

Case Report

Vieussens' arterial ring to main pulmonary artery fistula in patient with chest pain

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ABSTRACT

We report a rare case of Vieussens' arterial ring to main pulmonary artery fistula in patient with chest pain who underwent computed tomography (CT) coronary angiography. This case highlights a rare type of coronary-pulmonary artery fistula and role of CT angiography in detecting such anomalies which carry important hemodynamic implications. It is imperative to treat such complicated cardiovascular anomalies to avoid possible long-term clinical consequences.

Keywords: Computed tomography angiography, Coronary artery fistulae, Angina, Vieussens' arterial ring

INTRODUCTION

Vieussens' arterial ring (VAR) is a collateral pathway between the left anterior descending (LAD) and right coronary arteries (RCA) which often acts as lifesaving pathway in cases of obstructive coronary artery disease maintaining adequate myocardial perfusion. This channel undergoes dilatation when a critical occlusion is present in proximal LAD or RCA providing myocardial blood flow. Coronary artery fistulae (CAF) are rare congenital or acquired connections between the coronary arteries and the cardiovascular structures, such as the cardiac chambers, superior or inferior vena cava, the pulmonary arteries or its branches, or the pulmonary veins. CAF has important hemodynamic implications as it would lead to left to right shunt leading to increased right heart preload and coronary steal. Both these pathophysiological processes may lead to heart failure, or death. Treatment of CPAF is mandated in patients with signs of myocardial ischemia using endovascular or surgical techniques depending on type of fistula (simple/complex), location of fistula (proximal/distal) and number of drainage sites. We report a rare case of CAF with arterial feeders from VAR making it an exceedingly rare entity detected in an elderly male undergoing coronary CT angiography for evaluation of coronary arteries in view of chest pain.

CASE REPORT

A 62-year-old male with stable chest pain was referred for computed tomography angiography (CTA) for evaluation of coronary arteries. His baseline electrocardiogram and echocardiography were unremarkable. CTA demonstrated presence of dilated serpentine vascular channels anterior to the right ventricular outflow tract connecting the Vieussens' arterial ring (VAR) to main pulmonary artery with small additional contributing vessels suggesting complex type of coronary artery to pulmonary artery fistula (CPAF) [Figures 1-3]. There was no obstructive coronary artery disease.

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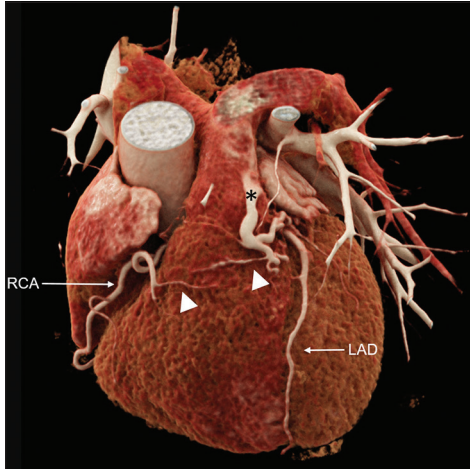


Figure 1: Cinematic 3D rendering of heart and coronary vessels shows fistula to the main pulmonary artery (*) with Vieussens' ring branches (white arrowheads) communicating with fistula. LAD: Left anterior descending artery, RCA: Right coronary artery



Figure 3: Supplementary image axial oblique computed tomography angiography showing tortuous Vieussens' ring.

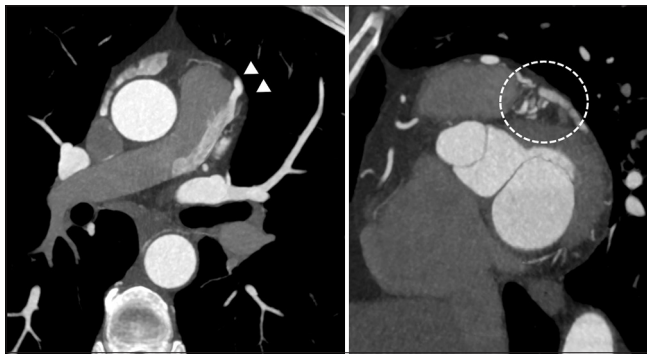


Figure 2: Maximum intensity projection reconstructions of the coronary computed tomography angiography show the site of fistula to main pulmonary artery (white arrowheads) and multiple fistulous vessels around the left anterior descending artery (dashed circle).

