Coronary Artery Fistulas Mimic and Misdiagnosis with Ruptured Sinus of Valsalva: 3 Cases Report and the Role of Imaging Modalities for Diagnosis

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ABSTRACT:

Coronary arterial fistulas (CAFs) are uncommon, although they are one of the most prevalent coronary artery anomalies and typically present with no symptoms in young patients. We report three cases of CAFs in which 2 cases were diagnosed with ruptured sinus of Valsalva aneurysm (RSVA) before surgery. Coronary computer tomography angiography (CCTA) is useful for evaluating CAFs because it requires less acquisition time and produces better temporal and spatial resolution than other imaging.

Keywords: *coronary arterial fistula, computer tomography angiography, cardiac anomaly*

BACKGROUND

Congenital coronary artery fistula was first identified by Krause in 1865, but the first successful surgical therapy was not recorded until 1958 by Fell and associates (1). In patients undergoing coronary angiography, the incidence of coronary artery fistulas (CAFs) is remarkably low, ranging between 0.13% and 0.22% (2,3). Despite their rarity, CAFs represent a significant coronary abnormality (4). This report will present some cases of CAFs with different approaches and include some key points in the diagnosis as well as a brief review of the condition management.

Case presentation

We hereafter provide a summary of the presentation and treatment of three cases involving CAFs originating from the coronary sinus of Valsalva to the heart chambers. The first two patients were a 15-year-old female and a 46-year-old female; both were admitted to the emergency department with symptoms of fatigue and dyspnea.

The 15-year-old girl presented with a murmur sound on examination and a positive "windsock sign" on echocardiography, suggesting a rupture of the right coronary sinus. Other modalities, including X-ray and electrocardiogram, showed limited diagnostic data. She was later transferred to the heart surgery department. The total delayed time before surgery was just three days. The following operation

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revealed that the right coronary sinus of Valsalva (RCS) was dilated and gave way to a fistula connecting the right ventricle of the patient. No rupture was spotted, and the fistula was successfully closed by patch and ligature. She followed the postoperative monitoring more than 5 years later to ensure complete resolution of the abnormalities.

The second patient had the condition diagnosed at a late age, at 46. Symptoms reported were fairly similar, with mild shortness of breath and vague fatigue. The physical examination and general screenings revealed no specific findings, except for a continuous murmur sound at the upper right sternal border. Echocardiography also recognized a dilated right coronary sinus of Valsalva with a positive windsock sign and a high-velocity Doppler flow through a discontinuity of the RCS wall to the right atrium. Those findings led to a preoperative diagnosis of a rupture in the sinus of Valsalva. Surgery was performed, although the operation planning was not thoroughly discussed due to the urgent course of the assumed diagnosis of Valsalva sinus rupture. Unexpectedly, rupture of the coronary sinus was excluded. Instead, the surgeons confirmed the presence of a coronary cameral fistula in the right atrium, which they later completely occluded with a patch and ligature. She also had to follow a long monitoring period.

Despite being successfully managed, both patients were misdiagnosed with coronary sinus of Valsalva ruptures in the initial assessment, where the signs and symptoms were non-specific and echocardiography was the main modality used to establish the diagnosis.

The misinterpretation of the fistula as an urgent rupture of the RCS despite patients' stable presentation could unnecessarily activate the emergent surgery protocol, with inappropriate preparation of the operation facilities, causing stress to the hospital capacity and patients' mental health.

The final case in this report was a female, 31 years old, with no history of cardiovascular disease. She presented to the outpatient clinic of our hospital with nonspecific symptoms, including mild dyspnea on exertion. The physical examination barely reported abnormalities, with normal heart rate and blood pressure and no edema. The examiner doubted there was a mild murmur radiating from the right border of the sternum to the other side.

X-ray showed enlargement of the right-sided mogul, suggesting expanded right atrium size. Electrocardiograms did not indicate any particular causes of the patient's symptoms. Transthoracic echocardiogram (TTE) recorded a sac-like structure expanding from the left coronary sinus at the left coronary artery origin location. The distant tip of the aneurysm was not feasibly assessed by TTE.

