CASE REPORT

Anomalous aortic origin of a coronary artery (AAOCA): A case report

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SUMMARY

Anomalous Aortic Origin of a Coronary Artery (AAOCA) is a rare anomaly of the coronary artery with a considerable risk of sudden cardiac death due to ischaemia of the heart. Symptoms may include chest pain on exertion, breathlessness or dizziness. We encountered a case of a 46-year-old female who complained of exertional chest pain with a positive-stress test and subsequently diagnosed with AAOCA through CT angiography (CTA). She successfully underwent a coronary artery bypass graft (CABG) surgery using a saphenous vein graft with uneventful recovery. Right internal mammary artery (RIMA) was not used as it was flimsy and the flow was very poor.

INTRODUCTION

Anomalous Aortic Origin of a Coronary Artery (AAOCA) is a rare anomaly of the coronary artery. The prevalence of anomalous coronary artery originating from the opposite sinus of Valsalva (ACAOS) was reported to be about 0.84%. Of all the ACAOS, the most common anomaly is the left circumflex artery originating from the right sinus of Valsalva followed by right coronary artery from left sinus of Valsalva.[1] Although rare, AAOCA can be life-threatening as there is an increased risk of sudden cardiac death. We present a case report an anomalous right coronary artery from the left sinus of Valsalva with an interarterial course in a 46-year-old female who presented with chest pain on exertion and shortness of breath.

CASE REPORT

A 46-year-old female netball player presented to us with exertional chest pain for the past two months. It was intermittent and mainly aggravated by strenuous exercise, associated with palpitations. She had had an episode of syncope at the age of 15 years while playing sports in school. However, there was no further follow-up or treatment. She does not have any other medical illness and there was no family history of congenital heart disease. Her physical examination was unremarkable with normal blood pressure, normal jugular venous pressure and normal heart sounds.

Her resting ECG showed sinus rhythm with frequent premature ventricular contractions (PVCs). The treadmill stress test was positive for ST segment depression in the inferior leads. Echocardiography was normal with an ejection fraction of 70%.

A cardiac catherisation was subsequently performed which showed common origin of both left and right coronary arteries from the left sinus of Valsalva [Fig 1]. Percutaneous coronary intervention (PCI) was attempted but the RCA could not be cannulated. A detailed anatomical analysis using the computed tomography (CT) angiography was done which confirmed the angiogram findings and the proximal segment of the right coronary artery was found to course in between the aorta and the pulmonary artery [Fig 2]. There was no obvious stenosis in the mid and distal segments of RCA.

She was referred to our centre for further management of her condition. She successfully underwent a coronary artery bypass grafting (CABG) surgery with a saphenous vein grafted to the mid-right coronary artery (RCA). The right internal mammary artery (RIMA) was initially harvested but unfortunately, it was flimsy with very poor blood flow and hence it was not suitable to be used as a conduit. The postoperative recovery was uneventful, and she was discharged on day 9 postoperative.

CASE DISCUSSION

The clinical presentation of AAOCA varies depending on the anatomical course of the affected coronary artery. Unfortunately, sudden cardiac death may be the first and only clinical presentation in patients who are asymptomatic. A study by Maron BJ et al in 2003 reported that the incidence of AAOCA was found to be 13% among 286 athletes who died of cardiovascular disease (CVD).[2] The risk of SCD is even higher if the coronary artery takes an interarterial course which was reported to be about 87% among patients with AAOCA.[3] The pathophysiology of sudden cardiac death in these patients remain unknown but several mechanisms have been proposed. In an interarterial course, there is vascular compression between the root of aorta and pulmonary artery, especially during the systolic phase due to dilatation of the two large vessels. This can reduce the blood flow to the heart, leading to cardiac ischaemia. Some patients may present with CAD-like symptoms such as chest pain, shortness of breath, syncope or dizziness and these symptoms are markedly present during exercise as there is an increase in the cardiac output, leading to greater dilatation of both

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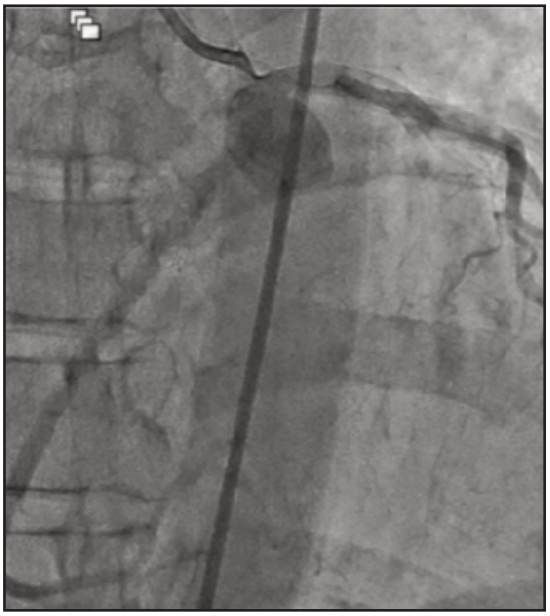

Fig. 1: Coronary angiogram showing both left and right coronary arteries arising from the left sinus of Valsalva.

vessels. Other mechanisms include intramural course of the anomalous coronary artery in which there is lateral compression from the contractions of myocardium, resulting in smaller diameter of the lumen as well as severe angulation of a retroaortic coronary artery.

