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Anomalous origin of left coronary artery from the pulmonary artery syndrome of an 18-year-old Palestinian male: case report

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ABSTRACT

Background: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital cardiac anomaly. The majority of patients die within the first year of life if left untreated. However, a few patients can be asymptomatic and survive into adulthood due to well-developed collateral between the left coronary artery and the dilated right coronary artery. **Case presentation:** We reported a case of a male who has been having nonspecific symptoms since infancy which were misdiagnosed as symptoms of heart failure. At the age of 18 years old, he presented with a sudden death, and his autopsy revealed ALCAPA syndrome with a dilated right coronary artery. Histopathological studies demonstrated myxoid degeneration of media of the proximal left coronary artery (LCA), right coronary artery (RCA) collaterals, the proximal RCA and the leaflet of mitral valve. In addition, interstitial fibrosis of the papillary muscle of the the mitral valve and leaflet of mitral valve, distal LCA, and in the left ventricle endocardium was observed. **Conclusions:** Aperiodic diagnosis and follow-up for the cardiovascular system is recommended for similar cases. In addition to the current recommended approaches for ALCAPA treatment, further studies emerge the need for determining reliable, noninvasive diagnostic biomarkers for early screening this congenial anomaly of the heart during infancy to avoid a tragic ending.

Keywords: ALCAPA; Congenital anomaly; Dilated right coronary artery (RCA); Palestine; Sudden death.

INTRODUCTION

Anomalous origin of the left coronary artery (LCA) from the pulmonary artery (ALCAPA) is a rare congenital anomaly that commonly presents in infancy, occurring in approximately 1/ 300,000 live births, which if untreated timely has up to 90% mortality (1,2). Most deaths occur before the first year of age as a result of complications related to heart failure (1,3). Some patients remain asymptomatic likely secondary to an extensive network of collateral vessels between the right and left coronary system (1–3). Common clinical manifestations include dilated cardiomyopathy, dyspnea, angina pectoris, signs of ischemia on the electrocardiogram, heart failure, sudden death, and death in childhood (4).

This case report discusses the imaging findings of ALCAPA in an 18-year-old man and performed postmortem histological studies of cardiac tissues.

Case description

On October 20th, 2016, an 18-year-old young male went to his work at the quarry where he started to work since 2 years. At (10 am), while he was taking his breakfast together with his workmates, he suddenly collapsed and fell to the ground. Immediately, he was transported to the nearest emergency room. Resuscitation efforts were made, but unfortunately, all had been unsuccessful and his death was determined. Subsequently, the body was transported to the forensic medical institute for further investigation.

The history of O.H. dates to his infancy period when he had an episode of cyanosis and dyspnea at the age of 7 months, and since that time he was misdiagnosed with unspecified cardiomyopathy and medical treatment was prescribed for him with echocardiography follow-up. According to his family, he took all medications as prescribed, and did not suffer from any complains, there was no chest pain, dyspnea, orthopnea, or syncope. He has 3 years' history of smoking. There is no family history of cardiac disease. In his medical records, the echocardiography at 2012 showed dilated left ventricle, mild mitral regurgitation, and poor left ventricular ejection fraction (32%).

Findings of the autopsy the general appearance of the corpse is of an 18-year-old young male, whose muscular structure and nutritional status were normal during life. His length was 175 cm and weight 75 kg. There was bluish discoloration around the injection puncture on the dorsum of his left hand, and there were signs of cardiopulmonary resuscitation (CPR), burns on the chest skin due to medical intervention. The face, neck, and upper chest were congested. There were no signs of injury or violence, especially around the mouth orifice, nose, nostrils, anterior and lateral sides of the neck, and the inguinal region. After dissection, we found pathological changes and congenital anomalies in the heart and coronary arteries. The heart weight was 400 g. The left coronary artery (LCA) arose from the pulmonary artery that was dilated and running in normal course (Fig 1). There was cardiomegaly with left ventricular hypertrophy (the thickness of the left ventricle wall was 2 cm). There was also left ventricle dilatation with extensive diffuse subendocardial fibrosis. The right coronary artery (RCA) arose from the orifice of the RCA above the right cusp of the aortic valve, which showed marked dilatation and ran a normal course, and gave many collaterals (branches) to the left ventricle (Fig 2).

