2**), pulmonary artery dilatation (PA dimension 34mm) and a prominent left main coronary artery (**Supplementary Video S1**). Incidentally, extensive bilateral pulmonary emboli in the main branch pulmonary arteries as well as segmental arteries were identified as the true cause of troponin elevation (**Supplementary Figure S4**), with no contrast reflux into the inferior vena cava.

The patient remained pain-free from admission and was commenced on rivaroxaban (15 mg twice daily for 21 days, 20mg once daily thereafter), and antiplatelet agents were stopped. Inpatient cardiac MRI confirmed an absence of myocardial infarction on late gadolinium enhancement imaging, with good global left ventricular systolic function. Pertinently, right ventricular free-wall oedema was identified via T2 STIR (**Supplementary Figure S5**), associated with mild right ventricular dilatation according to indexed volume, right ventricular hypertrophy (maximum wall thickness 6mm) and low-normal right ventricular ejection fraction (52%); these findings correlated with the right precordial T wave inversion and pulmonary embolic findings, consistent with right heart strain.

Follow-up

The patient remained well throughout admission and was discharged on rivaroxaban. Thrombophilia screening was unremarkable (including CT-thorax-abdomen-pelvis for malignancy; antiphospholipid antibodies, lupus anticoagulant, protein C, protein S and Factor V Leiden).

As per ESC adult congenital heart disease (ACHD) guidelines (1), anomalous origin of the coronary arteries are separated into anomalous left coronary artery from the pulmonary artery (ALCAPA) and anomalous right coronary artery from the pulmonary artery (ARCAPA). ALCAPA is associated with high mortality in infancy; and when diagnosed in adulthood, is more likely than ARCAPA to be pathologically significant (2). Patients may present with symptoms of myocardial ischaemia, ventricular arrhythmia, syncope or heart failure due to left-to-right shunting, and even sudden death (1-3). The ESC recommends surgery (Class I, level of evidence C) in patients with ALCAPA. However, incidental identification of asymptomatic ALCAPA in the elderly is not uncommon (2), and as clinicians we must always adopt a balanced approach to patient care.

Conversely, ARCAPA is commonly a benign condition that is incidentally diagnosed. Surgery for ARCAPA is recommended when symptoms are demonstrably attributable to the coronary anomaly (Class I), whereas there is a Class IIa recommendation for surgery in those with associated ventricular dysfunction or imaging evidence of ischaemia (1).

During follow-up, our patient underwent a normal outpatient treadmill exercise stress test, reaching heart rate targets after exercising for 10 minutes on the Bruce protocol without ischaemic symptoms. Our case was discussed at the regional ACHD multidisciplinary meeting, and the patient has currently opted for a conservative management approach.

Learning points

The ESC 2020 ACHD guidelines recommends for all ACHD patients to be reviewed once in a specialist centre regardless of the complexity of the heart defect. ARCAPA is a rare condition with a reported incidence of 0.002% (4). When compared to ALCAPA, ARCAPA is more likely to be diagnosed incidentally in older patients, and surgery is only indicated if there is evidence of symptoms, LV dysfunction or inducible ischaemia.

The current case perfectly illustrates the incidental nature of ARCAPA, as well as the unique roles multimodality imaging can play in the evaluation of patients with ACHD. A bedside transthoracic echocardiogram may first hint at a coronary anomaly with features as per our case. CTCA is well poised for the elucidation of anomalous coronary anatomy, and the superiority of cardiac MRI over standard transthoracic echocardiography for the structural and functional assessment of the right heart, as well as its tissue characterisation capabilities. Where feasible, exercise stress testing is deemed more physiological and has a Class IC recommendation for assessing ischaemia in patients with anomalous coronary anatomy (1). However, direct comparison between pharmacological and non-pharmacological stress are lacking, with some limited experience on the use of pharmacological stress in children and adults with anomalous coronary anatomy (5, 6).

Novel teaching points:

- An anomalous coronary artery from the pulmonary artery may either be an anomalous left coronary artery arising from the pulmonary artery (ALCAPA) or an anomalous right coronary artery arising from the pulmonary artery (ARCAPA).

- ALCAPA is commonly associated with high infant mortality, whereas ARCAPA tends to be incidentally diagnosed in adults.

- Invasive coronary angiography, cardiac MRI, CT and echocardiography each offer a unique perspective in the evaluation of this condition.

- Surgery for ARCAPA is recommended in the presence of symptoms, ventricular dysfunction, or inducible ischaemia.

- Stress testing (exercise preferred over pharmacological stress) is key in the risk stratification of asymptomatic ARCAPA patients.

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Video 1. Right radial approach diagnostic coronary angiogram, right-anterior-oblique cranial projection. Contrast injected into left coronary artery is seen draining via epicardial connections to the right coronary artery into the pulmonary artery.

Video 2. Echocardiography colour Doppler imaging from the apical-4-chamber and parasternal-short-axis views, demonstrating the presence of prominent transeptal collateral vessels that are characteristic for an anomalous origin of a coronary artery arising from the main PA.