Case Report

Bileaflet mitral valve prolapse in an elderly patient with of anomalous left coronary artery from the pulmonary artery

ABSTRACT

Anomalous origin of the left coronary artery from the pulmonary artery with bileaflet mitral valve prolapse is rare and seldom reported. We report a case of an elderly man who presented with typical angina symptom and a positive exercise stress test. Coronary angiogram failed to identify arterial course due to the presence of extensive collaterals. Images from cardiac computed tomography angiography provided the correct diagnosis and guided treatment. He was treated with medical therapy and remained asymptomatic on follow-up.

Keywords: Angina, anomalous left coronary artery from the pulmonary artery, cardiac imaging, coronary anomaly, mitral regurgitation, mitral valve prolapse

INTRODUCTION

Anomalous left coronary artery from the pulmonary artery (ALCAPA) is a rare anomaly and survival until 60 years of age is not commonly reported. This is the first case of ALCAPA reported from Malaysia.

CASE REPORT

A 62-year-old male presented with worsening angina over 6 months. He has a background history of hypertension and chronic atrial fibrillation. Clinical evaluation revealed a soft pansystolic murmur over the mitral area. Echocardiography revealed the presence of bileaflet mitral valve prolapse with severe mitral regurgitation and mild pulmonary hypertension [Figure 1a and b]. Left ventricular ejection fraction was 60% with normal chamber size, left ventricular hypertrophy with septal thickness of 12 mm as well as left atrial dilatation of 86 mL. An exercise treadmill test showed evidence of ischemia with 2 mm ST-segment depression in leads II, III, aVF, V4, V5, and V6.

The left main coronary was absent during angiography. Injection of the right system revealed multiple collaterals and incomplete filling of the distal artery [Figure 1c and d].

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A cardiac CT angiogram [Figure 2] confirming the diagnosis of anomalous left coronary artery from the pulmonary artery (ALCAPA). Subsequent cardiac magnetic resonance imaging demonstrated no evidence of subendocardial late gadolinium enhancement. There was some late gadolinium enhancement at the inferior right ventricular insertion point to the interventricular septum [Figure 3].

DISCUSSION

ALCAPA is usually detected in the pediatric population. The incidence in adulthood is estimated at 15%.[1] Symptoms commonly reported include angina, dyspnea,

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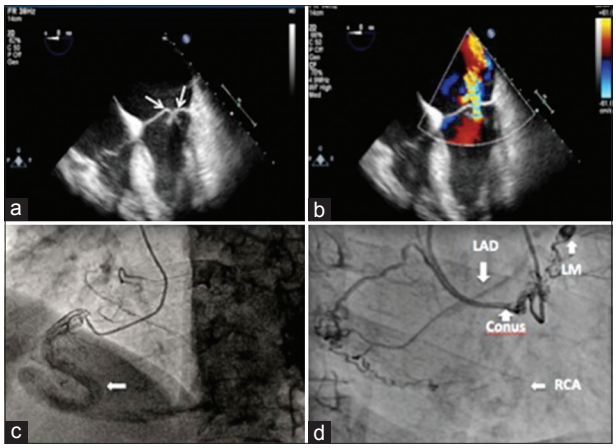

Figure 1: (a) Transesophageal echocardiography in mid esophageal view showing bileaflet mitral valve prolapse (arrow) and (b) showing severe mitral regurgitation. (c) Coronary angiography showing faint filling of the right coronary artery (arrow). (d) Conus branch from the right coronary artery supplying the right atrium. There is also small collateral from the proximal conus branch feeding into the left main which then fills up the left anterior descending artery

