Case Report

Rare Anomaly of a Single Coronary Artery Arising from the Right Aortic Sinus of Valsalva: A Case Report

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Abstract

Background: Single Coronary Artery (SCA) is a rare, most commonly asymptomatic, congenital anomaly which is usually discovered incidentally during investigations. This entity consists of a single coronary artery supplying the entire myocardium. It may be classified based on the origin and the course of the artery. Depending on the patient's presentation and its severity, medical or invasive interventions may be warranted.

Case Report: A 65-year-old female patient known to have diabetes and hypertension presented to our emergency department with chest pain and discomfort. Blood tests and electrocardiography (ECG) showed no significant abnormalities, but on coronary angiography, we suspected an anatomical variation. Computed tomography angiography (CTA) confirmed our suspicion revealing a single coronary artery supplying the entire myocardium and arising from the right sinus of Valsalva. The patient received medical therapy and was discharged home.

Conclusion: We presented this case of Single Coronary Artery due to the rarity of this diagnosis. Presentation, treatment plan, and prognosis typically vary depending on the subtype of the anomaly. Early recognition is very important, especially in young adults.

Keywords: Single coronary artery, congenital heart disease, left circumflex artery, left anterior descending artery, right ostium, chest pain, case report

Background

SCAs are rare congenital anomalies usually identified incidentally during coronary angiography (CA), upon presentation with different kinds of symptoms, or postmortem after sudden cardiac arrests in young athletic adults. Such anomalies were first described over a century ago. They consist of a unique coronary artery supplying the entire myocardium from a single coronary ostium. Incidence varies among different populations, from 0.014 to 0.066% in

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populations undergoing angiography, and up to 0.3% in autopsy series [1-4].

We report the case of an elderly lady presenting with angina that turned out to be associated with a single right coronary artery supplying the entire myocardium.

**Case Presentation**

A 65-year-old female known to have hypertension and type 2 diabetes mellitus, presented for a two-week history of retrosternal chest pain associated with severe dyspnea on mild exertion. Vital signs were normal and physical examination was significant for bilateral lower limb pitting edema. The electrocardiography (ECG) showed evidence of a left bundle branch block (LBBB) with ST-depression in leads I, aVL, V5, and V6. There were no new changes in comparison with previous ECGs. Cardiac markers (Creatine phosphokinase CPK, creatine kinase myocardial band CPK-MB, and troponin I) were negative on serial measurements. Subsequently, acute coronary syndrome treatment was not given. Chest radiography showed no relevant findings. Coronary angiography showed significant stenosis at the level of the second segment of the right coronary artery (RCA) and the origin of the posterior interventricular artery, without opacification of the left coronary circulation, despite multiple attempts (Fig. 1).

Echocardiography revealed a left ventricular ejection fraction (LVEF) of 35%. Computed Tomography Angiography (CTA) showed aberrant coronary anatomy with the left main coronary artery (LMCA) arising from the right ostium, passing anterior to the pulmonary trunk, and dividing into 2 branches: The Left Anterior Descending artery (LAD) and Left Circumflex Artery (LCX) (Fig. 2-3). Percutaneous Coronary Intervention (PCI) was deemed risky and the patient was instead managed with dual antiplatelet therapy, anti-anginal medications, and a statin. Her clinical status stabilized, and she was discharged home.

**Discussion**

SCAs are rare congenital anomalies that go unnoticed until adulthood and in some cases until the fifth or sixth decades of life. They consist of a single coronary artery supplying the entire myocardium. The reported incidence of SCAs is variable and ranges between 0.014 and 0.066% of the population [5-11]. *Turkmen et al.* reviewed 215,140 patients referred for angiography and found that 0.031% had SCAs [8]. *Yamanaka et al.* reported an incidence of 0.045% out of 126,595 patients [10], and *Tuncer et al.* reported an incidence of 0.014% out of 70,850 patients (Table 1) [7]. *Desmet et al.* reviewed 50,000 patients and found thirty-three cases of SCAs or an incidence of 0.066% [3].
SCAs may be associated with other cardiac abnormalities including Tetralogy of Fallot, pulmonary artery defects, coronary arteriovenous fistulae, transposition of the great arteries, bicuspid aorta, and patent ductus arteriosus [12-13]. Kervancıoğlu et al. found that SCAs were present in 3.5% of patients with Tetralogy of Fallot [5]. There is no gender preference with SCAs, however, most large series review patients presenting for coronary angiography, and therefore, the reported incidence may be different from the general population. The most dramatic picture of SCAs is when young athletes present with sudden death during strenuous exercise [14-16].

