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Asymptomatic Case of Anomalous Left Coronary Artery Originating from Pulmonary Artery: An Incidental Diagnosis

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ABSTRACT

Background and Objective: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare but well-described cardiac anomaly with a frequency of 0.25-0.5% among all cardiac anomalies. It is associated with a high mortality rate (90%) in infants (within 1 year of birth) due to inadequate collateral circulation development. However, if good collateral circulation to the left side of the heart is established, the patient may survive into adulthood without complications.

Case report: The present report describes an unusual case of a 14-year-old boy who was incidentally diagnosed with ALCAPA and was completely asymptomatic with normal left ventricular ejection fraction. The patient was treated with conservative medical therapy and recommended for regular clinical follow-up.

Conclusion: Although our patient's parents were not convinced about surgical repair, he could be further treated with conservative medical therapy. However, surgical repair should be performed in all cases of ALCAPA to prevent the progression of ischemia, arrhythmia, congestive heart failure, and even sudden death.

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Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), also known as Bland-White-Garland-syndrome, is a rare but well-described coronary anomaly. Its prevalence is 0.25-0.5% (1 of 300000 live births) among all congenital heart disease. Generally, ALCAPA is an isolated entity; however, it is encountered with other congenital cardiac anomalies. This anomaly of left coronary artery represents two clinical variants which are reported in infants (associated with high mortality rate in earlier months of life) and in older individuals [1-3]. During utero development, ALCAPA is not problematic as the pulmonary artery pressure equals systemic pressure providing proper myocardial perfusion. However, after birth, the pulmonary arterial pressure rapidly falls down the systemic pressure increasing the demand of oxygen reach blood to left ventricle. This creates coronary artery steal phenomenon causing myocardial ischemia by compromising left ventricular function which manifest into heart enlargement and congestive heart failure. This is the reason of early death in most of the cases of infant variant of ALCAPA within first year of life [3-5].

Here, we report an uncommon case of 14-year-old boy who was serendipitously diagnosed with ALCAPA during routine health check-up in school and has successfully survived 14 years of life without any symptoms with normal left ventricular ejection fraction (uncommon in ALCAPA).

Case Presentation

A 14-year-old boy was noticed with pansystolic murmur in left parasternal area in a routine school screening program and was referred to a tertiary care center in India, to rule out ventricular septal defect. The child did not report chest pain, dyspnea, exercise intolerance or respiratory infection. His demographic details include body weight - 23 kg, height - 134.5 cm and body mass index - 12.7 kg/m^2. The boy had normal heart rate (80 beats/minute) and systemic blood pressure (110/76 mmHg).

All routine blood tests were also normal and cardiac biomarkers (troponin and creatinine kinase) were within the normal range. Electrocardiogram demonstrated sinus rhythm with normal QRS duration, and no ST-segment or T-wave abnormalities. Chest X-ray showed clear lung fields and no cardiomegaly. Two-dimensional echocardiogram showed good left ventricular function (ejection fraction-60%) but also revealed dilated right coronary artery (~6 mm) and emerging left coronary artery from pulmonary artery forming a left-to-right shunt (**Figure 1**).

A subsequent cardiac catheterization confirmed the diagnosis of anomalous origin of left coronary artery from pulmonary artery showing huge collateral arising from the enlarged right coronary artery (**Figure 2**) and pronounced left-to-right shunting from the left main coronary artery into the main pulmonary artery trunk. Surprisingly, the child was not only alive but also totally asymptomatic with ALCAPA till 14 years of age though mortality rates are very high in first few months of life.

The patient was advised for a surgical repair of ALCAPA, but his parents refused the surgery, even after evidential counselling, as the child was asymptomatic with normal left ventricular function. Hence, the patient was discharged with conservative medical treatment (aspirin-75 mg and clopidogrel-75 mg) and advised for a regular clinical follow-up.

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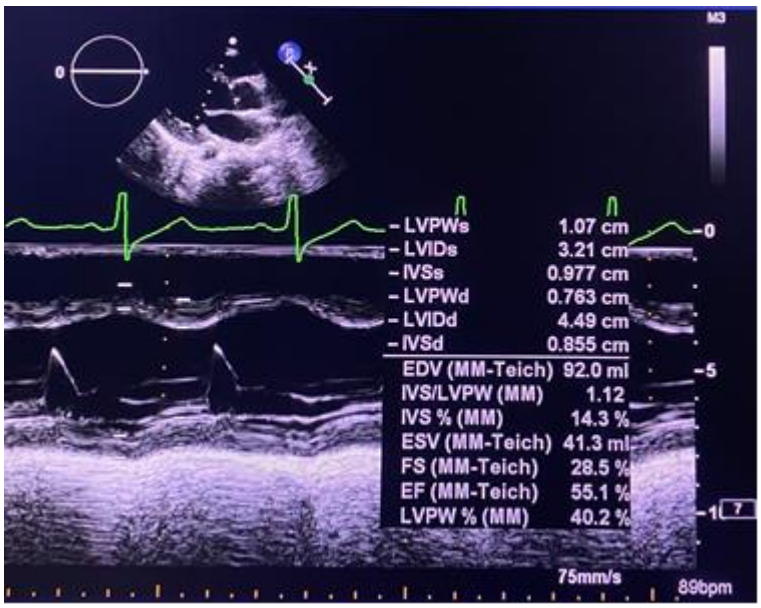

