CASE REPORT

Double trouble: hypertrophic cardiomyopathy coexistent with malignant anomalous right coronary artery

Stephen Liddy,¹ Andrew K Roy,² Charles McCreery³

¹Department of Cardiology, St James's Hospital, Dublin, Ireland

²Department of Cardiology, Mater Misericordiae University Hospital, Dublin, Ireland

³St Vincent's University Hospital, Dublin, Ireland

Correspondence to

Dr Stephen Liddy, stephenliddy@gmail.com

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SUMMARY

A 40-year-old man with no cardiac history was admitted for evaluation of atypical chest pain of 1-month duration. On investigation he was found to have hypertrophic cardiomyopathy and an anomalous right coronary artery arising from the left sinus of Valsalva. This is of great clinical significance as both these conditions are independently associated with sudden cardiac death. Following extensive risk assessment and detailed discussion with the patient, he was discharged on medical therapy, comprising bisoprolol, aspirin and rosuvastatin. He has remained free of symptoms for 1 year. This report describes the rare coexistence of these two distinct clinical entities, examines the various treatment options and provides support for medical management as an acceptable treatment strategy in appropriately selected cases.

BACKGROUND

Owing to the rarity of this unusual coexistence, there are limited data available to guide its management. This case provides support for medical management as an acceptable treatment modality in appropriately selected cases following risk assessment.

CASE PRESENTATION

A 40-year-old male smoker was admitted to our hospital for evaluation of atypical chest pain of 1-month duration. He had no personal and family history of heart conditions. He described a history of severe, central chest pain that was unrelated to exertion and usually lasted 5–10 min. It was not associated with dyspnoea, presyncope or palpitations. On admission, his blood pressure was 124/88 mm Hg and his pulse rate was 80 bpm. A tele-systolic murmur, heard loudest over the lower left sternal edge, was noted.

INVESTIGATIONS

Troponin levels were normal and remained normal throughout his admission. ECG on admission showed 1 mm upsloping ST segment elevation in leads V1 and V2, and T-wave inversion in leads V5 and V6. Two-dimensional echocardiography revealed asymmetric septal hypertrophy (interventricular septum 2.1 cm, posterior wall 1.2 cm) resulting in a peak instantaneous left ventricular outflow tract gradient of 121 mm Hg and a mean gradient of 30 mm Hg. In addition, moderate-to-

severe mitral regurgitation due to systolic anterior leaflet motion was also noted.

Cardiac catheterisation was performed 36 h post admission. The left ventricular end-diastolic pressure was 29 mm Hg and the pull-back gradient between the left ventricle and the subaortic outflow tract was 40 mm Hg. Coronary angiography revealed an anomalous right coronary artery (RCA) originating from the left sinus of Valsalva ([figure 1](#), [video 1](#)). The left anterior descending and circumflex arteries were normal in origin and distribution, with a 70% lesion at the origin of the first diagonal branch. The coronary arteries were otherwise unobstructed. On exercise stress testing, the patient reached his target heart rate in 12 min without symptoms or ECG changes suggestive of ischaemia. In addition, ambulatory ECG monitoring throughout admission revealed no arrhythmia.

MRI demonstrated findings consistent with hypertrophic cardiomyopathy (HCM) and confirmed the presence of an anomalous RCA arising from the left sinus of Valsalva and coursing between the aorta and main pulmonary artery before assuming its normal configuration in the right atrioventricular groove ([figure 2](#)). In addition, there were small punctate areas of fibrosis within the basal anteroseptal segment occupying 25% of the myocardial thickness. There was no evidence of myocardial bridging and no regional wall motion abnormalities were detected.

Figure 1 Coronary angiogram showing an anomalous right coronary artery arising from the left sinus of Valsalva. The left coronary cusp is non-selectively engaged with a 5 French Tiger catheter.

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Video 1 Coronary angiogram showing an anomalous right coronary artery arising from the left sinus of Valsalva. The left coronary cusp is non-selectively engaged with a 5 French Tiger catheter.

TREATMENT

Given the abnormal findings on the patient's admission ECG, consideration was given to stent insertion to the RCA. However, in light of the reassuring exercise stress test and ambulatory ECG recordings, in addition to the patient's preference for non-intervention, the authors have no plans for stent insertion at this time. He was discharged on bisoprolol, aspirin and rosuvastatin, but with strict advice to avoid strenuous physical activity.

