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CASE REPORT

Congenital atresia of left main coronary artery

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ABSTRACT

Congenital atresia of the left main coronary artery is a very rare form of coronary anomalies with poor clinical outcomes if left untreated. Patients require surgical correction by coronary bypass grafting after diagnosis. Here we report a case of congenital left main atresia in a 36 years old woman who had previous heart surgery with this anomaly having gone undetected.

1. Introduction

Left main coronary artery atresia (LMCAA) is a rare coronary anomaly in which the left coronary ostium is absent or has a blind aortic end. The blood flow to the left heart chambers is provided by collaterals from the right coronary artery. The prognosis is poor due to occurrence of chronic myocardial ischemia and reultant myocardial dysfunction.[1,2] Management in adults consists of surgical revascularization with arterial and venous bypass grafts placed on left anterior descending (LAD) and left circumflex (LCX) arteries. In the present case, we report a patient with congenital atresia of the left main coronary artery accompanied by subvalvular aortic stenosis not detected at the time her childhood cardiac surgery.[3,4]

2. Case report

A 36 year old woman with history of heart surgery presented to the adult congenital heart disease clinic for follow up. She complained of atypical chest pain episodes that had aggravated through the past year. Previous surgery had been performed at childhood for repair of subvalvular aortic stenosis and there was no record of coronary anomalies detected then. On physical examination there was a III/VI systolic murmur in the left sternal border. Echocardiographic evaluation revealed recurrence of severe subvalvular stenosis and severe aortic regurgitation. As echocardiographic views for precise measurement of sub-aortic gradients was not optimal, the patient underwent cardiac catheterization for the purpose of hemodynamic study. During aortic root injection, an ectatic and tortuous right coronary artery filling left system retrogradely was visualized and attempts to engage the left main coronary failed (Figs. 1 and 2). Coronary computed tomography-angiography (CTA) with 3-dimensional reconstruction was performed and showed that the left main ostium is atretic and separate from the aortic root confirming the diagnosis of LMCAA (Figs. 3 and 4).

The patient was scheduled for cardiac surgery. LM atresia was confirmed during the surgery and the patient received a saphenous vein graft on LCX and internal mammary artery graft on LAD. Subvalvular stenosis was also repaired and aortic valve was replaced. The patient had an uneventful recovery.

3. Discussion

Clinical diagnosis of LMCAA might be neglected due to the wide range of nonspecific presenting symptoms including chest pain in a young patient, dyspnea, syncope or aborted sudden cardiac death.[2,5–7] Patients could remain asymptomatic for a long time due to a well development collateral system. LMCAA is often an isolated anomaly but association with other congenital heart defects such as pulmonary stenosis, bicuspid aortic valve, supravalvular aortic stenosis and ventricular septal defects has been described.[2,5] Selective coronary angiography is helpful but multislice CTA would provide more precise details in a less invasive manner and is therefore recommended for evaluation

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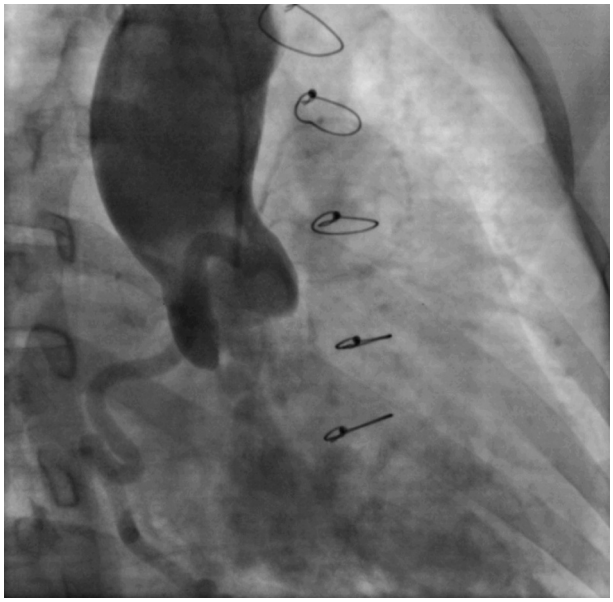

Fig. 1. No filling of left system during aortic root injection.