We later proceeded to perform coronary computed tomographic angiography (CCTA) on the patient. Imaging findings revealed an enlarged left coronary artery with a diameter of approximately 15 mm in the proximal segment. There was a fistula originating from the left main coronary artery, right before the bifurcation of the anterior descending and circumflex arteries (Fig. 1a, b, f).

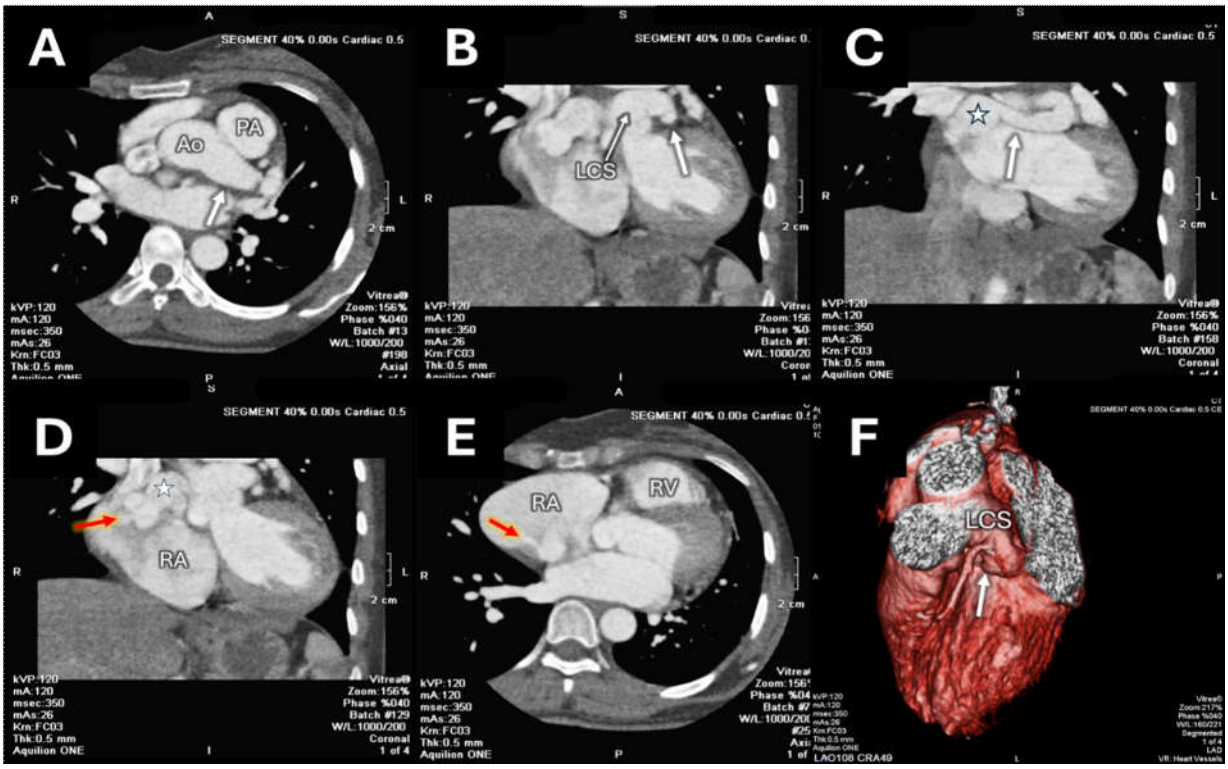
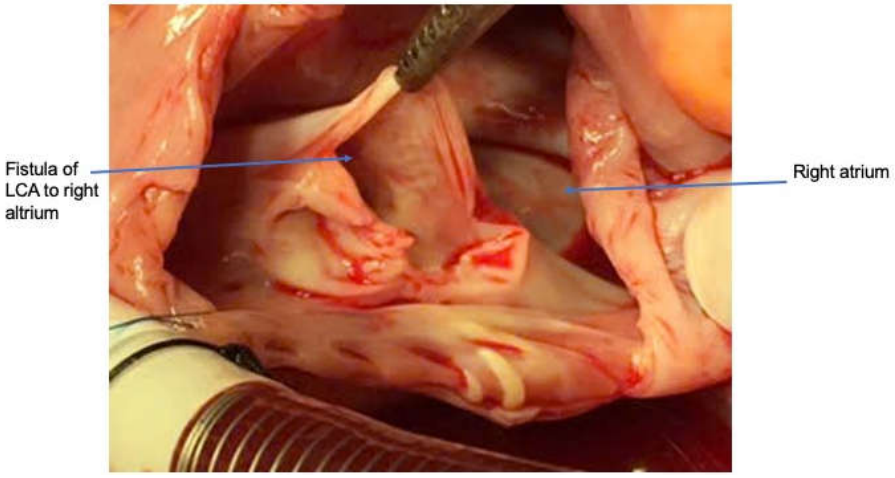