Portions of the heart autopsy were taken and embedded in paraffin for histopathological studies. H&E staining was performed and showed myxoid degeneration of the media of the proximal LCA (Fig 3), proximal RCA, right coronary artery collaterals, and the leaflet of mitral valve. Interstitial fibrosis was observed in the myocardium, the left ventricle, endocardium, papillary muscle of the mitral valve, and the leaflet of mitral valve (Fig 4).

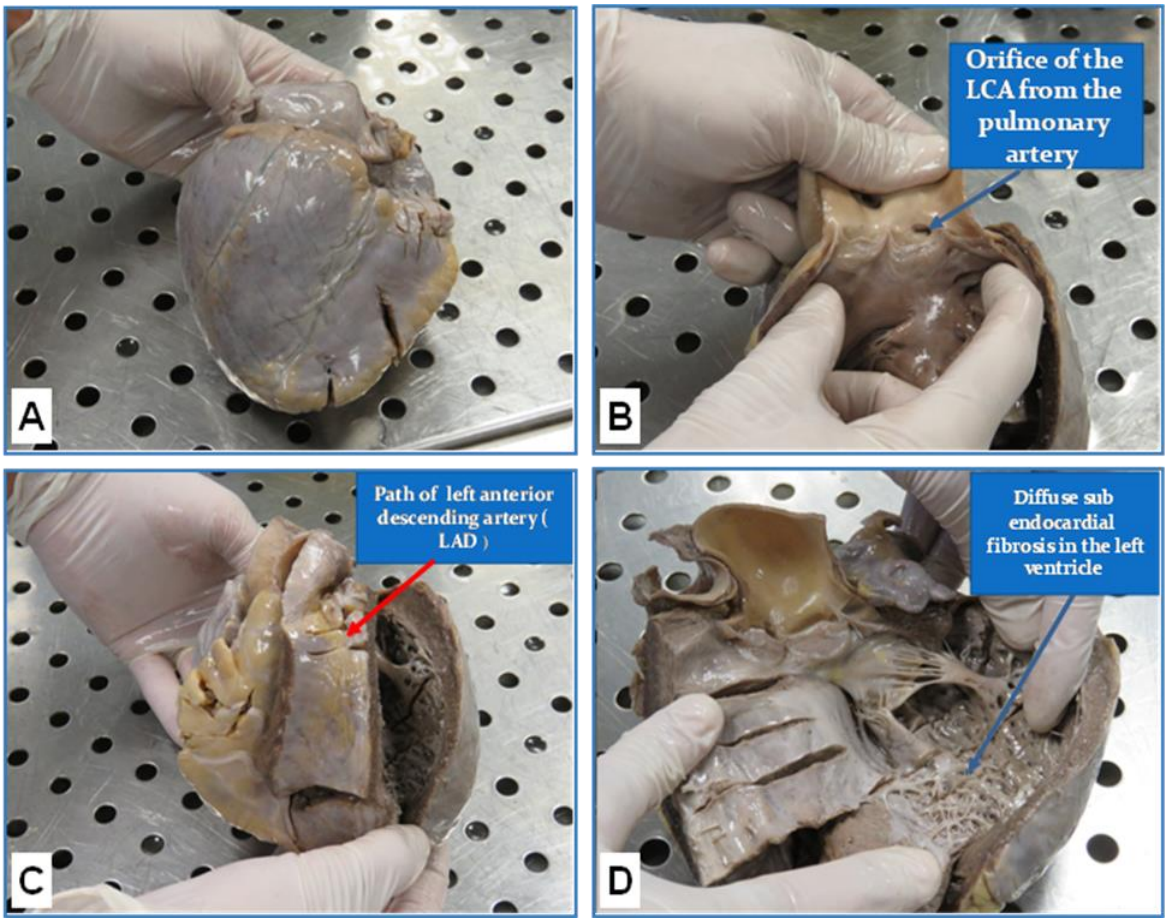

Figure (1): Macroscopic examination of a heart diagnosed as ALCAPA syndrome in an 18-year-old man. A: Overall view of the post-mortem heart**; B:** Anatomy of the heart shows the orifice of the left coronary artery (LCA) from the pulmonary artery; C: Dissection of the heart to show the path of the left anterior descending artery (LAD)**;** D: The arrow points at diffuse sub endocardial fibrosis in the left ventricle.

