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Case Report

Beyond Benign or Malignant: A Complex Case of Anomalous Left Main Coronary Artery with Transseptal Course

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Abstract

Anomalous coronary arteries (ACA) represent a heterogeneous group of anatomical variants with a wide range of clinical implications. While many are benign, specific courses may predispose to ischemia, myocardial infarction, heart failure, or sudden cardiac death. We present a rare case of a 62-year-old man with an anomalous left main (LM) coronary artery originating from a shared right coronary ostium, demonstrating a mixed interarterial and transseptal course. Multimodality imaging with cardiac CT angiography (CTA), cardiac magnetic resonance imaging (CMR), and invasive coronary angiography with intravascular ultrasound (IVUS) and physiologic assessment revealed dynamic systolic compression of a prominent first septal perforator, associated with mid-myocardial fibrofatty infiltration and fibrosis. Resting hemodynamics were normal; however, provocative maneuvers elicited significant ischemia as evidenced by abnormal instantaneous wave-free ratio (iFR) changes. Despite the anatomical complexity and hemodynamic findings, the patient opted for conservative management following a detailed discussion of surgical revascularization options. This case highlights the nuanced spectrum of clinical significance in anomalous LM coronary arteries, challenging the traditional benign versus malignant classification. Comprehensive anatomical and physiological evaluation is critical, especially in mixed-course anomalies. Our findings underscore the role of multimodal imaging modalities and functional testing in guiding individualized patient management in rare coronary anomalies.

Keywords

Anomalous Coronary Artery, Transeptal Course, Coronary Computed Tomographic Angiography, Exertional Chest Pain, Instantaneous Wave-free Ratio

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1. Introduction

Anomalous coronary arteries (ACA) encompass a wide range of morphologic variants with differential clinical implications. Their prevalence has been reported as high as 5.8% in cardiac CT series [1]. The majority of these are thought to have no major clinical repercussions and do not require intervention. Nonetheless, others can intermittently hamper myocardial perfusion, leading to angina, myocardial infarction (MI), heart failure (HF), ventricular aneurysm, or sudden cardiac death (SCD) [2].

2. Case Presentation

A 62-year-old man consulted his primary care provider due to episodes of transient vision loss, suspected to be symptoms of a transient ischemic attack (TIA), which resolved within minutes. His pertinent medical history included hypertensive heart disease, dyslipidemia, obesity, and a history of tobacco use. Given his cardiovascular risk factors, several cardiovascular evaluations were ordered.

A two-week Holter monitor was recommended to assess for potential cardiac arrhythmias, such as paroxysmal atrial fibrillation or flutter. The results indicated some isolated supraventricular ectopy, with patient-initiated events correlating with periods of normal sinus rhythm. There was no significant bradycardia or tachycardia, and the daily circadian rhythm variability appeared to be appropriate. Furthermore, a myocardial perfusion study revealed abnormalities in the distal apical and inferolateral regions, corresponding to the distal left anterior descending and left circumflex coronary arteries.

A cardiac computed tomographic angiography was also ordered for the exclusion of an intracardiac embolic source and evaluation of the coronaries. In Figure 1, a 3D-CTA reconstruction revealed an anomalous left main (LM) coronary artery sharing the right coronary artery (RCA) ostium. The LM coursed caudal to the pulmonary valve and appeared as a stretched, long vessel that coursed anterolateral and contained a transseptal segment, after which a prominent proximal septal perforator coursing through the interventricular septum was noted (Figure 2A, 2C, 2D). Significantly, fat-equivalent hypoattenuation was focused on the interventricular septum, most consistent with intramyocardial fat (Figure 2B). The LM ostium was described as usual in caliber and patent. The proximal take-off was of normal angulation, with no slit-like segment. The ejection fraction (EF) was normal at 64%, with a mildly globally dilated left ventricle (LV).

