Rare disease

Resolution of ischemia with β -blocker in a patient with anomalous right coronary arterySanjay Kumar,¹ Sashank Kaushik,² Alain Vaval,¹ Qian Zhao,¹ Jason M Lazar,¹ Richard Grodman¹¹Cardiovascular Medicine Department, State University of New York Downstate Medical Center, Brooklyn, New York, United States;²Internal Medicine Department, Brookdale University Hospital and Medical Center, Brooklyn, New York, United States**Correspondence to** Dr Sashank Kaushik, dr.skaushik@gmail.com**Summary**

Anomalous origin of right coronary artery (RCA) has been associated with angina, myocardial infarction and sudden death. Often these patients are referred for surgery. However, long-term β -blocker therapy instead of surgery has been used in some cases with success. The authors report a case of anomalous RCA where resolution of ischemia with β -blocker was objectively demonstrated on serial stress testing.

BACKGROUND

The prevalence of coronary artery anomalies (CAA) in patients undergoing coronary angiogram can be up to 5.64%.¹ The majority of these anomalies are benign; however, others are associated with significant morbidity. We report the case of a patient where the evaluation of chest pain revealed an anomalous right coronary artery (RCA) originating from the left main ostium.

CASE PRESENTATION

A male patient in his late 50s from Caribbean islands, presented with complaint of chest pain associated with emotional stress and sometimes with exertion. He also noted shortness of breath with exertion. These symptoms started insidiously 2–3 months prior when he had to climb stairs multiple times. He felt relief quickly with rest. He denied dizziness or passing out. There is no history of diabetes, hypertension or hyperlipidemia. He was admitted to the telemetry ward and started on aspirin, enoxaparin and a β blocker (metoprolol). His ECG showed sinus rhythm with right bundle branch block (RBBB). His cardiac enzymes were negative.

INVESTIGATIONS

The next day he underwent an exercise stress test. He walked on the treadmill for 12 min and complained of chest pain immediately upon stopping. This responded to one sublingual nitroglycerine rather quickly. The ECG showed >2 mm ST depression in the anterior leads. The next day selective coronary angiogram was performed that showed anomalous origin of RCA from the left main ostium. Its course could not be reliably appreciated on angiogram. The patient was continued to be monitored on telemetry without incident.

CT angiography of the coronary arteries was performed that confirmed the anomalous origin of RCA (anRCA). The RCA took an inter-arterial course (ie, between the ascending aorta and main pulmonary artery sometimes referred as malignant course) and appeared to be narrowed in the proximal part (figure 1). The mid and distal RCA had

normal calibre. In the mean time, the patient was apprehensive about the prospect of cardiac surgery. On day 7, a nuclear stress test was performed in order to demonstrate ischaemia. The patient again walked on a treadmill (Bruce protocol) for almost 12 min (12.9 metabolic equivalents (METs)). There was only borderline ST depression in the anterior leads this time (figure 2). Nuclear images showed attenuation artefact in inferior wall but no reversible ischaemia (figure 3). Due to the borderline reporting of the nuclear stress test, the patient underwent exercise stress echocardiography on the 9th day. He again walked for almost 12 min (12.9 METs) and denied chest pain. His ECG was negative for ischaemia (figure 2).

DIFFERENTIAL DIAGNOSIS

- ▶ Acute myocardial infarction
- ▶ Acute coronary syndrome (non-ST-elevation myocardial infarction, unstable angina).

TREATMENT

He was continued on metoprolol 25 mg orally twice daily and was advised to avoid heavy exertion.

OUTCOME AND FOLLOW-UP

He was reported being asymptomatic during his follow-up office visit over 6 months.

DISCUSSION

The prevalence of coronary artery anomalies (CAA) in patients undergoing coronary angiogram can be up to 5.64%.¹ The majority of these anomalies are benign; however, others are associated with significant morbidity. We report the case of a patient where the evaluation of chest pain revealed an anomalous RCA originating from the left main ostium. The patient subsequently found relief in symptoms with a β blocker. Abatement of ischemia was observed on serial stress testing. Often such cases are referred for coronary artery bypass surgery but incidental resolution of symptoms with β blockers has been reported.² We discuss the clinical importance of

Figure 1 CT angiogram of heart. LA, left atrium; LM, left main branch; RCA, right coronary artery; RVOT, right ventricular outflow tract.

Figure 2 ECG changes during stress testing.

Figure 3 Nuclear single-photon emission CT images.

