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Case Report

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Abstract

Introduction: ALCAPA – 'Anomalous Left Coronary Artery from the Pulmonary Artery', is a rare congenital cardiovascular anomaly characterized by the 'left coronary artery' originating from the 'pulmonary artery' in place of its normal origin, the aorta. Failing to treat this critical condition might result in myocardial ischemia, left ventricular dysfunction, and the occurrence of sudden cardiac death.**Clinical findings:** Exertional chest pain and fatigue for 6 months. **Interventions, and outcomes:** The diagnosis was established with the help of transoesophageal echocardiography and a coronary angiogram. Surgical repair by Left Coronary Artery reimplantation was done with a successful outcome. **Conclusion:** ALCAPA may rarely be present in adulthood. The prognosis in these cases is significantly better than in patients presenting in early childhood.

Keywords: ALCAPA, congenital cardiovascular anomaly, myocardial ischemia, left ventricular dysfunction, case report.

Introduction

ALCAPA, which stands for 'Anomalous Left Coronary Artery from the Pulmonary Artery' alternatively referred to as 'Bland-White-Garland Syndrome', is an uncommon congenital cardiac anomaly. The condition involves the 'Left Coronary Artery' (LCA) rising from the 'Pulmonary Artery' (PA) in place of its normal origin, the aorta. ALCAPA occurs in approximately one out of every 3,00,000 live births and represents about 0.25 to 5% of all congenital heart abnormalities (Cankurt et al., 2017; Li et al., 2015).

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ALCAPA rarely presents in adults and can be a significant cause of sudden cardiac arrest (Lardhi, 2010). The common clinical manifestations of ALCAPA include dilated cardiomyopathy, dyspnoea, angina pectoris, symptoms of ischemia on the electrocardiogram (ECG), heart failure, and unexpected death (Kreutzer et al., 1998). The condition causes left ventricular (LV) failure in about 90% of patients, resulting in death within the first year of life (Maeder et al., 2004).

Advanced cardiac imaging has increased ALCAPA diagnoses, even in asymptomatic adults. The outcomes depend on whether there is collateral circulation between the LCA and the 'Right Coronary Artery' (RCA). Sufficient collateral supply mitigates hypoxic damage to the left ventricle and enhances overall survival (Bhandari et al., 2019). We present an undiagnosed case of ALCAPA, in a middle-aged woman who presented with angina.

The case of a middle-aged woman with undiagnosed ALCAPA presenting with angina is particularly unique and instructive. Its rarity in adults highlights potential compensatory mechanisms, underscores the diagnostic challenges, and emphasizes the need for complex

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treatment strategies. This case offers rare insights into the long-term prognosis when ALCAPA is treated in adulthood and serves as a vital tool to enhance clinicians' awareness about this potentially fatal condition that could be overlooked in patients with unexplained ischemic symptoms.

Patient Information

a) De-identified patient-specific information

A 45-year-old woman of South Asian ethnicity working as a housekeeping staff.

b) Primary concerns and symptoms of the patient

Exertional chest pain and fatigue for six months, which was initially classified as New York Heart Association (NYHA) functional class 2, but with worsening of symptoms in the past few weeks.

c) Medical, family, psychosocial history, and genetic information

She had no prior comorbidities and no history of any major medical or surgical illness in the past. Her family history was unremarkable, with no reported congenital heart defects, early cardiac deaths, or genetic disorders. She is a widow living with her children with strong social support from relatives and friends.

d) Relevant past interventions with outcomes

There is no history of any prior interventions.

Clinical Findings

She had a Blood Pressure (BP) reading - 110/70 mm Hg, a heart rate - 82 bpm, and a saturation of oxygen (SPO_2) - 99% on room air. Her physical examination showed normal findings, and the laboratory investigations did not show any remarkable results (Figure 1).

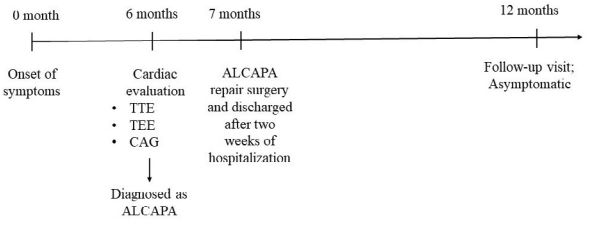

Figure 1: *Timeline of events*

TTE - Transthoracic Echocardiogram; TEE - Transoesophageal Echocardiography, CAG - Coronary Angiography; ALCAPA - Anomalous Left Coronary Artery from the Pulmonary Artery

Diagnostic Assessment

The chest X-ray (posteroanterior view) was normal. An ECG displayed a normal sinus rhythm and no ST-T changes. The Transthoracic Echocardiogram (TTE) revealed normal biventricular function, no wall motion abnormality, and normal valvular function. The treadmill test (TMT) was positive (7 METs).

The Coronary Angiography (CAG) showed these findings: dilated, tortuous RCA, and filling of 'Left Circumflex coronary artery' (LCx) and 'Left Anterior Descending coronary artery' (LAD) via abundant collaterals. LAD and LCx were traced from the 'Left Main Coronary Artery' (LMCA) to the 'Pulmonary Artery' (PA). No stenotic lesion seen (Figure 1). The Posterior Descending Artery (PDA) and Posterolateral Vein (PLV) were normal.