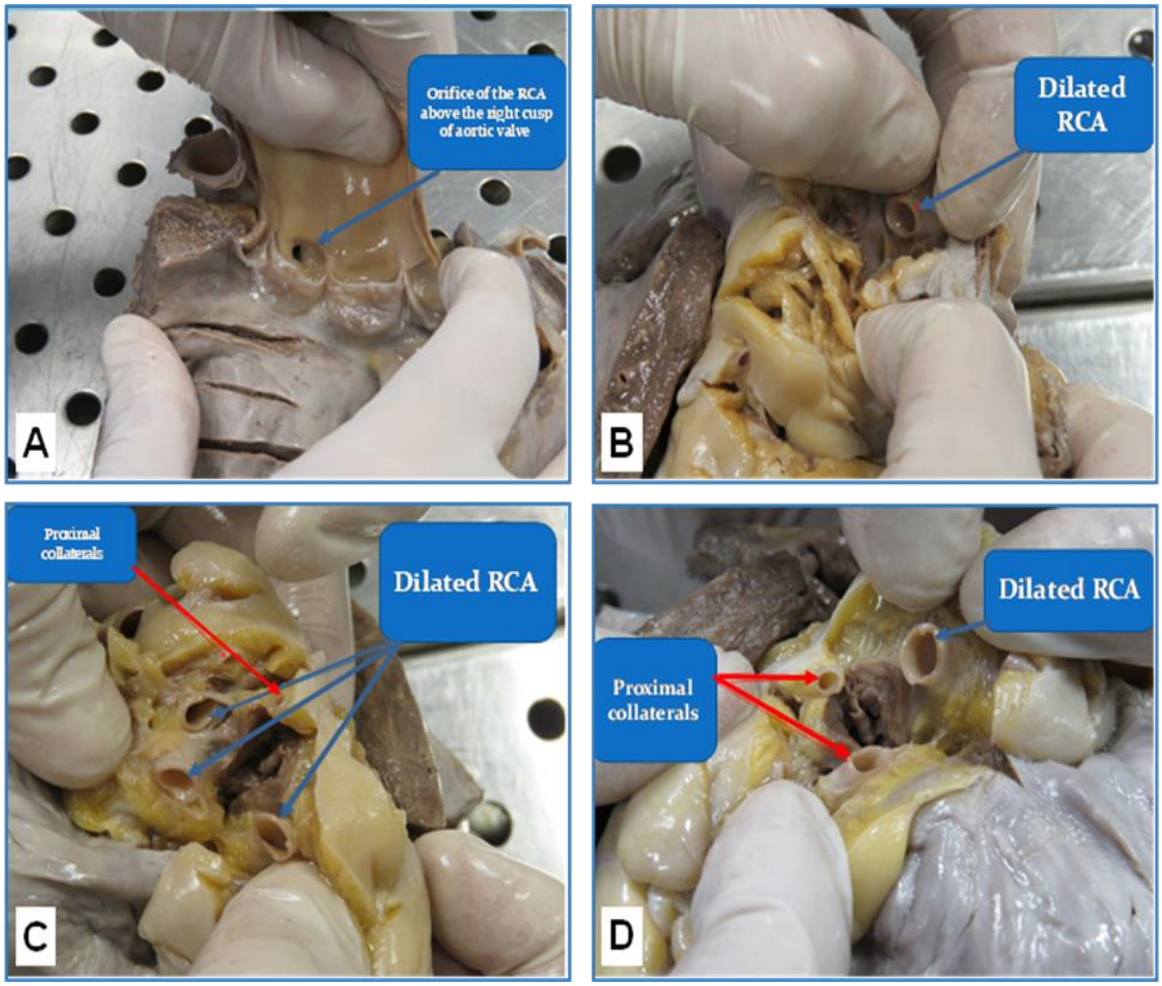

Figure (2): Macroscopic anatomical examination of the right coronary artery (RCA) of ALCAPA syndrome of 18-year-old male with sudden death. A: The arrow indicates the orifice of the RCA above the right cusp of the aortic valve; B: Dilated RCA (arrow); C: The red arrows point at the opening of proximal collaterals, and the blue arrows show dilated RCAs; D: Another view of the proximal collaterals (red arrows) and prominent dilated RCA.

