Congenital Atresia of the Left Main coronary artery: new presentation in a septuagenarian

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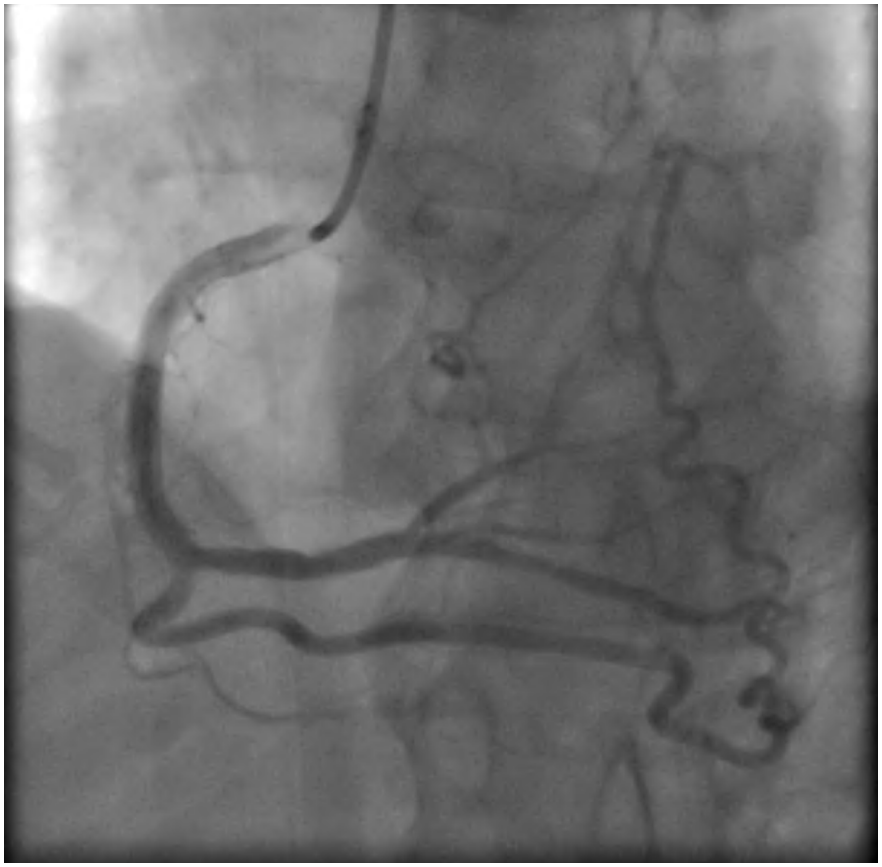
Abstract

Congenital anomalous anatomy and course of coronary arteries are common however atresia of LMS is extremely a rare condition. Fewer than 50 cases of congenital atresia of left main stem coronary artery have been reported in the literature. The ostium or trunk of the left coronary artery is atretic, and the left coronary artery and its branches are supplied by the right coronary artery via coronary collaterals in a retrograde direction. The pattern of presentation varies depending on the age and anatomy of the patient. According to the literature these cases present at a very early age with signs of heart failure whereas adults may present with angina or shortness of breath. Its clinical signs and symptoms often resemble anomalous origin of the left coronary artery from the pulmonary artery and endomyocardial fibrosis. In literature there are only five adult cases reported with congenital atresia of LMS and an interesting fact is that one out of those five cases was diagnosed on autopsy to have atresia of LMS. That patient was a female at age of 76 years who died of reasons other than heart symptoms. Therefore it may be inferred that the range of symptoms vary from being completely asymptomatic to overt heart failure.



INTRODUCTION

Variations in coronary anatomy are extremely common, including alterations in left/right dominance, dual anatomy and recessive vessels. For the left main stem (LMS), the commonest anomaly is either a short vessel or absent vessel (i.e. the LAD and circumflex emerge as a bifurcation at the left coronary ostia).

However, atresia of LMS is an extremely a rare condition: here, the left system is supplied completely by the right coronary artery (RCA) via retrograde collateral circulation. According to the literature these cases present at a very early age with signs of heart failure, and rarely in adults.

CASE REPORT

We report a case of congenital atresia of LMS in a 75-year-old male, presenting with acute severe chest pain on exertion, without prior cardiac symptoms whatsoever. His risk factors included hypertension, hypercholesterolemia and smoking.

