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Anomalous Origin of Left Main Coronary Artery from the Right Sinus of Valsalva: A Case Series-Based Review

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Established Facts

1. Anomalies of origin of Coronary arteries is an incidental finding occurring in 0.24 to 1.3% of the patients underdoing angiography.

2. The great majority of such anomalies are benign (anatomic variants of normal, with normal function).

3. Most common coronary anomaly is Left circumflex artery (LCX) originating from Right sinus of Valsalva (RSV), followed by a single coronary artery from left sinus of Valsalva.

4. Anomalous origin of Left Main Coronary Artery (LMCA) from Right sinus of Valsalva with an intramural course carries highest risk for sudden cardiac death and usually requires surgical intervention.

Novel Insights

1. Management of patients with coronary artery anomalies involving left main should be irrespective of ischemic symptoms or documented myocardial ischemia.

2. In coronary angiograms performed on symptomatic patients with coronary artery anomalies, the degree of stenosis does not typically correlate with the clinical presentation.

3. Due to lack of data and inability to predict sudden cardiac death, we agree with the latest AHA/ACC guidelines which recommend surgical intervention for all patients with LMCA from RSV, regardless of ischemia or ischemic symptoms.

4. If a malignant or intramural course is discovered incidentally, surgery should be planned to prevent sudden cardiac death in such patients.

Keywords: Left circumflex artery • Valsalva • Ischemic stroke • Left Main Coronary Artery

Introduction

Any coronary artery morphology which is not observed in >1% of a general population is morphologically defined as anomalous.Congenital anomalies of origin of coronary arteries are rare and usually an incidental finding.The Incidence variesfrom 0.24% to 1.3% of the cases [1,2]. The most common coronary artery anomaly is left circumflex artery (LCX) arising from the right sinus of Valsalva(RSV) or right coronary artery (RCA). Other anomalies include a single coronary artery from the left sinus of Valsalva, both coronary arteries from RSV and left anterior descending coronary artery (LAD) from RSV [3]. Majority of these anomalies are benign; however, few are potentially fatal like anomalous origin of left main from RSV or from pulmonary artery. We report a case series of three patients, who presented with acute coronary syndrome and were found to have anomalousleft main coronary artery (LMCA) arising from the RSV on diagnostic angiogram. We alsodescribe the review of literature of similar cases with an emphasis on managementand prognosis of this rare anomaly.

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Case Presentation

Case 1

A 78-year-old African American malepresented to the emergency department (ED) complaining of nausea, vomiting, and substernal chest pain for one day. He was a chronic smoker and had pertinent medical history of hypertension, hyperlipidemia, ischemic stroke and peripheral vascular disease. Initial electrocardiogram (ECG) showed ST elevation with Q waves in inferior leads suggesting acute inferior infarct. The patient was hemodynamically stable and had resolution of chest pain after initial medical therapy. An urgent coronary angiogram as part of early invasive strategy revealed anomalous origin of LMCA from the right coronary cusp (see Figure1). The RCA had 100% occlusion in mid segment with left-to-right collaterals. Decision was made to proceed with a coronary Computerized Tomography Angiogram (CTA) to rule out malignant course. Coronary CTA confirmed an anomalous common trunk origin of right and left coronary artery from the RSV with an inter-arterialand intramural course of LMCA across the wall of the ascending aorta and behind the pulmonary artery (see Figure 2). Cardiothoracic surgery (CTS) consultation was obtained at this point and Coronary Artery Bypass Grafting (CABG) was recommendeddue to malignant course of left main and high-grade stenosis in RCA. The patient subsequently underwentquintuple bypass withright internal mammary artery (RIMA) to LAD, Left IMA to ramus, Saphenous Vein Graft (SVG) to diagonal, obtuse marginal (OM) and RCA. Patient had an uneventfulpost-op hospital course and was discharged without complications. It has been about 3 years since his CABG and his last cardiac catheterization was performed about 2 years ago showing patent grafts with minimalnon-obstructivestenosis (Figures 1 & 2).