OUTCOME AND FOLLOW-UP

The patient was initially followed-up at three monthly intervals with regular reassessment by echocardiography. At 12-month follow-up the patient remained well and reported no recurrence of his symptoms. The findings on the initial ECG have persisted and the mean outflow tract gradient has remained unchanged.

Given the uncertainty surrounding the magnitude of risk in this case, lifelong follow-up is required. The authors will also keep a low threshold for repeating the cardiac MRI should there be any change in the patient's clinical status.

DISCUSSION

The coexistence of HCM and a malignant anomalous RCA is uncommon with only four previously published case reports. Efthimiadis *et al*¹ described a 45-year-old man with a known history of HCM who was admitted with atypical chest pain and was found to have an anomalous RCA. Moza *et al*² reported a 29-year-old man who presented with chest pain on a background of recurrent syncopal episodes. Tyczyński *et al* described a 65-year-old man with an anomalous RCA who underwent corrective surgery for HCM.³ Finally, Dermengiu *et al*⁴ described a 20-year-old man who died suddenly during exercise, although in this case postmortem analysis revealed the presence of multiple congenital cardiac anomalies in addition to HCM and an anomalous RCA.

In normal patients, the RCA arises from the right sinus of Valsalva and courses between the pulmonary trunk and right auricle before descending to the right atrioventricular groove. Anomalies of origin of the RCA are a heterogeneous group of conditions. However, it is of particular concern when the RCA arises from the left sinus of Valsalva as it almost invariably tracks between the aorta and pulmonary artery, which can lead to extrinsic compression during systole. Moreover, it is regularly associated with an acute takeoff angle and a slit-like ostium. It is suggested that when the heart rate increases and diastolic filling times reduce, these features can lead to reduced coronary blood flow, myocardial ischaemia, arrhythmias and sudden death, particularly during exercise.⁵

The treatment options for HCM have been well described.⁶ β -Blockers or verapamil are the mainstay of medical therapy and act to slow the heart rate, thereby allowing for greater diastolic filling in addition to mitigating exercise-induced outflow tract gradients. For patients with symptoms of heart failure despite optimal medical management, and who have an outflow tract gradient of greater than 50 mm Hg, surgical septal myectomy is the preferred treatment option, with percutaneous alcohol septal ablation considered a viable alternative. For patients

Figure 2 (A) MR angiogram showing the right coronary artery arising from the left sinus of Valsalva (arrow). It demonstrates a typical hyperacute angle at its takeoff and passes between the aorta and pulmonary artery before assuming its normal configuration in the right atrioventricular groove. (B) Paraseptal long-axis MRI demonstrating significant left ventricular outflow tract obstruction due to asymmetric septal hypertrophy. The shape and size of the left ventricle are within normal limits. (A, aorta; AA, ascending aorta; LA, left atrium; LV, left ventricle; PA, pulmonary artery; RA, right atrium; RCA, right coronary artery; S, septum).

deemed to be at increased risk of ventricular arrhythmias, implantable cardioverter-defibrillator implantation is the most effective way of preventing sudden cardiac death.

In contrast to HCM, the optimal treatment for patients with anomalous coronary arteries arising from the contralateral sinus remains unclear. Treatment strategies described include medical management, stent insertion and surgical correction; however, evidence supporting one treatment modality over another is lacking. One study of 56 patients treated with β -blockers reported no sudden deaths over a follow-up period of 5 years.⁷ In a second study, 16 patients treated with β -blockers and ACE inhibitors were all alive and symptom free after 34 months.⁸ In 2000, Doorey *et al*⁹ reported a series of 14 patients who were successfully treated with stent insertion. Several authors have analysed outcomes following surgical correction; however, these studies are limited by small patient numbers and short follow-up periods.⁵ Nevertheless, surgical revascularisation remains the most widely advocated treatment strategy and is supported by the joint guidelines of the American College of Cardiology and American Heart Association for cases of anomalous right coronary arteries with evidence of ischaemia.¹⁰

Learning points

- ▶ Coronary artery anomalies are a diverse group of conditions but the anomalous origin of the right coronary artery (RCA) from the left sinus of Valsalva is of particular concern.
- ▶ The coexistence of hypertrophic cardiomyopathy and an anomalous RCA is of great clinical significance as both these conditions are independently associated with sudden cardiac death.
- ▶ Optimal management of anomalous coronary arteries arising from the contralateral sinus remains unclear. Treatment strategies described include medical management, stent insertion and surgical revascularisation.
- ▶ This report provides further support for medical management as an acceptable treatment strategy in appropriately selected cases.

Competing interests None.

Patient consent Obtained.

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