The electrocardiogram demonstrated ST elevation in the anterior leads, and cardiac enzymes were positive, leading to emergent coronary angiogram. The latter demonstrated what was initially thought to be severe LMS stenosis with retrograde filling through right coronary artery (Figure 1). There were failed attempts to engage the LMS during aortic root injection. An ectatic right coronary artery was visualised which filled the the left side retrogradely.

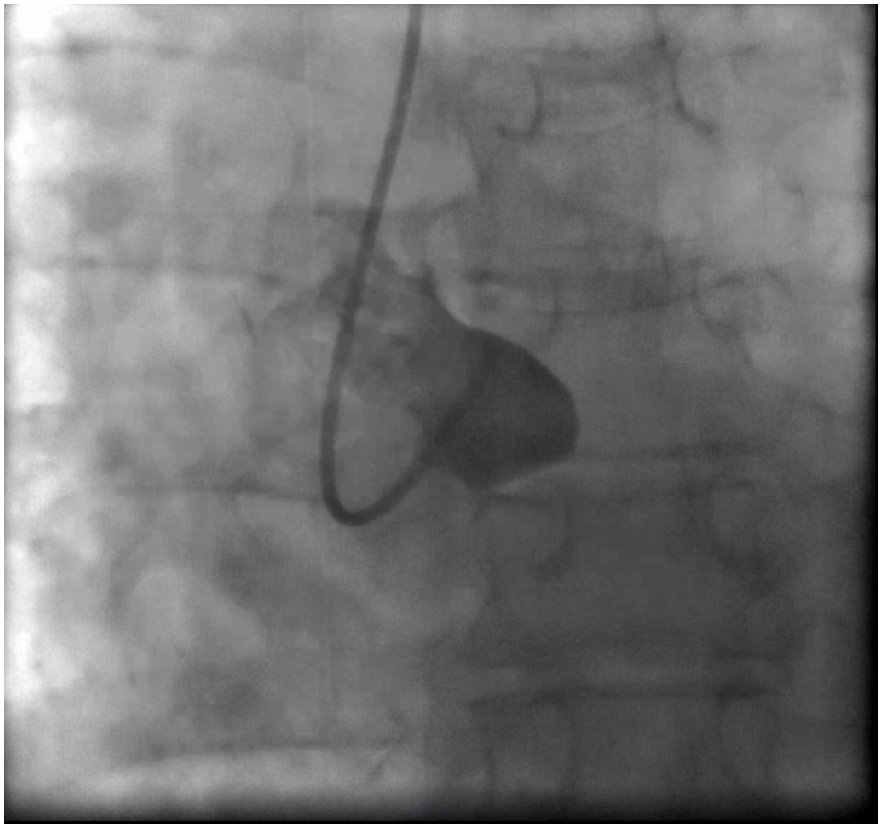


Fig 1. No filling of the LMS during aortic root injection.

There was suspicion of a separate narrowing in the collateralizing vessel and disease in the posterior descending artery. The computerized tomography (CT) coronary angiogram refuted the initial diagnosis, and instead confirmed the absence the LMS stump. A gated angiogram and a 3D complete reconstruction confirmed atresia of the LMS. The most interesting finding was that the LAD was supported by a conus branch of the RCA antegradely, while a collateral from the left ventricular branch of the RCA was feeding the LAD from apex of the heart. There was a tight narrowing in the antegrade feeding vessel and a calcified lesion at the junction of collaterals with the LAD.

