CASE REPORT

Late presentation of anomalous left coronary artery arising from pulmonary artery with acute coronary syndrome

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ABSTRACT

Congenital anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA) 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 asymptomatic. The extent of collateral circulation that develops between the right coronary artery (RCA) and left coronary artery (LCA) determines the outcomes. We present a case of hitherto undiagnosed case of ALCAPA, with first presentation as acute coronary syndrome (ACS) in young male.

Key words: Acute coronary syndrome, Adulthood, Congenital anomaly, Survival

INTRODUCTION

Congenital anomalous origin of the left coronary artery (LCA) from the pulmonary artery (PA) (ALCAPA) 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 asymptomatic. The extent of collateral circulation that develops between the right coronary artery (RCA) and LCA determines the outcomes. Adequate collateral supply leads to less hypoxic damage of left ventricle (LV) in infancy and an improved survival in adulthood. We present a case of hitherto undiagnosed case of ALCAPA, with first presentation as acute coronary syndrome (ACS) in a young man.

CASE SUMMARY

A 35-year-old man with no significant medical history was transferred to our facility for further management after he had an ACS. He had been admitted in another hospital with a history of sudden-onset chest pain 5 days before admission to our hospital. He had a new-onset and transient right bundle branch block during that episode of ACS-unstable angina. He also underwent a treadmill test, which was positive, so he was referred to us for cardiac catheterization.

The electrocardiogram at presentation to our hospital was unremarkable. Coronary angiography showed aneurysmally dilated RCA with empty left aortic sinus. RCA was providing large collateral supply to left coronary system [Figure 1]. On the basis of these findings, a suspicion of ALCAPA was made, and subsequent pulmonary angiography showed the LCA originating from the PA [Figure 1]. Cardiac computed tomographic image confirmed the origin of the LCA from the PA [Figure 2]. Echocardiogram also showed dilated RCA with the origin of LCA from the PA with abnormal color flow signals from collaterals. LV strain analysis revealed reduced global longitudinal strain in the apex and the apical septum [Figure 3]. The left ventricular ejection fraction was normal

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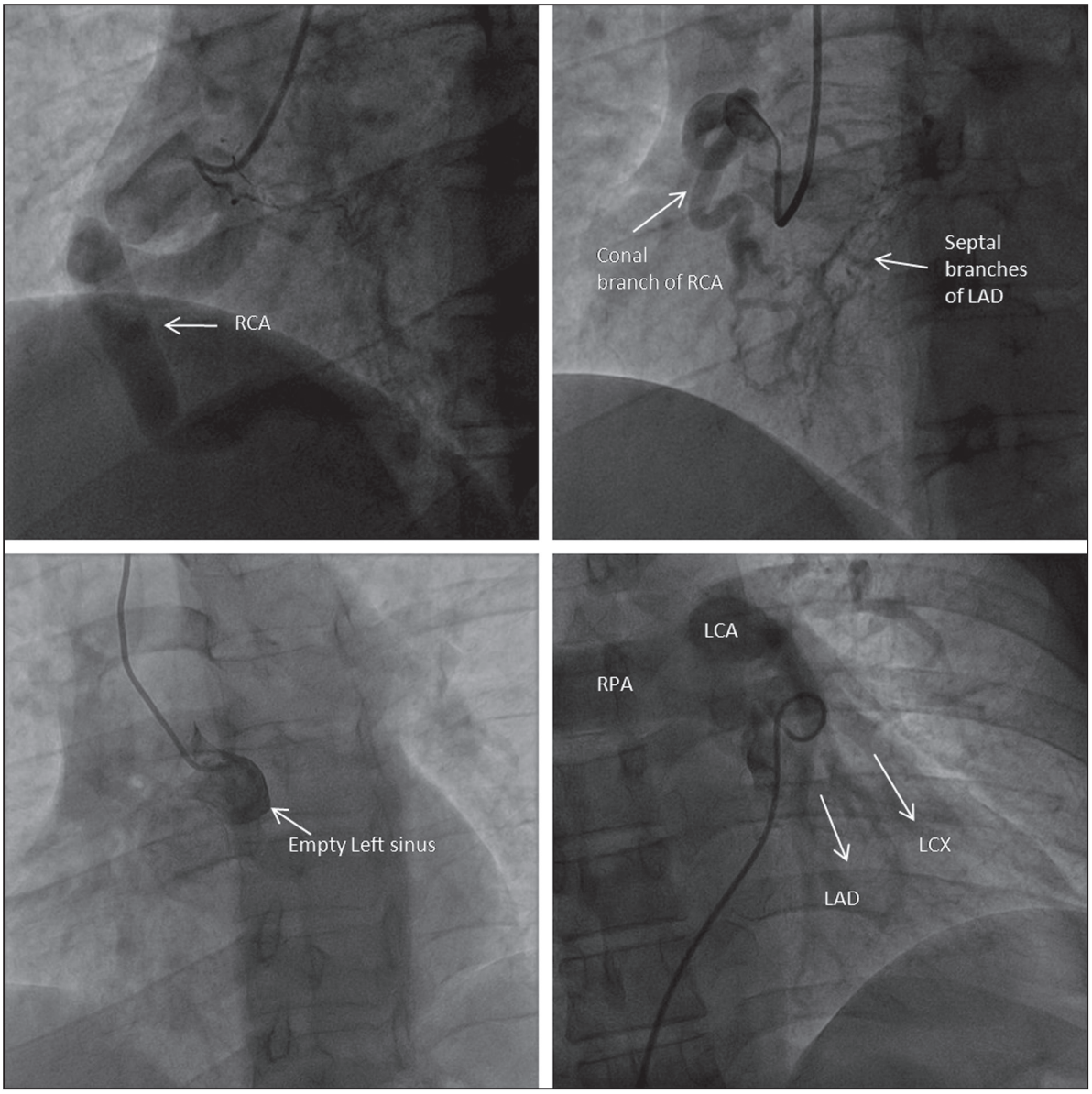

