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ALCAPA in Elderly: A Case Report

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Abstract:

The left coronary artery normally arises from the aorta. ALCAPA (Anomalous origin of Left Coronary Artery from the Pulmonary Artery) is a rare congenital condition of the heart in which the left coronary artery abnormally originates from the pulmonary artery. Consequently, the heart muscles are devoid of enough oxygen and begin to die off. The condition, therefore, presents typically in the early infancy. However, we came across a case of ALCAPA in an elderly female who presented with atypical chest pain for 3 months.

Keywords: ALCAPA, Bland White Garland syndrome, left coronary artery, pulmonary artery

Introduction

ALCAPA syndrome (Anomalous origin of Left Coronary Artery from the Pulmonary Artery), also known as Bland White Garland syndrome, is a rare congenital anomaly wherein the left coronary artery abnormally originates from the pulmonary artery instead of the aorta. It usually presents in infancy, within 1 to 2 months of birth, with symptoms of dyspnoea, failure to thrive and pallor. There is left to right shunting of blood, from the high pressure left coronary artery to the low pressure pulmonary artery causing "coronary steal" and impaired myocardial perfusion. It is extremely unusual for these patients to survive till adulthood if left untreated. The mortality rate among untreated patients is high due to massive myocardial injury and decompensated heart failure. Those who survive into adulthood may present with a spectrum of clinical presentations which include arrhythmias, myocardial infarction, mitral regurgitation, severe pulmonary artery hypertension, congestive heart failure and sudden cardiac death [1]. A more favourable prognosis is expected if there is an extensive collateral circulation between the right and left coronary arteries. Here we present the case of an elderly female with ALCAPA who managed to thrive till adulthood without any symptoms.

Case Presentation

An elderly female presented to the Cardiology outpatient department with history of occasional exertional chest pain, which was diffuse with no radiation, and associated with palpitation for the past 3 months. She was a diabetic (since 2 months) on oral antidiabetic medications (glimiperide 1

mg once daily and metformin 500 mg once daily). She did not have any past history of cardiac or lung disease. There were no congenital or genetic disorders in the family.

On examination, her pulse rate was 100/ minute and blood pressure 110/70 mmHg. She had a systolic murmur (grade II/V) at the cardiac apex. Her electrocardiogram was in sinus rhythm with ischemic changes in anterolateral leads (**Figure 1**). Her echocardiography was suggestive of ALCAPA with regional wall motion abnormality, moderate mitral regurgitation, moderate left ventricular dysfunction (ejection fraction 48%) and moderate pulmonary artery hypertension. Routine blood investigations were within normal limits. Computerized tomography coronary angiogram was deferred due to persistent tachycardia.

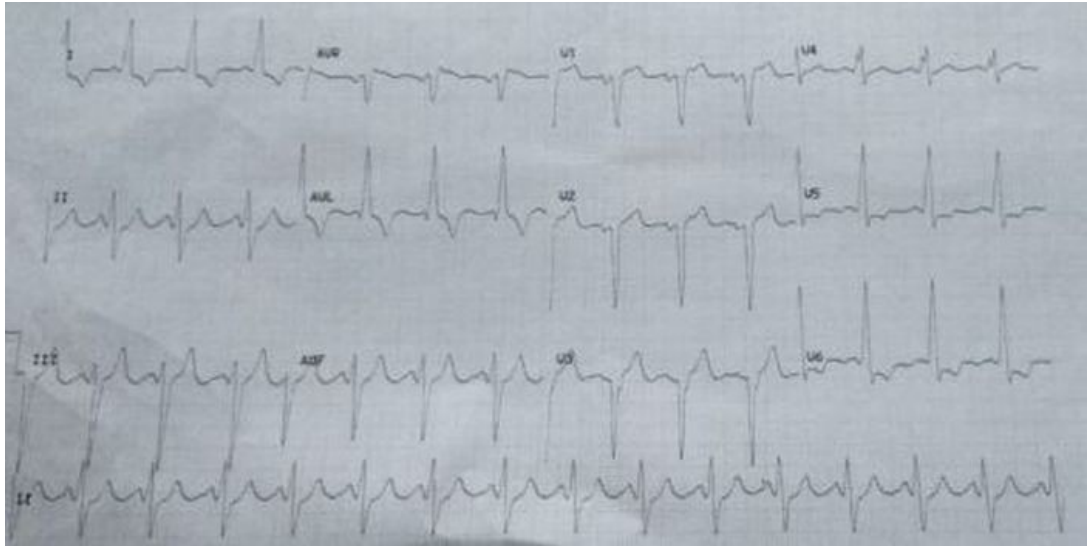


Figure 1: Electrocardiogram showing anterolateral ischemia

Coronary angiography showed anomalous origin of the left coronary artery from the pulmonary artery, with grossly ecstatic left anterior descending and left circumflex coronary artery filling through abundant collaterals from the right coronary artery. The right coronary artery was also ecstatic with no flow limiting lesions and giving abundant collaterals to the left coronary artery (**Figure 2 and 3**).