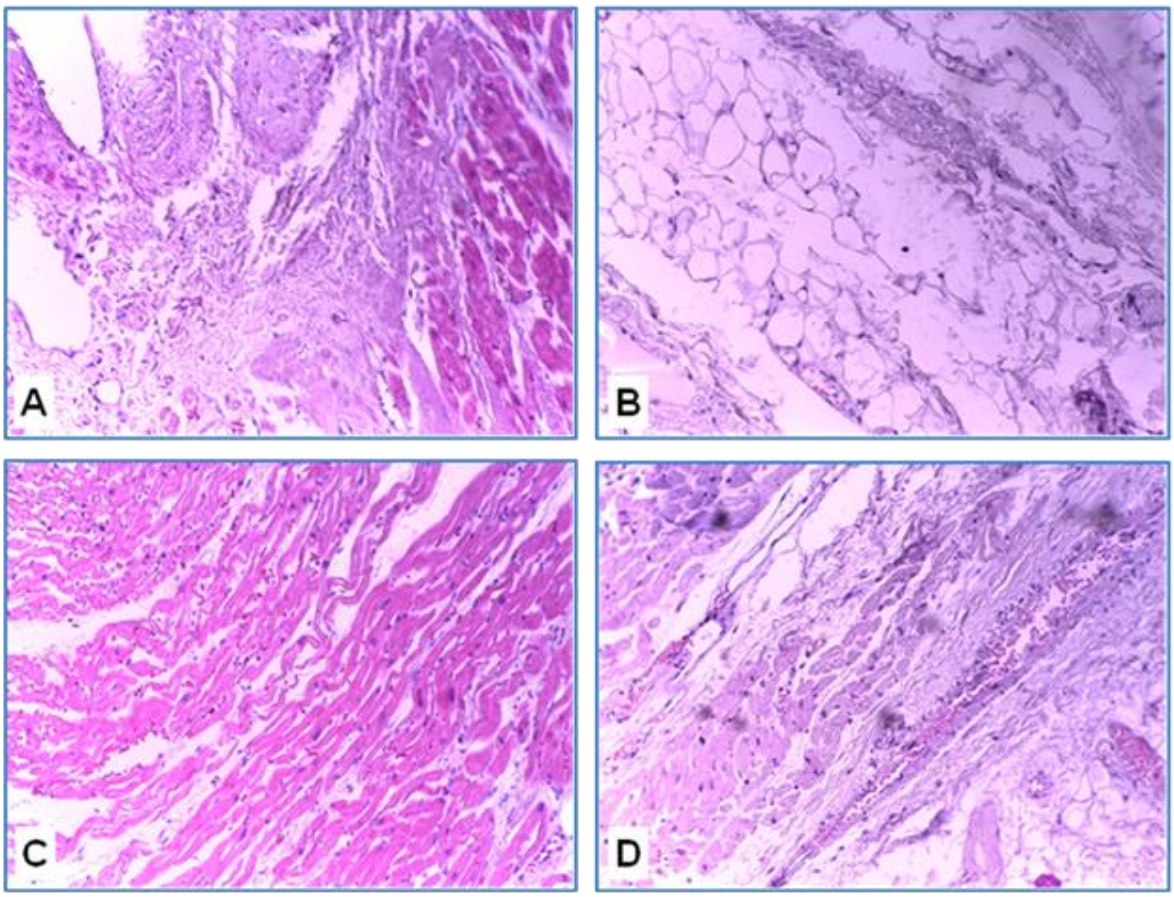

Figure (3): Histopathology studies of cardiac muscle tissue including coronary arteries. Specimens of cardiac tissue were embedded in paraffin for routine H&E staining (×100). A. Micrograph shows myxoid degeneration of media of the proximal left coronary artery (LCA), and myocardium: interstitial; B: Right coronary artery collaterals with myxoid degeneration. C: The image demonstrates myxoid degeneration in the proximal right coronary artery (RCA); D: Distal RCA: Normal myocardium.