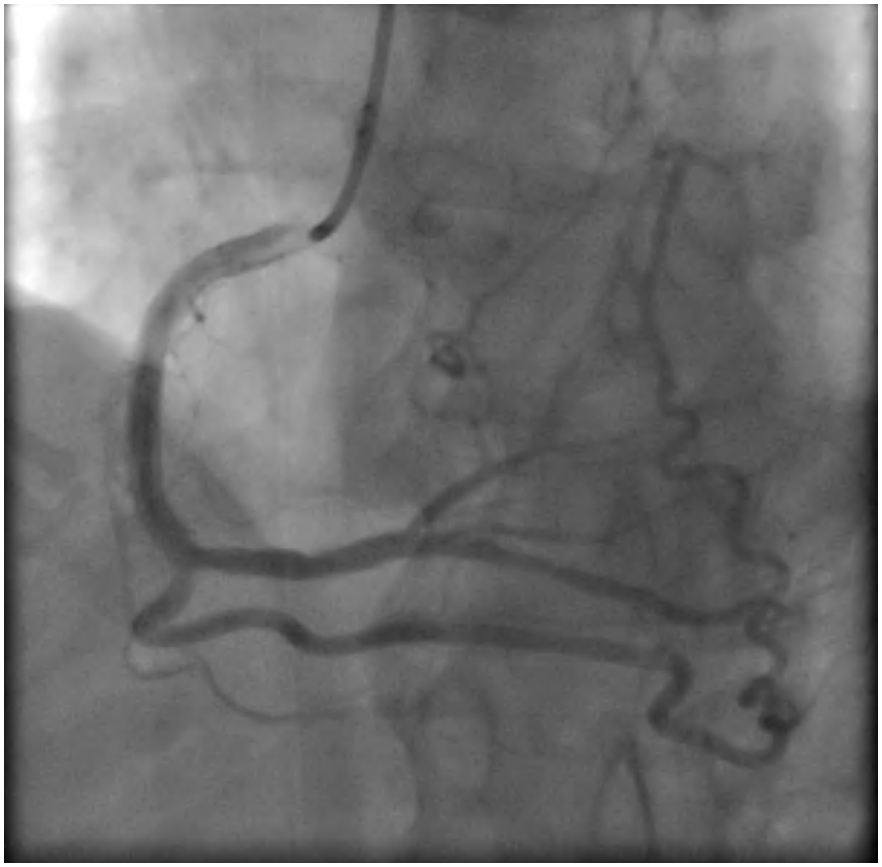


Fig 2. Retrograde filling of LAD by RCA injection, through collaterals.

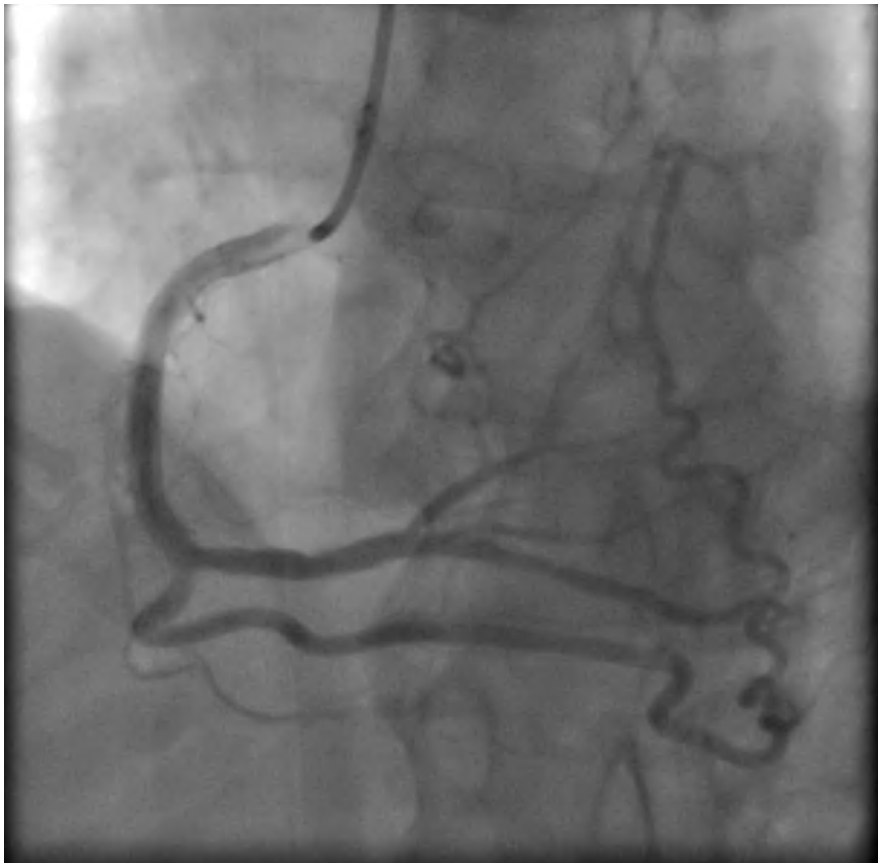


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Fig 3. Blind end of Left main coronary artery on CT angiography

The patient remained completely asymptomatic in the hospital which raised concerns whether chest pain was due to LMS atresia or one of the potentially culprit lesions in this complex coronary system. To confirm LAD territory ischemia, dobutamine stress echocardiography was conducted, identifying a remarkable decline in contractility of the anterior wall on stress and dilatation in left ventricular dimensions.