Figure 1. Selective coronary angiography of the right coronary artery revealing common origin of the RCA and LMCA along with 100% stenosis in mid RCA with TIMI-0 flow distally (arrow).



Figure 2. Coronary CTA image showing the RCA and LMCA originating from the right coronary cusp.

Case 2

A 47-year-old Hispanic femalepresented to ED with two days history of intermittent substernal chest pressure increasing in frequency and intensity. She had a significant family history of premature coronary artery disease (CAD) including her biological brother requiring CABG at 34 years of age. She also had a history of hypertension and was chronic smoker. Her initial ECG showed T-wave abnormality in the inferolateral leads. Subsequently, cardiac catheterization was performed for unstable angina whichrevealedanomalous origin of LMCA from the RSV with common ostia of the RCA and LMCA. She did not have any obstructive disease in the coronary arteriesperse (Figure 3). Decision was made to obtain coronary CTA to further delineate the course of left main; however, the patient started having active chest pain with worsening of ECG changes post cardiac catheterization. CTS was consulted, and patient was taken emergently to the operating room. Intraoperatively, the LMCA was noted to be originating from the extreme right side of the RSV with an intramural course in its proximal segment, a likely cause of compression of LMCA ostium leading to myocardial ischemia and given presentation. The LMCA was successfully re-implanted on the anterior surface of the aorta. Patienthad an unremarkable post-operative hospital course and was discharged 4 days after. She was apparently readmitted one week later for a large pericardial effusionrequiring drainage and eventual pericardial window. On follow-up, she has continued to do well (Figure 3).

Case 3

A 67-year-oldCaucasian woman presented to ED with complaints of recurrentand changing pattern of sub-sternal chest pressureradiating to the back and upper jaw. She had history of chronicsmoking. The initial ECG showed old inferior infarct and new T wave abnormality suggestive of ischemia. Coronary angiography was performed due to ongoing symptoms whichrevealed anomalous origin of LMCA from the right coronary cusp. There



Figure 3. Selective coronary angiography of the right coronary artery revealing common ostia of the RCA and LMCA with TIMI-3 flow.

was 90% stenosis of the mid LCX and 50% stenosis in LAD. RCA had 100% occlusion in mid segment with faint left to right collaterals (Figure 4). CTS was consulted who recommended urgent CABG given high grade stenosis in RCA and LCX. Patientunderwent triple vesselbypass with Left IMA to LAD, Right IMA to OM, and SVG graft to RCA. Patient had an unremarkable post-operative hospital course and was discharged home in a stable condition. One year later, she presented with acute coronary syndrome/NSTEMI. This time, her coronary angiography showed patent SVG-RCA graft with atreticleft and right IMA grafts. The LCX was 90% occluded and underwent successful PCI with drug-eluting stent placement. Medical therapy was continued, and patient continues to be symptom free on subsequent follow-up visits (Figures $4 \otimes 5$).



Figure 4. Selective coronary angiography of the right coronary artery revealing common origin of the RCA and LMCA from the RSV along with 100% mid occlusion of the RCA with TIMI-0 flow distally (arrow).



Figure 5. Selective angiography of the left main coronary artery showing a 90% stenosis in the mid LCX (arrow) and a 50% stenosis in the mid LAD with TIMI-3 flow.

Discussion

The majority of congenital anomalies involving origin of coronary arteries are benign and asymptomatic in 80% of the cases [1,2]. They are mostly foundincidentally on routine coronary angiography. The most common coronary anomaly is LCX originating from RSV, followed by a single coronary artery from left sinus of Valsalva and both coronary arteries from RSV and LAD from RSV [3]. In the largest angiographic series comprising 1,26,595 coronary angiograms; 1686 coronary anomalies were detected with a prevalence of 1.3% [2]. There were 22 cases with an anomalous LMCA arising from RSV (0.01%).