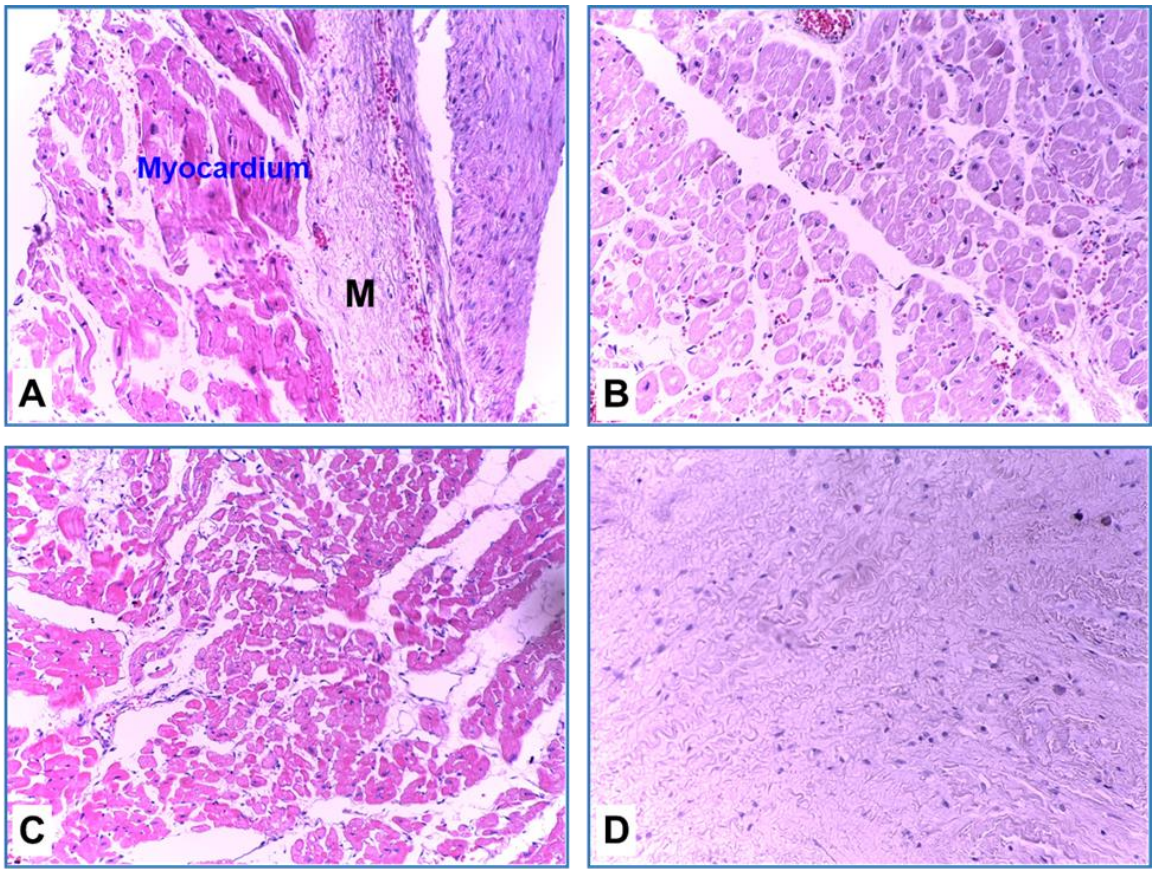

Figure (4): Microphotographs show fibrosis of the heart tissue and coronary arteries stained by H&E (×100). A: Myocardial biopsy showed interstitial fibrosis in the distal LCA, but there was no cardiomyopathic change. The media (M) contains many elastic lamellae and elastic fibers alternating with layers of smooth muscle; B: Fibrosis in the left ventricle endocardium; C: Mild interstitial fibrosis in the papillary muscle of the mitral valve. D: The micrograph shows fibrosis in the leaflet of the mitral valve and myxoid degeneration.

