A rare cause of ischemic heart failure in a neonate

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ABSTRACT

Herein, we present a case of ischemic heart failure that occurred immediately after birth in a neonate due to coronary artery fistula (CAF) from the left main coronary artery to the left atrial appendage associated with high pulmonary artery pressure. Ischemic heart failure in a neonate with a structurally normal heart is rare. Furthermore, CAF resulting in ischemic heart failure is very rare in neonates. We believe that the small CAF caused symptoms during the first few days of life due to moderate pulmonary hypertension which resulted in a low cardiac output. The coronary perfusion improved after the normalization of the pulmonary blood pressure and improvement of the cardiac output. Echocardiography is helpful when a CAF is suspected and can be confirmed using a cardiac computed tomography scan. Small CAFs are unlikely to cause symptoms in infants, provided there are no other factors affecting the cardiac output status.

Keywords: Coronary artery fistula, coronary artery steal, ischemic heart failure, pulmonary hypertension

INTRODUCTION

Ischemic heart failure in neonates is very rare and occurs in the anomalous origin of left coronary artery from the pulmonary artery (ALCAPA) and in some rare congenital heart diseases. However, in ALCAPA, heart failure presents at 3–4 weeks of age and not at birth. We are present a rare cause of ischemic heart failure in the early neonatal period.

CLINICAL SUMMARY

A full-term male baby weighing 3.3 kg was born via normal vaginal delivery with no known antenatal risk factors. The baby developed grunting and respiratory distress shortly after birth. Blood oxygen saturation was 90% on room air, and the baby had respiratory distress with moderate chest recessions. The saturations picked up to 100% using noninvasive positive pressure ventilation (FiO_2 of 40%.) The baby had tachycardia, poor perfusion, and a 2/6 pansystolic murmur. The blood pressure and urine output were normal. A chest X-ray showed cardiomegaly and congested lungs. Arterial blood gas analysis showed mixed respiratory and metabolic acidosis. The baby was started on first-line antibiotics to cover sepsis, and respiratory support was increased via intubation and ventilation.

His 12 lead electrocardiogram (ECG) showed sinus tachycardia and pathological Q wave in Lead I and aVL, pathognomonic of ischemia in the distribution of left descending coronary artery [Figure 1] The transthoracic echocardiogram showed a small patent foramen ovale with a left to right shunt. The cardiac function was moderately impaired (fraction shortening 23%, ejection fraction 47%). The left ventricle and right ventricle (RV) were dilated. The estimated RV systolic pressure was 2/3 systemic. There was a normal origin of

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both coronary arteries, and a dilated left coronary artery was noted [Figure 2]. There was an unusual flow in the diastolic phase at the left main coronary artery (LMCA) region on echocardiogram [Video 1 and Figure 3]. A tiny patent ductus arteriosus was also noted, with a 10 mmHg shunt gradient from left to right suggesting pulmonary hypertension. Cardiac enzymes were significantly elevated.

The baby required multiple inotropic support. Intravenous furosemide was started to help combat cardiac dysfunction and pulmonary congestion. High pulmonary FiO_2 was maintained to reduce pulmonary artery vasoconstriction and pulmonary artery pressures (PAP).

Given the dilated left coronary artery, a computed tomography (CT) coronary angiogram was performed. The CT showed a left coronary artery fistula (CAF) left of the atrial appendage. The fistula was dilated and elongated. The left anterior descending and circumflex artery were both dilated as well [Videos 2 and 3].

The PAP, cardiac function, hemodynamics, ECG, and blood gases improved over the next few days with supportive management. Ten days after birth, cardiac function and pulmonary artery pressure normalized, and the patient was discharged home on diuretics and captopril.

The baby was followed up in the clinic, and all medications were stopped after 8 months of initial presentation, at which point the echocardiogram and ECG were normal [Figure 4].

DISCUSSION

A CAF is an abnormal communication between a typically originating coronary artery and another cardiovascular structure. Although CAFs are rare, they represent 14% of coronary artery abnormalities[1] and 0.4% of congenital heart diseases.[2] They are unilateral in the majority of cases (80%) and are rarely bilateral.[3] In addition, CAFs are usually isolated. However, they may be associated with other cardiac defects in 5%–30% of cases.[4] Spontaneous closure of the CAF is unlikely (1%–2%),[5] and the majority of coronary fistulas arise from the right coronary artery (60%) and often drain into the right heart (80%).[1] Most pediatric patients are diagnosed incidentally due to

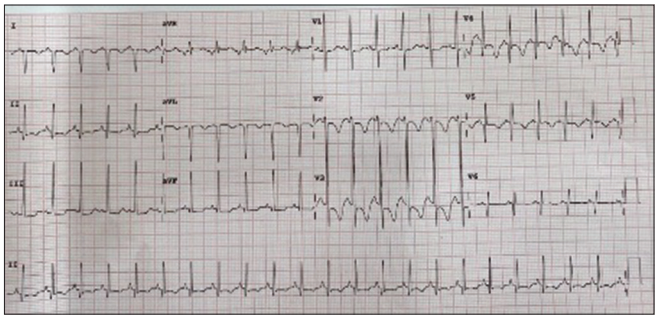

Figure 1: Electrocardiogram at presentation showed sinus tachycardia, T-wave negative in lateral leads, pathological Q wave in Lead I and a VL, suggestive of ischemia in the distribution of left descending coronary artery territory

