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Case Report

Anomalous origin of the left coronary artery from pulmonary artery a late presentation—Case report and review of literature

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ABSTRACT

An otherwise healthy 26-year-old female presented with sudden cardiac arrest. She was resuscitated with unsynchronized cardioversion for ventricular fibrillation. A left heart cardiac catheterization showed anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA). Cardiac computed tomographic angiography confirmed this abnormality. She underwent direct translocation of the left main coronary artery to the aorta, and after a stormy postoperative course was discharged home.

ALCAPA is a rare congenital abnormality of the coronary system that is associated with early infant mortality and adult sudden death. The use of advanced cardiac imaging has resulted in an increase in the number of diagnosed ALCAPA cases, especially in the adult population, sometimes even in the asymptomatic. The extent of collateral circulation that develops between the right coronary artery and the left coronary artery determines the outcome; the more collateral circulation there is, the less the hypoxic damage to the ventricle. This not only informs us that people survive ALCAPA into adulthood but also highlights the importance for adult cardiologists to be aware of this interesting disease. Corrective surgery remains the treatment of choice. We present a rare case of ALCAPA, with first presentation in adulthood in the form of a malignant ventricular arrhythmia.

<Learning objective: Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital abnormality of the coronary system that is associated with early infant mortality and adult sudden death. Hence we feel it is most important to be aware of this entity, especially as the incidence of diagnosed ALCAPA is on the rise in adulthood. Also the images provided and the discussion have high educational value for generalists and cardiologists alike.>

Introduction

Anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) is a rare congenital abnormality of the coronary system that is associated with early infant mortality and adult sudden death. Its incidence is estimated to be approximately 1 in every 300,000 live births [1]. Previous studies have reported that up to 90% of the infants die without treatment in the first year [2]. Use of advanced cardiac imaging has increasingly identified ALCAPA in adults suggesting that not all patients with ALCAPA have early mortality. ALCAPA is no more a disease of only infants and young children and adult cardiologists need to be aware of this interesting congenital heart disease. We present a case of ALCAPA with its first presentation in adulthood.

Case report

A 26-year-old female with no significant past medical history was transferred to our facility for further management of sudden cardiac arrest. She was a receptionist at the emergency room in another facility. On the day of presentation, while at work she had complained of sudden onset palpitations, which was immediately followed by syncope. The emergency room team noted her to be in ventricular fibrillation. She was promptly resuscitated with direct unsynchronized cardioversion and lidocaine therapy and transferred to our facility. She was otherwise healthy and had an uneventful childhood except that she had noticed one episode of chest discomfort while playing at the age of 10 years. She was seen by her pediatrician and was found to have a heart murmur.

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Subsequent transthoracic echocardiography (TTE) was apparently unremarkable. She was able to participate in volleyball and softball during high school without any untoward incidents. She was on no medications and was not a smoker. She did not drink excessive alcohol or use drugs, her review of systems was unremarkable for any intercurrent illness.

Her examination was unremarkable except for a II to VI systolic murmur heard along the sternal border, which seemed to increase with the Valsalva maneuver. Initial laboratory work at our facility did not show any abnormalities except a mild elevation in white cell count at 11,700 cells/ml, her creatinine kinase was normal at 98 IU/l with a troponin of 0.25 ng/ml which did not rise. Her electrocardiogram (ECG) at the outside hospital had showed ventricular fibrillation (Fig. 1). ECGs in our hospital showed left ventricular hypertrophy (LVH)-type pattern, T wave inversions in I and L, poor R wave progression anteriorly with no R waves until V3, a short PR but no clear delta wave, and a normal QT interval. A 2D TTE showed that the left ventricular systolic function was at the lower limits of normal, with enhanced epicardial blood flow, appearing as if the 'myocardium was in flames' (Fig. 2), and LVH was not detected. Left heart cardiac catheterization showed ALCAPA. It also showed significantly dilated right coronary artery (RCA) with extensive collateral inter-coronary arteries supplying all of the territory of the left ventricle (Figs. 3 and 4). Cardiac computed tomographic angiography (Figs. 5 and 6) was performed to better define the coronary anatomy, which corroborated that the left coronary artery (LCA) was arising from the posterior wall of the pulmonary artery just prior to its bifurcation into left and right main pulmonary artery segments. A transesophageal echocardiogram confirmed the abnormal origin of the LCA.

