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Coronary-Cameral Fistula from RCA to RA Masquerading as RSOV: A Case Report

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ABSTRACT

Introduction: Coronary-cameral fistulas (CCFs) are rare congenital anomalies characterized by abnormal communications between a coronary artery and a cardiac chamber. These fistulas can pose significant diagnostic challenges when they mimic other cardiac pathologies such as ruptured sinus of Valsalva (RSOV), especially given the overlapping clinical presentations and hemodynamic implications.

Case Report: We present the case of a 24-year-old female who experienced chronic chest pain and intermittent dyspnea, initially suspected to have RSOV into the right atrium (RA) based on transthoracic echocardiography. Further evaluation with computed tomography angiography revealed a dilated and tortuous right coronary artery (RCA) terminating directly into the RA, confirming a diagnosis of coronary-cameral fistula. Intraoperative assessment corroborated the imaging findings, revealing a large single fistulous ostium from the RCA to the RA without evidence of RSOV, congenital septal defects, or coronary ostial abnormalities. The fistula was surgically ligated, and the patient had an uneventful postoperative recovery with symptom resolution.

Conclusion: This case highlights the diagnostic complexity in differentiating CCFs from RSOVs, particularly when advanced imaging is not initially employed. The congenital nature, anatomical location, and absence of associated structural anomalies underscore the unique presentation of this fistula. Multimodality imaging, including cardiac CT and intraoperative confirmation, proved crucial in delineating the precise anatomy and guiding appropriate surgical management. CCFs, although rare, must be considered in the differential diagnosis of RSOV. Accurate anatomical delineation through advanced imaging techniques is essential for effective management. This case reinforces the importance of a multidisciplinary approach and high-resolution imaging in the evaluation of complex congenital coronary anomalies.

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Introduction

Coronary-cameral fistulas (CCFs) are abnormal connections between a coronary artery and a cardiac chamber, typically identified during diagnostic cardiac catheterization or advanced imaging (1). These rare congenital malformations can lead to a variety of clinical presentations, from asymptomatic detection to severe hemodynamic compromise, depending on the size, location, and flow dynamics of the fistula (2). While often benign, larger fistulas can cause significant left-to-right shunting, potentially resulting in volume overload, myocardial ischemia, and even heart failure

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(3). It is crucial to distinguish these fistulas from other cardiac anomalies, such as ruptured sinus of Valsalva (RSOV) aneurysms, as their clinical manifestations can overlap significantly, creating diagnostic challenges that require comprehensive imaging and hemodynamic assessment (4).

This report describes a rare case of a coronary-cameral fistula originating from the right coronary artery (RCA) and ending in the right atrium (RA). Initially presenting with symptoms suggestive of a RSOV, thereby underscoring the diagnostic complexities inherent in such cardiac anomalies. A RSOV aneurysm, more prevalent in Asian populations and males, typically involves the right coronary sinus draining into the right ventricle or right atrium (5). Differentiating these conditions is critical, as coronary ostial atresia or stenosis, whether congenital or acquired, presents a unique set of management challenges distinct from those of a typical RSOV (6,7). Congenital coronary artery anomalies, like ostial atresia, present a broad spectrum of anatomical variations with diverse pathophysiological implications that can mimic other cardiac conditions (8,9). This case highlights the diagnostic dilemmas encountered when a coronary-cameral fistula mimics a RSOV aneurysm, emphasizing the utility of advanced imaging modalities for accurate differentiation and timely intervention (10).

Case Presentation

A 24-year-old female presented with a 12-month history of left-sided chest pain and intermittent breathlessness. She denied palpitations, syncope, or orthopnea. Her medical, surgical, and family histories were unremarkable. She was not on any regular medications and had no known allergies. Physical examination revealed normal vital signs with no peripheral edema, cyanosis, or jugular venous distension. Cardiac auscultation revealed no murmurs, rubs, or gallops.

Initial laboratory investigations including complete blood count, renal and liver function tests, thyroid profile, and cardiac biomarkers like Troponin I, Pro BNP, CPK-MB were within normal limits. Chest radiography showed a normal cardiac silhouette with clear lung fields. Electrocardiogram revealed normal sinus rhythm without ST-T segment abnormalities. Transthoracic echocardiography showed mild right atrial enlargement and suggested an abnormal color flow pattern from the aortic root to the right atrium with Raised PAH, raising suspicion for a RSOV into the right atrium.

To confirm the diagnosis and further delineate anatomy, cardiac CT angiography was performed. It revealed a tortuous, dilated RCA originating normally from the right coronary sinus but terminating in a direct fistulous communication with the right atrium. No features of RSOV, such as aneurysmal dilation or rupture of the aortic sinus, were identified. Cardiac MRI was subsequently performed to assess shunt volume and ventricular function. It confirmed the presence of a high-flow coronary-cameral fistula without any myocardial fibrosis or ischemia.

Given these findings, the patient was referred for surgical correction. Preoperative coronary angiography confirmed the anatomical course and flow dynamics of the fistula, aiding in surgical planning.

