Infantile Heart Failure, Exotic Diagnosis but Dismal Prognosis

LIJO VARGHESE PUTHENMADOM
Department of Cardiology,
CHRISTIAN MEDICAL COLLEGE

Abstract:
Infantile heart failure, though most often due to idiopathic dilated cardiomyopathy or viral myocarditis, it could be due to plethora of causes like endomyocardial fibrosis, large shunt lesions, ventricular outflow tract obstruction, Kawasaki disease, genetic abnormality, rhabdomyolysis or ALCAPA. Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) is a rare congenital coronary anomaly which can lead to ischemic dilated cardiomyopathy with very high morbidity and mortality in infancy. In infants, it manifests as congestive cardiac failure and survival into adulthood is rare. The diagnosis can be reliably made by typical electrocardiogram and echocardiography findings. Our patient presented with gradually worsening of heart failure since 1 month of age. She was evaluated elsewhere as idiopathic dilated cardiomyopathy. Her electrocardiogram (ECG) showed typical pathological Q waves in lateral leads and echocardiography (ECHO) showed characteristic dilated chambers with regional wall motion abnormalities, mild mitral regurgitation, extensive intramyocardial collaterals and anomalous origin of left coronary artery from pulmonary artery. She was given symptomatic treatment and offered definitive surgery.

Keyword:
ALCAPA, congestive cardiac failure, Right and left coronary artery, pulmonary artery, echocardiography, electrocardiogram

Introduction:
Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) is a rare congenital defect with reported incidence of 1 in 3 lakh live births accounting for 0.25% of congenital heart diseases [1]. This condition is associated with high mortality in children (90% present in infancy and 70-80% die by one year of age), most common cause of death being congestive...
cardiac failure, arrhythmias and sudden cardiac death [2].

**Clinical History:**

Five month old child presented with failure to thrive along with history of interrupted-feeding and cold perspiration on crying and defecation. Physical examination revealed tachypnea, tachycardia, hepatomegaly and facial puffiness- all suggestive of congestive cardiac failure (CCF). The child was initially stabilized with intravenous diuretics, vasodilators, inotropes and broad spectrum antibiotics for a probable lower respiratory tract infection leading to worsening of heart failure.

ECG showed features of left ventricular (LV) hypertrophy with deep Q waves in leads I, aVL and V5, V6 as shown in figure 1.

**Figure 1: ECG- Pathological Q in 1, aVL, V2 to V6 with evidence of left ventricular hypertrophy**

Chest X-ray showed gross cardiomegaly with LV apical configuration, as shown in figure 2.

**Figure 2: Chest Xray- Gross cardiomegaly, LV apical contour, and pulmonary venous hypertension**

Echocardiography was done elsewhere at 3 months of age, which showed features of dilated cardiomyopathy with severe LV dysfunction and was started on anti-failure treatment without much benefit. The echocardiography done in our hospital also showed dilated LV with severe LV dysfunction with the LV ejection fraction being 25% (Fig.3). There was regional wall motion abnormality of LV (severe hypokinesia of mid and distal septum, apex and anterolateral segments) associated with mild mitral regurgitation (Fig.3 & 7). The pulmonary artery was also dilated along with LA and LV (probably due to left to right shunt at PA level) There were no features of RV cardiomyopathy or non-compaction of ventricles. The right coronary artery (RCA) was dilated, tortuous and arising from the right coronary sinus (Fig. 6).The left coronary sinus did not give rise to any coronary artery. The left coronary artery (LCA) was seen arising from the pulmonary artery and splits immediately into left anterior descending (LAD) and left circumflex (LCX) arteries (Fig. 4).On colour doppler, retrograde flow was demonstrated through the left coronary artery into the pulmonary artery (PA) (Fig. 5).
There were extensive intramyocardial collaterals connecting the right to left coronary arteries (Fig. 8). Hence, a diagnosis of ALCA PA was made and the child was referred for surgery after stabilization.

Figure 3: Dilated LV with RWMA on 2D

Figure 4: No coronary artery could be visualized arising from the left coronary sinus, on the contrary LCA is originating from PA (Red arrow)

Figure 5: Retrograde flow through LCA into PA
Figure 6: Dilated and tortuous RCA with torrential flow through it, arising from right coronary sinus of aorta

ECHO

Figure 4: No coronary artery could be visualized arising from the left coronary sinus, on the contrary LCA is originating from PA (Red arrow)
Course in hospital:
Intraoperative findings during corrective surgery were as follows: there was cardiomegaly, the left atrium and left ventricle were dilated. The LCA was filling retrogradely and emptying into the left posterior sinus of the PA, causing the PA to be dilated. Since translocation of the coronary artery was anatomically difficult, surgeons did Takeuchi surgery (intrapulmonary tunneling of LCA). Post op, the child showed initial improvement, but subsequently developed nosocomial infection with sepsis. Blood cultures showed coagulase negative staphylococcus and enterococcus. She was started on broad spectrum antibiotics but developed septic shock with multiorgan dysfunction. Child developed refractory ventricular fibrillation and died on 17th post op day.