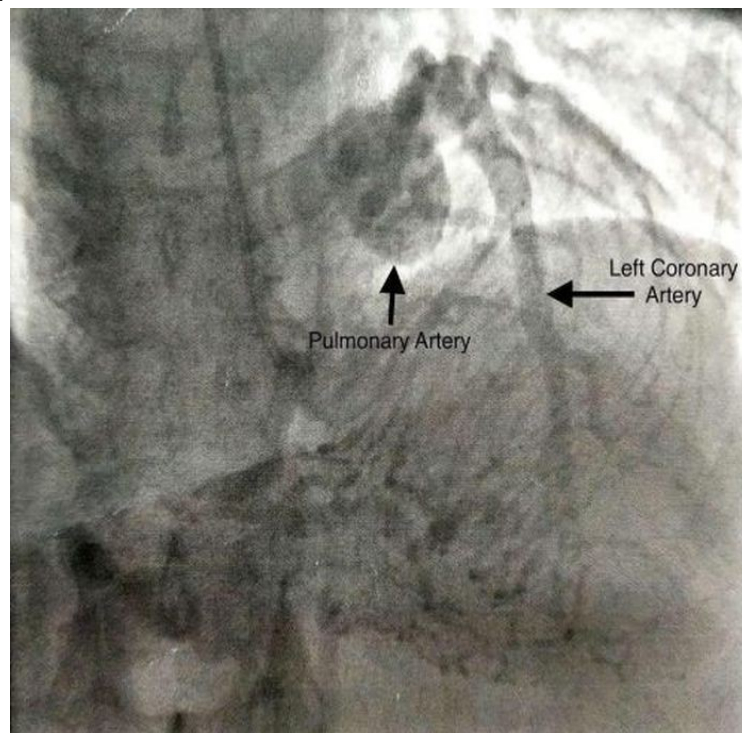


Figure 2: Anomalous origin of left coronary artery from pulmonary artery

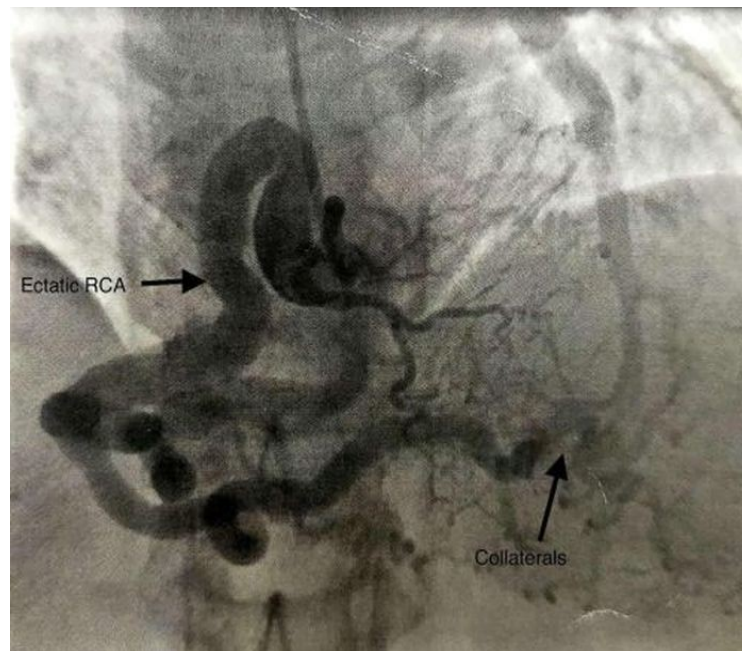


Figure 3: Ectatic right coronary artery with abundant collaterals to left anterior descending artery

A surgical repair was advised, which she refused. Considering that her current presentation of myocardial damage was secondary to lack of oxygenated blood during increased demand at the time of tachycardia, a rate control strategy was adopted (metoprolol 25 mg once daily) and advised regular follow-up.