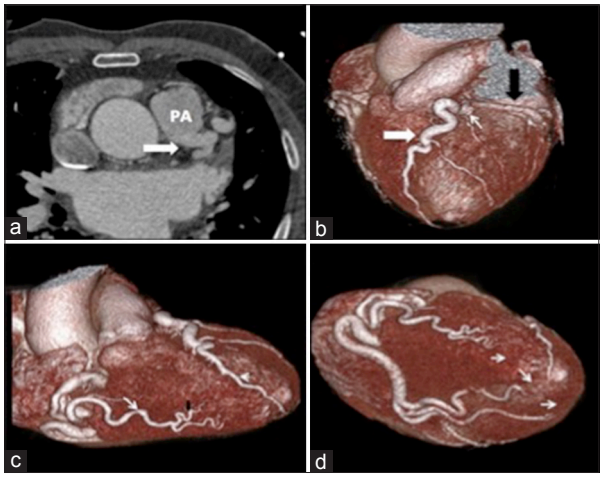

Figure 2: (a) Multislice computed tomography showing the origin of the left coronary artery (white arrow) from the pulmonary artery in axial view. (b) Three-dimensional volume-rendered multislice computed tomography showing origin of the left coronary artery (arrowhead). The left anterior descending (white arrow) courses in the interventricular groove. The left circumflex (black arrow) is seen in the left atrioventricular groove. (c) A dilated intercoronary collaterals (black arrow), which connect the tortuous right coronary artery (long arrowhead) to the left anterior descending (short arrowhead). (d) Collaterals between the large posterior descending artery to the distal left anterior descending

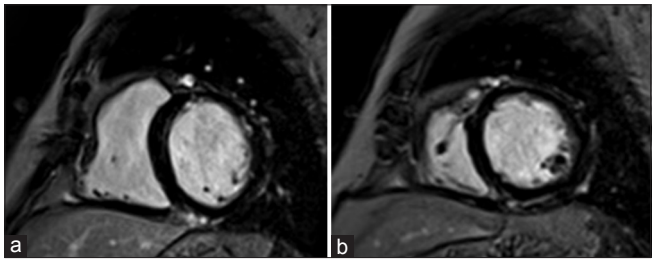

Figure 3: Cardiac magnetic resonance images in short-axis views (a and b) demonstrating some late gadolinium enhancement at the right ventricular insertion point to the inferior interventricular septum

palpitation, and fatigue.[2] Patients with ALCAPA have an expected lifespan of 94.5% at 20 years when corrected surgically in childhood.[3] A fifth of those who survived to adulthood face the risk of malignant arrhythmias or sudden death.

The survival of uncorrected ALCAPA is made possible by extensive collateral formation between the right coronary artery (RCA) and left anterior descending (LAD) artery. This group would remain asymptomatic with thirty percent of them manifesting symptoms beyond the age of fifty.[2] Symptom onset can be due to either flow-limiting coronary stenosis of the contributing vessel or coronary steal syndrome.[4]

The coronary steal syndrome occurs as retrograde LAD flow from distal RCA collaterals preferentially empties into a low-pressure pulmonary artery system as opposed the high resistance coronary circulation.[4] This leads to hypoperfusion of the subtended myocardium with ensuing ischemia and subsequent symptoms in situation of increased myocardial oxygen demand.

Patients with uncorrected ALCAPA have a tendency to develop left ventricular diastolic overload as a result of the left to right shunt.[5] This phenomenon may contribute to the left ventricular hypertrophy and mitral regurgitation seen in reported cases in the literature.[1,2]

Myocardial flow reserve plays a major role in maintaining perfusion in collateralized vessels.[6] Patients with ALCAPA have been shown to have reduced myocardial flow reserve despite extensive collateral formations.[7] Despite having vessels free of flow-limiting stenosis, our patient developed ischemia on exertion most likely due to demand perfusion mismatch as described above.

Mitral regurgitation in association with ALCAPA is not rare with an incidence of 32%–44%.[8] The reported etiology is thought to be due to either active ischemia or functional mitral regurgitation as a result of a dilated ventricle. Our patient has ALCAPA with primary degenerative mitral regurgitation which to the best of our knowledge has never been published.

A third of adult ALCAPA diagnosed above the age of 50 years old are managed medically.[3] Long-term results following reestablishment of a dual coronary system postsurgery are excellent. Data pertaining outcome in relation to medical therapy is not known.[2] Our patient remains asymptomatic and active at 1-year follow-up on optimal medical therapy.

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Conflicts of interest

There are no conflicts of interest.

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