

Anomalous left coronary artery originating from the pulmonary artery

***Prabhas Prasun Giri¹, Agnisekhar Saha²**

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Introduction

Cardiac disorders such as coarctation of aorta, aortic stenosis and anomalous origin of the left coronary artery from pulmonary artery (ALCAPA) can mimic sepsis and septic shock early in the life^{1,2}. Prompt identification of these non-infectious causes and rapid treatment can be lifesaving. Here our index case was a case of ALCAPA and initially she was thought to be having sepsis.

Case report

A 49 day old female baby was admitted with a history of poor feeding, refusal to suck and excessive lethargy for last 2 days followed by respiratory distress and oliguria. On admission, the baby had severe respiratory distress, tachycardia and weak femoral pulses. The capillary refill time was prolonged. Chest auscultation revealed good bilateral air entry and audible heart sounds without any evidence of gallop rhythm. Liver was palpable 3 cm below the right costal margin. The baby was immediately shifted to the paediatric intensive care unit where she was intubated and ventilated due to increased respiratory efforts, falling saturation and metabolic derangements. A provisional diagnosis of sepsis was made and the baby was started on intravenous cefotaxime. Chest x-ray revealed bilateral whiteout lungs (Figure 1) and arterial blood gases (ABG) revealed a mixed acidosis.

The total leucocyte count was 6,500/cu mm (N58, L37), the platelet count 360,000/cu mm, and the haemoglobin level 9.6g%. The C-reactive protein was normal. The prothrombin time was 38.5 seconds and the activated partial thromboplastin time 61.4 seconds. The blood urea was 42 mg/dl

¹*RMO cum Clinical Tutor, Institute Of Child Health, ²Consultant Paediatric Intensivist, Fortis Hospital, Kolkata, India*

*Correspondence: dr.prabhas@yahoo.co.in

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and the serum creatinine 1.1 mg/dl. The serum electrolytes were normal. The aspartate transaminase level was 402 IU and the alanine transaminase level 160 IU.



Figure 1: CXR showing bilateral whiteout lungs

After one hour of mechanical ventilation the blood pressure started falling and the perfusion worsened. Dopamine and dobutamine infusions were added. Due to persistent hypotension, features of poor perfusion and metabolic acidosis, echocardiography was done and this revealed a dilated left atrium and ventricle, severe systolic dysfunction with a left ventricular ejection fraction of 20% with moderate mitral regurgitation and classical anomalous left coronary artery from pulmonary artery (ALCAPA) with reversal of flow. A 12 lead electrocardiogram (ECG) revealed ST and T wave changes with deep Q waves in chest leads (Figure 2).

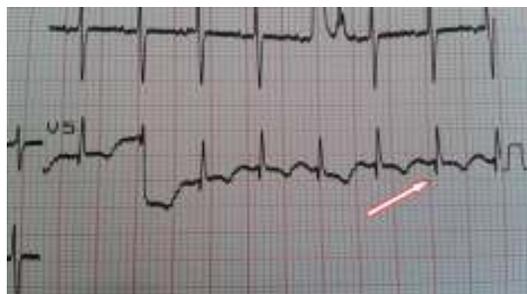


Figure 3: Deep Q waves in chest leads

A final diagnosis of ALCAPA was made. She was stabilised with inotropes and frusemide and transferred to a regional cardiology centre for further management.

Discussion

ALCAPA is a rare congenital abnormality affecting 1 in 300,000 live births³. It results in the “coronary steal” phenomenon, in which a left-to-right shunt leads to abnormal left ventricular perfusion and is the commonest cause of myocardial ischaemia and infarction in children⁴. In the neonatal period, the baby is asymptomatic because there is anterograde flow of desaturated blood from the pulmonary artery to the left coronary artery. As pulmonary arterial pressure drops, the combination of low flow and desaturated blood causes myocardial ischaemia, especially during exertion. Collateral vessels develop between the right and left coronary arteries. Further decreases in pulmonary arterial pressure result in reversal of flow, as the left coronary artery drains from the right coronary artery, through collaterals, into the pulmonary artery. This is known as myocardial steal; hence the nickname of Al Capone of coronary vessels⁵.

Onset of symptoms usually occurs about 8 weeks after birth. Babies usually present with paroxysms of irritability, excessive crying and overall pictures of distress correlating with episodes of angina. Rarely, it can mimic the soft signs of sepsis like poor feeding, lethargy like our index case. Features of shock like poor perfusion, tachycardia, weak femoral pulse, persistent hypotension and oliguria also occurs due to cardiac failure⁶. Significant heart murmur is usually absent, with a rare exception of a heart murmur of mitral regurgitation secondary to myocardial infarction. ECG shows an anterolateral myocardial infarction pattern consisting of abnormally deep and wide Q waves, inverted T waves and ST segment shifts in leads I and aVL and the precordial leads⁷.

Chest radiography may demonstrate left ventricular hypertrophy and dilation secondary to chronic myocardial ischaemia. Main differential diagnosis

at this stage is cardiomyopathy, which is excluded on echocardiography. Bilateral white out lungs, as in our index case, are suggestive of cardiogenic pulmonary oedema. Echocardiography shows the anomalous left coronary artery with Doppler flow confirming the reversal of flow⁸. Coronary artery angiography will additionally demonstrate dilated tortuous vessels and reversal of flow into the pulmonary artery on delayed images. CT coronary angiography has the benefit of being non-invasive and will also demonstrate the origin of the left coronary artery from the pulmonary artery.

Medical management involves treatment of cardiac failure and once patients are stable surgical correction to re-establish a two-coronary artery perfusion system is the treatment of choice. Among the many methods available, intrapulmonary tunnel operation (Taeuchi Repair) is the most popular one⁹. But in critically ill infants, palliative surgery like ligation of the anomalous left coronary artery close to its origin from the PA may be performed to prevent steal into the PA.

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