Discussion

ALCAPA is also known as Bland-White-Garland syndrome, after the landmark publication by Bland, White and Garland in 1933. It is a rare congenital heart disease, seen in approximately 1 of 300,000 new-borns in the USA. It accounts for 0.25 to 0.5% of congenital heart disease in children. About 90% of untreated patients die during the first year of life, due to myocardial ischemia and heart failure. About 10-15% of patients with this congenital heart disease reach adulthood [1-3]. Though it is an isolated cardiac anomaly, about 5% of the cases have been described with coarctation of aorta, atrial or ventricular septal defects [4].

The anomalous left coronary artery arises from the right pulmonary sinus, adjacent to the left aortic sinus. It may also originate from the other pulmonary sinuses or in the main or proximal branch pulmonary arteries. The pathophysiology depends on the direction of blood flow in the left coronary artery. During the foetal period, the systemic and pulmonary arterial pressures, as well as the oxygen saturation are equal. After birth, the pulmonary artery contains de-saturated blood and its pressures are lower than systemic pressures. There is also a decrease in the pulmonary vascular resistance from the second month of life. The flow from the left coronary artery reverses with the closure of ductus arteriosus (steal phenomenon), thereby reducing the perfusion pressure of the ectopic left coronary artery. At low pressures, the left ventricle is perfused with desaturated blood, and myocardial ischemia with left ventricular dysfunction may arise in the absence of a well-developed collateral circulation between the right and left coronary artery. It is this collateral circulation that determines the clinical symptoms of the patient as well as the area of myocardial ischemia [5].

ALCAPA, during infancy, can be fatal and patients present with myocardial infarction, mitral regurgitation, left ventricular dysfunction and silent myocardial ischemia, which can lead to sudden cardiac death. Clinically, tachypnoea, tachycardia, displaced apical beat, S3 and S4 heart sounds and systolic murmur of mitral regurgitation are present [6]. Adult patients may be asymptomatic or present with dyspnea, angina, syncope, arrhythmia, myocardial infarction or sudden cardiac death [3].

Coronary angiogram is the gold standard method for the diagnosis of ALCAPA. Dilated and tortuous right coronary artery with collateral filling of the left coronary artery, and variable degrees of shunting to the pulmonary artery has been observed [7]. The electrocardiogram of a baby with ALCAPA may either be normal or show abnormal Q waves with transient ST changes in anterolateral leads [8]. The diagnosis of coronary artery anomalies by transthoracic echocardiography, unlike new-borns, can be difficult in adults because it may not always be possible to show the origins of the coronary arteries [9]. Computerized tomography coronary angiogram is useful in demonstrating anomalous coronary arteries, with an accurate and detailed description of the origin and course of the coronary arteries, thereby providing a prognosis for the anomalous coronary arteries [10].

Surgical intervention is the mainstay of ALCAPA treatment, without which majority of patients die within the first year of life. Surgery aims at restoring normal coronary circulation and improving myocardial perfusion. This may be done by reimplantation or bypass of the left main coronary with left internal mammary artery graft. Another technique involves the formation of a tunnel with intrapulmonary baffle [11]. Conservative medical therapy in asymptomatic elderly patients might be an alternative option [3]. However, even if the patient is asymptomatic, with no ventricular arrhythmia or significant left-to-right shunt or risk of death, surgery should still be advised. Ventricular tachycardia occur either due to acute local ischemia caused by coronary steal, or by a re-entry circuit in the border zone of myocardial infarction, or due to electrical instability by endocardial fibrosis [12]. This may be prevented by surgical correction rather than implantable cardioverter defibrillator which also holds the risk of infection.

Yau et al. reviewed 151 adult cases of ALCAPA, and found that the average age at the time of diagnosis was 41 years. The incidence was more among females, with a 2:1 ratio. Around 66% of the patients had symptoms of angina, dyspnoea or palpitations at the time of presentation. Ventricular arrhythmia, syncope, or sudden death was the initial manifestation in 17% of the cases, and 14% were asymptomatic. The average age of patients who presented with life-threatening events were 33 years +/- 14 years; and 62% of them did not have any symptoms in the past [7].

Our patient was an elderly female, who presented with occasional exertional angina and palpitation. She was diagnosed to have ALPACA. Though surgical intervention was advised, she wanted to go ahead with conservative management. This might be one among the oldest asymptomatic ALPACA cases reported till now.

Conclusion

The number of ALCAPA cases reported in patients over the age of 50 years has increased over the past two decades. This can be attributed to the introduction of new diagnostic modalities. According to the natural course of ALCAPA, the incidence of sudden death during childhood and early adulthood is very high. But these events seem to decline after 50 years of age despite the low numbers of surgical correction. Surgical intervention continues to be the treatment of choice in order to prevent lethal events in this untreated population.

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