CASE REPORT

Anomalous origin of the circumflex coronary artery presenting with ventricular fibrillation cardiac arrest

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SUMMARY

We report a case of an incidental finding of an anomalous left circumflex coronary artery arising from the right pulmonary artery that effectuated a ventricular fibrillation cardiac arrest in a woman aged 34 years. This rarity was detected during routine work-up to delineate the cause of this arrhythmia. Our patient had a background of double-outlet right ventricle and a ventricular septal defect, which was repaired with a Dacron patch and a left ventricle patch over to the aorta at age 14 months. Angiographic study at the time of her presentation showed anomalous origin of the left circumflex artery originating from the right pulmonary artery; this was discussed in multispecialty team meeting and surgical intervention was recommended; eventually, surgery was performed with reimplantation of the anomalous circumflex artery into the ascending aorta. We highlight the importance of early angiographic studies in patients with known congenital heart defects and emphasise the optimal strategy of treatment.

BACKGROUND

Coronary artery anomalies are a large group of congenital origins, which may present with different signs and symptoms depending on the location of the anomaly and severity of the involvement of the artery.¹ The incidence of coronary artery anomaly rate is 5.6% globally, with 0.92% of anomalous involving the origin of the right coronary artery and 0.15% anomalous origin of the left coronary artery.¹ Presentations vary from being symptomatic as early as first week of life to being asymptomatic until late age of life or presenting with sudden death, all depending on the presence of collaterals supplying the region of the anomalous origin of the artery.² However, while the most common presenting symptom is exertional dyspnoea,³ it has been rarely described in that literature that the anomalous circumflex artery can present with sudden cardiac arrest.

CASE PRESENTATION

We report a case of a woman with anomalous origin of the left circumflex coronary artery from the right pulmonary artery presenting with ventricular fibrillation cardiac arrest suffered while attending a family party was admitted to our centre in 2016. Immediate bystander cardiopulmonary resuscitation was initiated and restoration of spontaneous circulation was achieved within 11 min after two shocks being delivered.

Her medical history included a double-outlet right ventricle and a ventricular septal defect, which

was repaired with a Dacron patch and a left ventricle patch over to the aorta at age 14 months. In addition, she had pulmonary artery stenosis and a bilateral superior vena cava (SVC) with the left SVC draining into the coronary sinus. On admission, the transthoracic echocardiography showed good biventricular function with moderate mitral regurgitation. A coronary angiogram revealed dilated left main stem coronary artery, dilated and tortuous left anterior descending coronary artery; very tortuous right coronary artery arising from the right coronary sinus with extensive coronary collaterals; however, the circumflex coronary artery was originating from the right pulmonary artery (figure 1 and video 1). This anomaly was further confirmed on cardiac MRI (figure 2), her cardiac MRI also showed inducible subendocardial perfusion defect in the basal and midlateral wall (video 2). A single-chamber implantable cardioverter defibrillator was inserted as a preventive measure for her arrhythmia and her case proceeded to be discussed in the joint cardiology and cardiothoracic surgery meeting at our centre. A decision was made to proceed to surgical reimplantation of the circumflex artery onto the aorta. Surgery was performed via median sternotomy with establishment of cardiopulmonary bypass via femoral cannulations; the heart was arrested using cold cardioplegia, cardiopulmonary bypass time was 188 min and aortic cross-clamp time of 63 min. Postoperatively, she was transferred to the intensive care unit and extubated on day 1. Her recovery was unremarkable and she was discharged home on day 9 postsurgery. At 3 months follow-up postdischarge from hospital, the patient denies any symptoms.

Figure 1 Cx, circumflex artery originating from the right pulmonary artery; LAD, left anterior descending artery originating from the left coronary cusp.

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Video 1 Coronary angiogram demonstrating the origin of circumflex coronary artery and the extensive coronary artery collaterals.

Figure 2 Cardiac MRI showing the circumflex coronary artery (Cx) originating from the right pulmonary artery (RPA).

Video 2 Cardiac MRI showing an inducible subendocardial perfusion defect in the basal and midlateral wall.