Because of her symptomatic presentation of ALCAPA, in the form of malignant ventricular arrhythmia, and evidence of left ventricular dysfunction, in consultation with the cardiothoracic surgeons and the pediatric cardiologists it was deemed that she would be best served by corrective surgery. She underwent direct translocation of the LCA to the aorta. Her postoperative course was complicated by cardiac arrest and cardiogenic shock. An implantable cardiac defibrillator was put in her for sustained ventricular tachycardia, postoperatively. She recovered well and was discharged home. We present a rare case of an otherwise healthy female with ALCAPA,

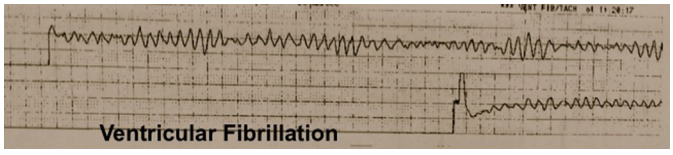

Fig. 1. Telemetry strip from the scene of cardiac arrest showing ventricular fibrillation.

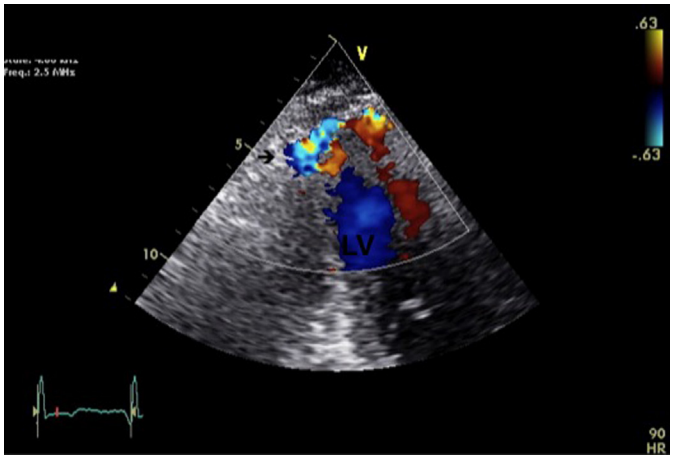

Fig. 2. Transthoracic echocardiogram showing increased epicardial blood flow – appearing as if the 'myocardium was in flames' LV, left ventricle.

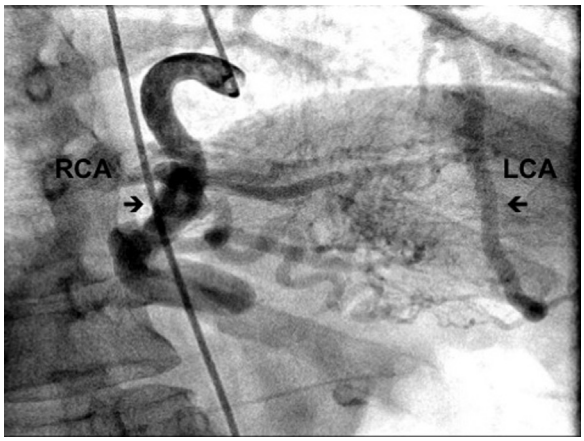

Fig. 3. Selective angiogram of the right coronary artery (RCA) showing dilated RCA with collaterals filling the left coronary artery (LCA).

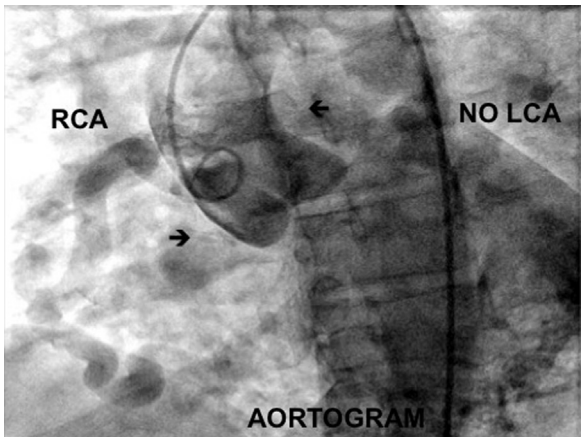

Fig. 4. Aortogram showing the origin of right coronary artery (RCA) only. Note smooth contour of the left side of the ascending aorta, and absence of the left coronary artery (LCA) at its usual origin.