Intraoperative Findings

Intraoperative exploration confirmed a significantly dilated RCA (Figure 1) with a single large fistulous opening into the right atrium. The fistula was located at the level of the atrial wall near the tricuspid valve annulus. There was no evidence of RSOV, aortic root dilation, or dissection. The coronary ostia were normally situated, and no other congenital cardiac anomalies were observed. The tricuspid valve and surrounding structures appeared intact and functional.

Surgical management involved direct ligation of the fistula at its atrial entry point while preserving the native course of the RCA. Intraoperative transesophageal echocardiography confirmed successful closure with no residual shunting or new valvar regurgitation.

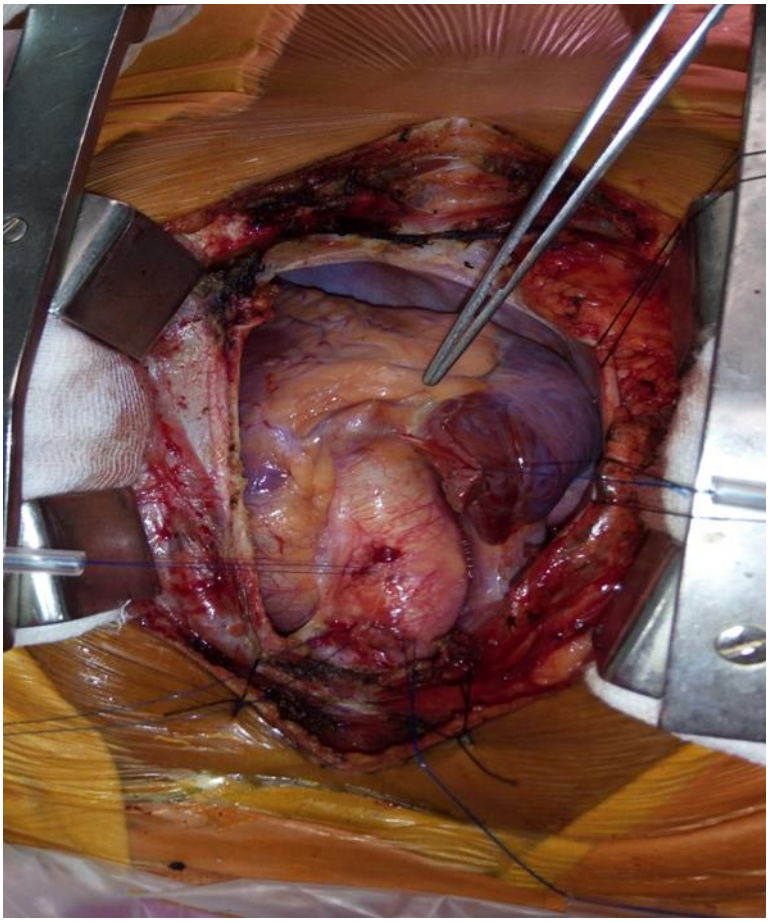

Figure 1. Intra Operative image showing Coronary (RCA) to cameral (RA) Fistulous tract.

The patient was extubated in the operating room and then transferred to the cardiac intensive care unit for monitoring.

Postoperative Course

The patient experienced an uneventful postoperative recovery. Follow-up echocardiography revealed resolution of right atrial dilation and no residual shunt. She was discharged on the sixth postoperative day with instructions for routine cardiac follow-up. During the three-month review, the patient reported complete resolution of chest pain and breathlessness. Repeat imaging confirmed durable closure of the fistula and preserved biventricular function.

Discussion

CCFs are rare anomalies, accounting for less than 0.2% of congenital cardiac defects (1, 2). These connections most commonly involve the RCA draining into the right heart chambers, particularly the right atrium or ventricle (2, 3). The pathophysiological consequences of such fistulas depend on their size, location, and associated anomalies. Small fistulas may remain asymptomatic, while large ones can lead to significant left-to-right shunting, volume overload, and myocardial ischemia due to the coronary steal phenomenon (4,5).

In the present case, the fistula's drainage into the RA resulted in chronic volume overload, explaining the patient's exertional symptoms. The absence of a murmur highlights the diagnostic difficulty in such cases, where subtle hemodynamic changes may occur without auscultatory findings. The initial suspicion of RSOV was reasonable given the echocardiographic findings; however, advanced imaging modalities were critical in refining the diagnosis.

Multimodal imaging plays a pivotal role in the evaluation of coronary fistulas. Echocardiography can identify abnormal flows, but its spatial resolution may be insufficient to accurately define fistula anatomy, especially when differentiating from RSOV (6, 7). Cardiac CT provides excellent visualization of coronary artery anatomy and fistulous connections (8).

Cardiac MRI complements this by assessing flow dynamics and ventricular function, which are essential for determining the clinical impact of the shunt (9).

The surgical approach aims to eliminate the abnormal communication while preserving myocardial perfusion. Direct ligation is preferred when feasible, though transcatheter embolization is an option in select cases (10, 11). Our patient underwent successful surgical repair with symptom resolution and no evidence of ischemia or recurrence.

