Case Report

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Coronary cameral fistula to right atrium in a 10 year old male: a case report

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ABSTRACT

A coronary cameral fistula is a rare cardiac anomaly and involves an abnormal communication between one of the coronary arteries and a cardiac chamber. In most of cases, it is a congenital lesion and is asymptomatic in early life. Once symptoms appear, patient warrants early evaluation and treatment. Presently, CT Angiography is the best investigation for diagnosing the disease and the best treatment modality is surgical closure. We present a case of a 10 year old male with large coronary cameral fistula from right coronary artery to right atrium, who was symptomatic and was operated for the same.

Keywords: Coronary artery fistula, Coronary cameral fistula, Epicardial ligation

INTRODUCTION

Coronary artery fistulae (CAF) are uncommon lesions, seen only in 0.002% of the general population. Right coronary artery (RCA) is the most common site of origin (60%) of such fistulae and mostly terminate (90%) in the right side of the heart. They are usually asymptomatic in younger patients unless they are very large and haemodynamically significant. As age increases, symptoms begin to appear, and the incidence of complication rises.

CT angiography remains the best investigation for diagnosing the disease.² Treatment is by transcatheter embolisation (in selected cases) or surgical closure. Surgery gives the best results and should be performed early.³ We present a case of a 10 year old male with coronary cameral fistula from proximal RCA to rare site right atrium (RA) who was symptomatic and was operated for the same.

CASE REPORT

A ten year old male patient presented with complaints of dyspnoea on exertion and palpitation of eight months duration. Cardiovascular examination continuous murmur in the precordium. The rest of physical examination was normal. The electrocardiogram showed normal sinus rhythm with no ST/T wave changes. The chest radiograph showed cardiomegaly. All routine laboratory parameters were normal and transthoracic echocardiography revealed good left ventricular function. The RCA was dilated and a fistula could be traced from the RCA to the RA. Cardiac catheterization (Figure 1) revealed aneurysmally dilated RCA with a fistulous tract terminating in the right atrium, with the ratio of pulmonary to systemic flow (Qp/Qs) of 1.6.

Electrocardiogram gated contrast enhanced (CE) CT showed an enlarged and tortuous proximal RCA in the right atrio ventricular groove with large solitary fistulous

communication between proximal part of vertical segment of RCA and posterior wall of RA thus forming intracardiac left to right shunt consistent with a coronary cameral fistula. RA was markedly dilated with mildly dilated left ventricle (LV), other chambers were of normal size (Figure 2).

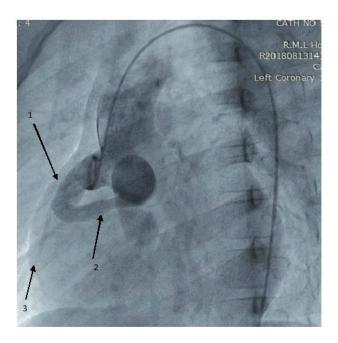


Figure 1: Coronary angiogram showing dilated portion of RCA proximal to fistula. (1) Proximal dilated RCA. (2) Fistula connecting RCA and RA. (3) Normal caliber RCA distal to fistula.

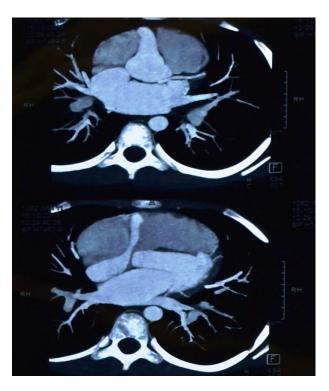


Figure 2: MDCT of heart showing dilated RCA and fistulous tract communicating with RA.

Patient was planned for surgical closure of the fistula. Chest opened through vertical sternotomy and after mobilising thymus pericardium was opened. On inspection RA was found dilated along with dilated and tortuous proximal RCA with fistulous tract of approximately 8mm diameter seems to entering RA. RCA distal to origin of fistula was of normal calibre. The fistula was mobilised epicardially in the beating heart. A ligature was placed around the fistula and was temporarily occluded (Figure 3).

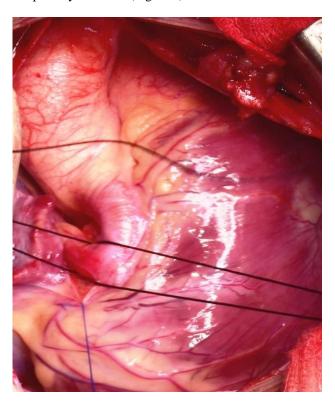


Figure 3: Intraoperative view showing dissected and looped fistulous tract.

The heart was observed for signs of ischemia, and the ECG monitored. As no sign of ischemia was evident, the ligature was tied. A second suture ligature was also placed to ensure obliteration. The chest was closed in standard manner. The patient was extubated after two hours in the intensive care unit. Patient required no ionotropic support.

Postoperative recovery was uneventful with the disappearance of symptoms and the heart murmur and the patient was subsequently discharged after two days. At discharge there was no fistula detected on transthoracic echocardiogram.

DISCUSSION

Coronary artery fistulae (CAF) are uncommon lesions, seen only in 0.002% of the general population and reported in 0.25% of patients undergoing coronary angiography. 1

Myocardial stealing distal to the site of connection is the pathophysiological mechanism responsible for symptoms in patients of coronary fistula.⁴ Since most CAFs are small, thereby myocardial blood flow is not compromised and patients are asymptomatic. As the child grows with age, fistula size increases and symptoms appear. As symptoms appear, early evaluation surgical correction is indicated.

Two-dimensional echocardiography usually establishes the diagnosis, demonstrating the origin and drainage site, or provide clues such as coronary dilation or chamber enlargement. Magnetic resonance imaging (MRI) and computed tomography (CT) cardiac coronary angiography are useful, noninvasive, and accurate imaging techniques for the diagnosing majority of such anomalies.

The anaesthetic management of these patients involves prevention of coronary steal and perioperative myocardial ischaemia, which can occur with an increased left to right shunt. Intraoperative ECG monitoring is a useful guide to detect perioperative ischaemia.⁵ TEE can detect fresh regional wall motion abnormalities intraoperatively and is being used with increasing frequency.⁶ Adequate perfusion to the myocardium beyond the fistula is to be maintained during CPB.

Although there are numerous reports of transcatheter coil embolization of these fistulae, the surgical closure of the fistula remains the gold standard until now.³ Many fistulas can be closed even without the use of CPB; however, CPB must always be available. About 50% of fistulas can be exposed and ligated or divided epicardially in the beating heart.⁷ Complications of surgery include myocardial ischaemia (3%) and recurrence (0-4%).

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