

Surgical repair of congenital coronary artery fistula: A case report

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Abstract

Coronary artery fistula is an exceedingly uncommon congenital disorder characterized by fistulous communication of a coronary artery with asystemic or pulmonary vessel or a heart chamber. Small fistulas are asymptomatic and benign while large fistulas present in a variety of ways like heart failure, arrhythmias or endocarditis. We report here a rare case of eight years old girl who presented to us with history of shortness of breath and palpitation. Her echocardiography revealed a moderately large right coronary artery fistula that was opening into right ventricle. Angiography later confirmed its presence. We surgically closed it with a pericardial patch on cardiopulmonary bypass. The girl showed uneventful recovery and her postoperative echocardiography revealed no residual fistula. She was discharged on 5th postoperative day and her follow up was also unremarkable.

Keywords: Coronary artery fistula, congenital disorder, cardiopulmonary bypass.

Introduction

Coronary artery fistula, though a non-ubiquitous condition is considered a major congenital anomaly. It is characterized by fistulous communication of a coronary artery with either a systemic or pulmonary vessel or a heart chamber.¹ The frequency of Coronary Artery Fistulas (CAF) is 0.002% in the general population.² Small fistulas usually go undiagnosed and are benign. They are unexpected findings during routine echocardiography or angiography. Nevertheless, large fistulas present in a variety of ways like heart failure, disturbed cardiac rhythms, bacterial endocarditis or even angina pectoris.^{3,4} Because of vast variety of intracardiac communications, they behave differently i.e. may appear as atrial septal defect if opening into the right atrium and as patent ductus arteriosus if opening into pulmonary artery, may result in volume overload of left atrium exactly acting like mitral regurgitation and even left ventricular overload⁵ and behaving like aortic regurgitation. Krause was first to describe the condition in 1865⁶ and Biorck and Crafoord

were the first to close it surgically.⁷ We present here first ever reported case of right coronary artery (AV) fistula in Pakistan.

This Case is being reported after taking signed consent from parents of the patient. Approval was obtained from Ethical Committee of Hospital as well.

Case Report

In November 2015, an 8-year-old girl was referred to our outpatient department of Chaudhary Pervaiz Elahi Institute of Cardiology Multan, for surgical consultation with history of shortness of breath and palpitation. General physical examination detected no abnormality except a thrill on apex of heart. Auscultation revealed a continuous murmur. Her echocardiography report showed a moderately large right coronary artery fistula opening into right ventricle and moderate tricuspid regurgitation. Her angiography showed gigantic right coronary artery and runoff of dye into the right ventricle thus confirming the fistula (Figure-1). Paediatric

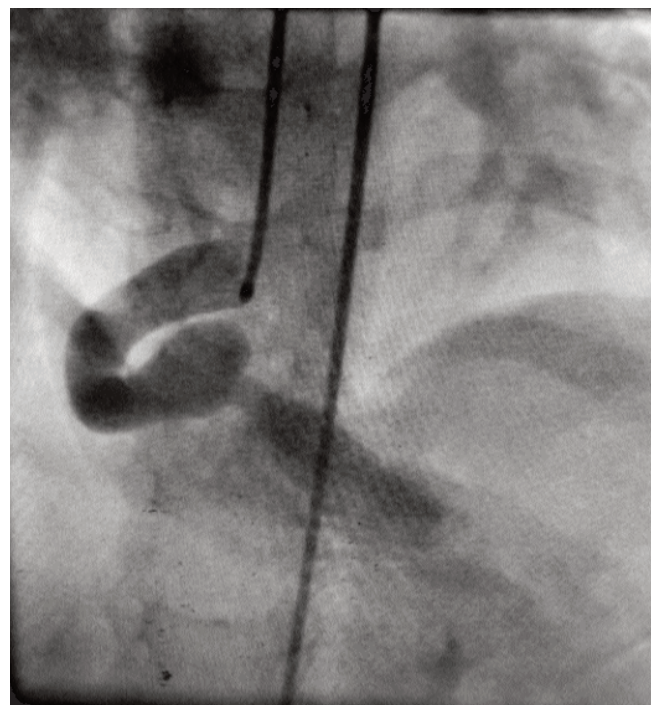


Figure-1: Selective right coronary artery angiography. Extravasation of dye from a gigantic artery beautifully illustrating the fistula.

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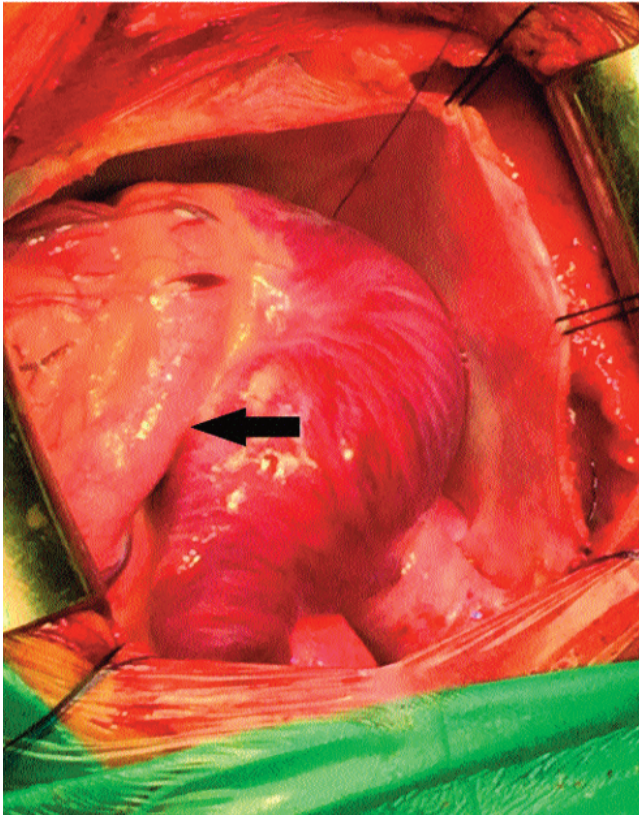