Figure 1. Preoperative CCTA findings. A, B: CCTA showed enlargement of the left coronary sinus of Valsalva (white arrow), protruding downward of the LCS; C: The tortuous fistula ran backward and to the right. Many aneurysms (white star) spotted along the fistula; D, E: The anastomosis (red arrow) where the fistula flowed into RA; F: The aneurysm at the origin of the fistula viewed on volume rendering of the patient's heart. CCTA: coronary computed tomographic angiography; LCS: left coronary sinus; RA: right atrium; RV: right ventricle; Ao: aorta; PA: pulmonary artery.

A)



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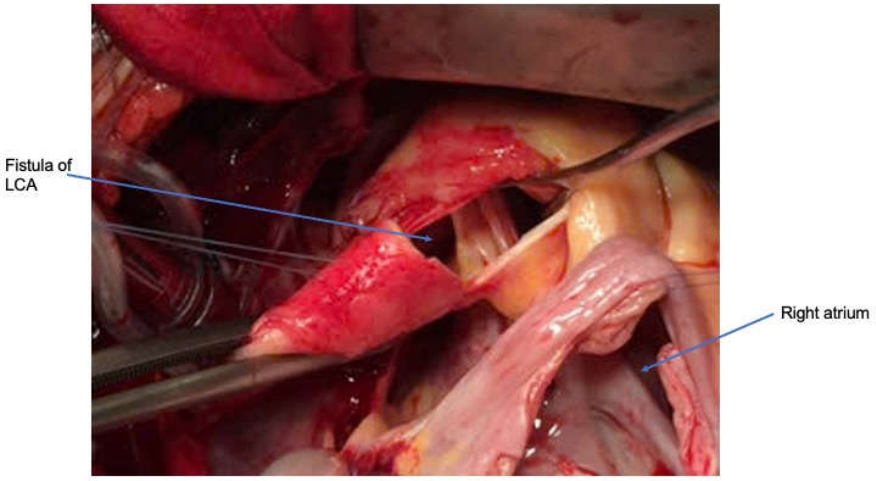

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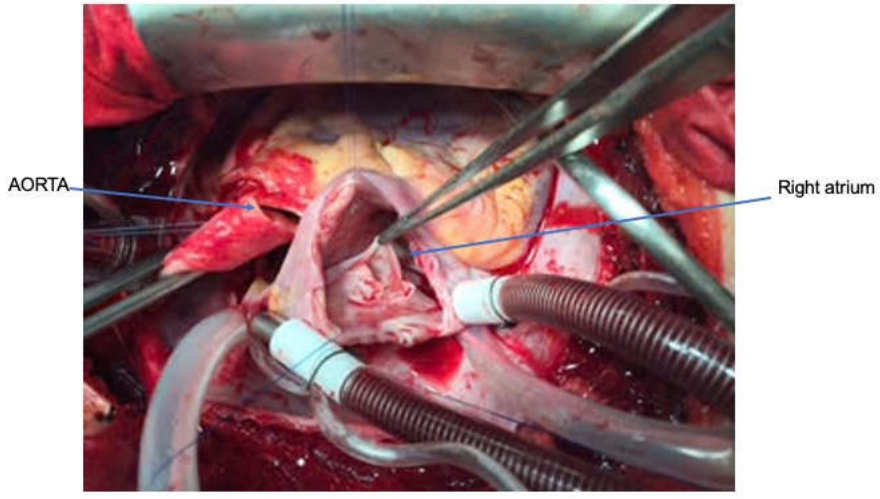