A cardiac MRI (CMR) was performed to confirm the suspected focus of interventricular septal intramyocardial fat and to exclude fibrosis (Figure 3A). The area of septal hypoattenuation shown by computed tomography was confirmed as fatty infiltration. Additionally, an associated rim of fibrosis was evidenced by a thin peripheral layer of late gadolinium enhancement (LGE). Additionally, subendocardial LGE was noted over the mid-LV and proximal antero-apical walls (Figure 3B). No epicardial or pericardial abnormalities were seen. The study also showed a dilated LV (226 mL), normal EF (63%), and preserved cardiac output and index, while redemonstrating the anomalous LM. Invasive coronary angiography with Intravascular ultrasound revealed minimal epicardial atherosclerosis with a short region, less than 5 mm in length, where the LM had an oval shape corresponding with the transseptal course. The LM proximal to the transseptal segment revealed no overt compressed segments (Figure 4). The transseptal portion displayed some degree of systolic compression. In turn, invasive iFR of the left anterior descending (LAD) at rest was normal at 0.96. The iFR changed to a range of 0.8-0.86 with provocation maneuvers using IV normal saline, dobutamine infusion, and atropine (Figure 5). Pullback iFR displayed a step-up in the proximal LM segment. With dobutamine and atropine, the patient experienced a run of supraventricular tachycardia, which resolved spontaneously.

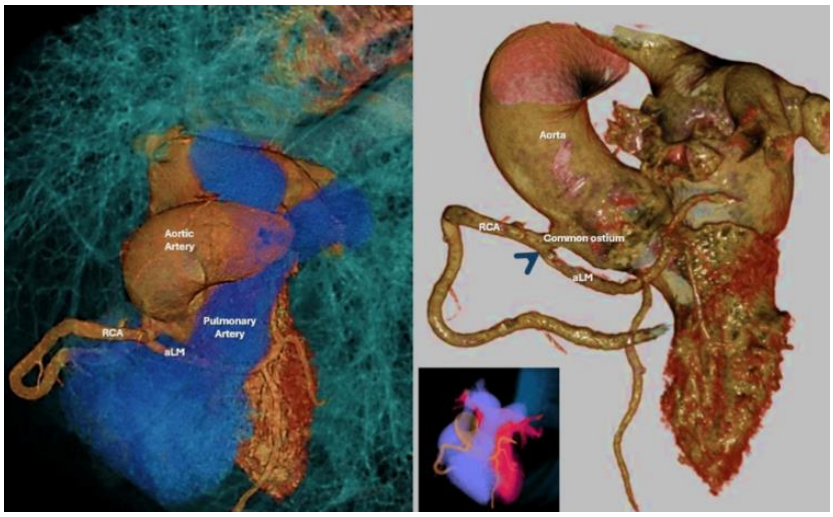

Figure 1. *Coronary CTA 3D reconstruction. The blue arrow shows the left main coronary artery has a common ostium with right coronary artery with a transeptal course running in between the aortic and pulmonary arteries.*