Figure 4 Illustrations of take off angle in a normal right coronary artery (RCA) origin and in anomalous RCA origin. anRCA, anomalous origin of RCA; LM, left main branch; L, left coronary cusp; LCX, left circumflex; LAD, left anterior descending; NC, non-coronary cusp; R, right coronary cusp.

this anomaly and review the current literature regarding therapy.

In earlier studies, anRCA patients did not experience death.^{3 4} However, now it has been established that anRCA is associated with sudden or exercise related deaths albeit less likely than anomalous left coronary artery.⁵ AnRCA can have a variable take off and course before it supplies the intended myocardium. One particular type where the anRCA has an acute angle take off (<45 degrees) and an inter-arterial course between aorta and pulmonary artery, is associated with mortality (figure 4). Recent studies have shown that the anRCA exits the aorta at an acute angle and may run through the aortic wall (intramural) for few centimetres before taking a normal round calibre distally. Other

than this anatomy, young age (<30 years), male gender, exercise and symptoms are associated with worse prognosis.^{5 6}

Based on the cumulative data, Paolo Angelini has suggested intervention (stenting or surgery) for anRCA for patients with 1) disabling symptoms or high risk of sudden death, 2) area stenosis more severe than 50% with respect to the distal vessel seen on intravascular ultrasound, 3) a large dependent myocardial territory (more than a third of the total) and 4) reversible ischaemia on nuclear stress test.¹

Our patient did have an acute angle origin with some area narrowing in proximal segment but it remains uncertain whether the proximal segment is intramural at its origin due to slice thickness of modified discrete cosine transform. Fortunately, patient's stress nuclear test showed no definite sign of ischaemia. His age (58 years) was also a favourable prognostic predictor.

Medical therapy encompassing observation, avoidance of severe exertion and β -blocker has been used especially in cases without poor risk predictors. It is suspected that β -blockers are probably as effective as avoidance of severe exertion. Kaku *et al* reported medical management of 56 cases of CAA (79% anRCA). These patients were middle aged to older and 12 received medications including β -blockers. During the follow-up period from 2 months to 15 years, no deaths were directly attributable to CAA.⁷

β blockers have been shown to reduce mortality in patients with coronary artery disease. The mechanism of action appears to be attenuation of exercise induced tachycardia and reduction in systolic blood pressure. In this case the β -blocker was started on admission. It had sufficient time (half life 3–7 h) to reach steady levels during the hospitalisation. In successive stress tests, there was a reduction of peak heart rate and double product (heart rate $\times$ systolic blood pressure) at similar work loads. This would be attributable to the β -blocker (figure 3). β -blockers have been shown to improve measures of exercise duration, maximum ST depression and time to symptomatic and asymptomatic ischemia in patients with angina.⁸ Whether the β -blocker provided protection in CAA by similar mechanism cannot be proven in this case study.

Additionally, we cannot be fully sure whether the β -blocker was the sole reason for ischaemia resolution. The patient could have had paroxysmal coronary artery vascular spasm that was only evident on the first stress testing.

Rather, vascular spasm may worsen with β -blockers. The ST depression in anterior leads on first testing could have been entirely due to RBBB however, the gradual regression of ST segment depression in subsequent ECG argues against it.

Unique to this report is that patients with anRCA may have some but not all the poor risk predictors. Advanced age, absence of ischaemia in nuclear imaging and absence of severe disabling symptoms could favour medical therapy over angioplasty or surgery. In our knowledge, the first time resolution of ischemia on successive stress testing has been documented in a patient with anRCA.

Learning points

- ▶ It has been established that anRCA is associated with sudden or exercise related deaths albeit less likely than anomalous left coronary artery.
- ▶ One particular type where the anRCA has an acute angle take off (<45 degrees) and an inter-arterial course between aorta and pulmonary artery, is associated with mortality.
- ▶ Other than this anatomy, young age (<30 years), male gender, exercise and symptoms are associated with worse prognosis.
- ▶ Based on the cumulative data, Paolo Angelini has suggested intervention (stenting or surgery) for anRCA for patients with 1) disabling symptoms or high risk of sudden death, 2) area stenosis more severe than 50% with respect to the distal vessel seen on intravascular ultrasound, 3) a large dependent myocardial territory (more than a third of the total) and 4) reversible ischemia on nuclear stress test.
- ▶ In this case reduction of peak heart rate, double product, improved exercise duration in successive stress tests could be attributable to β -blockers, however, we cannot be completely sure.

Competing interests None.

Patient consent Not obtained.

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