Figure 2. Intraoperative findings. A: Confirmation of dilated LCA; B: Fistula confirmed beneath the left coronary artery; C: Connection to the RA identified. (LCA left coronary artery; RA right atrium).

The fistula ran downward and backward to form a tortuous pathway between two atria, with many aneurysmal segments (Fig. 1c), before penetrating into the right atrium with a 10 mm anastomosis (Fig. 1d, e). We performed sternotomy, with proper planning time, and confirmed the diagnosis, consistent with CCTA findings, with the large left coronary sinus of Valsalva and the connection to the right atrium of the fistula (Fig. 2).

The pathway was occluded using a patch and ligation of the aneurysm. Perioperative TEE showed no residual blood flow through the fistula into the right atrium. After the operation, the patient witnessed an improvement in symptoms, with her NYHA score shifting from III to I. She was discharged 10 days later and was monitored for more than 3 years with no recurrence of symptoms.

DISCUSSION

CAFs are uncommon congenital defects, accounting for 0.8% of coronary anomalies. In the overall population, CAFs incidence is merely 0.002%. In the embryologic development, the primitive myocardium is nourished by sinusoids connected to the tubular heart; failure of these sinusoids to regress can result in persistent fistulous connections (5,6). Most pediatric CAF patients are asymptomatic, with only 20% of this age group exhibiting symptoms (5). The symptom prevalence increases with age, with only 40% of adults remaining asymptomatic (7). Common symptoms in older patients include exertional dyspnea, fatigue, and angina, signaling the myocardial ischemia caused by coronary steal (5,8).

Examination findings in CAF patients also pose poor specificity. Signs could be more vivid when complications such as congestive heart failure, pulmonary hypertension, or atrial arrhythmias emerge (9). A continuous murmur, from persistent blood flow between high-pressure coronary arteries and low-pressure cardiac chambers throughout the cardiac cycle (6). Differential diagnoses for continuous murmurs include ruptured sinus of Valsalva aneurysm, patent ductus arteriosus, and pulmonary arteriovenous malformations (10). Those cases mentioned in our report shared the same tone of mild and nonspecified symptoms, posing the need for appropriate tools to establish a diagnosis.

Approximately half of patients undergoing surgical correction for CAFs exhibit normal electrocardiograms (ECGs). Chamber hypertrophy—either left- or right-sided—due to chronic volume overload could be recorded in some CAF patients (5). In older individuals, especially those with right atrial fistulas, atrial fibrillation is common, secondary to atrial dilation. Ischemic ECG patterns could sometimes appear following the steal physiology (11,12). In our three patients presentations, ECGs did not provide any supplemental information for the final diagnosis, despite their ages ranging from adolescents to middle-aged women.

Color Doppler echocardiography (EC) is still highly valuable in screening for the presence of CAFs, thanks to its accessibility and versatility. Besides transthoracic echocardiography (TTE), transesophageal echocardiography (TEE) could enhance visualization of fistulous tracts, facilitate localization of drainage sites, and provide dynamic assessment of wall motion abnormalities (12). Nevertheless, echocardiography can only exactly confirm about 20% of CAFs throughout studies (13).

Definite diagnosis could be really challenging, with CAFs mimicking more prevalent conditions such as patent ductus arteriosus, ruptured sinus of Valsalva aneurysm, ventricular septal defect, or arteriovenous malformations of systemic vessels. Imaging tools, such as CCTA and MRI, are increasingly recommended to improve diagnostic accuracy and guide therapeutic planning (14). In particular, in our third case with a complex and tortuous fistula pathway, the direction and anastomosis were clueless on EC images, and various overlapping abnormalities could not be excluded, which highlighted the necessity of a better diagnosing modality.