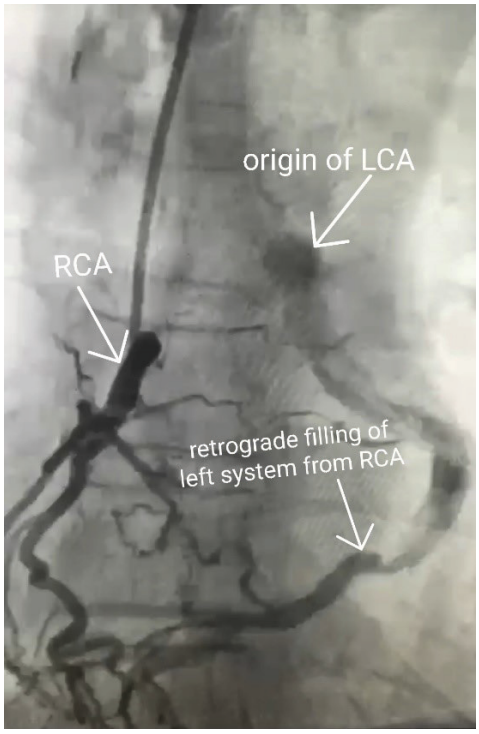

Figure 2: *CAG showing ALCAPA*

RCA - Right Coronary Artery; LCA - Left Coronary Artery; CAG - Coronary Angiography; ALCAPA - Anomalous Left Coronary Artery from the Pulmonary Artery

Transoesophageal Echocardiography showed ALCAPA, RCA dilated 7 mm, 6-7 mm LMCA origin seen in the Main Pulmonary Artery (MPA), 2.5 cm above the pulmonary valve, retrograde colour flow in LMCA to PA, minor septal blush present, and good LV function.

Therapeutic Intervention

She was advised surgical correction and underwent a successful ALCAPA repair. Intraoperative findings confirmed ALCAPA. The LCA was detached from the PA and reimplanted into the aorta. The LCA reimplantation surgery was done under normothermic cardiopulmonary bypass (CPB). Intra-operatively and postoperatively, the patient's vital signs (heart rate, respiration rate, blood pressure, oxygen saturation, etc.) and hemodynamic remained stable. She was extubated postoperatively after 24 hours of mechanical ventilatory support and shifted to the ward after four days in the intensive care unit. The patient's postoperative progress was uneventful, and the patient showed significant improvement in symptoms. Managing the patient's pain levels post-surgery, monitoring for signs of cardiac failure or arrhythmia, managing potential complications such as infection, and providing education and emotional support to the patient's family were the major nursing problems encountered. Two weeks after the surgery, the patient was discharged from the hospital. She was prescribed a single antiplatelet and a statin.

Follow-up and Outcomes

A follow-up cardiac CT revealed normal coronary blood flow and no evidence of stenosis at the anastomotic site, indicating that she is still asymptomatic.

Discussion

Chest pain frequently presents as a primary concern in outpatient settings, often caused by atherosclerotic coronary heart disease leading to cardiac ischemia (Malik et al., 2013). To determine the underlying cause, physicians conduct a comprehensive assessment involving medical history, physical examination, and additional tests like ECG, TTE, and stress tests. However, diagnosing chest pain originating from rare conditions like ALCAPA poses a significant challenge for clinicians.

ALCAPA is an uncommon congenital cardiovascular anomaly that may also manifest during adulthood. The extent of myocardial ischemia is determined by the presence of collateral circulation between the RCA and LCA. Consequently, patients who possess well-developed collateral vessels are classified as having the adult type, whereas those who lack collateral vessels are classified as having the infantile type (Barbetakis et al., 2005). Due to the left to right shunting of blood from LCA to PA, pulmonary hypertension may also manifest in certain patients with adult-type ALCAPA. Surgical intervention is necessary to restore normal coronary circulation, preventing complications like myocardial ischemia and left ventricular dysfunction. Early intervention is vital for favourable outcomes (Mishra, 2021). Unexpected cardiac death affects up to 90% of patients, usually manifesting around the average age of 35 years (Fierens, 2000).

The diagnosis of ALCAPA is often made during CAG. Non-invasive studies may also provide clues to the diagnosis. An ECG can display signs of hypertrophy of the left ventricle, left axis deviation, Poor R Wave Progression (PRWP), and abnormal Q waves in lead I and aVL (Barbetakis et al., 2005). Echocardiography can detect the LCA's abnormal origin from the PA (Chang & Allada, 2001). CT angiography and cardiac MRI are additional imaging techniques that can provide a direct visualization of the LCA arising from the PA. Moreover, MRI can depict the reverse flow from the LCA into the PA through cine imaging (Khanna et al., 2005; Pena et al., 2009).

The initial technique involved ligating the LCA due to collateral circulation from the RCA. Current surgical interventions aim to create a two-coronary artery system through surgical reconstruction or the use of bypass grafting (Ozer et al., 2008). This case report emphasizes the role of keeping ALCAPA in the differential diagnosis of adults who present with angina and the importance of detailed evaluation to prevent misdiagnosis.

Conclusion

ALCAPA can rarely manifest in adulthood, with a notably improved prognosis compared to early childhood presentation, which stems from the study of this rare adult case. Despite the delayed diagnosis, this patient's survival into adulthood underscores potential compensatory mechanisms. The critical lesson from this case report is to not overlook rare conditions like ALCAPA when diagnosing adult patients presenting with effort angina, underscoring the importance of comprehensive diagnostic scrutiny for timely treatment and improved outcomes.

Patient Perspective:

Navigating life with undiagnosed ALCAPA until middle age was likely a confusing and exhausting journey for this patient, marked by unexplained chest pain, fatigue, and declining physical capacity. The onset of angina exacerbated her distress, leading to an extended diagnostic journey filled with uncertainty and worry. The unexpected diagnosis of a heart condition that is typically paediatric and rare, along with the ensuing corrective surgery, proved overwhelming, eliciting mixed feelings of fear, relief, and apprehension about future health. Despite these challenges, being part of a case report gave her a sense of contributing to medical science, fuelling hopes for earlier diagnosis and treatment in others facing similar conditions.

Informed Consent: The patient provided written consent to publish her clinical information.

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