The importance of differentiating CCFs from other entities such as RSOV, coronary artery aneurysms, and anomalous coronary origins cannot be overstated. Misdiagnosis can lead to inappropriate management, delayed treatment, or missed complications. Therefore, accurate anatomical delineation using advanced imaging and a multidisciplinary approach involving cardiologists, radiologists, and cardiothoracic surgeons is essential (12).

The prognosis following closure of isolated congenital CCFs is excellent, particularly when diagnosed and treated before the onset of irreversible myocardial damage or chamber dilation. Long-term follow-up is recommended to monitor for recurrence, residual shunting, or late complications (13-15).

Conclusion

This case highlights the diagnostic and therapeutic challenges posed by CCFs, particularly when mimicking more common conditions such as RSOV. Accurate diagnosis hinges on comprehensive imaging and clinical evaluation. Surgical ligation remains a definitive and effective treatment in anatomically suitable cases. Timely identification and intervention can result in excellent clinical outcomes with symptom resolution and preserved cardiac function.

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Ethical Approval and Informed Consent

The Institutional Review Board (IRB) or equivalent ethics committee of GCS Medical College, Hospital, and Research Centre provid approval. Research approval was not applicable as this is a case report/study. The subject provided informed written consent for the publication of the study data.

Conflict of Interest

The author declared no potential conflicts of interest with respect to the research, authorship, and/or publication of this article.

Availability of Data and Materials

The datasets used and/or analyzed during the current study are available from the corresponding author upon reasonable request.

References

1. Nunn DB, Doberneck RC, Barila TG, Dixon RB. Experimental coronary arteriovenous fistula as a stimulus for increasing intercoronary collaterals. Journal of Surgical Research. 1963 Jun 1;3(4):220-4.
2. Sathienchoak P, Surakiatchanukul S, Yothasamud C, Siriratana U. Coronary arteriovenous fistula. Circ. 1978;61:235.
3. Gorbachevsky SV, Shmaltz AA, Zaets SB. Poor left ventricular performance in a child with coronary sinus stenosis: successful surgical repair. The Annals of Thoracic Surgery. 2015 Sep 1;100(3):1096-9.
4. Calkins Jr JB, Talley JD, Kim NH. Iatrogenic aorto-coronary venous fistula as a complication of coronary artery bypass surgery: Patient report and review of the literature. Catheterization and cardiovascular diagnosis. 1996 Jan;37(1):55-9.
5. Vamsidhar A, Rajasekhar D, Vanajakshamma V. Transcatheter device closure of multiple defects in ruptured sinus of Valsalva aneurysm. Indian Heart Journal. 2015 Dec 1;67:S74-7.
6. Sharma D, Prashar A. Coronary ostial acquired occlusion or congenital atresia: an ongoing discussion. Texas Heart Institute Journal. 2023 Jul 28;50(4).
7. Angelini P. Congenital coronary artery ostial disease: a spectrum of anatomic variants with different pathophysiologies and prognoses. Texas Heart Institute Journal. 2012;39(1):55.
8. Bajwa A, Dhoot J, Gupta S. Coronary Sinus Lead Placement in Patients With Coronary Sinus Ostial Atresia: An Innovative Approach. Case Reports. 2021 Apr 1;3(4):614-8.

9. Reid K. The anatomy of the sinus of Valsalva. Thorax. 1970 Jan 1;25(1):79-85.
10. Alsaddique AA, Elsaegh MM, Fouda MA. Aortic valve replacement in a patient with an aberrant left coronary artery. Asian Cardiovascular and Thoracic Annals. 2003 Jun;11(2):169-70.
11. Kang SR, Park WK, Kwon BS, Ko JK, Goo HW, Park JJ. Management of Coronary Sinus Ostial Atresia during a Staged Operation of a Functional Single Ventricle. The Korean Journal of Thoracic and Cardiovascular Surgery. 2018 Apr 5;51(2):130.
12. Porepa M, Benson L, Manson DE, Friedman JN. True blue: a puzzling case of persistent cyanosis in a young child. CMAJ. 2009 Mar 31;180(7):734-7.
13. Esposito A, Francone M, Andreini D, Buffa V, Cademartiri F, Carbone I, et al. SIRM–SIC appropriateness criteria for the use of Cardiac Computed Tomography. Part 1: Congenital heart diseases, primary prevention, risk assessment before surgery, suspected CAD in symptomatic patients, plaque and epicardial adipose tissue characterization, and functional assessment of stenosis. La radiologia medica. 2021 Sep;126(9):1236-48.
14. Marcu CB, Beek AM, Van Rossum AC. Clinical applications of cardiovascular magnetic resonance imaging. Cmaj. 2006 Oct 10;175(8):911-7.
15. Moir TW, Pritchard WH. Study of the Hemodynamic Effects of the Aorto-Coronary Sinus Graft Operation in Patients with Coronary Artery Disease. Circulation. 1957 Dec;16(6):1070-6.