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CASE REPORT

Aborted Sudden Cardiac Death in a Young Male with Anomalous Left Coronary Artery Arising from the Pulmonary Artery

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Anomalous left coronary artery arising from the pulmonary artery (ALCAPA) is a rare type of congenital coronary abnormality that may be associated with early infant mortality and sudden adult cardiac death. We report a case regarding a 23-year-old male who collapsed during a marathon race and was resuscitated with cardiopulmonary resuscitation. Subsequent workups verified the diagnosis of ALCAPA. The patient underwent surgical intervention with obliteration of the ALCAPA orifice and coronary artery bypass grafting with left internal mammary artery to left anterior descending coronary artery. The procedure was done smoothly, and he was discharged uneventfully.

Key words: Aborted sudden cardiac death, congenital coronary anomaly, Anomalous left coronary artery arising from the pulmonary artery

INTRODUCTION

Coronary artery anomalies are congenital abnormalities in the number, origin, course or termination of coronary arteries and are found in approximately 1% of the population.[1] Anomalous left coronary artery arising from the pulmonary artery (ALCAPA), or Bland-Garland-White syndrome is a rare congenital heart disease which accounts for about 0.25%–0.50% of congenital heart diseases.[2] ALCAPA is associated with high mortality in infants and is an uncommon diagnosis in adults. We report a case of an exercise-induced sudden cardiac death resulted from ALCAPA in a young male. The patient was rescued with immediate cardiopulmonary resuscitation (CPR), adequate defibrillation, and subsequent successful surgical intervention.

CASE REPORT

A healthy 23-year-old male, who was robust in the past with no significant previous medical history, was a regular runner and experienced dyspnea on exertion. Because of the exacerbated symptoms, he visited our cardiology section 2 years ago before this admission. At that time, echocardiography showed dilation on both left atria (44 mm) and left ventricle (59 mm in diastolic phase and 41 mm in systolic phase) without valve abnormality and an ejection fraction of about 57%. The treadmill exercise test was positive, and a Thallium 201 Myocardial Imaging depicted scintigraphy with decreased perfusion to the extensive anterior wall, apex and mild-sized inferior wall of the left ventricle. Coronary angiography (CAG) and computed tomographic coronary angiography (CTA) were performed and revealed that the orifice of the left main coronary artery (LMCA) originated from the pulmonary trunk; in addition, a giant RCA with significant collateral vessels to left anterior descending (LAD) and left circumflex (LCx) were also observed. The diagnosis of ALCAPA was impressed, and surgical treatment was advised. However, the patient refused surgery and follow-up appointments. After the diagnosis, the patient still continued the habit of regular running, with an average distance of 12–21 km a day.

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At present, the patient was admitted to our hospital after a witnessed cardiac arrest during a marathon race. He was found suddenly collapsed into what was described by his teammates as a "pulseless, tonic-clonic seizure," and total unconsciousness. Immediate CPR was performed. On the way to the local hospital, the patient was found to be in a ventricular fibrillation rhythm, and an automated external defibrillator was used that resulted in the spontaneous return of circulation and recovered consciousness. After the vital signs were stabilized in the local hospital, the patient was transferred to our hospital for further workup and management.

The initial laboratory workup showed elevated white blood cell count of 18,180 cells/mcL, mildly elevated aspartate transaminase of 103 U/L, elevated CK of 637 U/L, elevated troponin I of 1.518 ng/mL, elevated BNP of 164 pq/mL, while the electrolytes were within normal limits. The electrocardiogram in the emergency room showed a normal sinus rhythm. CAG and CTA revealed inflow from the right coronary artery (RCA) to the pulmonary artery (PA) through multiple collateral vessels and a retrograde flow of the LMCA that verified an ALCAPA diagnosis [Figure 1a and b].