Systematic reviews from many authors have confirmed that CCTA is the modality of choice in assessing coronary sinus and artery fistula (15,16). In contrast to TTE or TEE with their limitation in assessing small or complex fistulas, CCTA has shown diagnostic accuracy

rates exceeding 75% in large retrospective cohorts (17). The visualization of coronary arteries is pivotal for diagnosis, anatomical delineation, and surgical repair planning (18).

The proposed protocol for the best view of the coronary sinus of Valsalva should include a field of view (FoV) covering the chest, from above the aortic arch to the diaphragm; a bolus tracking or test bolus protocol to ensure optimal enhancement of the aortic and coronary lumen; biphasic contrast media injection (contrast followed by saline) to emphasize the distinguished attenuation of the left side and right side of the heart, enhancing the visualization of the fistulas' flow and anastomosis; and a prospective ECG gating to cover the cardiac cycle and dynamic of the flow in the fistulas (5). Review and interpretation of the CCTA images should be carefully discussed in the heart team to establish the most likely diagnosis and optimize the management.

Despite the fact that invasive angiography was the conventional gold standard to diagnose CAFs, electrocardiographically gated CCTA has become the preferred noninvasive alternative modality thanks to its short acquisition time, high spatial and temporal resolution, and better safety prognosis without invasive procedure adverse events. Additionally, the anatomy of CAFs, including their origin, drainage site, complexity, and the quantity and size of fistulous tracts, could be accurately assessed thanks to three-dimensional CT angiograms (18). Surgeons could leverage the knowing of those anatomical notes for intervention planning through multi-planar reformatted images and 3D reconstructions (19). In two of our three cases, the condition was incorrectly diagnosed prior to surgery. The third patient's preoperative diagnosis was exact with the utilization of CCTA.

The number of incidentally identified CAFs on multidetector CT has been increasing, and multidetector CT scanning is now frequently used to diagnose cardiovascular abnormalities (20). Besides CCTA, cardiac magnetic resonance imaging (CMR) is also a rising modality for identification of major coronary artery abnormalities (21).

Experts recommend early surgical correction for advanced CAFs with congestive heart failure, significant shunt (Qp-to-Qs ratio 1.5), or overt symptoms (20,22,23). The aim of surgery is to close the fistulous connection while preserving the anatomy of the coronary arteries. Performing surgery after complications have started increases the mortality and morbidity rates (24,25). Transcatheter closure is generally safe and effective, has a favorable clinical record, and has a manageable risk profile (24,26). A variety of occlusion devices are utilized in CAF closure, including: Coils (e.g., interlocking detachable coils); Vascular plugs (e.g., Amplatzer Vascular Plug II); Covered stents (e.g., PK Papyrus, Graftmaster); Patent ductus arteriosus (PDA) occluders; Detachable balloons and histoacryl resin (22,27,28). The choice of appropriate devices largely depends on the anatomy of coronary arteries and the size and site of the fistula anastomosis, highlighting the impact of exact diagnosis and planning by CCTA.

From the perspectives of the abovementioned cases of fistula from the coronary sinus of Valsalva to the heart chambers in our report, establishing the definite diagnosis is necessary for the condition's

following management because it influences the timely and suitable operation plan, patient compliance, and prognosis. Coronary computed tomographic angiography is the best modality for evaluating this kind of complex shunt in the heart system since it definitely provides the benefit of spatial visibility, particularly in cases with tortuous and obscure abnormalities.

CONCLUSION:

Most infants and children with CAFs are asymptomatic. In adulthood, CAFs can cause several problems, such as adult myocardial ischemia, heart failure, arrhythmia, and infective endocarditis. CT angiography is the modality of choice for diagnosis, management planning, and prognosis of CAFs.

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