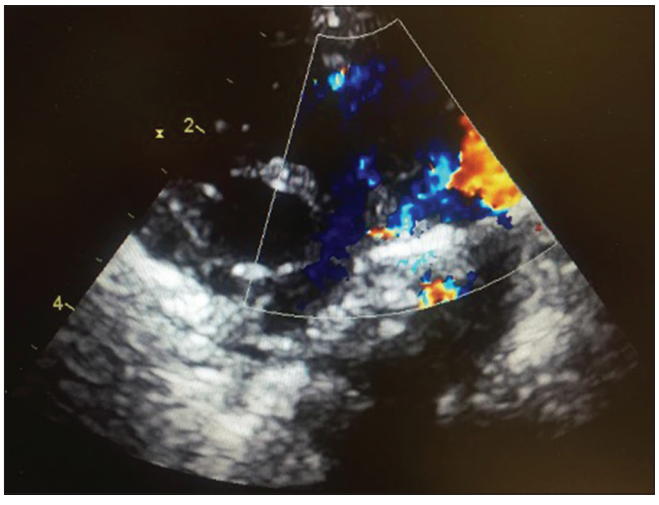

Figure 3: Color Doppler image showing an unusual diastolic flow

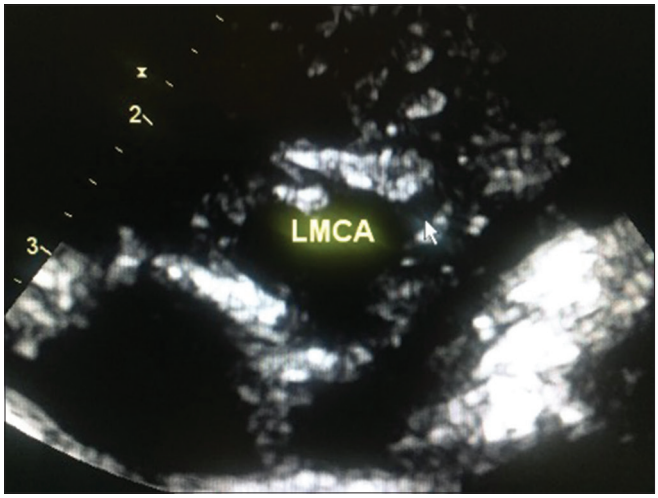

Figure 2: Echocardiogram image showing dilated left main coronary artery

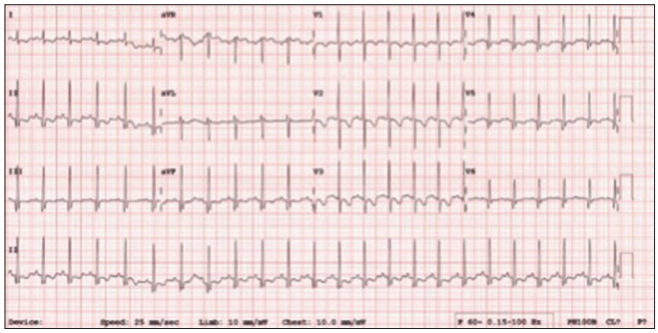

Figure 4: Electrocardiogram 1 month post presentation is showing complete resolution of the old ischemic changes, normal T wave, and ST segment

the presence of a continuous murmur on auscultation. Some patients may present with congestive heart failure due to volume overload; however, this is less common. Most adults above 25 years of age are symptomatic and may present with angina, congestive heart failure, dyspnea on exertion, or endocarditis.[1] Neonatal ischemic cardiac failure due to fistula is exceptionally rare.

In a neonate, transthoracic echocardiogram and color Doppler are valuable modalities for diagnosing CAF, as they can show dilatation of the coronary artery, drainage site in the heart, volume overload, and regional wall motion abnormalities.

The ECG can show volume overload with left or right chamber enlargement depending on the drainage site and features suggestive of myocardial ischemia. The chest X-ray can help in picking up cardiomegaly and pulmonary congestion.

Despite the numerous means to assess these patients, advanced imaging such as the CT scan is superior to echocardiography in its precision in delineating the anatomy of the coronary fistula, presence or absence of obstruction, and the drainage site.[6]

The present case shows the rare occurrence of ischemic heart failure immediately after birth due to a coronary fistula of the LMCA to the left atrial appendage. The high pulmonary artery pressure in the first few days of life can cause a low cardiac output state, especially in the context of a small intra-atrial communication.[7] The fistula size is small to cause symptoms in a neonate with normal hemodynamics. In our case, the presence of persistent pulmonary hypertension exacerbated the coronary artery steal due to fistula and made the ischemic changes more apparent. The ischemic changes improved after the resolution of pulmonary hypertension.

The standard of care in hemodynamically significant CAFs that present with ischemic heart failure during infancy is a catheter or surgical intervention.[8] However, we would like to highlight the importance of reassessing the coronary fistula after eliminating all other factors that can affect the hemodynamics, especially during the very early neonatal period when persistent pulmonary hypertension can be a real hurdle.

CONCLUSIONS

Ischemic heart failure in neonates is very rare and occurs in the anomalous origin of the left coronary artery (ALCAPA) and also during some rare congenital heart diseases. However, it is worth noting that in ALCAPA, ischemic heart failure presents at 3–4 weeks of age and not at birth. This case further underscores the need for physicians to identify the cause of heart failure as quickly as possible so that the treatment can be rapidly initiated.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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