Fig 1. M mode echocardiogram showing normal left ventricular function

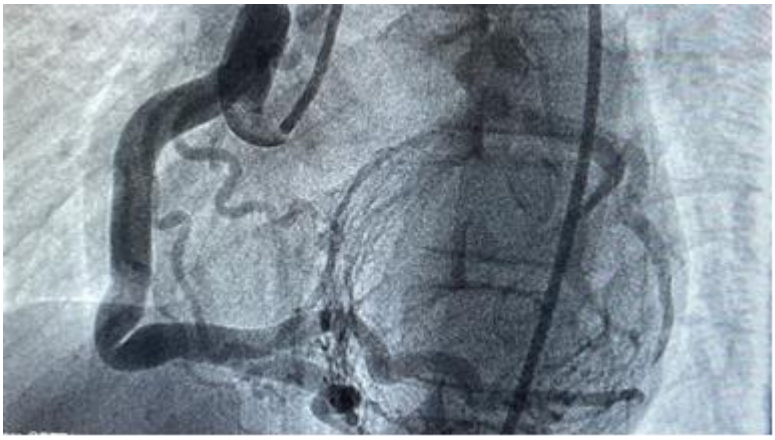

Fig 2. Dilated right coronary artery (RCA) giving collaterals to anomalous left coronary artery (LCA) and filling it retrogradely

Discussion

ALCAPA is a rare congenital cardiac anomaly with unknown true incidences due to misdiagnosis or sudden infant death. In adulthood, patients with ALCAPA most commonly present with syncope, chest pain, angina on exertion, silent or symptomatic myocardial infraction, left ventricular dysfunction and sudden death, but rarely remain asymptomatic [3]. But in present case, the patient was completely asymptomatic and was presented to our center due to murmur at the left parasternal border recorded during a routine health check-up at school which was found as common incidental finding of adult variant of ALCAPA. After multiple tests and thorough examinations, the diagnosis of ALCAPA was confirmed. Similarly, Jinmei Z et al. [5] also reported

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nine cases diagnosed in childhood or adolescence between 5 and 16 years of age. Among all nine cases, only one developed apsychia and no evidence of left ventricular dysfunction was reported in all the cases. Mitral regurgitation (mild to severe) was noted in six ALCAPA patient in that study, however, in our patient no mitral regurgitation was reported [2, 5]. Good collateral development providing adequate blood supply to left side of the heart might be the reason for no symptom development in current and previous reports. In contrast, Regeer MV et al. [6] reported a case of 19-year-old young adult who was diagnosed with ALCAPA due to ventricular fibrillation that caused out-of-hospital cardiac arrest. That patient was successfully managed with subcutaneous implantable cardioverter-defibrillator and was doing well.

Several cases have been reported with ALCAPA, however in very few cases patient remain asymptomatic at a younger age of life (14 years in present case). Compared to older patients (>50 years), younger patients experience more severe life-threating symptoms and have higher risk of sudden cardiac death. Gratefully, in present case, the patient was safe and asymptomatic till diagnosis [5, 7]. In infancy, surgical repair (preferred one is coronary button transfer) is the standard choice for the management of ALCAPA to ensure the patient's survival as rapid death ensues in up to 90% of cases within weeks or months of birth[3, 8]. Moreover, in adult variant of ALCAPA, patient may survive till adolescence or adulthood due to adequate collaterals development between the right coronary artery and left coronary artery. However, once diagnosed, the patient should undergo a surgical repair which includes Takeuchi procedure (pulmonary baffle up to coronary ostia), coronary artery bypass graft (putting venous graft in left anterior descending and ligating main artery) or coronary button transfer in selected cases. Similarly, when ALCAPA was diagnosed in our patient, though asymptomatic, surgical repair was suggested to prevent inevitable myocardial damage and fibrosis which may lead to sudden cardiac death. Even after evidential counselling, the patient's parents refused surgical repair as they did not find the child symptomatic. Hence, the patient was sent home on conservative medical treatment (aspirin and clopidogrel) and recommended for regular clinical follow-up.

Conclusion

The present case was one the rare cases of asymptomatic ALCAPA at younger age of life (14 years). Here, in this case we tried our best to convince patient's parents for surgical repair to avoid severe consequences in future, but they refused the same as the patient was completely asymptomatic. Thus, the patient was preserved on continues follow-up with conservative medical therapy. Surgery is the main and appropriate treatment for adult variant of ALCAPA; however, due to his parents' opposition, he underwent regular medical treatment and follow-up.

Funding

This study was self-funded.

Ethical consideration

We have taken patient's consent to publish the case.

Conflict of interest

There was no conflict of interest.

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