Pediatr Cardiol (2011) 32:815–817
DOI 10.1007/s00246-011-9938-3

CASE REPORT

Bilateral Coronary Artery Fistulas and Left Ventricle Noncompaction in a Neonate: Diagnosis and Management

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Received: 15 September 2010 / Accepted: 9 February 2011 / Published online: 24 February 2011

Abstract This report describes a neonate with heart failure resulting from bilateral coronary artery fistulas and left ventricle noncompaction. The patient underwent successful surgical ligation of the fistulas.

Keywords Coronary artery fistulas · Left ventricle noncompaction · Heart failure

Coronary artery fistula (CAF) represents a rare group of coronary anomalies with an incidence of 0.13% to 0.3% among congenital cardiac lesions. Findings show that CAFs result from persistent embryologic remnants during myocardial and coronary development. They can originate from any major coronary arteries, most commonly the right coronary artery (RCA). The drainage site most commonly is the right ventricle.

The natural history of CAFs depends on the size and number of fistulas [12]. Infants and children with CAFs often are asymptomatic, whereas older patients can present with fatigue, dyspnea, angina, myocardial infarction, heart failure, sudden death, and syncope [7]. Symptoms result from coronary steal or volume overload of cardiac chambers [5].

Complications of CAFs include endocarditis and coronary aneurysms or rupture [3]. When medical therapy fails to control heart failure, the clinician can choose closure or percutaneous occlusion with coils or devices [6] as another treatment option.

Left ventricular noncompaction (LVNC) is a rare type of cardiomyopathy resulting from abnormal arrest of endomyocardial morphogenesis. On color Doppler imaging, LVNC is characterized by deep intertrabecular recesses with a ratio of noncompacted-to-compacted layers of myocardium during end systole greater than 2 [9]. Most commonly, LVNC occurs in apical and inferolateral segments of the left ventricle (LV). The diagnosis is made using echocardiography, magnetic resonance imaging (MRI), or computed tomography (CT) [2].

The typical course of LVNC is characterized by progressive systolic or diastolic dysfunction. Morbidities and mortality are attributed to heart failure, arrhythmia, or thromboembolic complications. Among adults, the mortality rate can be as high as 35%. Patients with end-stage heart failure may require cardiac transplantation [9].

This report describes a neonate with heart failure resulting from bilateral CAFs and LVNC. To the best of our knowledge, the presentation of combined CAFs and LVNC is rare in adults and children based on previous case reports of coronary vessel anomalies, LVNC, or a combination of both anomalies [4, 11].

Case Report

A female infant was born at 35 weeks due to premature labor. Her birth weight was 3 kg. An abnormal cardiac

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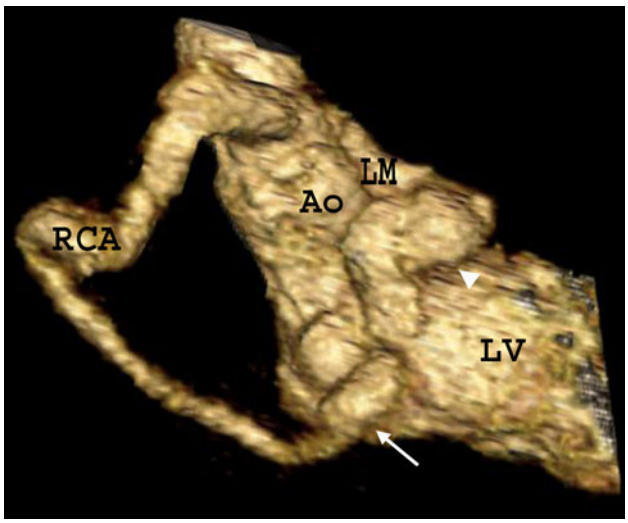

Fig. 1 Computed tomography of bilateral coronary artery fistulas. *Arrow* right coronary artery fistula. *Arrowhead* left coronary artery fistula. *RCA* right coronary artery, *Ao* aorta, *LM* left main artery, *LV* left ventricle

position shown on an obstetrics ultrasound led to an antenatal echocardiogram, which demonstrated high velocity flow from the coronary arteries into a dilated LV and diastolic flow reversal in the aortic arch. Postnatal echocardiogram, CT, and angiography demonstrated bilateral CAFs originating from the distal right coronary artery and distal left main coronary artery (Figs. 1, 2). The CAFs drained into a dilated LV with decreased systolic function, mild aortic insufficiency, and apical LVNC (Fig. 3).

