A rare congenital cardiovascular abnormality presenting as respiratory distress in an infant

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Abstract

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) is a rare congenital cardiovascular anomaly which presents in early infancy with congestive cardiac failure, sometimes precipitated by inter current respiratory tract infections. We report a case of an eight week old baby boy, with a short history of cough, difficulty in breathing, marked tachycardia and signs of respiratory distress. He was managed for bronchiolitis but eight hours later had major desaturation during feeding which needed intubation and mechanical ventilation. He improved over the next few hours and self extubated but after 12 hours needed reintubation and ventilation following a similar event of desaturation as observed previously. The chest x ray revealed cardiomegaly and diuretics were added in the on going management and he was gradually weaned off from the ventilator. The echocardiography examination revealed ALCAPA, resulting in myocardial ischaemia. Surgical revascularization was performed and baby was discharged after ten days on regular anti failure medication in a stable condition.

Keywords: ALCAPA, Congestive cardiac failure, Angina pectoris, Syncope, Mitral regurgitation.

Introduction

Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) accounts for 1 in 300,000 live births and comprises 0.5% of all congenital heart diseases.[1] ALCAPA is usually an isolated cardiac anomaly but in rare cases it may be associated with other congenital heart diseases. There is a wide spectrum of clinical presentation ranging from congestive cardiac failure in infancy to angina pectoris, syncope and sudden death in older children. The diagnosis is confirmed on two dimensional echocardiography and prognosis is excellent after successful surgical revascularization. We present a case of an 8 weeks old baby boy admitted with bronchiolitis but ultimately diagnosed as having ALCAPA resulting in congestive cardiac failure.

Case Report

An eight weeks old baby boy presented to our out patients in the month of January with four days history of cough and breathing difficulty. He had reluctance to feed with clear history of changing colour around the lips. However there was no history of fever or vomiting, He was initially seen in a neighboring hospital and was referred for further management to our hospital for his respiratory problem. He

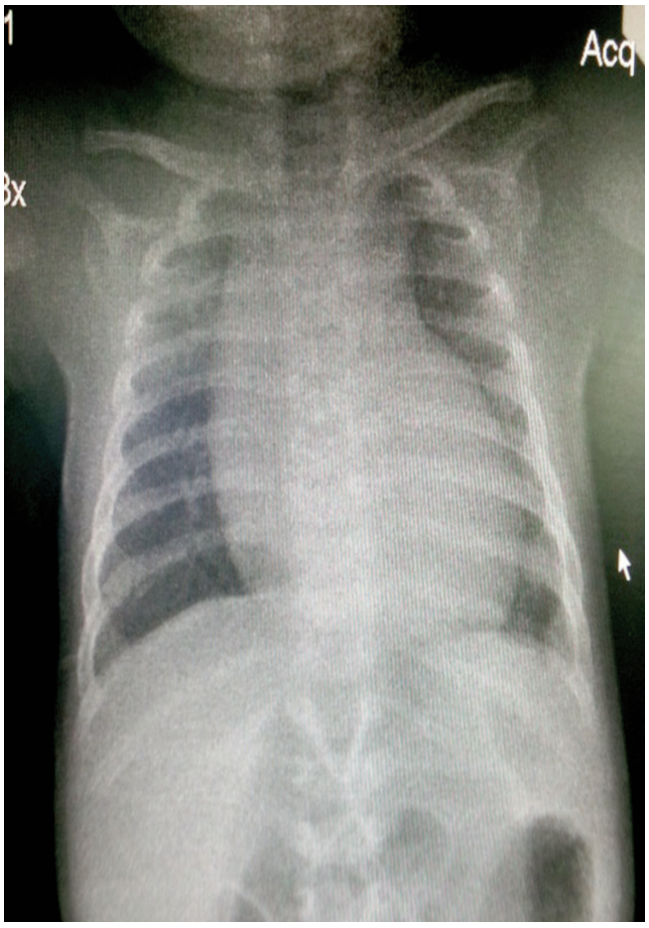

Figure-1: Chest x ray showing cardiomegaly.

