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A rare case of Vieussens ring fistula to pulmonary artery finding on cardiac magnetic resonance: A Case report and Literature Analysis.

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Abstract
A 61-year-old woman presents with functional decline due to dyspnea and angina. Cardiac Magnetic Resonance imaging reveals a Vieussens' arterial ring (VAR) with a fistula from the coronary artery to the main pulmonary artery trunk.

Keywords: Cardiac magnetic resonance; Coronary artery fistula; Vieussens' arterial ring; Cardiovascular imaging

Abbreviations: VAR, Vieussens' arterial ring; CAD, coronary artery disease; CMR, cardiac magnetic resonance; CAF, coronary artery fistula; PET, positron emission tomography; SPECT, single photon emission computed tomography; NMR, nuclear magnetic resonance; CCTA, coronary computed tomography angiography.

Introduction

Vieussens' arterial ring (VAR) is defined as an anastomosis between the cone branch of the right coronary artery and the left anterior descending coronary artery. This malformation persists from an embryonic conotruncal circle, rarely accompanied by a secondary arterial fistula where Vieussens' ring connects to the main pulmonary artery. Simultaneous presence of both defects is found in 14% of all reported coronary anomalies, affecting approximately 0.002% of the general population **(1)**. This collateral pathway provides a network directing blood flow to either the right or left arterial system, depending on flow demand, which is beneficial in coronary artery disease (CAD) **(1,3)**.

A coronary artery fistula (CAF) is an abnormal vascular communication with a cardiac chamber or any part of the circulation. Over 90% of CAFs are congenital, representing about 0.3% of congenital heart diseases. Symptomatology ranges from asymptomatic patients to acute coronary syndromes and heart failure. Approximately 15-30% of all CAFs are coronary-to-pulmonary artery fistulas, with increasing incidence due to widespread cardiovascular imaging use **(2)**.

Patients' average age is 56 years. Clinical course depends on fistula size; symptoms arise from hemodynamic changes secondary to pathology, clinically manifesting as chest discomfort, angina, heart failure, myocardial ischemia, dyspnea, and can also be asymptomatic **(1)**.

Several VAR cases are described, yet VAR combined with a pulmonary artery fistula is exceptional. Symptomatology varies, necessitating cardiovascular imaging for patient management. We analyze a case of VAR associated with a pulmonary artery fistula, confirmed by Cardiac Magnetic Resonance (CMR).

Presentation case

A 61-year-old woman with longstanding systemic hypertension and type 2 diabetes mellitus presents reduced functional capacity and angina on moderate exertion progressing to minimal effort. Physical examination is unremarkable. Electrocardiograms and cardiac markers are normal. Tomography locates atrioventricular and arterial ventricular connections, confirmed by CMR showing collateral vessel emergence from the proximal segment of the left anterior descending coronary artery (Vieussens' ring) connecting with the right coronary artery, specifically the pre-pulmonary pathway connecting to the anterior descending artery, generating fistulas in the pulmonary artery trunk **(Figure 1)**.

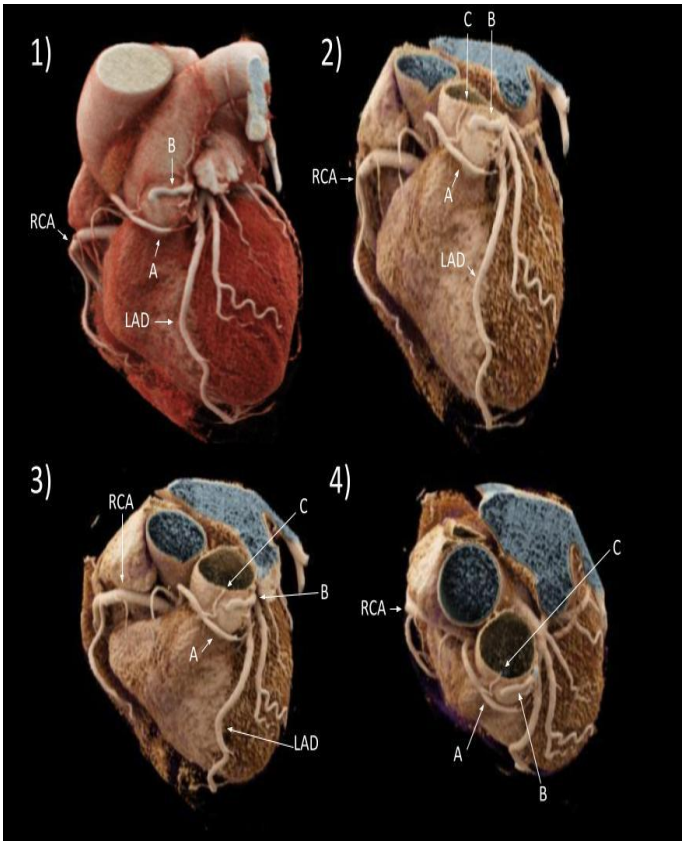

Figure 1: Cardiac CT 3D Volumetric reconstruction. 1, 2, 3) Frontal view; 4) Superior view.
A, Vieussens' arterial ring; B, coronary artery fistula; C, Perforating branch of the fistulous duct; RCA, Right Coronary artery; LAD, Left anterior descending

Discussion

Currently lacking an official classification, literature proposes categorization based on anatomical findings to standardize diagnosis and optimize treatment strategies. Christodoulou et al. suggest six VAR types based on conal artery origin and terminal segment location **(1)**. Similarly, Dogan et al. categorize VAR variants into three types considering collateral vessel origin and acquired secondary pathologies such as fistulas or aneurysms. In a study including 3443 symptomatic or suspected ischemia patients, 11 VAR cases were identified using coronary angiotomography, with 8 presenting as typical cases without additional anomalies (Type 1A), and no reported cases directly associated with fistula (Type 1B) **(3)**. Accordingly, our patient would classify as Type 1B per Dogan's scale.

Cardiovascular imaging aids early diagnosis of coronary anomalies; approximately 48% of the population has this condition, but detection via cardiovascular imaging remains low **(3)**. Initial approaches may include echocardiography **(4)**, but coronary angiography remains primary for evaluating coronary anatomy. Multidetector CT complements with detailed descriptions of course, dimensions, drainage site, depth, and anatomical relationships **(1)**. Nuclear medicine like PET or SPECT is essential for identifying ischemic areas in fistula patients, stratifying risk, planning intervention, and forecasting patient outcomes **(5)**. Cardiac MRI provides superior anatomical resolution and soft tissue characterization. Though not widely used for coronary anomaly diagnosis, multi-imaging approaches have shown MRI's utility in collateral pathway evaluation **(6)**.

Asymptomatic CAF treatment in adults without significant hemodynamic changes is controversial. Surgical and interventional procedures should be considered. Percutaneous transcatheter closure is suggested for mild fistula distortion, proximal origin, single drainage site, absence of multiple fistulas, and no other cardiovascular malformations **(2,7)**. Surgery suits large shunts, multiple fistulas, tortuous expansion, aneurysms, or patients unfit for interventional therapy. Surgical techniques include sole surgical ligation, coronary aneurysm resection, and surgical ligation with coronary artery bypass grafting **(8)**. Coronary computed tomography angiography (CCTA) plays a crucial role in diagnosis, treatment planning, and follow-up **(9)**, preferred for CAF diagnosis and monitoring **(8)**.

Conflicts of Interest

The authors declare they have no competing interests.

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Conclusion

This case highlights cardiovascular imaging's utility in initial coronary anomaly assessment, enabling optimal evaluation and personalized clinical intervention for patients.

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