Figure-2: Hugely dilated right coronary artery.

cardiologists tried to close the fistula twice but they could not have success so the patient was referred to us for surgical closure. The patient was operated through a standard median sternotomy incision. The opening of pericardium revealed a huge dilated right coronary artery (Figure-2). A thrill was felt on the coronary artery. Cardiopulmonary bypass was established through aortic and bicaval cannulation. Cardiac arrest was ensured through cardioplegia after cross clamping the aorta. Right atriotomy revealed opening of fistula into right ventricle under the tricuspid valve. Cardioplegia through the opening was confirmed the fistula opening was closed with a pericardial patch (Figure-3). Right atrium was closed and patient was weaned off from bypass smoothly. Thrill from coronary artery had disappeared. Total bypass time was 81 minutes, cross clamp time was 52 minutes and ICU stay was 24 hours. Patient showed uneventful recovery. She was extubated within 5 hours. Her post-operative echocardiography showed no residual flow in fistulous tract, mild to moderate tricuspid regurgitation and good biventricular systolic function. She was mobilized well and was sent home on 5th post-operative day in satisfactory condition. Her follow up was pleasant and unremarkable.

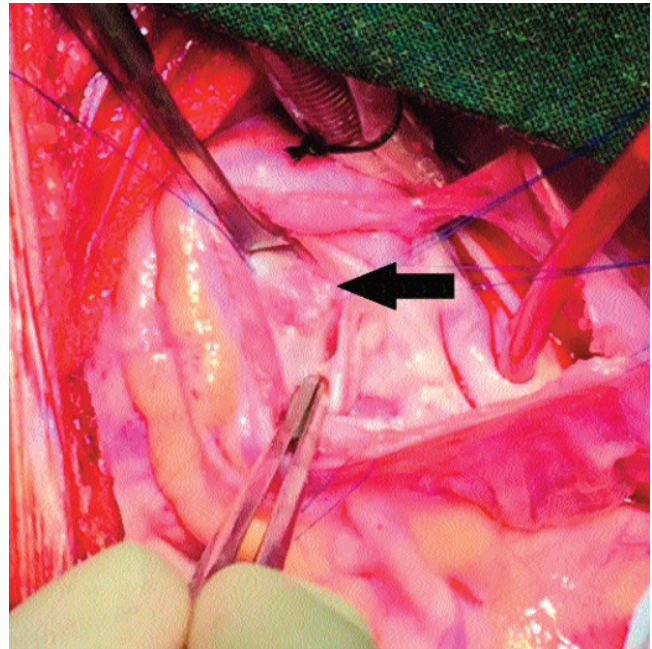


Figure-3: Pericardial patch applied on the opening of fistula.

Discussion

The incidence of coronary artery fistula although rare, is associated with a high level of morbidity. The right coronary artery is involved in 50 to 55% of cases while left coronary artery is in 35% cases. Both arteries are involved in 5% cases.⁸ Regarding the site of opening, more than ninety percent open in right-sided chambers.^{9,10} Most fistulas are found as isolated but some have other heart defects. Age of presentation is usually later in life, very few present in childhood. They rarely present in infancy. Small coronary artery fistulas do not produce any symptoms and neither require any treatment. Large fistulas mostly present as cases of shortness of breath and palpitation. Angina is a rare presentation and so also myocardial infarction is. Large fistulas can produce complications i.e. heart failure, infective endocarditis, myocardial ischaemia and severe mitral valve regurgitation.

Diagnosis is usually made by two-dimensional echocardiography and colour Doppler while coronary angiography and cardiac catheterization are necessary investigations to decide the exact location and thus the mode of intervention to close the fistula.¹¹ Indications and timing of intervention are not precise. Nevertheless, volume overload of any heart chamber producing secondary valve disease, heart failure, significant left to right shunt, disturbed cardiac rhythms and evidence of ischaemia necessitate early

closure. Indications for interventions in asymptomatic patients are controversial but presence of increased left atrial diameter and Qp/Qs ratio more than 1.3 need early interventions to prevent further complication.¹²

Two modalities of treatment are prevailing. Surgical closure with or without cardiopulmonary bypass (CPB) and trans-catheter closure. Surgical closure is the most widely used mode of treatment and is considered the gold standard. It achieves nearly 100% survival and no residual fistulous tract is acquired.¹³

When the artery is large, direct arriotomy and closure of fistulous opening by running sutures is possible followed by closure of artery. Alternatively, by opening the draining chamber and closure of fistula with or without help of pericardial patch is also possible. When the aneurysm involves most of the artery then there is no option other than closing the proximal artery distal to the sac. It must always be followed by performing bypass grafting (CABG) using internal mammary artery or cephalous vein which of course is not feasible if distal artery is small.

Hospital mortality in coronary artery fistula repair not involving aneurysm, approaches nearly zero percent while those with aneurysm had 2-4%, myocardial infarction 3.6% and fistula recurrence 4%.¹⁴

Trans catheter closure can be considered a reasonable alternative to surgical technique in selected patients. They should be opted depending upon morphology, association with aneurysm and the draining chamber. Among the various devices, few are detachable balloons, coils, controlled release coils and patent ductus arteriosus devices. Migration of devices and residual fistulous tract are the few bothersome complications of this mode of treatment for which ultimately surgery is required.

Conclusion

Coronary artery fistulas are rare congenital disorders.

Surgery is the gold standard treatment modality with excellent prognosis.

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Conflict of Interest: None to declare.

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