Case report

Undiagnosed coronary fistula causing low cardiac output syndrome after pediatric heart surgery

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Abstract

A patient with preoperative pulmonary hypertension and a large left-to-right intracardiac shunt underwent surgical correction of a complex cardiac anomaly and required extracorporeal life support for unexplained ventricular dysfunction following the procedure. Following recovery, a fistulous connection between the right coronary artery and main pulmonary artery was demonstrated. Implications and management strategies of unsuspected coronary fistula in pediatric heart surgery are discussed.

Keywords: Vascular fistula; Coronary vessel anomalies; Extracorporeal circulation

1. Introduction

A congenital coronary fistula is an abnormal communication between a coronary artery and any of the cardiac chambers. The reported incidence is less than 0.5%. Such fistulae can occur in isolation or in association with other congenital anomalies [1]. We present a case of an undiagnosed coronary fistula that caused reversible myocardial dysfunction requiring extracorporeal life support following an open-heart procedure.

2. Case report

A 34-week, 2-kg premature female presented with respiratory distress shortly after birth and was found by echocardiography to have a discrete juxtaductal aortic coarctation, proximal and distal aortic arch hypoplasia, a large perimembranous VSD, and a secundum ASD. There was a large left–right shunt and evidence of pulmonary hypertension with near systemic right ventricular pressure and moderate tricuspid insufficiency. She was transferred to our institute, intubated, and on a prostaglandin E1 infusion. Surgery was delayed by a urinary tract infection, Staphylococcus epidermidis septicemia and impaired renal function.

On day 27 she underwent surgical correction (ductus arteriosus ligation, aortic arch augmentation with a homograft patch, VSD and ASD closure) using cardiopulmonary bypass (CPB), deep hypothermia, antegrade cold blood cardioplegia, and selective antegrade cerebral perfusion. She was weaned from CPB for a short period after the repair during which time modified ultrafiltration and epicardial echocardiographic confirmation of adequacy of repair were accomplished. Several episodes of ventricular fibrillation with deterioration in hemodynamics and poor ventricular function (especially of the anterior wall) led to re-establishment of CPB, from which she could not be weaned because of persistent arrhythmias. The etiology of the ventricular dysfunction was unclear at this point. Extracorporeal membrane oxygenation (ECMO) was instituted.

During ECMO support she had intermittent episodes of wide-complex tachycardia, which were controlled with amiodarone. Serial echocardiograms showed gradual improvement in anterior wall function with resolution of the pulmonary hypertension. She was weaned from ECMO 72 h later on inhaled nitric oxide. On POD #5 she underwent delayed sternal closure and cardiac (right atrial appendage) and skeletal muscle biopsies to rule out possible metabolic or myopathic disorders. On POD #11 a pre-discharge echocardiogram showed continuous flow across a connection between the proximal right coronary artery and the main pulmonary artery (MPA) just above the pulmonary valve. The orifice of the fistula at its pulmonary termination was 2–2.5 mm, and there was continuous flow into the MPA.
Coronary angiography demonstrated many distal fistulae from the right coronary artery, which coursed through the anterior free wall, coalesced, and then entered the MPA just above the pulmonary valve anteriorly (Fig. 1). $Q_p/Q_s$ measured 1.2:1. The patient made an uneventful recovery without further surgical treatment. Both muscle biopsies were negative for any mitochondrial or metabolic myopathy.

3. Discussion

In infants and children, isolated coronary artery fistulae do not usually cause symptoms. Symptoms often begin at 20–25 years of age, and 21% of untreated fistulae will be complicated by congestive heart failure, myocardial infarction, rupture, endocarditis, or death (6%) [2]. Coronary angiography remains the gold standard for diagnosis. Transesophageal echocardiography provides useful information intraoperatively to detect residual connections and to assess ventricular wall motion. Although surgical closure of coronary artery fistulae in children is safe and effective with excellent long-term results, the majority of small fistulae can be safely closed by catheter emboli with coils or other devices [3]. There is general consensus that treatment in children is indicated in the presence of symptoms, complications, or a large shunt. Decision-making in asymptomatic children is still controversial [1,3–5], though the indications for closure may have been liberalized in the era of interventional cardiology. Acquired coronary artery fistulae created by muscle resection could result in inadequate myocardial protection during the subsequent administration of cardioplegia and may be an underestimated cause of myocardial dysfunction following repair of tetralogy of Fallot.

The patient we have described is unique in several respects. The coronary fistula could not be demonstrated preoperatively because flow through the fistula into the MPA was restricted by pulmonary hypertension. Preoperative on-table transesophageal echocardiography was precluded by low birth weight, and may also have failed to demonstrate the fistula for the same reason. During cardiopulmonary bypass the right heart was decompressed and consequently ‘coronary steal’ from both the right and left coronary arteries through the fistula into the MPA may have occurred during the administration of cardioplegia. The resulting suboptimal myocardial protection may explain the anterior ventricular wall dysfunction and the requirement for ECMO postoperatively. The successful use of ECMO for other forms of reversible myocardial dysfunction has been well described and it would be reasonable to assume that it could be quite effective in the current setting [6].

Though air embolism remains a possible explanation, the typical presentation of ST elevation and transient ventricular dysfunction that invariably recovers with pharmacologically induced hypertension and inotropic support with or without a preceding short period of CPB was not evident in this patient.

Coronary steal through fistulae has been shown to increase disproportionately with the increase in cardiac output even in the presence of a low pre-existing $Q_p/Q_s$ [7]. This suggests that even a small fistula can have serious hemodynamic implications in the presence of an existing supply/demand imbalance, as is not uncommon in congenital heart disease. We chose not to close the fistula because the patient eventually had a satisfactory surgical repair and was asymptomatic with complete recovery of ventricular function. The patient was too small for coil occlusion of the fistula at the time; however, future intervention cannot be ruled out and will certainly be undertaken should any further cardiac surgery become necessary, as the main problem with this small communication would be during cardioplegia administration.

4. Conclusion

Undiagnosed coronary fistulae should be considered in all cases of unexplained low cardiac output syndrome following cardioplegic arrest, especially in patients with pre-existing pulmonary hypertension or large left to right shunts. If identified preoperatively in patients undergoing surgery for other congenital cardiac anomalies, a case can be made for dealing with the fistula before cardioplegic arrest or using retrograde cardioplegic techniques.

References


Fig. 1. Cardiac catheterization demonstrating the fistulous connection between the proximal right coronary artery and the main pulmonary artery (white arrow).

