Anomalous origin of left coronary artery from pulmonary artery (ALCAPA) A missed opportunity

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Abstract:
Anomalous origin of coronary artery from pulmonary artery (ALCAPA) is an infrequent, well described, but important anomaly of coronary origin. Early diagnosis and prompt surgical treatment of the disease can be life saving. However, there are potential sources of error in the seemingly simple stereotype diagnosis. We report a case of ALCAPA and allude to some of the caveats in the diagnosis of this entity.

Keyword: ALCAPA, Mitral regurgitation, Left ventricular function, Coronary anomaly

CASE REPORT:
An 11 year old male child came to our institution for a routine follow up after a cardiac surgery which he has undergone at age the of 6 months. The child was symptomatic since the age of one month with complaints of feeding difficulties and fast breathing. Echo done elsewhere at that time showed severe mitral regurgitation (MR) with preserved left ventricular (LV) systolic function. Patient had recurrent episodes of lower respiratory tract infection (LRTI) and congestive heart failure (CHF) for next 5 months. So, he underwent mitral valve repair surgery at age of 6 months elsewhere. But patient continued to have recurrent episodes of CHF till 5 year of age. After 5 years of age patient’ effort tolerance gradually improved and became asymptomatic at 7 years of age. He continued to have local follow up with echocardiography. Now, patient was asymptomatic with good effort tolerance (NYHA class I). Clinically, there was cardiomegaly, soft S1 and continuous murmur in left lateral sternal border.
Chest roentgenogram (Image-1) revealed LV type cardiomegaly with left atrial (LA) enlargement.

Image 2
An electrocardiogram (ECG) showed (Image-2) left ventricular hypertrophy (LVH), left atrial enlargement and q wave in lead I and aVL (0.04 secs wide and 4 mm deep).

Image 3
Echocardiography showed a dilated LA and LV with moderate mitral regurgitation even after mitral valve repair. There was immobile posterior mitral leaflet and prolapsed anterior mitral leaflet. There was hyperechogenicity of anterolateral papillary muscle suggestive of old scarring (Image-3). There was no regional wall motion abnormality and LV systolic ejection fraction was normal.

Image 4
There was very prominent multiple intraseptal collaterals (Image-4) that increased suspicion of ALCAPA. These prominent collaterals was earlier reported as small multiple muscular ventricular septal defects (VSD). In parasternal short axis view, the left coronary artery (LCA) was clearly identified to originate from pulmonary artery with antegrade red flow (Image-5) and retrograde flow towards aorta (blue) (Image-6).
Right coronary artery was also dilated as seen in parasternal long axis view (Image-7).

Computed tomography (CT) coronary angiogram clearly showed dilated RCA with prominent collaterals arising from RCA and LCA originating from left posterior facing sinus (Images- 8)

**DISCUSSION**

ALCAPA is the abnormal origin of the LCA from the pulmonary artery, usually from left posterior facing sinus. The first report relating clinical and autopsy findings in a three month old boy was by Bland et al(1). The anomaly has thus been called as Bland-White-Garland syndrome. It is well tolerated in fetal life, because pressure and saturations are same in aorta and pulmonary artery (PA). After birth, PA contains desaturated blood at pressures that rapidly falls below systemic pressures. Therefore, the left ventricle is perfused with de-saturated blood with low pressures. Collateral flow is initially low. At first, ischemia is transient and occurs only with exertion such as feeding and crying, but further increase in demand lead to infarction of anterolateral wall. This causes congestive heart failure (CHF) which is often made worse by MR secondary to dilated mitral valve annulus and dysfunction of anterolateral papillary muscles. Collaterals vessel between RCA and LCA enlarge. This causes increased blood flow to LCA. So, RCA becomes dilated. However, because LCA is connected to low pressure pulmonary artery, the collateral flow tends to pass into pulmonary artery rather than higher resistance myocardial blood vessels. There is pulmonary-coronary steal with left to right shunt. The shunt is usually small in terms of cardiac output but relatively large in terms of coronary flow. In about 15% of these patients, myocardial blood flow can sustain myocardial function at rest or even during exercise because of well developed and large collaterals. These are the patients who reach adult life. **Electrocardiographic features:** Although the ECG is usually abnormal, a normal ECG does not rule out ALCAPA(2). Abnormal q waves in the lateral leads (I, aVL and V4-V6) are commonly seen in ALCAPA, which is extremely rare in normal infants. The morphology of the q wave is unique in that it is much deeper but narrower when compared to adult ischemic heart disease. Other features include poor R wave progression, ST-T changes in left precordial leads and left atrial enlargement.
**Echocardiographic diagnosis:** The diagnosis is often easy, but occasionally missed by even an experienced echocardiographer. In most cases, children are referred to echo lab to identify the cause for CHF. ALCAPA should be suspected in all young infants with ventricular dysfunction or mitral regurgitation as in this case. Classically, there is scarred and thinned out myocardium at the anterolateral wall while posterobasal part may show hypertrophy and hyperfunction. Features such as hypechogenicity (scarring) of papillary muscles, endocardial fibroelastosis of LV and MR are almost always present, which may be mild to severe. Dilated RCA and presence of myocardial collaterals especially in ventricular septum also suggest ALCAPA. These features may not be prominent in patients of severe LV systolic dysfunction as they have poor collaterals. In adult type of ALCAPA these features become more prominent as in this patient. Once ALCAPA is suspected because of clinical and echocardiographic clues, direct imaging of coronary origin should be done. CT coronary angiography scans have shown high resolution for defining coronary anatomy and origin in older patients. The main advantage of this technique is rapid acquisition time and high resolution especially with 64 slice CT. Invasive cardiac catheterization and angiography used only when results of noninvasive testing is uncertain. Of all children born with this rare anomaly, approximately 87% present in infancy(1), and of these, 65% to 85% die before one year of age from intractable CHF(3), usually after 2 months of age. 15% may improve spontaneously(4). These patients may or may not have symptoms, perhaps because of extensive collaterals and even a restrictive opening of LCA from PA. Nevertheless, even these patients also are at risk of sudden cardiac death(5), especially during exercise. Some present as adults with exercise induced angina(6) or with CHF owing to MR(7). Direct reimplantation of the LCA into aorta with a button of pulmonary artery around has been the standard treatment of choice both in infants and adults (even if patient is asymptomatic).

**CONCLUSION**

ALCAPA is an important diagnosis to be looked out for in infants presenting with LV systolic dysfunction or mitral regurgitation. A high index of suspicion and awareness of various pitfalls in the diagnosis of ALCAPA can help in achieving the right diagnosis.

**BIBLIOGRAPHY**


