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Collaterals of Life: Fistulous Communication of Vieussens' Arterial Ring

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Abstract

The Vieussens' arterial ring (VAR) is a rare coronary anomaly that may be associated with a pulmonary fistula, and has been linked to ischemia, myocardial infarction, heart failure, arrhythmias, and sudden death. We present the case of a 54-year-old man who was admitted to the emergency room with angina and dyspnea on moderate exertion, in the context of atrial fibrillation and decompensated heart failure with left ventricular systolic dysfunction. In the study of its etiology, coronary computed tomography angiography (CCTA) identified an anomalous conal artery outflow (type C) as part of the VAR and a fistula to the pulmonary artery trunk (variant 1b), with an arterial flow linking the conal artery and the middle third of the left anterior descending artery (LAD), which then fistulized to the pulmonary artery trunk. The hemodynamic and clinical repercussions presented with signs of ischemia in the LAD territory and acute heart failure. In this case, CCTA identified the coronary anomaly and its trajectory with great precision through volume rendering reconstruction, as well as the dynamics of coronary flow in the presence of the pulmonary fistula. This anatomical variant is rarely described, and CCTA is of great utility in the therapeutic decision for fistula closure.

Keywords

Vieussens' Arterial Ring, Pulmonary Fistula, Coronary Artery Anomaly

1. Introduction

The Vieussens' arterial ring is an embryological remnant that gains clinical significance as an intercoronary collateral vessel, first described by Raymond de Vieussens in 1706 [1]. One part of this remnant is the anatomical variant of the

conal artery, known as the "third coronary artery" [2], which communicates with the anterior descending artery. The VAR variant with pulmonary fistula is classified as 1b and is a rare presentation [3]. The case described presents an anatomical variant VAR type 1b, identified by CCTA, with clinical and hemodynamic repercussions associated with ischemia and heart failure, allowing for the definition of its therapeutic management.

2. Case Summary

A 54-year-old man with no history of pathology presented with palpitations and dyspnea for four years. He was admitted to the emergency department with marked limitation in activity, NYHA class III. Physical examination revealed basal bilateral pulmonary crackles, lower limb edema, cardiac arrhythmic and tachycardic sounds, and the ECG showed atrial fibrillation and ischemic-appearing changes (V2 to V5). Laboratory test results showed elevated transaminase levels (AST 893 U/L, ALT 940 U/L), NT-pro-BNP 5492 pg/ml, CRP 2.6 mg/dl, and Troponin T within the normal range. The transthoracic echocardiography showed an ejection fraction (EF) of 25%, segmental abnormalities of myocardial contractility in the left anterior descending (LAD) territory, and preserved systolic function in the right ventricle. The coronary angiography revealed coronary arteries without lesions and the presence of an anomalous outflow branch of the LAD with probable distal drainage into the pulmonary artery, showing TIMI III flow. The HITACHI Scenaria 128-slice tomograph was used for the CCTA. The calcium score analysis was 0 Hounsfield Units (HU). The 3D reconstruction (volume rendering) revealed the independent exit of the conal artery from the right coronary sinus, with an anterior route to the pulmonary artery. The conal artery communicated with the anterior descending artery in its medial segment and fistulized into the pulmonary trunk through a tortuous vessel with evidence of contrasted flow. The multiplanar reconstruction of the coronary arteries showed a decrease in coronary flow in the LAD following its union with the communicating vessel of the Vieussens' arterial ring, consistent with a physiology of coronary steal (see **Figure 1**). The study of the pulmonary artery and its branches showed dilation.

Based on the described findings, the case was diagnosed as acute decompensated heart failure with systolic dysfunction of ischemic etiology due to a congenital coronary anomaly. The cardiology team, considering the ischemic etiology and assessing risk and benefit, decided on percutaneous closure as the intervention for this case. It should be noted that the cardiac tomographic study provided important diagnostic aid by allowing clear identification of the Vieussens' arterial ring and demonstrating its fistulous trajectory, identifying the coronary steal in agreement with the findings of the coronary catheterization. Therefore, CCTA provided us with the advantages of a safe and fast study with clear images of the coronary and cardiac anatomy for an accurate diagnosis, as in the presented case.

