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Case Report

Congenital Left Main Coronary Artery Occlusion Presenting as Severe Left Ventricular Dysfunction in a 45-Day-Old Infant- A Case Report

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Abstract: Congenital anomalies of the coronary arteries are rare but clinically significant causes of myocardial ischemia in infants. We present the case of a 45-day-old female infant with complete occlusion of the left main coronary artery (LMCA) leading to severe left ventricular systolic dysfunction. The child presented with fever, cough, excessive forehead sweating, and feeding difficulties. Echocardiography revealed severe left ventricular dysfunction, while coronary angiography demonstrated complete occlusion of the LMCA with retrograde filling of the left anterior descending (LAD) artery through inter-coronary collaterals. Surgical angioplasty was advised, but definitive treatment was deferred due to financial constraints. This case highlights the importance of considering coronary artery anomalies in infants presenting with heart failure and ischemic changes on electrocardiography.

Keywords: congenital coronary artery anomaly, left main coronary artery occlusion, neonatal ischemic cardiomyopathy, infant heart failure, pediatric angiography, coronary angioplasty, consanguinity

INTRODUCTION

Congenital coronary artery anomalies represent a rare but clinically significant group of cardiac malformations, with an estimated incidence of 0.2–1.3% in the general population based on angiographic and autopsy studies [1]. While many anomalies remain clinically silent, certain variants can result in severe myocardial ischemia, ventricular dysfunction, arrhythmias, or sudden cardiac death, particularly when involving the left main coronary artery (LMCA) or anomalous origins of the coronary arteries [2,3].

Occlusion or atresia of the LMCA in neonates and infants is exceptionally rare, with only isolated case reports and small series described in the literature. The LMCA is responsible for perfusing a large portion of the myocardium, including the left anterior descending (LAD) and left circumflex (LCX) territories. Therefore, complete obstruction often leads to severe left ventricular (LV) dysfunction early in life [4]. Clinical manifestations in this age group can be subtle and nonspecific, commonly including tachypnea, poor feeding, excessive sweating, irritability, or recurrent respiratory infections [5]. These symptoms frequently mimic more common neonatal conditions such as pneumonia or sepsis, which contributes to delays in recognition and appropriate management.

Electrocardiographic findings such as ST-segment elevation and T-wave inversion, as well as elevated cardiac biomarkers, can provide important early clues to underlying myocardial ischemia in infants. Echocardiography plays a pivotal role in assessing ventricular function, although direct visualization of coronary artery abnormalities is often limited in neonates. Definitive diagnosis is usually achieved through coronary angiography, which remains the gold standard for delineating coronary anatomy and assessing obstructive lesions [6].

The management of congenital LMCA occlusion is challenging, particularly in resource-limited settings. Medical therapy with cardiac decongestants, diuretics, and afterload-reducing agents may provide temporary stabilization, but surgical intervention such as coronary angioplasty, bypass grafting, or reimplantation remains the definitive therapy [7-8]. Timely revascularization is crucial for preserving myocardial viability and improving long-term outcomes [7].

In this report, we present the case of a 45-day-old infant with congenital LMCA occlusion who presented with severe LV dysfunction and ischemic changes on electrocardiography. The case underscores the importance of maintaining a high index of suspicion for coronary anomalies in infants with unexplained cardiomyopathy and highlights the diagnostic utility of coronary angiography in such rare but life-threatening presentations.

CASE PRESENTATION

A 45-day-old term female neonate, born via normal vaginal delivery with a birth weight of 3 kg, presented with fever, cough, and increased work of breathing for three days. The infant was the fourth child of a second-degree consanguineous marriage. The mother had a

history of typhoid fever in the second trimester but reported no other significant antenatal complications. Parents described episodes of excessive forehead sweating and difficulty breastfeeding, with the infant frequently pausing during feeds due to tachypnea and fatigue. On admission to KLE's Dr. Prabhakar Kore Hospital and Medical Research Centre, Belagavi, the neonate appeared pale, lethargic, and tachypneic. Cardiovascular examination revealed a grade 3 pansystolic murmur with clinical features of heart failure.

RESULTS AND OBSERVATIONS:

Clinical Investigations

Laboratory investigations revealed a hemoglobin level of 11.0 g/dl with total leukocyte and platelet counts within normal limits. The inflammatory marker high-sensitivity C-reactive protein (hs-CRP) was elevated at 16 mg/L, suggesting an active inflammatory or ischemic process. Cardiac biomarkers showed raised CK-MB levels (24 U/L) and a positive troponin-T value (0.14 ng/ml), consistent with myocardial injury. Renal function parameters were within normal limits, with a serum creatinine of 0.32 mg/dl and normal blood urea nitrogen (BUN). Electrocardiography demonstrated ischemic changes with ST-segment elevation and T-wave inversion. Two-dimensional echocardiography revealed severe left ventricular systolic dysfunction with an ejection fraction of approximately 25%, and a pansystolic murmur corresponding to regurgitant flow on Doppler assessment. Cardiac catheterization findings confirmed complete occlusion of the left main coronary artery (LMCA) on aortic root angiography, while selective right coronary angiography (RCA) showed a normal course with retrograde filling of the left anterior descending (LAD) artery via collateral circulation. The final diagnosis was left main coronary artery obstruction with severe left ventricular dysfunction and ischemia involving the LAD and left circumflex (LCX) territories.

A 2D echocardiogram revealed severe left ventricular systolic dysfunction with an ejection fraction of 25%. Cardiac catheterization was performed under conscious sedation to evaluate coronary artery anatomy. **Aortic root angiography revealed complete occlusion of the left main coronary artery (LMCA), with no antegrade flow into the LAD and LCX territories (Figure 1).** Selective right coronary angiography demonstrated a normal RCA origin and course, with retrograde collateral filling of the LAD (Figure 2).