Discussion:
Although ALCAPA was first described in 1886 [3], it was in 1933 that Bland et al described the clinical features and along with autopsy correlation of this condition. Hence this anomaly has been also called as Bland White Garland Syndrome [4]. This anomaly results from either abnormal septation of conotruncus into aorta and pulmonary artery or persistence of coronary buds, which eventually form the coronary artery. Mostly it occurs as an isolated defect but has been associated with patent ductus arteriosus, ventricular septal defect, tetralogy of Fallot and coarctation of aorta.

The clinical expression of syndrome results from evolving morphological-functional alterations in pulmonary circulation that occur after birth [1]. Based on this there can be 3 different types of presentations: 1) Soon after birth, resistance of the pulmonary circulation is so high permitting antegrade flow from the pulmonary artery (PA) to left coronary artery (LCA), which perfuses the left ventricle. Therefore, occurrence of sudden death is extremely rare in this age group. But since the saturation in the pulmonary artery is lower, the left ventricle gets perfused by desaturated blood at low pressure. This predisposes to myocardial ischemia, especially on exertion such as feeding or crying [5]. 2) As the pulmonary resistance falls in following weeks, the blood flow from PA to LCA stops and left ventricular perfusion totally depends upon collaterals to LCA developed from right coronary artery (RCA), which becomes tortuous and dilated. This effectively causes a "coronary steal" from myocardium to PA, which contributes to ischemia and causes infarction of antero-lateral left ventricular free wall [6].
3) Death ensues if collaterals are poorly developed while on the other hand if collaterals enlarge after an initial period of decompensation, improvement and survival into adulthood occurs - so called adult type of ALCAPA. In infancy, they present with intractable CCF after 1-2 months of age (after PVR drops). They may have continuous murmur of intracoronary anastomosis or pansystolic murmur of mitral regurgitation. Later they may develop features of pulmonary hypertension.

Wesselhoeft et al has classified ALCAPA into following clinical syndromes: infantile syndrome, mitral regurgitation syndrome, continuous murmur syndrome and sudden death in adolescents [7].

The diagnosis can be made by non invasive tests in most cases. Chest x-ray shows cardiomegaly with LV contour and evidence of pulmonary edema. ECG shows evidence of antero-lateral myocardial infarction (QR with T inversion in leads I, aVL & V5, 6). Echocardiography with Doppler is diagnostic and has replaced cardiac catheterization in most cases as in our case. It shows LCA arising from PA and retrograde flow through LCA to PA, apart from quantifying the LV dysfunction and grade of mitral regurgitation [8]. Three diagnostic criteria are used (for echo or angiographic diagnosis: (a) Retrograde filling of LCA; (b) Connection of LCA with PA; (c) Absence of LCA origin from aortaExtensive intramyocardial collaterals may be seen in older children. Our patient satisfies all the above criteria on ECHO. Initial management involves antifailure treatment with vasodilators, diuretics, digoxin and inotropes. Once stabilized, definitive surgical treatment can be planned. Initially, the LCA was ligated thus reducing the steal but this converts the heart into a single vessel system (RCA supplies the whole heart) [9]. This was associated with higher post op mortality and ongoing ischemic damage to myocardium. Of late, re-implantation of LCA into aorta or subclavian artery, saphenous vein/left internal mammary graft to LAD and intrapulmonary tunnel operation (Takeuchi operation) have been done with variable success rates [10]. Peri-operative mortality is comparable and long term results were excellent in all these dual coronary arterial supply surgery.

**Conclusion:**

ALCAPA is a rare coronary anomaly and one should suspect it while treating refractory heart failure in infancy. A careful study of an ECG and detailed echo is diagnostic. Once diagnosed, they should be referred for early surgery to avoid irreversible LV dysfunction, scarring, malignant arrhythmias or sudden death. Long term prognosis is good with early surgery especially when aggressive measure are taken to treat stunned myocardium preoperatively.

**REFERENCES:**


3 Brooks H. 2 cases of abnormal coronary origin from PA. J Anat Physiol 1886;20;26-9

4 Bland EF, White PD, Garland J; Congenital coronary anomalies. Am J Heart 1933; 8; 787-801.


7 Wesselhoft et al. ALCAPA – clinical review of 140 cases. Circulation 1968;38;403-25.

8 King et al. Non invasive detection of ALCAPA by ECHO. AJC 1985; 55; 608-9.