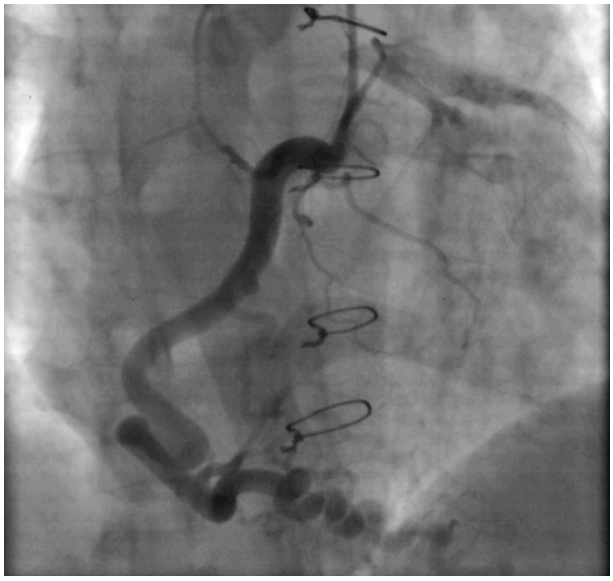

Fig. 2. Retrograde filling of left system by RCA injection.

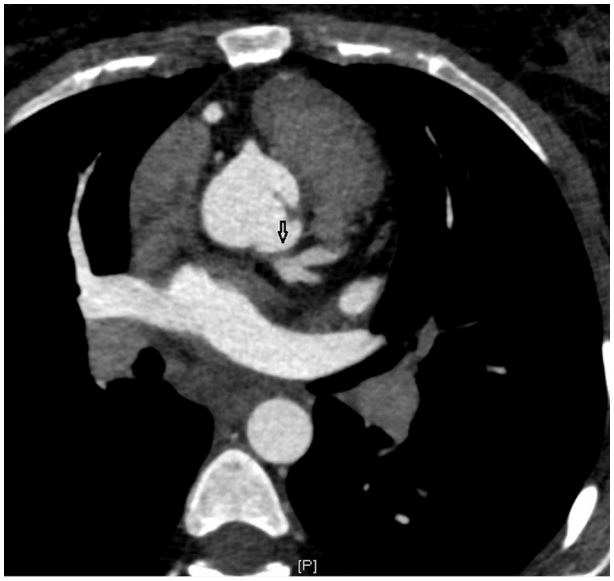

Fig. 3. Atretic and blind end of left main coronary in CT-angiography (arrow).

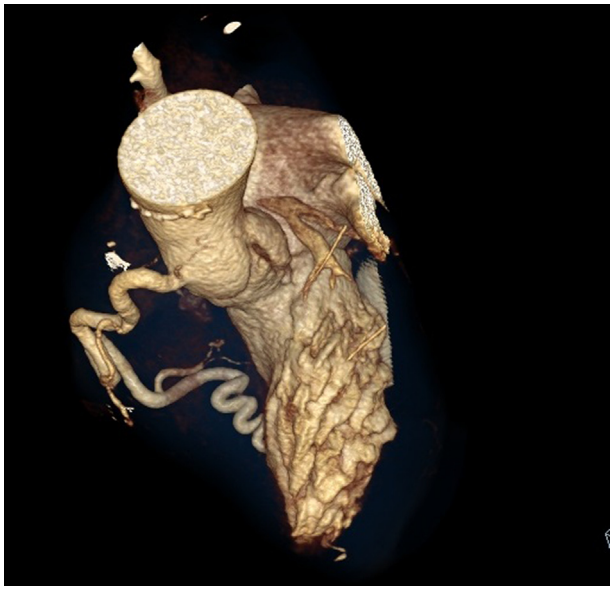

Fig. 4. 3D reconstruction depicting tortuous right coronary artery and atretic ostium of left main coronary artery.

of congenital coronary abnormalities.[8] In contrast to adults, in patients who undergo surgery as a child selective delineation of coronary anatomy is less often performed that might lead to neglected coronary anomalies. This case emphasizes that in patients with congenital heart disease subtle evaluation of coronary origin and course is absolutely necessary as coronary anomalies are relatively frequent association in this patient population. As pointed earlier patients with LMCAA should undergo surgical correction with restoration of the antegrade flow to the left coronary system by bypass grafting. However there are no guidelines on management due to the scarcity of the condition.[9]

Conflict of interest

There are no conflicts of interest in this article.

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