Figure 1: Coronary angiogram showing aneurysmally dilated right coronary artery (RCA) (upper left). Huge collateral supply from conal branch of RCA to left coronary artery (LCA) (upper right). Coronary angiogram showing empty left sinus (lower left). Pulmonary angiogram showing the origin of LCA from the pulmonary artery (PA). [LAD = left anterior descending artery, LCX = left circumflex artery, RPA = Right pulmonary artery.]

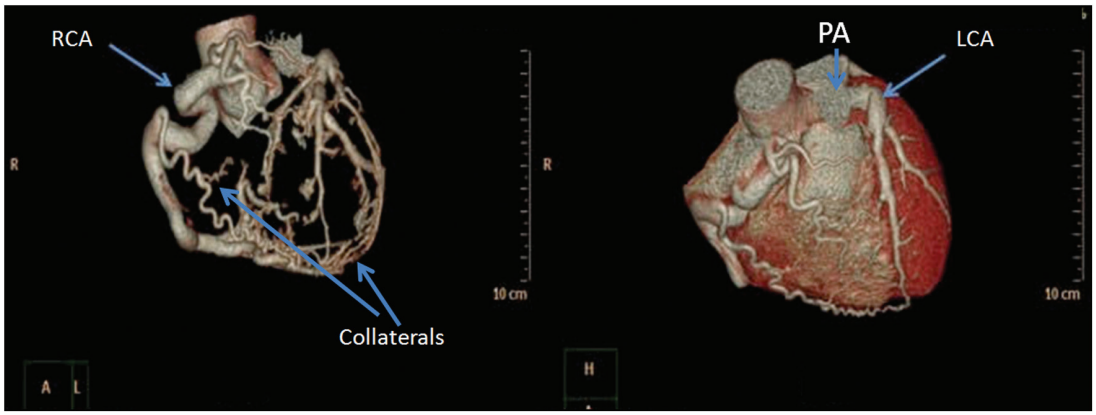

Figure 2: Computed tomographic angiogram showing aneurysmally dilated right coronary artery (RCA), giving rise to collaterals to left coronary artery (LCA) (arrow, left panel), and LCA arising from pulmonary artery (PA) (arrow, right panel).