DISCUSSION

ALCAPA is a rare congenital coronary artery malformation. Based on the Canadian heart registry, the incidence of ALCAPA is believed to be 1 per 300,000 live births (5) and it accounts for 0.25%–0.50% of all congenital heart diseases (5, 6). This estimate may not accurately reflect the true incidence, as many patients may be asymptomatic until their death and therefore remain undiagnosed (7).

ALCAPA was first described at 1911 by Abrikossoff on autopsy for a 5 months infants (7). However, the first clinical description of ALCAPA was in 1933 by Edward Bland, Paul Dudley White and Joseph Garland. When they reported for 3 months old the infant presented with symptoms of pallor, cold sweats, and grunting during feeding who unfortunately died at 3.5 months. The discovery of the origin of the LCA arising from the pulmonary artery was made during the autopsy. Therefore, ALCAPA is also known as Bland-White-Garland syndrome (3,6).

During fetal life, the pulmonary arterial pressure equals systemic pressure. Therefore, there is antegrade flow in the LCA and RCA with relatively equivalent oxygen concentration in both systemic and pulmonary circulation. Thus, at this point the patients with ALCAPA will have no symptoms. Shortly after birth, at one to two months of age, the pulmonary arterial pressure gradually starts to decrease. And the flow of LCA becomes retrograde with poor oxygenated blood. Along with the increase demand of myocardium during feeding and crying. Here, infants with ALCAPA start to have myocardial ischemia with ventricular systolic dysfunction and mitral regurgitation which leads to congestive heart failure (1,6). The extent and severity of that depend on the collaterals that have been developed between RCA and LCA and on the diameter of RCA (1,3,6).

According to these changes, ALCAPA can be classified into two types: infantile type, in which there are little collateral with mildly dilated RCA. Therefore, there is no enough blood supply to the left ventricular myocardium, which leads to myocardial ischemia. Infants start to develop symptoms such as pallor, cyanosis, failure to thrive, profuse sweating, dyspnoea, and chest pain, which mostly misdiagnosis as infantile colic. Finally, death can occur in this type in 90% of patients within the first year of life with no intervention (2,3,6). The second type of ALCAPA is adult type its less common accounting for 10%-15% of ALCAPA patients. (7) In this type, adequate collaterals have been developed between RCA and LCA with significant dilatation of RCA, the patient may survive to adulthood with nonspecific symptoms such as reduced exercise tolerance, dyspnoea, palpitation, fatigue, ventricular arrhythmia. Some patients may be asymptomatic and present with sudden cardiac death and the diagnosis was only made postmortem (2,3,6).

In the current report, a history of 18-years-old man who had an episode of cyanosis and dyspnea at the age of 7 months, and since that time he was misdiagnosed with unspecified cardiomyopathy. Unfortunately, the final outcome of this case was a sudden death, and postmortem diagnosis of a dissected heart revealed ALCAPA syndrome.

In our current case report, post-mortem histopathological studies demonstrated Myxoid degeneration of media of the proximal LCA, RCA collaterals, and the proximal RCA. As many as 151 adult cases of ALCAPA were reviewed, of which 12% were diagnosed at autopsy. The average reported age was 41 years and the oldest were 83 years old. There is a greater than 2:1 predominance of females. At the time of presentation, 66% had symptoms of angina, dyspnea, palpitations, or fatigue. 17%presented with ventricular arrhythmia, syncope, or sudden death. A minority (14%) were asymptomatic. The case we present here manifested a sudden death at 18 years old. Early autopsy studies indicated that the average age for sudden death in untreated ALCAPA was around 35 years. This led to all adults with a diagnosis of ALCAPA to undergo early surgical treatment/ correction, however, with the increased use of advanced cardiac imaging and frequent diagnosis of ALCAPA in adults, we are beginning to

understand that the true association between sudden death and ALCAPA may be lower, especially among older patients (8).