Atypical chest pain or non-specific symptoms, syncope, heart failure, arrhythmias, dyspnea on exertion, palpitations, ventricular tachycardia, and myocardial infarction or sudden cardiac death, have been reported as presenting symptoms. In the Turkmen et al. series, 4.5% of the patients presented with myocardial infarction. The majority of these patients did not turn out to have critically stenosed coronaries [8].

Significantly severe symptoms like angina, syncope, or sudden cardiac arrest are usually seen when the SCA courses between the aorta and pulmonary arteries where it is subjected to kinking and compression, especially during exertion when the arterial diameter grows [17, 18]. Heavy exercise is one of the main triggers of death in such patients. A study of 242 deceased patients with isolated congenital coronary anomalies showed that one-third suffered from sudden cardiac death, and half of these were exercise-related; patients younger than 30 years are more likely to suffer from sudden cardiac death during physical exertion [19].

Lipton et al. were the first to propose a classification of SCAs (Fig. 4) [2]. Other authors like Yamanaka et al. added some modifications to this classification (Table 2) [10]. The origin of the SCA from the ostia is designated with R for right ostium and L for left ostium. Since SCAs may have different origins and different courses, this letter is followed by a roman numeral that describes the course. Class I is when the SCA follows the anatomical course of the normal Right Coronary Artery (RCA) or Left Coronary Artery (LCA). Class II describes a normally arising SCA from the proximal part of the ostium that reinserts into the mid or distal LAD, feeding the proximal LAD and Circumflex arteries. In Class III, the LAD and LCX arise from separate coronary trunks. This Roman numeral is followed by a letter describing the course of the transverse branch as follows: (A) anterior to the large vessels, (B) between the Aorta and Pulmonary artery, (P) posterior to the large vessels, (S) passing over the interventricular septum, or (C) combined (Table 2) [2,10,16,20].

The majority of patients with SCAs are of the RI, LI, and RIIA types. These subtypes have a benign course with symptoms arising, at times, from coronary atherosclerotic lesions [21]. In cases where the SCAs originate from the contralateral sinuses and travel between the large trunks, such as in RIIIB, LIIIB, and RIII, more serious clinical courses can occur. The large trunks dilate during exercise, when the demand is high, which results in compression of the SCAs [21]. Therefore, the inter-arterial course (B) is the one associated with the worst outcomes. Additionally, slit-like orifices, unusual SCA take-off angles, and kinked compressible courses are predisposing features...
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<thead>
<tr>
<th>Classification</th>
<th>Description</th>
<th>Prognosis</th>
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<tbody>
<tr>
<td>RI</td>
<td>SCA originating from the RCC, following the course of RCA or LCA</td>
<td>Generally benign</td>
</tr>
<tr>
<td>LI</td>
<td>SCA originating from the LCC, following the course of RCA or LCA</td>
<td>Generally benign</td>
</tr>
<tr>
<td>RII</td>
<td>SCA arising from the RCC, feeding the mid and distal LAD and mid LCX</td>
<td>RIIA-Usually benign, asymptomatic RIIIB- Symptoms of ischemia and PVCs</td>
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<tr>
<td>LII</td>
<td>SCA arising from the LCC, feeding the mid and distal LAD and mid LCX arteries</td>
<td>RIIA-Usually benign, asymptomatic RIIIB- Symptoms of ischemia and PVCs</td>
</tr>
<tr>
<td>RIII</td>
<td>SCA arising from the RCC and feeding the LAD and LCX from separate coronary trunks</td>
<td>Usually associated with sudden death and symptoms during exercise</td>
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<tr>
<th>Course Designation</th>
<th>Description</th>
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<tr>
<td>A</td>
<td>Anterior to the large vessels, right ventricle</td>
</tr>
<tr>
<td>B</td>
<td>Between the aorta and pulmonary arteries</td>
</tr>
<tr>
<td>P</td>
<td>Posterior to the large vessels</td>
</tr>
<tr>
<td>S</td>
<td>Follows the interventricular septum</td>
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<tr>
<td>C</td>
<td>Combined type</td>
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Table 1: Summary of 3 large series on SCAs incidence [7, 8, 10]

