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Case Report

A rare cause of heart failure in adults you should not miss

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Introduction

The anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital abnormality, occurring in approximately 1 in 300,000 live births and accounting for 0.5% of all congenital heart diseases. It was first described in 1933 by Edward Bland, Paul Dudley White, and Joseph Garland while investigating a case of a three-month-old infant with angina pectoris. Consequently, it is also known as Bland-White-Garland syndrome [1].

ALCAPA can be classified into two subtypes—infant and adult—depending on the degree of collateral vessel formation. Infants typically present between two to three months of age with angina pectoris, heart failure, mitral regurgitation, and failure to thrive. Adults, on the other hand, present between the ages of 30 and 50 years with symptoms such as syncope, angina, fatigue, and sudden cardiac death [2].

Timely surgical correction is the treatment of choice, as infant mortality without intervention is as high as 90% within the first year of life [2]. In adults, surgical correction is preferred if the symptomatic benefits outweigh the surgical risks.

Here, we present the case of a 42-year-old female who was diagnosed with ALCAPA after presenting with heart failure and atrial fibrillation. She subsequently underwent successful surgical correction.

Case Presentation

A 42-year female, diagnosed patient with hypothyroidism for 2 years on levothyroxine 50micrograms daily, presented with progressive shortness of breath on exertion (NYHA II-III), chest pain on exertion and palpitation for 3 months. She did not complain of cough, wheezing, recent respiratory tract infections or symptoms suggestive of anemia. In the past she was apparently healthy and there was no history of ischaemic heart disease, no family history of atherosclerotic cardiovascular diseases and she was not diagnosed with diabetes mellitus, hypertension or dyslipidemia. She denies active or passive smoking or other psychoactive substance abuse. She has had an uneventful childhood and until the current presentation she had engaged in routine day to day chores as a housewife. She is a mother of two children where she had uncomplicated pregnancies, delivered vaginally.

Upon examination, she was thin built, not pale, not cyanosed, blood pressure was 100/70mmHg. Cardiovascular examination was normal but her pulse was irregularly irregular and heart rate was 65 beats per minute.

Her laboratory investigations were normal except for an elevated TSH with normal free T4 (Table.01). Her 12-lead electrocardiogram (ECG) showed atrial fibrillation and T inversions in V1 to V6 leads. Her transthoracic echocardiogram showed a dilated left ventricle (LV), a left atrium with global hypokinesia with an ejection fraction of 45%, mild mitral regurgitation and mild aortic regurgitation. A dilated RCA and abnormal flow draining to the pulmonary artery just distal to pulmonary valve was noted in a nonstandard view. There was no evidence of pulmonary hypertension in the transthoracic echo cardiogram. (Figure 01)

Table.01 Summary of laboratory investigations

Haemoglobin	12g/dl (12-15.5)
Serum creatinine	0.9mg/dl (0.5 – 1.1)
AST	20 U/l (8-33)
ALT	19 U/l (5-38)
Albumin	4.2 g/dl (3.5 – 5.5)
TSH	19.94 mIU/L (0.4 – 4.0)
Free T4	1.54 ng/dl (0.9 – 1.7)
FBS	84mg/dl (70 – 100)