DISCUSSION

VAR is a collateral pathway between the left anterior descending (LAD) and right coronary arteries (RCA) seen in up to 40–50% population.^[1] VAR is an embryonic conotruncal collateral that often acts as a lifesaving pathway in cases of obstructive coronary artery disease maintaining adequate myocardial perfusion. This channel undergoes dilatation when a critical occlusion is present in proximal LAD or RCA and provides a low resistance pathway to blood flow.^[2] Coronary artery fistulae are rare congenital or acquired connections between the coronary arteries and the cardiovascular structures, such as the cardiac chambers,

superior or inferior vena cava, the pulmonary arteries or its branches, or the pulmonary veins with a reported incidence of 0.1–0.2% in all patients undergoing coronary imaging.^[3] CPAF is a very rare entity with the most common presenting symptoms being dyspnea and chest pain. In a systematic review of 105 patients of CPAF published by Verdini *et al.*, the most common feeder artery was LAD in 84% cases and RCA in 38% cases.^[4] CPAF with arterial feeders from VAR is an extremely rare entity with only a few case reports described in literature. Hirzallah *et al.*^[5] reported case of a complex CPAF through an aneurysmal VAR. On cross-sectional imaging such as CTA, the CPAF fistulous channels appear as fusiform dilated channels with single/multiple ramifications and are seen communicating with main pulmonary artery. However, majority of CPAF are asymptomatic, which may result in left to right shunting, with larger shunts predisposing the myocardium to ischemia, arrhythmia, and dysfunction secondary to coronary steal phenomenon.^[6] There is no clear consensus regarding treatment strategies for CPAF by the American College of Cardiology/American Heart Association; however, generally simple fistulas arising from proximal coronary bed are preferred for endovascular embolization (coil or plug) whereas complex fistulas with multiple ramifications often require surgical closure.^[7]

CONCLUSION

CPAF can have important hemodynamic implications as it would lead to left to right shunt leading to increased right heart preload and coronary steal. Both these pathophysiological processes may lead to heart failure or death. Treatment of CPAF is mandated in patients with signs of myocardial ischemia using endovascular or surgical

techniques depending on the type of fistula (simple/complex), location of fistula (proximal/distal), and number of drainage sites. Our case highlights that the role of CTA in evaluation of such patients where detection of such important complex connections is important to understand anatomy and plan the treatment, either endovascular or surgical based on imaging findings.

TEACHING POINTS

1. Vieussens' arterial ring is an embryonic conotruncal collateral arcade formed between the conus branch of the right coronary artery and the left anterior descending artery in epicardial plane anterior to the right ventricular outflow tract, providing lifesaving blood flow pathway in cases of obstructive coronary artery disease.
2. Coronary to pulmonary artery fistulae are rare congenital or acquired connections between the coronary arteries and the cardiovascular structures, such as the cardiac chambers, superior or inferior vena cava, the pulmonary arteries or its branches, or the pulmonary veins leading to shunting of blood. Vieussens' arterial ring to pulmonary artery fistula is an exceedingly rare entity with left to right shunting of blood.

MCQs

1. Vieussens' arterial ring is a collateral pathway between which of following arteries?
 - a. Right coronary to left anterior descending artery
 - b. Left anterior descending to left circumflex artery
 - c. Ramus intermedius to sinoatrial node artery
 - d. Posterior descending artery to conal artery

Answer key: a

2. A symptomatic patient with coronary artery to pulmonary artery fistula will most likely present with which of following symptoms?
 - a. Pulmonary artery hypertension
 - b. Myocardial ischemia secondary to coronary steal phenomenon
 - c. Stroke
 - d. Pulmonary thromboembolism

Answer key: b

3. In clinical setting, for evaluating a patient with atypical chest pain, which of the following imaging modalities is most helpful?
 - a. MRI Angiography
 - b. CT Coronary angiography
 - c. IVUS-Intravascular ultrasound
 - d. Manometry

Answer key: b

Ethical approval

The Institutional Review Board has waived the ethical approval for this study.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

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