Table 2: Classification and prognosis of the SCAs as per Yamanaka et al. [10] SCA: Single Coronary Artery, RCC: Right Coronary Cusp, LCC: Left Coronary Cusp, RCA: Right Coronary Artery, LCA: Left Coronary Artery, LAD: Left Anterior Descending artery, LCX: Left Circumflex artery PVCs: Premature Ventricular Contractions.
to different symptoms of ischemia or sudden death. About 15% of patients with detectable exercise-induced ischemia may have those abnormalities [10, 22, 23]. Our case is of the RII-A subtype, where the SCA arises from the right sinus of Valsalva, courses anterior to the large vessels, and gives rise to the LMCA, which bifurcates into the LAD and LCX.

Cardiac Catheterization (CAG) remains the gold standard for accurate assessment of SCA anatomy. Additionally, multislice CTA scanning can generate 3-dimensional imaging showing the anomaly with the advantage of a less invasive method. Additional adjuncts for revealing the degree of ischemia during stress include: the adenosine stress cardiovascular Magnetic Resonance (MR), dobutamine stress MR, ECG gated single-photon emission computed tomography (SPECT), and echocardiography with stress-induced wall motion abnormalities [18-24-26].

Because of the rare incidence and the variable anatomy and symptoms encountered, no consensus or guidelines are available, and each case has a specially tailored therapeutic approach. Revascularization is not recommended as a systemic approach, except when it may reverse ischemia, correct a slit-like ostium, reimplant a type III coronary anatomy, or fix a kinked, compressible course.

For group I, medical therapy is generally recommended, as the clinical course is benign. In individual cases with significant atherosclerosis and symptoms, a coronary graft may be necessary. Percutaneous interventions (PCI) have a high risk of ostial dissection and thrombosis [27-30]. Young patients with symptoms of ischemia on exertion should undergo surgery due to the risk of sudden death associated with types II and III SCAs. The preferred surgery is CABG, along with re-implantation of the abnormal artery to the aorta. Stenotic atherosclerotic lesions also call for CABG to reverse ischemia. Coronary arteries arising from the contralateral coronary sinus without stenotic or slit-like lesions can be reversed surgically by reinsertion into the normal vessels where feasible [31-34].

**Conclusion**

SCA is a congenital anatomic abnormality in which only one coronary artery originates from a single coronary ostium in the aortic sinus and supplies the entire myocardium. It is important to recognize this anomaly early on, especially in young adults presenting with symptoms of syncope, weakness, chest pain, or other unexplained, recurrent symptoms. The significant difference in the reported incidence of each type is likely due to the numerous variations in different populations and different inclusion criteria of each study. The majority of the large series looked at patients presenting for coronary angiography with cardiac symptoms, and only a few reports looked at postmortem groups that succumbed to sudden death, particularly young athletes.

The different reports give us a clear idea about who is likely to develop ischemic, non-atherosclerotic symptoms. Those patients with RIIIB, LIIB, and RIII seem to be at high risk of developing symptoms during exertion due to compression of the coronary vessels and may benefit from corrective surgery if caught in time.

Cardiologists and cardiovascular surgeons should be aware of this rare condition, which carries a potential risk of sudden cardiac death, especially in young and active adults. The symptoms vary significantly, and some patients may live an uneventful life, while others may be
prone to catastrophic outcomes. A high index of suspicion and early recognition of a possibly eventful clinical course could save lives.

Acknowledgments

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References


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