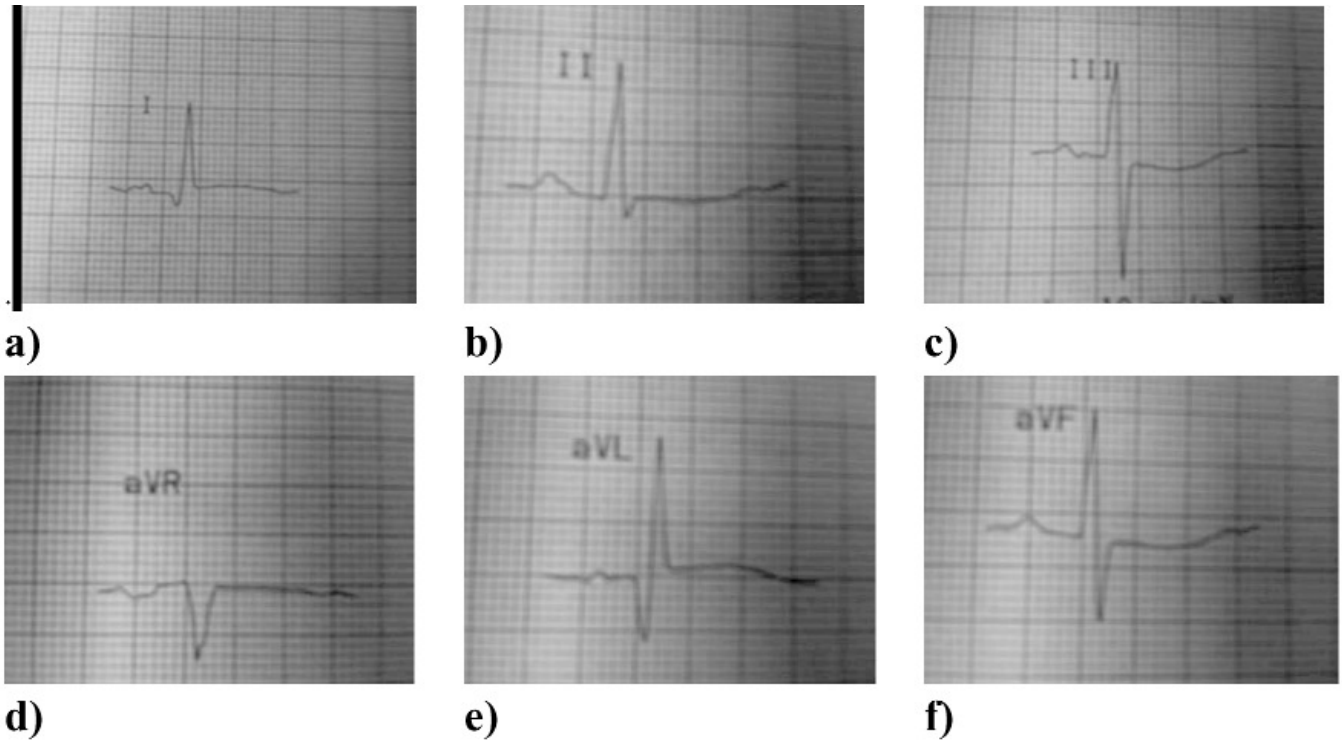

Figure-2: (a&e) ECG showing QR pattern, ST segment elevation and flat T wave in lead I and AvL. (b,c,d & f) Normal ECG pattern in lead II, III, aVR and aVF.

was the third child of a consanguineous marriage, born at term by spontaneous vertex delivery and had no perinatal complications. He was breast fed and his immunization was up to date. On examination he had signs of respiratory distress with respiratory rate of 65/ min and marked subcostal and intercostal recessions. He was tachycardic, with heart rate 140/ min and required 2L/ min of oxygen to maintain his saturation. On chest examination he had good air entry with scattered rhonchi all over the chest. His abdomen was soft with no hepatosplenomegaly and rest of the examination was unremarkable. The baby was admitted with provisional diagnosis of bronchiolitis or pneumonia and was managed with supplemental oxygen, intravenous antibiotics and fluids. The initial investigations revealed haemoglobin of 10.3 g/dl and WBC count of 7200/ µL with 50% polymorphonuclear cells. His blood chemistry was normal and arterial blood gases showed respiratory alkalosis with normal pO_2. His initial chest radiograph showed borderline cardiomegaly (Figure-1).

Eight hours after admission, the baby had an acute life threatening event with sudden onset of cyanosis and desaturation following feeding. He was resuscitated, intubated and was put on mechanical ventilation. He improved quickly and got self extubated few hours later and remained well with minimal supplementary oxygen but had persistent tachypnoea.