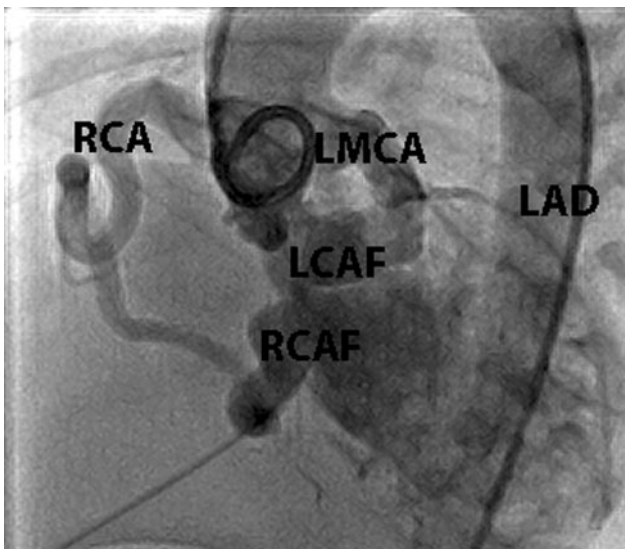

Fig. 2 Aortic root angiogram demonstrating the origins of coronary arteries and bilateral coronary artery fistulas. *LMCA* left main coronary artery, *LAD* left anterior descending artery, *RCA* right coronary artery, *LCAF* left coronary artery fistula, *RCAF* right coronary artery fistula

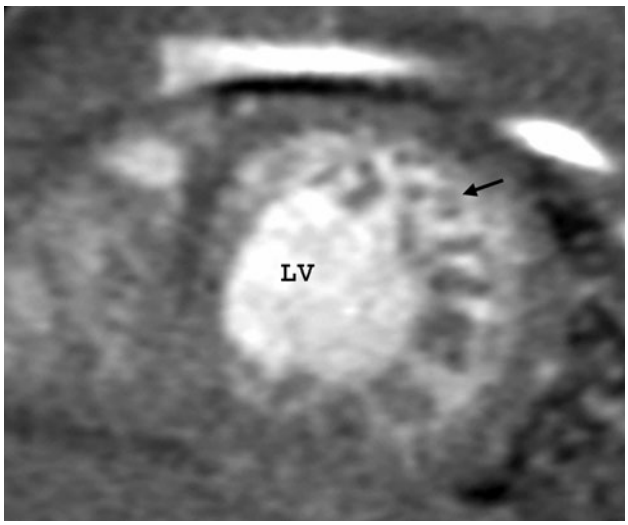

Fig. 3 Computed tomography (CT) short-axis image demonstrating left ventricle (LV) noncompaction. *Arrow* noncompacted left ventricle myocardium

The patient exhibited worsening heart failure symptoms requiring intermittent intravenous furosemide and continuous milrinone infusion. The patient maintained a natural airway. Biomarkers such as brain natriuretic peptide, troponin I, and electrocardiography (ECG) were used to track the progression of heart failure and myocardial ischemia. Nutritional support was entirely intravenous due to concerns of mesenteric hypoperfusion suggested by intolerance of enteral feeds and diastolic flow reversal in the abdominal aorta.

The patient underwent surgical ligation of CAFs at the age of 10 days based on persistent tachypnea, hyperdynamic precordium, continuous murmur, wide pulse pressure, decreasing urine output, and lack of improvement in myocardial function despite milrinone infusion and diuretics therapy. Intraoperative findings confirmed large bilateral CAFs draining into the LV (Fig. 4).