Physical examination rarely reveals any significance except if there is a concomitant congenital heart disease such as tetralogy of Fallot or transposition of the great vessels. Most cases of AAOCA are incidental findings on investigations. The first modality to detect an anomalous coronary artery would be an echocardiography but this can be very challenging due to its operator-dependency. The best choice of investigation to delineate the anatomical location and course of coronary artery would be coronary magnetic resonance angiography (CMRI) or CT coronary angiography but this depends on the availability of the CMRI and the cost incurred.

There is no definite guideline on the treatment of AAOCA but careful consideration must be undertaken due to high risk of sudden cardiac death in these patients. Asymptomatic patients may not need any intervention but they require regular follow-up for cardiac assessment. Some indications for intervention include a positive-stress test, an interarterial course and patients who have symptoms. These patients may be managed either by medical treatment, percutaneous coronary intervention (PCI) or surgical correction. However, there have been no studies on the benefits of one treatment over another. Medical treatment such as antianginal therapy, beta-blockers or angiotensin-converting enzyme inhibitors (ACE-I) may provide temporary symptomatic relief. Our patient received a trial of anti-anginal therapy which did not really help relieve her symptoms.

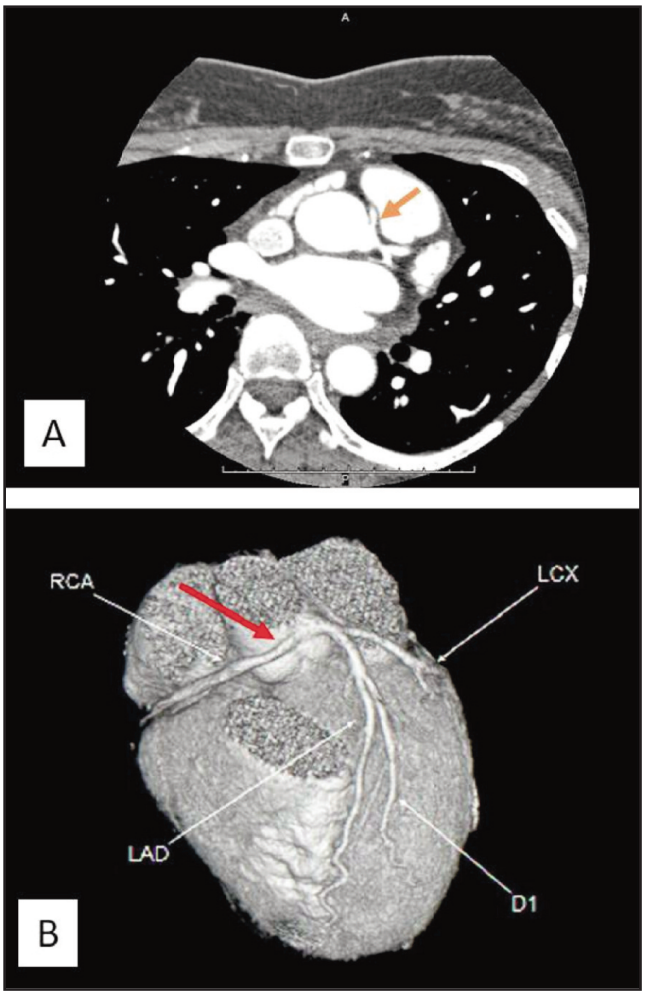

Fig. 2: (A) CT scan of thorax showing the RCA originating from the left sinus of Valsalva (B) Coronary Computed Tomography Angiogram (CCTA) showing the interarterial course of the proximal segment of right coronary between the aorta and pulmonary artery.

A prospective case study in 2015 reported the use of PCI stenting with 90% of success rate but it requires highly skilled interventional cardiologists to deploy the stent successfully as well as a proper choice of guide catheter.[4] However, PCI may be difficult in an interarterial course due to vascular compression such as in our case. Therefore, a surgical correction may be the only option and is usually needed to relieve symptoms, reduce ischaemia and prevent sudden cardiac death. There are various surgical techniques for AAOCA but according to a survey from cardiothoracic surgeons worldwide, the most preferred technique of surgical correction would be unroofing of coronary ostium (45%) followed by CABG using the either the RIMA (30%) or saphenous vein graft (10%) and coronary reimplantation as the least preferred option[5]. However, there have been no clinical studies on the outcomes of a particular choice of operation as it depends on the preference of the surgeons.

CONCLUSION

AAOCA remains an important congenital heart disease due to risk of sudden cardiac death. High index of suspicion is required in young patients who present with CAD-like symptoms. Surgical treatment remains the preferred choice of management to correct the anomaly. Further clinical trials and consensus guidelines are required in managing these patients and assessing the outcomes.

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