The initial clinical inclination was to offer the patient medical management, being asymptomatic, and then intervene at a later stage were symptoms to arise. However, following further investigation with magnetic resonance imaging (MRI), this identified a considerable loss of contractility of the anterior wall, septum and apical segments of the heart with full viability in all segments.

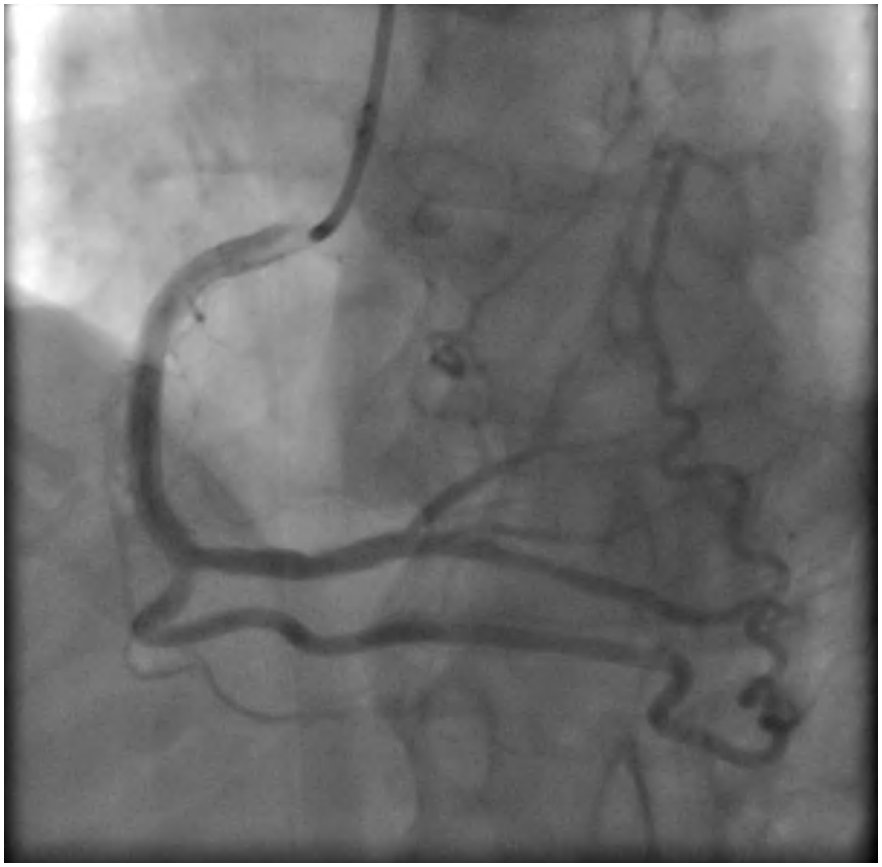

Fig 4. 3D reconstruction depicting atretic ostium left coronary artery and tortous left anterior descending

Following further multi-disciplinary discussion, the patient was offered off-pump coronary artery bypass grafting.

The patient received a left internal mammary-to-LAD graft and a saphenous vein graft to the posterior descending artery. The postoperative recovery of the patient was uneventful and he was discharged home with complete resolution of his angina symptoms.

DISCUSSION

Only five adult cases of congenital LMS atresia have been reported previously in the literature, and interestingly, one case was only diagnosed on autopsy[1 2]. This handful of adult cases have confirmed that strong collateralization of the arterial tree let them skip a turn up at an early age.[3, 4]The range of symptoms vary from being completely asymptomatic to overt heart failure.[5] Important differential diagnoses also include origin of the LMS from the pulmonary artery and endomyocardial fibrosis.[3,6]

Our case report depicts the chain of investigations which are required for diagnosis of coronary atresia and determination of the culprit lesions jeopardizing heart muscle function. Kate et.al emphasized the limited role of invasive coronary angiography in this important diagnosis. Cardiac MR angiography and multi-slice computed tomography (MSCT) gives a 3d visualization and are potentially more powerful modalities to identify this diagnosis. [7]

In the present case report, directed investigations played an essential role in committing to the correct treatment. Whereas gated CT angiography provided the actual coronary anatomy, stress echocardiography

determined the true origin of ischemia and guided subsequent therapy. This suggests that, in the case of demonstrable myocardial compromise, prompt revascularization is the optimal therapy for patients with LMS atresia.[1]

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