

Brief Report

The sinister course of an intramural right coronary artery

Deane L. S. Yim,¹ Mark C. K. Hamilton,² Robert M. R. Tulloh¹

¹Bristol Congenital Heart Centre; ²Department of Clinical Radiology, University Hospitals Bristol NHS Foundation Trust, Upper Maudlin Street, Bristol, United Kingdom

Abstract We report the case of an adolescent who was presented with long-standing exertional symptoms, and was diagnosed with an anomalous right coronary arterial origin arising above the commissural junction between the left and right aortic sinus, with inter-arterial and intramural compression. The precise origin of this lesion outside the aortic sinuses is unusual, and multi-detector computed tomography gave excellent definition and spatial resolution of the anomalous origin and course. It is crucial to have a high index of suspicion of exertional symptoms, as sudden death may be the first manifestation of an anomalous coronary artery.

Keywords: Anomalous coronary; sudden death; opposite sinus; computed tomography

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Case report

A 16-year-old boy presented with a long-standing history of chest discomfort, palpitations, and dizziness on exercise. He felt anxious when these events occurred and had limited his activity levels.

Cardiovascular examination was normal. A 12-lead electrocardiogram and holter monitor was unremarkable apart from the isolated monomorphic ventricular ectopics. Cross-sectional echocardiography reported good ventricular function, but the coronary arterial origins were not well visualised. On exercise testing, he remained asymptomatic and tolerated 20 minutes of exercise using the standard Bruce protocol. However, he was noted to have 1 millimetre ST depression in the inferior leads during recovery, which was resolved after 30 minutes of rest.

High-resolution computed tomography demonstrated a high take-off of the left coronary artery at the level of the sinutubular junction above the left aortic sinus. This was of a good size and was not obstructed. The right coronary arterial origin arose anomalously from above the commissure between

the left and right aortic sinus, and had an aberrant inter-arterial and intramural course with proximal compression in the side-to-side plane by the great vessels (Fig 1a and b).

The patient underwent surgical de-roofing of the anomalous intramural right coronary artery. He developed pericarditis post-operatively, but otherwise made an uneventful recovery. He continues to be well and symptom-free on short-term follow up.

Discussion

We report an interesting case of an anomalous right coronary arterial origin from the supra-commissural aspect between the left and right coronary sinus, and demonstrated that multi-detector computed tomography gave excellent definition and spatial resolution of the anomalous origin, intramural and inter-arterial course. It is crucial to have a high index of suspicion of exertional symptoms, as sudden death may be its first manifestation.

The incidence of coronary arterial anomalies in otherwise normal heart ranges from 0.2% of young patients undergoing echocardiography to 1.2% of patients at angiography.^{1,2} The incidence was much higher at 5.6% in a study of adults undergoing coronary angiography for investigation of coronary

Correspondence to: Dr R. M. R. Tulloh, DM, FRCP, FRCPC, Bristol Congenital Heart Centre, University Hospitals Bristol NHS Foundation Trust, Upper Maudlin Street, Bristol BS2 8BJ, United Kingdom. Tel: +44 117 342 8176; Fax: +44 117 342 8857; E-mail: Robert.Tulloh@uhbristol.nhs.uk