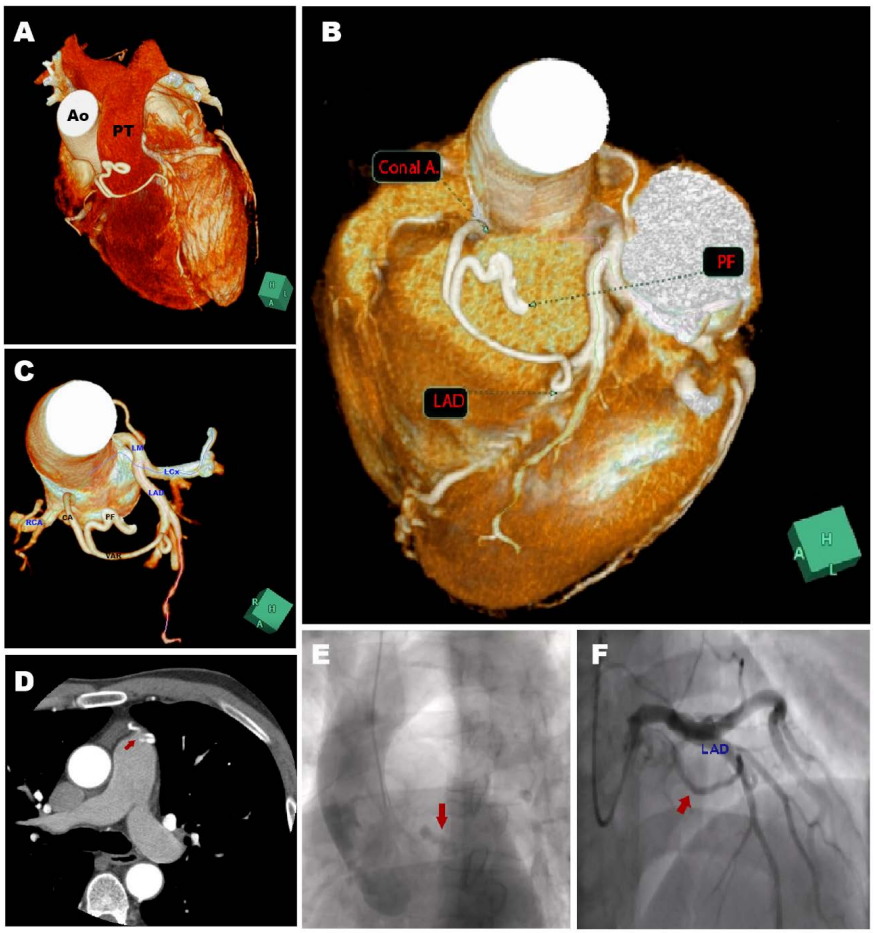

Figure 1. CCTA: volume rendering reconstruction (A-C) depicts the trajectory of the pulmonary fistula and the VAR circuit (the course of the conal artery towards the middle third of the LAD). A reduction in the distal caliber of the LAD is observed after its junction with the VAR. Contrast-enhanced axial image (D-E) shows the red arrow indicating the fistulous tract leading to the PT, as well as the trajectory of the VAR in relation to the LAD (F). Invasive coronary angiography (E-F). Abbreviations: Ao: aorta; PT: pulmonary trunk; Conal A: conal artery; PF: pulmonary fistula; LAD: left anterior descending artery; RCA: right coronary artery; LCx: left circumflex artery; VAR: vieussens' arterial ring.

3. Discussion

During embryonic development, endothelial cells grow outside the aortic wall and connect with the subepicardial vascular plexus. It has been shown that coronary arteries arise from a peritruncal ring and grow inwards towards the aortic sinuses. Arteries with tunica media and adventitia persist and end in three vascular circles: the atrioventricular circle, which gives rise to the right coronary artery and the circumflex artery; the interampullary circle, which gives rise to the anterior and posterior descending arteries; and the conotruncal circle, which persists as Vieussens' ring and communicates with the lumen of the truncus arteriosus and the other two circles [4]. Shahbaz *et al.* referred to a phenomenon as the "circle of life" when they reported a case of non-ST-elevation myocardial infarction (NSTEMI), in that case, they demonstrated VAR flow to the circumflex

artery, which acted as a salvage coronary circuit for the patient [5].