Subsequent open heart surgery was performed through a median sternotomy, and a cardiopulmonary bypass was established with cannulation from the ascending aorta and superior vena cava. The antegrade cardioplegic solution was infused through the ascending aorta with temporary occlusion of the ALCAPA orifice. During the operation, abundant collateral flow to the coronary arteries was noted, and it was difficult to arrest the heart for even 3 min after cross-clamping of the aorta and completed infusion of the cardioplegic solution. The orifice of LMCA over PA was obliterated and left internal mammary artery (LIMA) was anastomosed to LAD. The patient tolerated the procedure well and was discharged uneventfully. He is now doing well and has a regular running program 18 months after the operation.

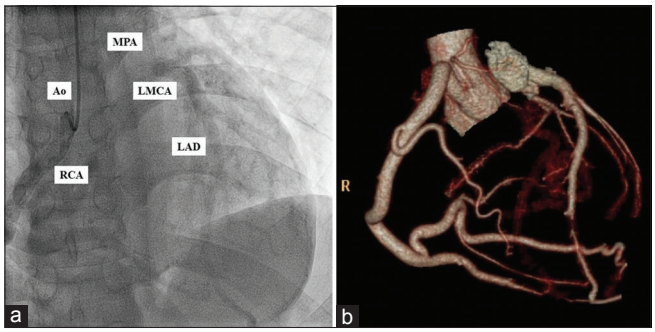

Figure 1: Coronary angiography (a) and computed tomographic angiography (b) showed a dilated and tortuous right coronary artery with retrograde filling of the left main coronary artery and main pulmonary artery through extensive posterior and apical collateralization

Follow-up echocardiography showed normal left ventricular dimensions and ejection fraction. No resting regional wall motion abnormalities were detected.

DISCUSSION

ALCAPA was first described in 1933 and has been reported in about 1/300,000 live births.[3,4] It is usually well tolerated at birth due to the elevated pressure in newborn pulmonary arteries, which provides an antegrade perfusion of myocardium through the left coronary artery. After the 1st year of life, pulmonary vascular resistance falls, which results in a progressive retrograde filling of the left main coronary and pulmonary arteries through the collateral vessels from the RCA. This result in a steal of blood flow from the myocardium, leading to myocardial ischemia, heart failure, and mitral regurgitation.[5] Meanwhile the left-to-right shunt also leads to ventricular overload and dilation. Without timely treatment, these complications are life-threatening and only about 10% of patients can have sufficient collateral circulation from the RCA and survive beyond infancy.[6] Asymptomatic ALCAPA patients may be found in 14% of them and diagnosed either incidentally or by biopsy. However, most patients present with symptoms of angina, palpitations, or fatigue. In about 17% of cases, life-threatening arrhythmias, syncope, and sudden cardiac death may develop due to inadequate myocardial perfusion, which triggers arrhythmias, especially under rigorous exercise as in this presented case.[7]

Surgical correction is the gold standard for the treatment of ALCAPA and has demonstrated excellent results in infants with a survival rate of about 95% after a 20-year follow-up.[8] In adult patients, who survived a sudden cardiac arrest and were treated surgically, available data reveal promising results as well. Most adult patients who were resuscitated from cardiac arrest secondary to ALCAPA were young, with an average age of 33 years and presented with cardiac arrest as the first symptom of this anomaly.[9] Several surgical options were reported to treat ALCAPA including reimplantation of the anomalous left coronary artery, a Takeuchi operation and closure or ligation of the LCA with or without coronary artery bypass graft (vein or LIMA). The management and long-term outcomes have not been adequately defined, but anatomical surgical correction remains the best treatment modality for this subset of patients.[10] No randomized control trials are yet available to evaluate long-term outcomes of different managements.

CONCLUSION

In summary, coronary artery anomalies are rare but should be suspected in young adults who have good exercise

capabilities but present with cardiac symptoms. Rigorous exercise may induce aborted sudden cardiac death in patients with ALCAPA. Early surgical interventions are advised to correct the coronary steal syndrome and myocardial ischemia, and therefore establish a two coronary perfusion system that is mandatory for long-term survival.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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