The fistulas were ligated at their entrance into LV cavity. Intraoperative transesophageal echocardiogram confirmed cessation of flow into the LV without regional wall motion abnormalities.

The patient was discharged from hospital on postoperative day 16. At the 6-month clinical follow-up visit, the patient was being fed orally, and her weight was in the 25th percentile. Her LV global function and dimension were normal. The heart failure regimen included enalapril.

Discussion

We report a case of bilateral large CAFs and LVNC causing heart failure in a neonate. Management of this patient required appreciation of pathophysiology associated

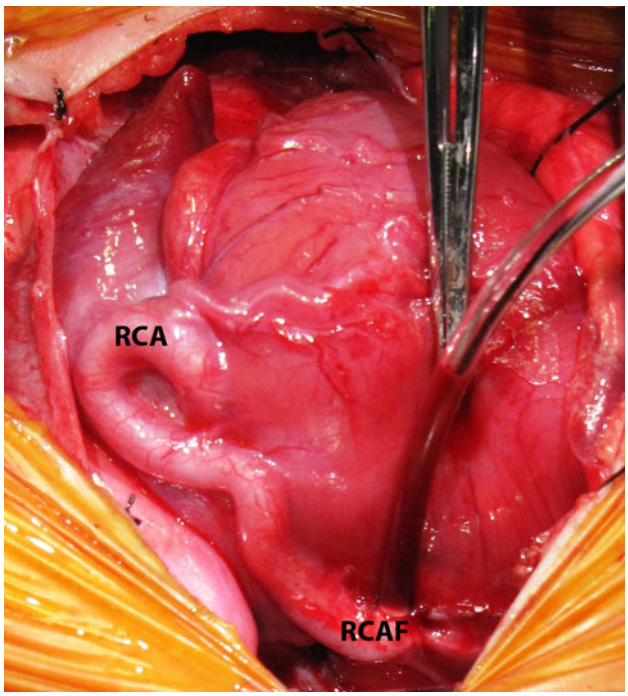

Fig. 4 Direct intraoperative visualization of the fistulas. *RCAF* right coronary artery fistula, *RCA* dilated right coronary artery

with CAFs and LVNC cardiomyopathy. The clinical course was characterized by progressive heart failure secondary to LV volume overload due to shunting from coronary arteries to LV, ongoing ischemia from coronary steal, mesenteric hypoperfusion from diastolic flow reversal, and dysfunction from LVNC cardiomyopathy. When medical therapy failed to control or reverse heart failure symptoms such as tachypnea, an ongoing need for inotropic support, and feeding intolerance, surgical intervention was deemed necessary.

The surgical literature suggests excellent late survival after ligation of CAFs. Mavroudis et al. [8] recommend intervention for all symptomatic patients and asymptomatic patients with significant clinical, ECG, and angiographic findings. Transcatheter closure with coils or device occluders also has demonstrated excellent midterm outcomes, with complete occlusion of fistulas, normalization of coronary size, and improved heart failure symptoms [6].

The factors that precluded percutaneous intervention for the reported patient were the presence of distal fistulas, the close proximity of adjacent coronary arteries, and young age. Whereas surgical and device closure of CAFs leads to normalization of chamber size and improvement in heart failure symptoms, the long-term outcome associated with LVNC cardiomyopathy is less clear. Some infants and children exhibit the undulating phenotype, with transient recovery of function in the early course of the disease. For a significant number of patients, worsening heart failure develops later in life [10]. Although it is encouraging to see early normalization of myocardial function and resolution of heart failure symptoms in the reported patient at this writing, the eventual clinical trajectory remains uncertain.

In conclusion, the combined findings of CAFs and LVNC in a neonate present additive risks for myocardial dysfunction and the development of heart failure. This report demonstrates safe and successful surgical ligation of CAFs in a neonate. We recommend longitudinal follow-up assessment to monitor the development of heart failure secondary to LVNC [1, 10].

Acknowledgment The authors express gratitude to Dr. David Wax for the cardiac catheterization image.

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