The VAR has been reported with a prevalence of 48%, but it is associated with very rare variations [1]. The origin of the coronary circulation as an early peritruncal structure is evidenced in the early stages of development [4]. Furthermore, the anomalous conal artery outflow as a segment of the VAR with a fistula to the pulmonary trunk is a very rare occurrence. The anomalous conal artery has been named the third coronary artery [2]. In the present case, the remnants of the VAR were the anomalous conal artery with anomalous outlet and the anterior descending artery in its middle third.

Several classifications have been proposed for the anomalous origin of the conal artery. Five patterns of conal artery outflow have been recognized, with the percentages for each type being as follows: Type A (32.1%), Type B (16%), Type C (40.3%), Type D (8%), and Type E (3.6%). Our case was classified as type C, which is characterized by the origin of the conal artery from the right coronary sinus with an independent exit [6]. According to Edwards *et al.* in 1981, they studied 305 necropsy specimens comparing fetal and adult life, finding an incidence of 20% in fetuses and 40% in adults. They stated that coronary patterns are not firmly established at birth [7].

The incidence of coronary artery fistula is 1/50,000 live births [1]. Individuals between 30 and 76 years of age may present with a clinical picture that depends on the shunt, time of evolution, presence of aneurysmal dilatations, and myocardial ischemia distal to the fistula, which is defined as "myocardial steal". Asymptomatic cases may also occur until the 5th or 6th decade of life, when left ventricular failure debuts [8]. In the case presented, myocardial ischemia data consistent with coronary steal was demonstrated by the absence of coronary lesions and the presence of dominant flow to the VAR and from the VAR to the pulmonary trunk; CCTA, volume rendering, and decreased distal LAD development were demonstrated, coinciding with reduced systolic function suggested by myocardial hibernation as a mechanism of myocardial adaptation.

CCTA has proven to be very useful in the assessment of coronary anomalies, especially those of the VAR type, and the study of collateral circulation, especially when there is discordance between coronary angiography findings and ventricular function [9], allowing them to be classified into 5 variants according to Nurullah *et al.* [3], our case belonging to type 1B.

According to Said Salah *et al.*, coronary angiography remains the gold standard for the detection of congenital coronary pulmonary fistulas. However, the development of CCTA as an adjuvant technique provides full anatomical detail of the fistulae [10], making use of 3D reconstruction of the coronary arteries, volume rendering and curved multiplanar reconstruction.

Regarding therapeutic management, Beneyto *et al.* propose expectant management if asymptomatic, and surgical management if symptomatic with other alterations. On the other hand, if the fistula is single, extensive, symptomatic with a large shunt, fistula ligation or embolization with an inflatable balloon or

embolization employing devices such as a coil [8]. In the present case, data compatible with myocardial ischemia were demonstrated by a myocardial perfusion test, which showed evidence of a reversible perfusion defect in the left anterior descending (LAD) territory, no evidence of coronary obstructive lesions was demonstrated by coronary catheterization and computed tomography angiography (CCTA), consistent with the alteration of segmental contractility in this territory by echocardiography. These findings support the coronary steal physiology. The clinical symptoms of heart failure are consistent with progressive left ventricular systolic dysfunction secondary to myocardial perfusion defect. Therefore, treatment would be related to the closure of the coronary fistula.

4. Conclusion

In conclusion, the presented case highlights the importance of computed tomography angiography in the diagnosis and therapeutic decision-making process of coronary steal syndrome. CCTA allowed for the rapid and noninvasive identification of the VAR and coronary-pulmonary fistula, which defined the etiology of heart failure and justified the therapeutic decision to close the fistula percutaneously. Further research is needed to provide a higher level of evidence for the primary approach to this type of pathology.

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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