Adult ALCAPA Syndrome As First Presentation With Atrial Fibrillation In A Marathon Runner

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Introduction

Anomalous left coronary artery arising from the pulmonary artery (ALCAPA) is also known as Bland-White-Garland syndrome, is a very rare congenital heart disease with high mortality rate near to 90% in the first year of life if not treated. Fewer patients survive beyond first year of life if adequate collateral circulation is present [1]. Similarly, diagnosis in living adult is rare as most information of ALCAPA syndrome is from case reports and patients who have been resuscitated and survived [2]. On the contrary, our patient presented with chest pain and palpitation secondary to atrial fibrillation observed on admission. Also, developed a stroke which resulted in left sided weakness the next day.

Case Presentation

A 38-year-old female with a past medical history of mitral regurgitation diagnosed at age 24 presented with a sudden onset of chest pain, palpitations and diaphoresis in the emergency department. The patient was a marathon runner and normally in good health. On presentation, the patient's vitals revealed an irregular heart rate in the 150s and a blood pressure of 88/47. Atrial fibrillation with a rapid ventricular response (Afib RVR) was observed on the electrocardiogram. The patient was calm and comfortable at that time with no subjective symptoms. Initial troponin was negative but 6 hour later it increased to 15.8 ng/mL and further increased to 18.5 ng/mL later on. Intravenous (IV) heparin therapy was administered, and the patient underwent urgent cardiac catheterization. The cardiac angiogram showed markedly ectatic right coronary artery (RCA) (Figure 1 and 2). Left main coronary artery (LMCA) appears to be anomalous. Left descending artery (LAD) and left circumflex artery (LCx) not visualized in usual position due to possibly occluded LAD. It appears to be fistula involving branch of LAD (Figure 3). Echocardiogram revealed ejection fraction (EF) 55%, moderate mitral regurgitation, mildly dilated left atrium, and right ventricle systolic pressure was estimated to be 52mmHg. There was no intervention performed during the cardiac catheterization due to anomalous of coronary arteries. The patient spontaneously converted to sinus rhythm, and heart rate and blood pressure improved. Local cardiothoracic surgeon recommended further evaluation by a congenital heart specialist in a tertiary health center and patient discharged with referral.

The patient presented the next day with left sided weakness and slurred speech, which CT brain without contrast showed right frontal 4.2 cm hematoma, no midline shift (Figure 4). The patient had successful evacuation of hematoma; left sided weakness started improving.

After complicated hospitalization, she followed up with cardiovascular surgery at outside facility. She had a coronary computed tomography angiogram (CCTA) done which was showing LMCA origination from main pulmonary artery (Figure 5). 3D configuration of CCTA also showed dilated and tortuous RCA with multiple collaterals extending to left ventricle (Figure 6). The diagnosis of ALCAPA made, and surgical options discussed with the patient. The patient recovered after a successful surgical repair of anomalous left main coronary artery with left main translocation to aorta.

Imaging

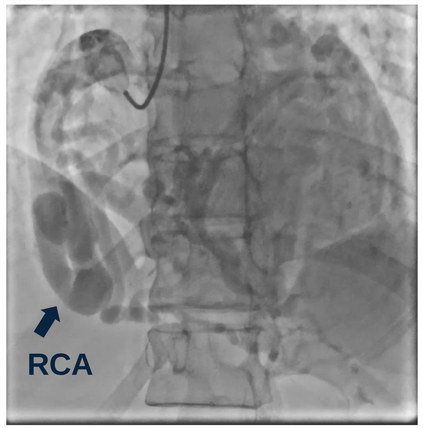

Figure 1: Coronary angiography showing right coronary artery (RCA) dilation and tortuosity, right circulation extending to the left ventricle (LV).

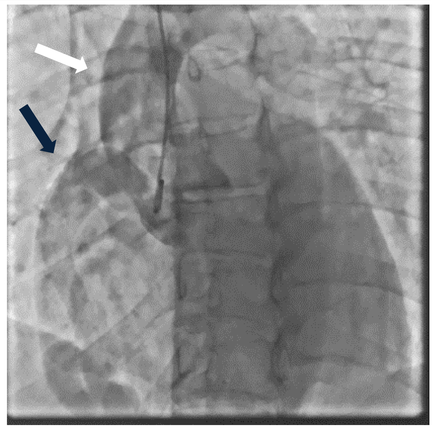

Figure 2: Coronary angiography showing right coronary artery (black arrow) almost the same size as the aorta (white arrow).