There are several diagnostic strategies to screen ALCAPA syndrome invasively and non-invasively. Until recently, conventional coronary angiography has been the gold standard diagnostic tool of choice for coronary anomalies. However, angiography is an invasive procedure with morbidity and mortality rates of 1.5% and 0.15%, respectively (9). Angiography will demonstrate a dilated tortuous right coronary artery with collateral filling of the left coronary system (10).

Multislice computed tomography angiography and magnetic resonance imaging are valuable noninvasive modalities that can be used to identify and define anomalous coronary arteries and their course with a very high accuracy MSCT is a noninvasive imaging technique which is fast and offers excellent spatial resolution, which is required to assess small vessels such as the coronary arteries. The short investigation time, relative non-invasiveness of the procedure, simple preparation, and minimal aftercare make MSCT coronary angiography advantageous over conventional coronary angiography. The main disadvantages of MSCT angiography are its relatively high radiation dose and its inability to assess flow (11). MR imaging does not use ionizing radiation. However, the main disadvantages of MR imaging in comparison with CT are its relatively long examination time and its low spatial resolution (11).

The standard treatment of ALCAPA is through surgical approaches, in which there will be improvement in ventricular function and chronic subendocardial ischemia, which reduces the risk of malignant dysrhythmias and sudden death. Medical therapy may be just used as supportive therapy and to stabilize the patients for surgery (3,7). During the last decades, there have been several surgical techniques used to repair ALCAPA. Ligation of the LCA at its pulmonary artery origin converts the circulation of the heart into one coronary system, so the heart perfusion becomes dependent only on RCA. It is associated with a lot of complications such as atherosclerosis, severe mitral regurgitation, subendocardial ischemia, angina, recanalization of the ALCAPA and sudden death. Therefore, it is no longer used. The other surgical techniques depend on the creation of two coronary systems that provides physiological anatomy, restore left ventricular systolic function and mitral function (3,7,12) includes: coronary button transfer, coronary artery bypass graft (CABG) with ligation of LCA origin and takeuchi procedure.

Coronary button transfer, in this technique, the anomalous LCA with a button of pulmonary artery is directly re-implanted into the aorta, it is considered the best among ALCAPA repairing surgery in restoring anatomy. Most commonly, it is being used in infants because in adults the distance between the aorta and left coronary Ostia is too large. Moreover, LCA with age become friable and less elastic, so there will be a risk of tearing, bleeding, and kinking (1,13). CABG with ligation of LCA origin, is the preferred method in adults in which a venous or arterial bypass graft is being placed from the aorta to proximal left anterior descending (LAD) artery along with ligation of the anomalous LCA at the pulmonary artery origin to avoid competitive flow. It has the risk of stenosis and occlusion of graft, especially vein graft (3,6). Takeuchi procedure that involves using the aortic pulmonary window to make an intrapulmonary tunnel that connects the ostium to the aorta (1,3). It can be used when the LCA origin is distant from the aorta, in case of excessive collateral around the LCA and rigid LCA. It has the risk of pulmonary stenosis and buffalo tearing or leak (3,14,15).

Histopathological examination of the autopsy of the present 18-year-old case showed interstitial fibrosis in the papillary muscle of the the mitral valve and leaflet of mitral valve, distal LCA, and in the left ventricle endocardium. Concomitant repair of the mitral valve during primary surgery for ALCAPA is still controversial. Most of the studies prefer of only primary repair of anomalous LCA in which postoperative follow-up showed that most cases have improvement in MR. As the cause of MR in

ALCAPA patients is secondary to papillary muscle dysfunction and LV dilatation (12,14). Studies recommended close and long-term follow-up for all ALCAPA repaired patients (3,13). In addition to already adopted approaches, there is an emerging demand to determine reliable biomarkers and screening tests for early diagnose of ALCAPA syndrome to treat these cases as early as feasible.

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COMPETING INTERESTS

The authors declare that they have no competing interests.

CONSENT FOR PUBLICATION

Written informed consent for publication of clinical details and/or clinical images was obtained from the parents of the patient. A copy of the consent form is available for review by the Editor of this journal from Dr. Rayyan Al Ali (Head of Forensic Medicine Institute at An-Najah National University). This case report was approved by the Institutional Review Ethical Committee at An-Najah National University.

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