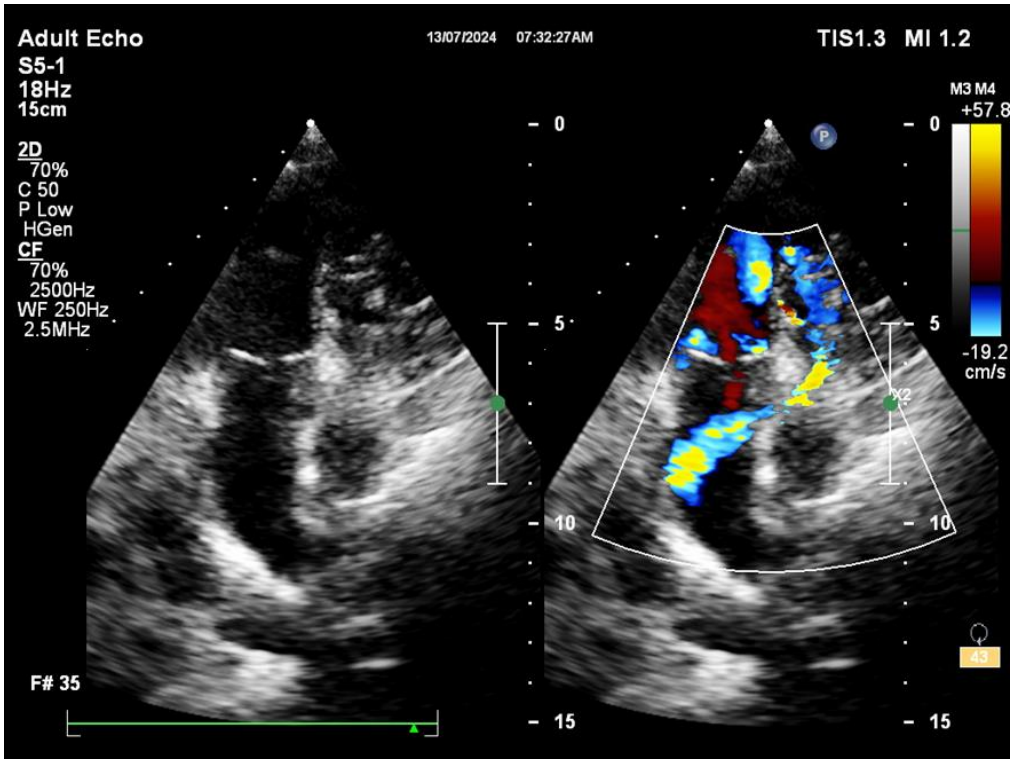

Figure 01 Transthoracic echocardiography showing blood flow draining towards pulmonary artery

She underwent a coronary angiogram which showed retrograde blood flow in the left coronary artery, draining into pulmonary artery. The right coronary artery (RCA) was dilated and tortuous with the acute marginal branch supplying collaterals for the left coronary system confirming the diagnosis of ALCAPA. (Figure 02)