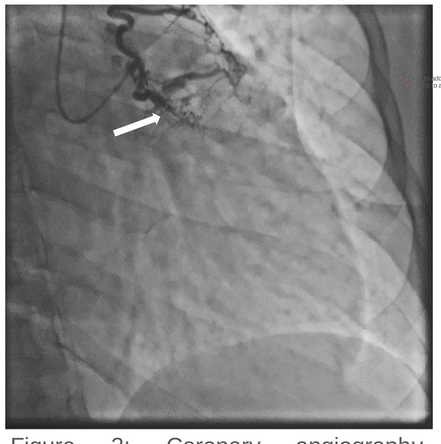

Figure 3: Coronary angiography demonstrating left coronary circulation. LAD is occluded (white arrow)

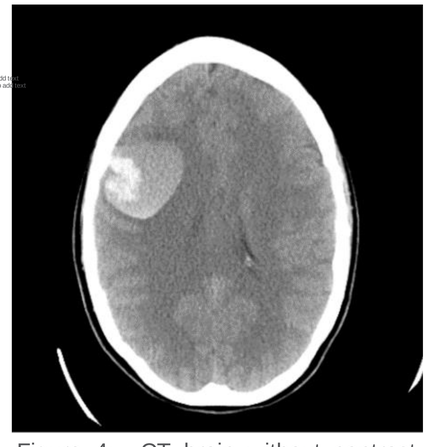

Figure 4: CT brain without contrast shows 4.2 cm hematoma in the right frontal lobe with no midline shift.

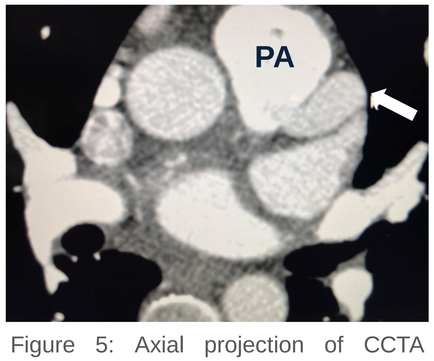

Figure 5: Axial projection of CCTA showing left main coronary artery (white arrow) originating from main pulmonary artery (PA).

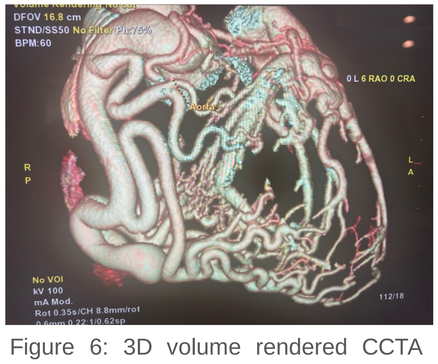

Figure 6: 3D volume rendered CCTA showing dilated and tortuous RCA with multiple extensive collaterals supplying to left ventricle, while left coronary circulation is very limited.

Discussion

ALCAPA syndrome is a rare disease involving the anomalous rising of the LCA from the Pulmonary artery, and its presentation in adulthood is even rarer. The disease can be classified into two types: the infant type and the adult type [3]. In infant type, there is no or very limited collateral development from RCA to LCA which leads to early signs and symptoms of heart failure, eventually results in serious outcomes including sudden cardiac death in 90% of the cases (1). Adult type ALCAPA seems to present with a wide variety of symptoms including dilated cardiomyopathy (DCM), mitral regurgitation, chest pain, dyspnea on exertion as well as acute coronary syndrome and ventricular arrhythmia (2).

Our patient is a marathon runner whose medical history consisted only of MR. The patient denied any history of rheumatic disease in childhood or symptoms suggestive of Kawasaki disease, which was an initial differential diagnosis in the setting of dilated RCA found on imaging. An echocardiogram was also negative for wall motion abnormalities or dilated cardiomyopathy. Though our patient did not have DCM and never received adequate workup for the cause of her mitral valve insufficiency, data suggests that ALCAPA syndrome should be considered in the differential diagnosis of a patient with the combination of dilated cardiomyopathy and mitral regurgitation [4]. For suspected ALCAPA syndrome, there are two main modalities used for diagnosis; transthoracic echocardiography (TTE) and coronary computed tomography angiography (CCTA); research found there was no statistical difference between two for accuracy [5].

The treatment of choice for ALCAPA syndrome is surgical repair to improve myocardial perfusion. Varying surgical approaches may be employed and can be classified into 2 groups: one-coronary-system and two-coronary-system repairs (preferred) [6].

Conclusion

We presented a case of a healthy young female with history of mild MR who presented with A-fib RVR and NSTEMI as first presentation of adult type ALCAPA. Before surgery could be performed, she developed a hemorrhagic stroke which still has no relation to her cardiac abnormality, as we know. In conclusion, we would like to emphasize that mitral valve insufficiency in a young healthy patient should raise concerns for further investigation.

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