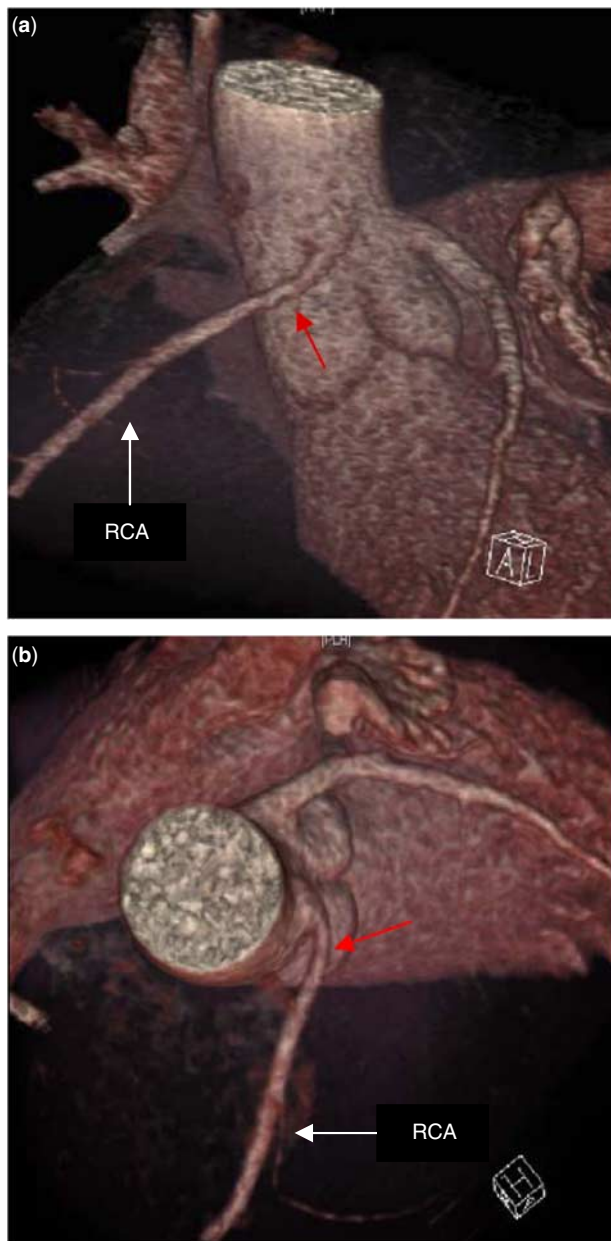


Figure 1.
 (a) Three-dimensional high resolution computed tomography reconstruction of the right coronary artery arising anomalously from the commissural margin between the right and left facing sinuses. (b) The right coronary artery has a tangential origin and follows an aberrant course between the great arteries with proximal compression (red arrow).

artery disease, and 1.1% of this cohort was found to have an ectopic right coronary arterial origin arising outside the borders of the right coronary sinus.³

Solanki et al³ precisely described an ectopic right coronary artery as one that originates outside the posterior two-thirds of the right coronary sinus. Using this definition, the most common origin of

an anomalous right coronary artery is within the anterior third of the right coronary sinus, followed by an anomalous origin from the left-facing sinus. It rarely originates from the non-coronary sinus, left anterior descending or circumflex arteries, pulmonary artery or descending aorta.^{4,5}

Our patient was interestingly found to have an anomalous location of the right coronary ostium from outside the sinuses, arising from the supra-commissural margin between the left and right coronary sinuses. This anomaly has not been specifically defined in Angelini's extensive classification of coronary anomalies,² nor has it been reported in large series as a cause of an ectopic coronary arterial origin. The reasons for this are unclear – either the lesion described in our case is exceedingly rare, or it is grouped by authors to be a form of an anomalous right coronary arterial origin from the left or right sinus of Valsava.

An anomalous coronary artery gains significant clinical importance if the arterial origin takes an intramural and/or inter-arterial course, potentially leading to coronary compression, myocardial ischaemia, and sudden death. This is mainly reported in the setting of an anomalous origin from the opposite-facing coronary sinus. A slit-like ostium, oblique angulation or high take-off are associated with an anomalous coronary arterial origin, and may further interrupt coronary flow and cause arterial vasospasm.

Our patient had chronic exertional symptoms with an unremarkable baseline work-up. Computed tomography was arranged due to the suspicious history and the fact that coronary arterial arrangement was not adequately seen on echocardiography. It reflects the importance of taking a careful and thorough history, despite symptoms being a feature in only 38% of individuals who have a malignant coronary arterial abnormality. Younger patients are more likely to die suddenly or during exercise compared with older patients aged greater than 30 years of age, and symptoms are more common shortly before sudden death.^{6,7} Although the presence of symptoms do not predict the risk of sudden death,⁷ it is crucial to have a high index of suspicion for cardiac disease as sudden death may be an avoidable event.

Indeed, coronary arterial anomalies are the second most common cause of sudden cardiac death in competitive athletes,^{6,7} and has been reported to account for 19% of sudden death in young athletes.⁸ The high-incidence of cardiac events in athletes may relate to arterial compression of the anomalous coronary artery and interference in diastolic coronary perfusion in increased cardiac output states.⁹

The diagnostic yield of echocardiography is high, provided the clinician is confident with defining

coronary arterial origins and its course.¹⁰ Further non-invasive imaging is required if there is a high suspicion based on symptoms, or if the coronary arteries are not well defined on echocardiography.

Multi-detector computed tomography gave excellent two- and three-dimensional delineation of the anomalous right coronary arterial origin and course. It not only assisted with the diagnosis, but also provided the surgeon with a clear visual image of the precise right coronary arterial origin between the left and right sinuses. Focused coronary imaging endeavours to reduce the amount of contrast used and the dose of ionising radiation. Coronary magnetic resonance angiography requires little to no contrast or radiation, however it may yield less spatial resolution than computed tomography and cannot be used with pacemaker devices.

The management of a symptomatic patient with an anomalous coronary arterial origin includes unroofing of the anomalous artery, translocation, ostioplasty or bypass grafting. There are good short-to-medium term but no long-term outcomes for unroofing and bypass grafting.² Many authors favour surgical correction even in asymptomatic cases, over medical therapy or discouraging an active lifestyle, in order to prevent a catastrophic event.^{1,2}

In summary, it is imperative to high index of suspicion of exertional symptoms and to investigate thoroughly, so that a catastrophic event may be avoided. We found that multi-detector computed tomography gave excellent definition and spatial resolution of the unusual origin and intramural right coronary arterial course.

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