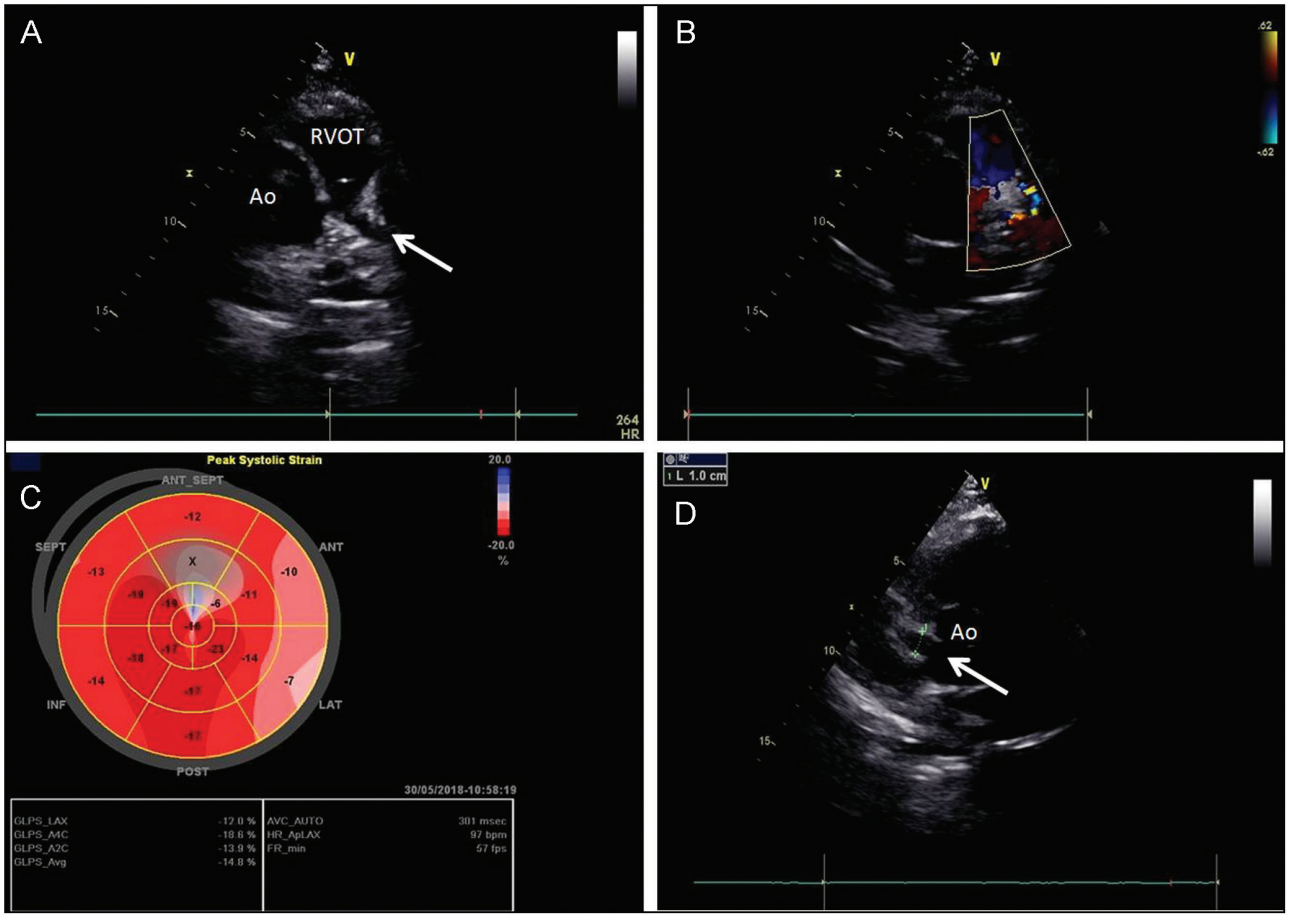

Figure 3: Two-dimensional echocardiography images showing: (A) left coronary artery (LCA) arising from pulmonary artery (PA) (arrow, upper left). (B) Anomalous diastolic color flow of collaterals (upper right). (C) Reduced global longitudinal strain at apex and apical septum (lower left). (D) A dilated right coronary artery (RCA) is seen in aortic short-axis view (arrow, lower right). [Ao = Aorta; RVOT = Right ventricular outflow tract].

(EF = 55%) with no regional wall motion abnormalities. After confirming diagnosis, the patient was referred for corrective surgery. The surgical plan decided for the case was ligation of anomalous artery coupled with bypass grafting. However, he refused immediate surgery and opted for medical management instead. At first follow-up, he was asymptomatic on guideline-directed medical therapy.

DISCUSSION

ALCAPA is well tolerated *in utero* and in infancy because of antegrade flow of oxygenated blood from PA to LCA. After birth, the PA saturation gradually declines and with subsequent decline in PA pressure, there is retrograde flow from RCA through collaterals into PA. This leads to coronary steal and hypoxic damage of LV. The survival into adulthood (10%–15%) depends on the extent collaterals from RCA to LCA.[1] Adequate collateral supply leads to a less hypoxic damage of LV in infancy and an improved survival in adulthood.

Patients may be asymptomatic or present with mitral insufficiency, ischemic cardiomyopathy, or with malignant ventricular dysrhythmias in whom sudden cardiac death (SCD) occurs in 80%–90% of cases.[2] Approximately two-thirds have symptoms of angina, dyspnea, palpitations, or fatigue and approximately 62% have life-threatening presentations.[3,4]

Increasing availability of cardiovascular imaging has resulted in increase in rate of diagnosis of ALCAPA. The incidence remains low at 0.24%–0.65% of all congenital heart diseases.[3] Early autopsy studies indicated that the average age for SCD in untreated ALCAPA was around 35 years. Surgical therapy is definitive and remains the treatment of choice in patients with symptoms or left ventricular dysfunction. Common surgical options include bypass grafting with concomitant ligation of anomalous artery, reimplantation of anomalous artery into aortic sinus, and finally, a channel connection between aortopulmonary window and coronary artery (Takeuchi procedure). Long-term surgical outcomes are excellent with a 98% survival at 20 years.[5]

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Conflicts of interest

There are no conflicts of interest.

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