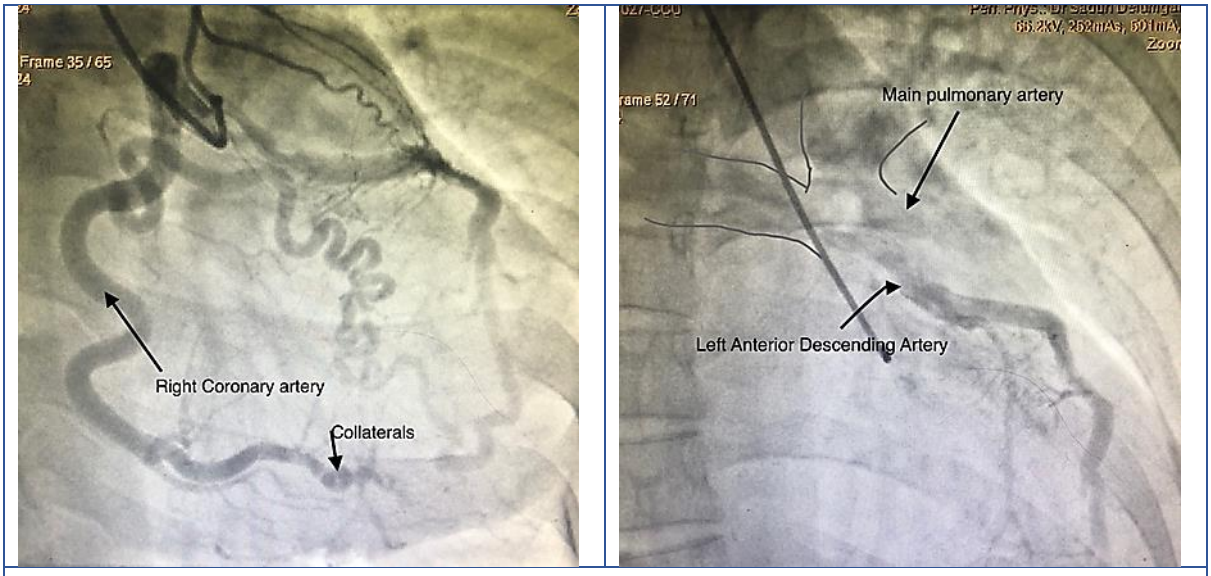

Figure 02: Coronary angiogram showing the dilated tortuous RCA with collaterals

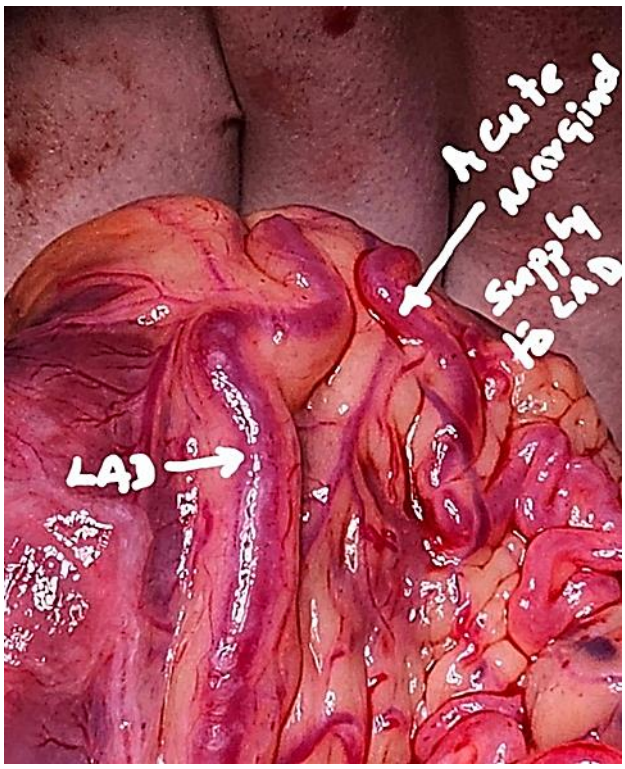

Figure 03: Intra operative demonstration of extensive collateral blood supply to the left coronary artery

Following the coronary angiogram, our patient was referred to a cardio thoracic surgeon for definitive corrective surgery. She underwent grafting of the LAD (Left anterior descending artery) with a great saphenous vein graft after ligating both the anomalous origin from the pulmonary artery and the communicating branch with RCA (Figure 03) Her post operative period was uneventful. She was discharged after 11 days of in-patient care. She was reviewed after one month at the cardiology clinic where she revealed that her symptoms were remarkably resolved and she could engage in her regular household activities without any difficulty. Her repeat transthoracic echocardiogram showed mild improved LV function, ejection fraction 50%, mild mitral regurgitation and dilated left atrium. We discharged her on long term oral anticoagulation and beta blocker therapy for paroxysmal atrial fibrillation and guideline-directed medical therapy for heart failure of mildly reduced ejection fraction. She was planned to follow up in the cardiology clinic.

Discussion

In patients with ALCAPA, the embryological development of the capillary plexus fails to reach the normal coronary ostial locations at the aorta and pulmonary artery due to a deficiency of endothelial growth factor-C. This results in the anomalous origin of the coronary arteries [2]. In utero, because the pressures in the pulmonary artery and aorta are equal, and due to right-to-left shunting through the foramen ovale and ductus arteriosus, normal coronary perfusion is maintained. However, around two months after

birth, as pulmonary artery pressure drops and right-to-left shunts close, blood flow in the left coronary artery diminishes [3]. This leads to hypoperfusion of the myocardium supplied by the left coronary artery, resulting in left ventricular failure, arrhythmias, and sudden death. When intercoronary collaterals develop, blood flows retrogradely from the left coronary artery into the pulmonary artery, causing a left-to-right shunt and the coronary steal phenomenon.

ALCAPA is associated with other congenital heart diseases in approximately 5% of cases, including atrial and ventricular septal defects and coarctation of the aorta [3].

The clinical presentation depends primarily on the degree of collateral formation between the right and left coronary arteries. The severity of the disease is partly mitigated by right coronary artery dominance, minimal coronary steal from the pulmonary artery due to left main ostial disease, restricted left main opening from the pulmonary artery, and the development of additional collaterals from bronchial arteries, allowing some patients to survive into adulthood [2].

Infant-type ALCAPA typically presents with dyspnoea, tachypnoea, poor feeding, pallor, and diaphoresis due to left ventricular dysfunction and mitral regurgitation, which may result from either a dilated mitral annulus or dysfunctional papillary muscles. Infants usually present at around 2–3 months of age due to inadequate collateral formation between the right and left coronary arteries.

Adult-type ALCAPA, which accounts for approximately 15% of total cases and is less common than the infant type, typically presents with dyspnoea, angina pectoris, fatigue, dysrhythmias, syncope or sudden death, though many patients remain asymptomatic. It is characterized by left ventricular failure and mitral regurgitation. However, the risk of sudden death decreases with advancing age (2).