INVESTIGATIONS

- **Coronary angiography:** showed dilated left main stem coronary artery, dilated and tortuous left anterior descending artery, dilated right coronary artery arising from the right coronary sinus, and the left circumflex artery (LCX) originating from the right pulmonary artery. Extensive coronary collaterals.
- **Cardiac MRI:** confirmed origin of the circumflex artery from the right pulmonary artery, it also demonstrated an inducible subendocardial perfusion defect in the basal and midlateral wall.

DIFFERENTIAL DIAGNOSIS

The main differential diagnosis initially was thought to be ventricular arrhythmia as the main reason of her cardiac arrest; however, during the admission work-up and the identification of the anomalous origin of LCX from the right pulmonary artery, it is thought that this had contributed significantly to her sudden cardiac arrest.

TREATMENT

Our patient underwent surgical reimplantation of the anomalous circumflex artery into the ascending aorta using cardiopulmonary bypass. This resulted in very good outcome with the patient being discharged home on postoperative day 9 in good health.

OUTCOME AND FOLLOW-UP

Very good outcome was obtained, and the patient discharged home on postoperative day 9 in good health. At 3-month follow-up postdischarge, the patient denies any cardiac symptoms.

DISCUSSION

Normal coronary anatomy consists of the right and left main coronary arteries originating from the sinuses of Valsalva in the aortic root. The left main coronary artery branches into the left anterior descending and left circumflex coronary arteries. However, anomalous origins of the left coronary artery have been reported in the literature and are originating from the pulmonary artery. In that sense, anomalous origin of the LCX was first described in 1992 by Garcia *et al.*,⁴ and it is considered extremely a rare form of coronary artery anomalies. Clinical presentation of coronary anomalies are divided into two main types, those that cause significant haemodynamic instability and present at early age of life and as such necessitate early surgical intervention; or anomalies that are asymptomatic until later age of life and are unidentified unless presented with other cardiac symptoms or found incidentally, as described by Shriki *et al.*⁵ Hence, patients with anomalous origins of the circumflex artery usually present in childhood due to the association with other congenital cardiac abnormalities such as coarctation of the aorta and patent ductus arteriosus;⁶ however, the presence of isolated anomalous LCX without congenital heart defects is very rare. The most common form of presentation of such anomalies in adult life is exertional dyspnoea, sign and symptoms of ischaemic heart disease, and rarely could present with sudden cardiac arrest.³ The initial myocardial ischaemia is tolerated depending on the collateral circulations between the left and right coronary arteries, and therefore, sudden cardiac arrest usually comes after an acute myocardial ischaemia during exercise or ventricular arrhythmias generated from myocardial scar tissue.⁷

The literature at large describes only seven adult cases, with only two of the cases presenting with sudden cardiac arrest³ making our case report the third, presenting with sudden cardiac arrest. The most common form of successful treatment for such anomaly is surgical ligation and reimplantation of the anomalous coronary artery to the ascending aorta or bypass grafting, this usually gives satisfactory results with good outcome;⁸ as in our case, surgical ligation was performed and the anomalous LCX reimplanted to the ascending aorta and the patient was discharged in good health.

Patient's perspective

I am very grateful to Professor Hsia and his team, I feel very lucky for surviving such major cardiac event and I would like to thank every team member who looked after me, you saved my life and my family.

Learning points

- ▶ Anomalous origin of the left circumflex coronary artery could present in early life as part of complex structural heart defects with severe heart failure symptoms or could stay asymptomatic as late as third decade of adult life.
- ▶ We recommend that patients presenting with congenital heart disease to have early coronary angiogram to assess coronary anatomy and avoid potential sudden cardiac arrest.
- ▶ Gold standard method for diagnosis of coronary anatomy is coronary angiogram.

Coronary artery anomalies presentations are variable, as these anomalies are very rare, presentation ranges from severe heart failure symptoms at the time of birth to being asymptomatic until adulthood. The presence of congenital heart defects should raise the presence of coronary anomalies. Multidisciplinary team collaboration between a cardiologist, electrophysiologist and cardiothoracic surgeon will assist in early diagnosis of coronary anomalies and treating them at early age averts future sudden cardiac arrest.

Contributors AH and MG drafted the case report, reviewed, amended and approved by MB. TYH is the responsible cardiac surgeon who operated on the patient.

Competing interests None declared.

Patient consent Obtained.

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