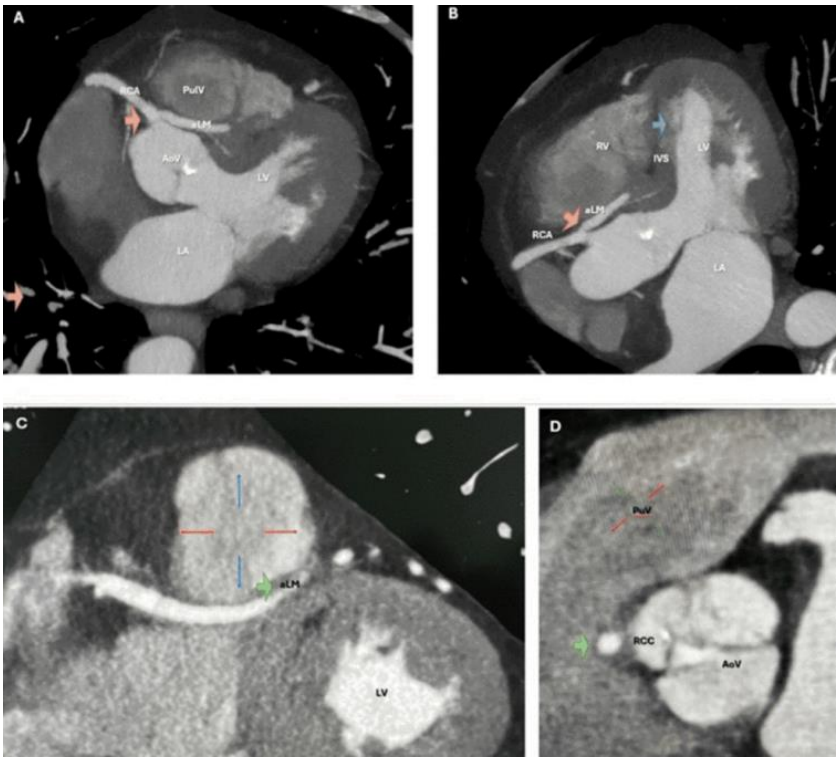

Figure 2. *Cardiac CTA performed to exclude intracardiac embolic. On figure 1A, 1C, and 1D the LM and RCA share a common ostium (orange arrow) with LM running through a caudal course towards the pulmonary valve. On figure 1B, demonstrates a focus of fat-equivalent hypoattenuation in the interventricular septum, most consistent with intramyocardial fat (blue arrow).*

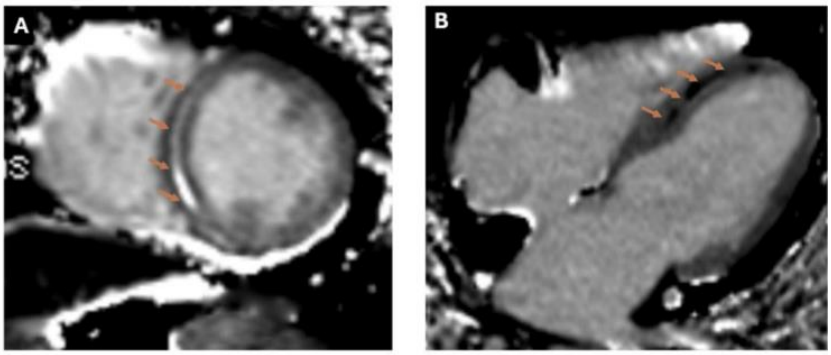

Figure 3. *Cardiac magnetic resonance. 3A LGE-SAX (orange arrows) demonstrating fat saturation and 2B Four-chamber-LGE (orange arrows) disclosing areas of fibrofatty infiltration of the interventricular septum.*

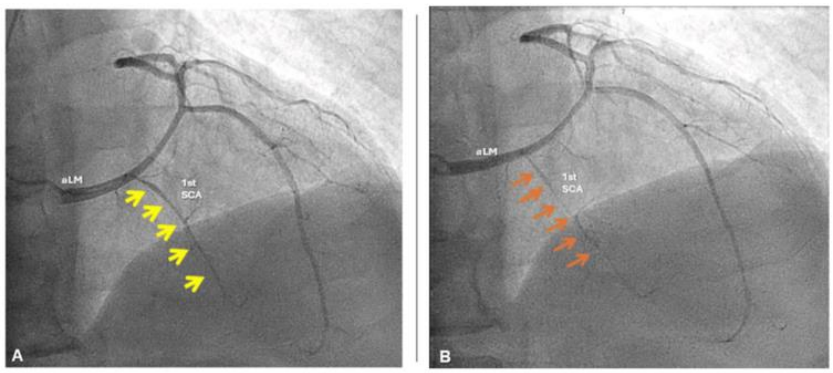

Figure 4. *A. Coronary angiography with aLM giving LAD with patent S1 artery during diastole (yellow arrows) and systolic occlusion (orange arrows).*