Anomalous LMCA arising from RSV is rare but a critical diagnosis as it can lead to sudden cardiac death, especially during exercise. Based on the course of left main, this anomaly can be further classified into four subcategories [4]:

- 1. Inter-arterial: LMCA traversesbetween aorta and pulmonary artery behind the right ventricular outflow tract before coursing anteriorly
- Intramyocardial or Intramural: The initial segment of left main traverses through the subendocardial or intramyocardial region of ascending aorta before it comes out at mid-septum to bifurcate into LAD and LCX
- 3. Retro-aortic: LMCA originates right of RCA and courses posterior to aortic root
- 4. Anterior: The LMCA passes anteriorly over the right ventricular outflow tract

Of these, LMCA with an initial intramural course in wall of ascending aorta carries highest riskfor sudden cardiac death [5]. The exact mechanism of coronary ischemia in these patients is not well understood but severalhypotheseshas been proposed. The compression of smaller and slit-like ostium of anomalous left main arising with anacute angulation is the most common etiology of ischemia or infarction in the supplied myocardium. Other potential cause includescompression of left main between pulmonary artery and aorta during or post exercise from dilatation and stretching of aortic root and pulmonary truck. Other proposed theories are superimposed vasospasm from endothelial dysfunction, compression of intra-mural segment due to hypertension and an ostial'ridge' which obstructs the blood flow during increased flow states [6-8]. The follow-up studies with semi-quantitative analysis of individual case severity has shown that no single factor is associated with sudden cardiac death in these patients [9].

The clinical presentation in patients with anomalous LMCA from RSV varies. On review of literature, only 20% of the patients presentwith symptoms mainly including exertional angina, dyspnea or syncope [10-12]. Unfortunately, invast number of cases, sudden cardiac death is the initial and frequently terminal presentation [10]. Electrocardiographic findings in symptomatic patients are nonspecific and could vary from ST-wave abnormalities suggesting ischemia [13] to ventricular tachycardia or fibrillation [14]. Two of our patients had ST-T abnormality suggestive of ischemia while the third had an inferior infarct. Echocardiogram is typically obtained to assess left ventricular systolic function, but it does not provide any additional diagnostic information. Trans-esophageal echocardiography has been used in some cases to identify the origin of anomalous arteries although is usually not the best modality to delineate the origin and course of the coronary arteries [15]. Coronary angiography is the gold standard test to establish diagnosis [1,2]. Coronary CTA is most commonly used imaging modality to rule out intramural and inter-arterial course of the artery.

The management of patients with an anomalous left mainis irrespective of documentedischemia or ischemic symptoms. The 2018 AHA/ACC guidelines (Figure 6), released in April 2019, asserted that there is a lack of data to prove that any management strategy can prevent sudden cardiac death (SCD) in these patients (Figure 6).

Surgical intervention is class-I recommendation for patients with established

Anomalous Aortic Origin of the Coronary Artery



*Surgical intervention to involve unroofing or coronary revascularization for patients with concomitant fixed obstruction.

Figure 6. 2018 AHA/ACC Guidelines for the Management of Adults with Congenital Heart Disease.

diagnosis. Most recently, a working group provided the first consensus statement on practice guidelines for AAOCA [16].

There are different methods of surgical correction depending upon anatomical variabilities as well as expertise of the surgeon [17-22]. The latest and most commonly used method now-a-days is called "Unroofing" and involves an anterior aortotomy with incision of the common wall between the aorta and intramural segment of the anomalous coronary artery [21]. It recently gained traction as the procedure of choice for young patients with an intraarterial or intramural malignant course [22,23]. Benefits include elimination of intramural portion of the artery, enlargement of orifice and relocation of functional orifice to appropriate sinus. Potential pitfalls include damage to the intracoronary commissure which may cause aortic insufficiency, exposure of layers of aortic wall to systemic pressure at site of neo-ostium which may create a localised dissection sometimes requiring repair or aggressive over roofing beyond the intramural segment which can result in aggressive bleeding. Aortic insufficiency may be avoided by un-roofing only that portion of coronary which is not behind the commissure [21] but careful dissection and prevention is clearly the best way.