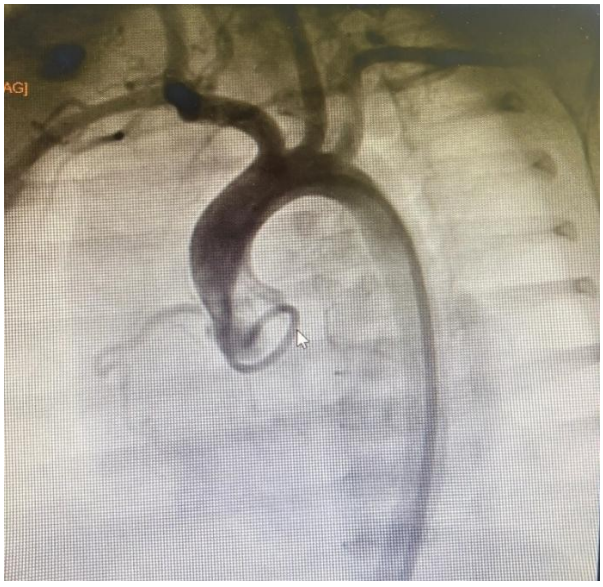

Figure 1. Aortic root angiography

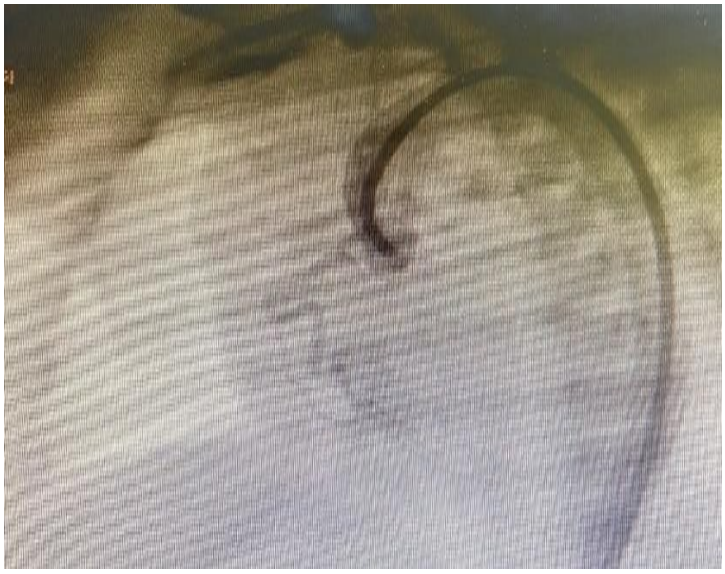

Figure 2. Selective right coronary artery angiography

MANAGEMENT

The infant was initially stabilized with oxygen supplementation to improve arterial oxygen saturation and relieve hypoxemia. Intravenous fluids were administered to ensure adequate hydration and circulatory support. Empirical intravenous broad-spectrum antibiotics were initiated in view of fever and possible infection. The infant was managed initially with stabilization measures and supportive therapy, followed by cardiac-directed medical treatment. Cardiac-specific therapy included digoxin to enhance myocardial contractility, spironolactone as a potassium-sparing diuretic to reduce preload and relieve congestion, and enalapril as an ACE inhibitor to lower afterload and improve cardiac output. Diagnostic coronary angiography, including aortic root and right coronary artery injections, demonstrated complete occlusion of the left main coronary artery (LMCA) with retrograde filling of the left anterior descending artery via right coronary collaterals. A surgical angioplasty was advised as a definitive intervention to achieve revascularization and support recovery of left ventricular function; however, it was deferred due to financial constraints. The infant was discharged on optimized medical therapy with close outpatient follow-up for continued monitoring and management.

DISCUSSION

Congenital coronary artery anomalies are rare, with an incidence of less than 1% in the general population, but they carry high clinical significance when they involve the left coronary system [1]. The left main coronary artery (LMCA) supplies a major portion of the myocardium, and its obstruction in neonates or infants can rapidly lead to ischemia, severe left ventricular dysfunction, or sudden death [2].

Our patient presented with non-specific symptoms—fever, cough, excessive sweating, and difficulty feeding—that are frequently misattributed to infections. In neonates, feeding difficulties and diaphoresis are classical red flags for cardiac dysfunction [3]. The presence of a pansystolic murmur, ischemic ECG changes, and elevated cardiac biomarkers provided important diagnostic clues, later confirmed by coronary angiography. Similar cases in the literature describe delays in diagnosis due to overlap with common neonatal illnesses [4].

Echocardiography is useful for assessing ventricular function but is often limited in visualizing coronary origins. In this case, angiography revealed complete LMCA occlusion with collateral filling of the LAD from the right coronary artery. Although collaterals provide partial compensation, they are rarely sufficient to sustain normal myocardial perfusion in infants [5].

Medical therapy with digoxin, spironolactone, and enalapril was initiated for stabilization, but definitive treatment requires surgical revascularization, such as angioplasty or bypass grafting. Studies suggest significant recovery of ventricular function following early surgical correction [6]. Without intervention, prognosis is poor, with progressive heart failure and high mortality in the first year of life [7].

This case emphasizes the importance of considering coronary anomalies in infants with unexplained LV dysfunction. Early coronary angiography is essential when ischemic features are present, and timely surgical revascularization remains the cornerstone of management.

CONCLUSION

Congenital occlusion of the LMCA is a rare but critical cause of ischemic cardiomyopathy in neonates and infants. This case emphasizes the importance of considering coronary artery anomalies in infants with unexplained heart failure and ischemic changes on ECG. Prompt diagnosis through echocardiography and coronary angiography is essential, and surgical revascularization offers the best chance for survival and recovery of ventricular function.

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