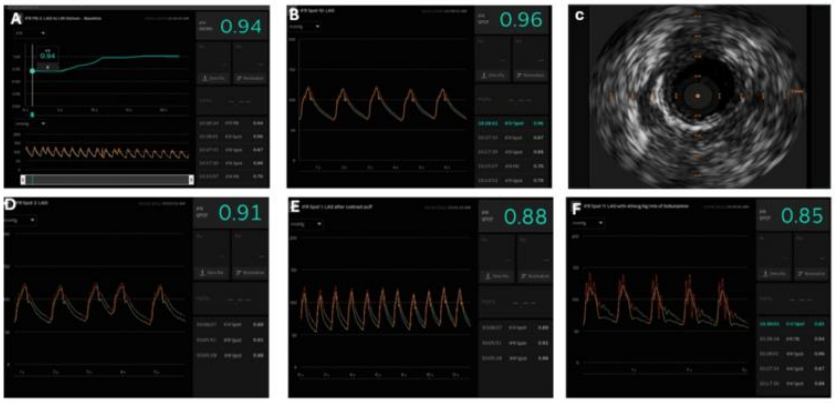

Figure 5. *Flow reserve study with IFR/FFR run.*

3. Discussion

We present a challenging and rare case of an anomalous LM from a common single coronary ostium from the right sinus of Valsalva. The proximal segment of the vessel courses between the aorta and right ventricular infundibulum, and subsequently, a distal transseptal segment. The initial cardiac CTA did not reveal an interarterial course, acute angle take-off, slit-like ostium, valve-like mechanism due to an ostial ridge, or coronary proximity to the aorta. All of these are known to predispose to adverse cardiovascular events from associated coronary kinking during exercise [3], or scissor-like effect on the proximal vessel from associated aortic and pulmonary vasodilation during exercise [2, 4].

The clinical significance and potential complications depend upon the anomalous artery's course. There are four main anomalous LM anatomic categories: precardiac, retro aortic, interarterial, and septal [5]. The precardiac and retro aortic variants are considered benign [6]. Conversely, the interarterial variant has been considered malignant, particularly if accompanied by features suggestive of intramurality [4]. However, there is conflicting evidence regarding the transseptal course [7], with some describing adverse events [8]. This opinion is not universally shared in the literature, with some suggesting that anatomical course alone may not suffice to determine clinical relevance or guide treatment decisions. In one of the most extensive retrospective studies evaluating anomalous LM coronary arteries, Torres et al. hypothesize that there is a clinical spectrum of anomalous courses. These may account for some of the outcomes' differences, and a dualistic interarterial vs. transseptal classification may be too simplistic [6]. Thus, comprehensive non-invasive and invasive re-stratification leading to patient-centered decisions is preferred.

Our CTA revealed structural LV changes with fibrofatty replacement of the myocardium in the septum in an atypical pattern. Pathologic and physiologic myocardial fatty infiltrate discrimination has been described in cardiac CT literature. It is generally accepted that intramyocardial fatty deposition may be a physiological response to aging, a healed MI, among other conditions. Discrimination among these entities depends on the distribution, location, and whether corresponding to the territory of a culprit artery [9, 10]. Myocardial fat replacement from a healed MI is thin, curvilinear, subendocardial, and corresponds to a coronary perfusion territory [11]. Mid-myocardial or subepicardial ischemic fatty replacement is rarely observed, given the natural ischemic wave progression starting in the subendocardial [11, 12]. In this case, we demonstrate the relatively infrequent ischemic mid-myocardial fat infiltrate. We hypothesize this finding stems from chronic ischemia in the territory of the sizable first septal perforator (which was shown to compress dynamically during the cardiac cycle) and, to a lesser extent, the LAD.

Cardiac MR is very sensitive in detecting prior MI due to the characteristic late gadolinium enhancement (LGE). Double and triple inversion CMR phases are useful in tissue characterization to note fibrofatty replacement of normal myocardium from ischemic insults [13]. A possible discrepancy in fat detection by CMR vs. CTA has been described. Goldfarb et al. noted some ischemic MI-related fatty deposition in the mid-myocardial and epicardial walls. However, this finding was contrasted with subendocardial findings on CTA [14]. This discrepancy may result from the CMR's ability to accurately delineate all myocardial layers and classify histologically [13]. Our case represents an accurate correlation between these two dissimilar imaging modalities. Another salient point is that such histopathological myocardial compositional changes correspond to chronic rather than acute insults, thus supporting our theory of indolent subclinical events, in this case, from the systolic milking of the prominent septal perforator.