The diagnosis of ALCAPA relies primarily on either invasive or noninvasive imaging modalities, with a high degree of clinical suspicion. Transthoracic echocardiography is the preliminary non-invasive imaging modality which shows impaired LV function, absent left coronary origin in aortic sinus, dilated RCA etc. Invasive coronary angiography (ICA) remains the standard diagnostic tool for ALCAPA. However, it is increasingly being replaced by noninvasive modalities such as coronary computed tomography angiography (CCTA), and coronary magnetic resonance angiography (MRA). Common findings of each imaging modality are summarized in Table 2.

Table 02: Comparison of findings of imaging modalities in diagnosis of ALCAPA.

Imaging modality	Findings
TTE	• Visualization of LCA origin from pulmonary artery. • Retrograde flow in LCA towards PA. • Lack of LCA origin from aortic sinus • Dilated RCA and collaterals (lower Nyquist limit). [Dilated collaterals may be mistaken as multiple trabecular VSDs, but continuous flow in collateral vessels with pulse Doppler (PW) helps to differentiate from VSDs which have pulsatile flow] • In addition, mitral regurgitation and LV dysfunction and increased echogenicity of papillary muscles can be visualized.
ICA	Dilated tortuous RCA with collateral filling of LCA which is flowing towards pulmonary artery.
CCTA	Direct visualization of LCA origin from PA. Dilated RCA with extensive collaterals along epicardial surface and ventricular septum. Dilated bronchial vessels.
MRA	Origin of LCA from PA Retrograde flow from LCA to PA LV dysfunction, regional wall motion abnormality, LV hypertrophy. Late sub-endocardial enhancement.

(LCA: left coronary artery, RCA: Right coronary artery, PA: Pulmonary artery, VSD: Ventricular septal defect, LV: left Ventricle)

CCTA is the preferred modality of imaging in adult patients with ALCAPA due to its non-invasive nature and high visualization of surrounding structures with good spatial resolution. MRA is preferred in infants due to its noninvasiveness, high temporal resolution, non-use of ionizing radiation and convenience to utilize in patients with high heart rates like infants.

The typical ECG findings of ALCAPA include poor R wave progression in I, aVL, V4-V6, pathological Q waves (50% of cases) and inverted T waves, LV hypertrophy pattern (28% of cases) and normal in 4% of cases. In addition, stress ECG and stress imaging tests will be positive for ischaemia in 85% and 87% of cases, respectively [4].

The natural course of ALCAPA often leads to sudden death during childhood and early adulthood. However, the risk appears to decline after the age of 50, despite less frequent surgical interventions [4]. Surgery is the recommended treatment for patients with ALCAPA, even if they are asymptomatic, due to the risk of sudden cardiac death and ventricular arrhythmias. However, in elderly patients over 50 years of age, conservative management is often preferred due to the high surgical risk and the potentially lower risk of sudden cardiac death with advancing age [5].

Several surgical procedures have been suggested based on the localization of the abnormal coronary artery ostium. These include reimplantation of the left coronary artery (LCA) into the aorta, LCA bypass using the left internal mammary artery, and the Takeuchi procedure which involves the formation of a tunnel with an intrapulmonary

baffle. Another technique, described by Laks et al., is also among the surgical options [5]. Reimplantation is technically challenging in adults due to the friability of dilated coronary arteries and the risk of excessive stretching during the procedure [6]. Therefore, LCA bypass grafting and the Takeuchi procedure are generally preferred in adult patients undergoing surgery.

Medical management of ALCAPA focuses on treating left ventricular failure and arrhythmias while preventing sudden cardiac death. In asymptomatic adults over 50 years of age, medical management is often favored over surgical intervention. However, the decision for conservative management should be individualized, taking into account surgical risk, the risk of sudden cardiac death, and patient-specific factors as robust data on long-term outcomes remain limited.

Conclusion

ALCAPA is a rare congenital abnormality which has a bimodal age distribution with the first peak occurring in early infancy and the second peak occurring later in adulthood. Our patient with ALCAPA who was apparently healthy, presented at the age of 42 years with symptomatic heart failure and atrial fibrillation underwent LAD grafting after ligating the anomalous origin from the pulmonary artery. She had a remarkable improvement in her symptoms following corrective surgery of ALCAPA.

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