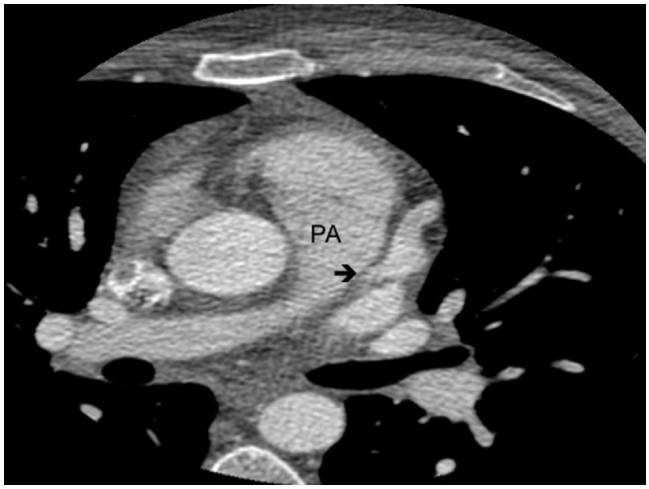

Fig. 5. Coronary angiography with computed tomography showing origin of the left coronary artery from the pulmonary artery (PA).

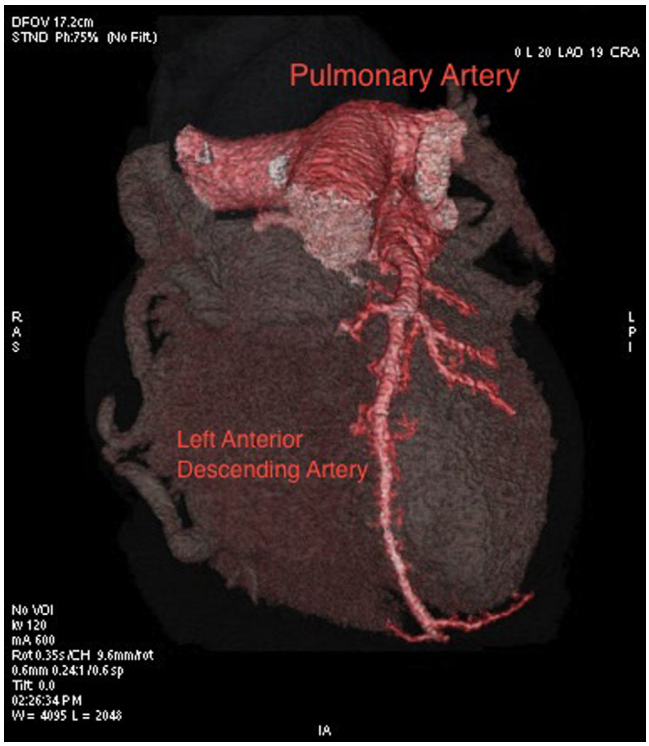

Fig. 6. 3D reconstruction of computed tomography of the heart showing origin of the left coronary artery from the posterior aspect of the pulmonary artery.

with her first presentation in adulthood in the form of a malignant ventricular arrhythmia.

Discussion

ALCAPA results either from an abnormal septation of conotruncus into aorta and pulmonary artery or from the persistence of the pulmonary buds combined with involution of the aortic buds that form the coronary arteries [3]. It usually manifests alone, but associated abnormalities like atrial septal defect, ventricular septal defect, and coarctation of aorta have been described [4].

ALCAPA is well tolerated in-utero and in infancy because pulmonary arterial pressure equals the systemic pressure, this leads to antegrade flow in both the anomalous LCA and the normal RCA. Also, the saturation difference between these two circulations is not significant. Even in the early stages of discovery of ALCAPA, it was evident that the symptom onset coincides with the closure of the ductus arteriosus [5]. The illustration of a 3-month old infant with cardiac ischemia after a quiescent period of 2 months came to be known as Bland–White–Garland syndrome [6].

The pulmonary arterial pressure and saturations gradually decrease after birth, leading to poor perfusion with hypoxic blood of the anomalous LCA. Total separation of the pulmonary and the arterial systems after the closure of ductus arteriosus leads to the desaturated low-pressure pulmonary artery perfusion of the LCA, leading to chronic hypoxemia of the left ventricle. Subsequently, flow through the LCA reverses and the oxygenated blood is shunted from high-resistance myocardial circulation to low-pressure pulmonary circulation. This shunt, known as the 'steal phenomenon', leads to failure of LCA to supply the myocardium. The extent of collateral circulation that develops between the RCA and LCA during this time determines the extent of myocardial injury and hence, the outcome. This probably is the difference between the patients who survive the infancy to adulthood and patients who do not. Those who survive to adulthood, which accounts for about 10–15% of patients, have shown compensation by an enlarged RCA with extensive collateral inter-coronary arteries [7]. Minimal coronary steal from the pulmonary artery due to ostial stenosis of the LCA or a restrictive opening into the pulmonary artery limiting the left-to-right shunt and development of systemic blood supply to the LCA via bronchial artery collateral vessels have also been attributed to increased survival [8]. However, as with other collateral circulations, this is often not sufficient leading to chronic left ventricular subendocardial ischemia.