Pulmonary artery translocation is the second method of surgical correction and may be used if the anomalous artery is not intramural and compressed between the great vessels [16]. This procedure involves mobilisation of the main pulmonary root through meticulous dissection and moving its course thus preventing it from compressing the anomalous coronary. An alternative version is dividing the right pulmonary artery at its origin, transposing it anterior to the aorta and re anastomosis to the original site.

If the left and right coronary orifices are separated with little or no intramural segment, then re-implantation of ostia may be the best option as demonstrated by Erez et al. [24]. The preferred approach in this setting is coronary re-implantation into the correct aortic sinus followed by bovine pericardial patch closure of the defect in the aortic wall thereby recreating a normal coronary anatomy. Risks included potential damage to aortic wall commissure causing insufficiency due to its proximity requiring long term follow-up [24,25]. This is also a somewhat challenging technique as it involves mobilisation of full coronary ostia to avoid kinking as well as precise patching and re-implantation.

The most technically challenging procedure, an 'anatomical repair' involves creation of a neo-ostium for the anomalous vessel in the sinus from where it would normally have exited [16,26-28]. In this procedure, an incision is made in the coronary sinus with corresponding incision in the coronary artery away from the aorta. These incisions are then joined with augmentation of open areas with a patch. It may be used in all anatomical variants with or without an intramural course. Initial results are satisfactory but long term

outcomes need to be determined. Also the surgical expertise required for this kind of procedure makes it less likely the procedure of choice in the future [28]. Standard coronary artery bypass grafting (CABG) with a saphenous or internal mammary vein grafting is an option used in many centers for rerouting blood flow around the intramural segments, however, performing a bypass graft without ligating the native vessel can lead to occlusion of the patent native vessels from competitive flow [29-31]. Using internal mammary arteries limits revascularization options for these patients in the future and because of competitive flow from native coronaries, puts them at risk for atrophy and disuse occlusion as happened in case 3. CABG should be limited to only those patients in which the anomaly is accompanied by significant atherosclerotic narrowing andwhere alternative procedures produced unsatisfactory results [16,29-31].

To summarize, the unroofing is usually the procedure of choice in coronaries with intramural course whereas re-implantation or ostial reconstruction is used if there is little to no intramural course. Pulmonary artery translocation may be used to augment any primary procedure above. CABG should only be limited to special circumstances involving coronary atherosclerosis or failure of alternate therapy.In our first patient (Case 1), coronary angiography revealed LMCA from the right coronary cusp with a malignant course detected on coronary CTA. There was high grade stenosis in RCA and LAD as well.In our second patient (Case 2), angiography revealed non-obstructive stenosis in the coronary arteries with a malignant course of LMCA from the right cusp which required relocation of ostium. In our last patient (Case 3), the angiogram revealed non-stenosed LMCA originating from RSV with course of LCX posterior to the aorta. RCA, in this case, was completely occluded. She underwent triple coronary artery bypass.

It is important to mention that despite of stenosis being angiographically nonsignificant in the coronary arteries per se, two of our three patients presented with symptoms of chest pain and acute coronary syndrome. The possible mechanism of this presentation is compression of LMCA coursing in an inter-arterial fashion between aorta and pulmonary artery. Also, the slit like coronary ostia can get compressed within the wall of aorta in case of intramural origin [5-7].

Till date, there is no screening test indicated to elucidate coronary artery anomalies at birth. Due to its extremely low incidence in the general population and majority of affected patientsbeing asymptomatic, coronary angiogram should not be performed in healthy asymptomatic patients, unless otherwise medically indicated. However, if a malignant course is discovered incidentally, surgery should be planned to prevent sudden cardiac death in such patients.

To the best of our knowledge, no comprehensive study describing long term outcomes in anomalous LMCA from RSV exists.

Conclusion

Left main coronary artery from the right sinus of Valsalva is a rare, but often fatal anomaly. Due to a lack of data and inability to predict risk of sudden cardiac death, major society guidelines recommend surgical intervention for all patients regardless of ischemia or symptoms.

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