In this case, the CMR was also helpful in corroborating the normal EF, describing the lack of wall motion abnormalities, and describing the pattern of LGE. Mid-myocardial septal LGE has been associated with a nonischemic cardiomyopathy pattern and a worse prognosis [15, 16]. Thus, this case likely represents an atypical LGE pattern stemming from a series of chronic ischemic events from the systolic compression of the prominent first septal perforator and, to a lesser extent, that of

the transseptal portion of the vessel. Current literature suggests that the significance of myocardial bridging may be related to length, depth, and degree of systolic compression. However, other series have failed to prove these relevant points even for extreme cases of myocardial tunneling in asymptomatic patients with no significant imaging findings [6].

Invasive coronary angiography with IVUS confirmed the CTA interpretation. iFR has been used to evaluate the hemodynamic significance of anomalous ectopic coronaries [17]. In our case, resting hemodynamics were normal; however, such information is limited to resting conditions. Giuseppe et al. published their anatomic vs. physiological experience in tunneled coronary segments [18]. The authors argue that physiological analysis always trumps anatomical analysis in evaluating hemodynamic significance in such cases. They employed ionotropic infusions and other provocative maneuvers to simulate a physiological response for optimization. In this case, we share many of their findings, including abnormal iFR readings after volume and inotrope infusion, along with atropine to achieve a targeted HR. Another shared result is the abnormal step-up through the tunneled LM segment on pressure wire pullback. Importantly, their results and ours may further validate the extension of iFR beyond resting conditions-i.e., with significant tachycardia, coronary hyperemia, or any physiological condition that increases myocardial oxygen demands. One limitation of their small prospective study is the lack of invasive and non-invasive correlation. In this study, we provide that link, albeit limited to this patient's case. Nonetheless, abnormalities noted in cardiac CTA and MR appear to correlate closely with invasive hemodynamic data. However, larger prospective studies are warranted, including complex cases such as the one presented.

The complexity of our case-i.e., a single coronary ostium without separate coronary ostial buds, along with the mixed anatomic course of the anomalous LM, the first septal perforator proximal to the LAD ostium with demonstrable dynamic compression, among others- posed multiple revascularization challenges. Several surgical therapeutic options are available, including unroofing, CABG, and coronary reimplantation. Unroofing only applies to patients with an intramural component (absent in this case). Patients with an interarterial course undergo CABG or unroofing [19]. A different approach involves transferring and reimplanting the coronary ostium into the appropriate sinus [20]. These technical approaches and the potential complications were extensively discussed with the patient, who ultimately declined any intervention and elected to continue conservative medical management.

4. Conclusions

In this case, we present a rare, complex clinical scenario of a patient with a mixed anomalous LM with a transseptal course. Current literature shows growing evidence regarding the significance of certain anomalous coronary anomalies and the optimal diagnostic risk stratification studies. Our case reveals consistent findings among cardiac CTA, MR, invasive catheter angiography, and hemodynamic studies. We also support the previously published concept that anomalous LM is a complex entity that should not be dichotomized into benign vs. malignant but evaluated and understood within the spectrum of clinical presentations it represents. In transseptal course anomalous coronaries, there should always be concern for muscular compression of the intramyocardial ectopic segment. As such, therapeutic interventions must be guided by multipronged, comprehensive diagnostic evaluations; nonetheless, this remains beyond the scope of our study and must be addressed by larger investigations. The latter may be challenging, given the infrequent presentation of this entity.

Abbreviations

ACA	Anomalous Coronary Arteries (ACA)
MI	Myocardial Infarction
HF	Heart Failure
SCD	Sudden Cardiac Death
TIA	Transient Ischemic Attack
LMCA	Left Main Coronary Artery
LAD	Left Anterior Descending
RCA	Right Coronary Artery
EF	Ejection Fraction
LV	Left Ventricle
CMR	Cardiac Magnetic Resonance
iFR	Instantaneous Wave-free Ratio

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Conflicts of Interest

The authors declare no conflicts of interest.

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