Patients may be asymptomatic or present with mitral insufficiency, ischemic cardiomyopathy, or with malignant ventricular dysrhythmias in whom sudden cardiac death occurs in 80–90% of cases [9]. About two thirds had symptoms of angina, dyspnea, palpitations, or fatigue and about 62% with life-threatening presentations did not have antecedent symptoms [1]. The effects of exercise on the dynamics of such circulation have not been well investigated; malignant arrhythmias during exercise implicating acute ischemia and coronary steal phenomenon have been reported [10]. Yau et al. [1], in their review of the adult ALCAPA cases, reported that, among the 151 cases, 12% were diagnosed at autopsy, with the average age of life-threatening presentations at 33 years old $\pm$ 14 years with a 2:1 female predominance. They also found that on examination about 87% of the patients had murmurs with 28% having a continuous or "to-and-fro" murmur predominantly localized to the left sternal border or apical region. On ECG, Q-waves were noted in 50%, LVH in 28%, and left axis deviation in 15%.

Although, coronary angiography remains the traditional standard for diagnosis, cardiac CT and cardiac magnetic resonance imaging (MRI) have been increasingly employed for identification of ALCAPA. They all clearly show the abnormal origin of the LCA from the pulmonary artery (commonly on the left inferolateral side of the main pulmonary artery just after the pulmonic valve), increased collateral coronary flow and a mega RCA. Frequent echocardiographic findings include abnormal origin of the LCA, dilated RCA, retrograde filling of the anomalous coronary, abnormal diastolic flow in the main pulmonary artery, and more importantly abnormal septal or epicardial color flow signals from the collaterals [11]. Global hypokinesis, mitral valve prolapse, fibrotic papillary muscles and chordae have also been reported. MRI has an added advantage in that it provides information about the viability of the myocardium. Other causes of coronary dilatation such as Kawasaki disease, Takayasus arteritis, polyarteritis nodosa, atherosclerotic coronary ectasia, coronary fistulas, Ehlers–Danlos syndrome, scleroderma, hereditary telangectasia among others should be considered in the differential diagnosis [12].

Early autopsy studies indicated that the average age for sudden death in untreated ALCAPA was around 35 years [13]. This led to all adults with a diagnosis of ALCAPA to undergo early surgical treatment, 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 [1]. Surgical treatment is definitive. Surgeries aimed at establishment of dual coronary supply are superior to surgeries that provide only a single coronary artery, as in ligation of the anomalous LCA alone [14]. Two-coronary system surgeries include coronary button transfer or transpulmonary baffle (Takeuchi procedure) or coronary artery bypass graft placement with ligation of the ALCAPA [8]. Orthoptic cardiac transplant [15] and percutaneous closure with vascular plug [16] have also been reported as treatment modalities. Management of patients with incidental discoveries of ALCAPA among asymptomatic patients is controversial, and a conservative approach has been adopted in the elderly due to seemingly lack of benefit from surgery [17]. Some have suggested use of viability and functional data obtained by a cardiac MRI to guide appropriate therapy [18]. Long-term results following establishment of

dual coronary systems are excellent [19], but are lacking in non-surgically managed patients. In the study by Yau et al. [1], of the 7 medically managed patients 5 were doing well up to 5 years after diagnosis, while 1 died of cancer and the other relented to surgery. The implantation of defibrillator following surgery remains controversial; it is not conclusive that treatment of ischemia abolishes the risk of malignant arrhythmias in these patients, as ischemia may not be the sole cause of malignant arrhythmias [20].

Conclusion

ALCAPA is a rare, yet life-threatening, congenital abnormality of the LCA arising from the pulmonary artery. About 10–15% of patients survive to adulthood. Increasing availability of cardiovascular imaging has resulted in increasing incidence of ALCAPA. Surgical therapy is definitive, and remains the treatment of choice in patients with symptoms or left ventricular dysfunction. We present a rare case of ALCAPA presenting as fatal ventricular arrhythmia in adulthood, which was aborted in a timely manner. She underwent successful coronary button transfer. She was doing well upon discharge.

Conflict of interest

Authors declare no conflict of interest.

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