Twelve hours later, the baby had another acute episode of desaturation and had to be re intubated and ventilated. By then he had developed tender and firm hepatomegaly. However the examination of cardiovascular system was normal with no parasternal heave or thrill. His heart sounds were normal and there was no murmur. A repeat chest radiograph confirmed cardiomegaly. Along with other management, diuretics (furosemide) were added and his fluids were restricted to two third of daily maintenance. Gradual clinical improvement was noted over the next 24 hours and he was successfully weaned from the ventilator. Over the next couple of days he improved and his respiratory distress settled.

However it was noted that he had persistent tachypnoea, subcostal recessions and wheezing which exacerbated during crying and feeding. Further serial chest radiographs revealed cardiomegaly with prominent upper lobe vessels. At this point (4th day of admission) it was decided to have cardiac opinion to rule out congenital heart disease. His ECG showed QR pattern in lead I and AvL, as well as ST elevation was noted with flattened T wave indicating myocardial ischaemia (Figure-2). Finally the echocardiography revealed left coronary artery arising from pulmonary artery with retrograde flow of blood. Left ventricle was noted to be dilated with hypokinetic septum and there was grade I mitral regurgitation. The cardiac enzyme CK MB was found to be normal (2.4 ng/ml, normal value upto 7.2 ng/ml).

The child was referred for corrective cardiac surgery

(6th day of admission) at Paediatric Cardiac surgical unit of Armed Forces Institute of Cardiology where he had coronary angiography followed by a successful surgical revascularization. He was discharged home 10 days after his surgery in a stable condition on regular diureties.

Discussion

As the blood flow and oxygen concentration is high in the pulmonary circulation during the foetal life resulting in normal myocardial perfusion, therefore ALCAPA does not present prenatally.

After birth, the decrease in pulmonary artery resistance and the oxygen content of pulmonary blood results in relative myocardial ischaemia during periods of stress such as crying or feeding. With further increase in myocardial oxygen requirements, there may be infarction of the anterolateral wall of the myocardium. Collaterals may develop between the right and left coronary system resulting in reversal of left coronary artery flow on account of low pulmonary vascular resistance which is often referred as coronary steal phenomenon. Ultimately the left ventricular dysfunction and associated mitral insufficiency may result in congestive heart failure (CCF).

Rarely the children may stabilize following myocardial infarction and present later in childhood with mitral regurgitation.[2] Angina pectoris, syncope or sudden death may occur in late childhood or adulthood. These patients may also decompensate during acute illness. Mc Clard et al report a child admitted with Respiratory Syncitial Virus (RSV) infection and was subsequently diagnosed with ALCAPA and underwent successful surgery following resolution of RSV infection.[3] Same was the case with our patient, who was previously asymptomatic but decompensated following an episode of bronchiolitis.

The diagnosis is confirmed by two dimensional echocardiography with Doppler colour flow mapping which identifies the abnormal origin of left coronary artery. Mitral regurgitation and left ventricular dysfunction may also be identified. The same findings were also noted in our patient as well. Recently researchers in China have described multidetector-row computed tomography (MDCT) as a valuable alternative tool to confirm the diagnosis of ALCAPA syndrome.[1]

The initial management is supportive, to stabilize the patient for surgery and comprises very careful use of oxygen, diuretics, afterload reduction with medications and inotropes. It is important to remember that all these medications must be used with close supervision as they may be deleterious in this condition. High flow oxygen may reduce pulmonary vascular resistance and magnify the coronary steal phenomenon. Similarly with aggressive afterload reduction, right coronary artery perfusion may be reduced resulting in decreased left coronary blood flow. The inotropes, in high doses, increase the myocardial oxygen consumption, thus worsening myocardial ischaemia. In our case, the patient was given diuretics to reduce preload and was referred for surgery as soon as he was clinically stabilized.

Once the patient is stabilized, surgical revascularization is performed. A dual coronary system is created either by direct coronary transfer or by coronary extension techniques if anomalous coronary artery arises at a long distance from the aorta.[4]

Following successful surgery, prognosis remains excellent. Our patient also quickly stabilized following successful surgical revascularization and was discharged home in a stable condition.Ojala et al followed 29 patients with ALCAPA who underwent successful surgery for a period of 11 years. Early deaths (< 30 days following surgery) were reported in 17% patients. Among the survivors, left ventricular function had returned to normal by 1 year and 80% of patients were classified in NYHA Class I.[5]

Conclusion

Infants and young children who present with lower respiratory tract infection but have an atypical course of illness should be evaluated for congenital heart disease.

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