VIEUSSENS'S RING AND NONISCHEMIC CARDIOMYOPATHY: WHEN THE HEART STEALS FROM ITSELF

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INTRODUCTION

- Vieussens' arterial ring (VAR) is a rare embryonic remnant that represents a congenital collateral pathway between the conus branch of the right coronary artery (or a separate aortic ostium) and the proximal left anterior descending artery (LAD).
- It was first described by Raymond de Vieussens in 1706 and is normally obliterated during fetal development.
- When persistent, VAR is usually benign and hemodynamically insignificant under normal conditions because of balanced pressures in both coronary systems.
- However, when complicated by aneurysm formation or fistulization (Dogan Type 1B), VAR can produce a coronary steal phenomenon, leading to chronic myocardial hypoperfusion and progressive heart failure despite minimal or absent atherosclerotic disease.

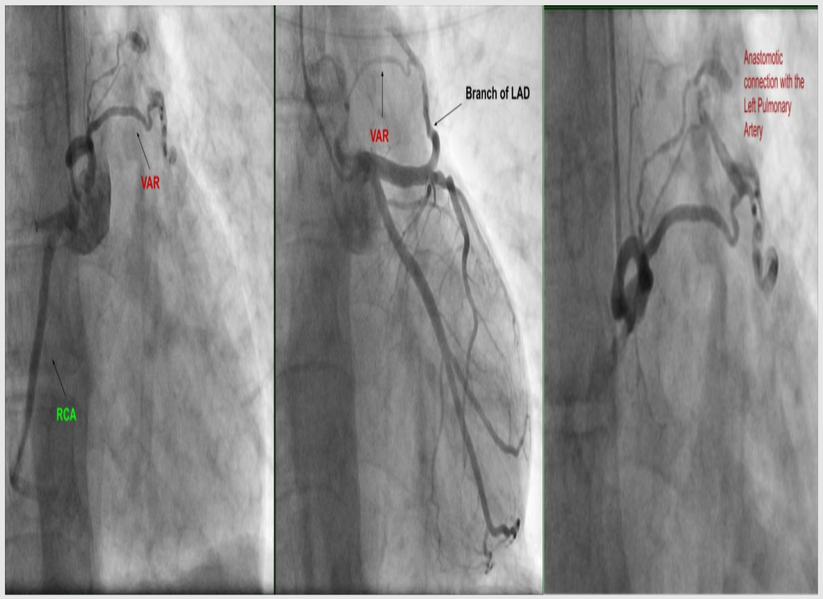

Image 1: Coronary angiography showing Type 1B Vieussens' arterial ring (VAR) and coronary-pulmonary artery fistula

CASE PRESENTATION

• A 62-year-old male with a past medical history of hypertension, hyperlipidemia, COPD, and prior systolic heart failure (LVEF 40–45% documented in 2022) presented in October 2025 with fever, cough, generalized weakness, and progressive dyspnea.

• On admission he was noted to have tachycardia (heart rate 128 bpm), borderline hypotension (blood pressure 107/70 mmHg), and hypoxia (SpO_2 91% on room air).

• **Laboratory evaluation** revealed marked leukocytosis (WBC 30.6 × 10^9/L), elevated lactate (2.3 mmol/L), elevated BNP (521 pg/mL), and mild troponin elevation (27–32 ng/L).

• Electrocardiogram demonstrated sinus tachycardia with inferolateral ST-segment depression and frequent unifocal premature ventricular contractions.

• **Chest CT angiography** confirmed multilobar pneumonia as the cause of severe sepsis; incidentally, it revealed a vessel originating from the anterior aorta just superior and medial to the right coronary artery that coursed in a serpiginous manner anterior to the main pulmonary artery and communicated with a branch of the left anterior descending artery as a 1.4 × 1 cm aneurysmal fistula — consistent with Vieussens' arterial ring.

• **Transthoracic echocardiogram** showed a reduced left ventricular ejection fraction of 36% with global hypokinesis.

• **Left-heart catheterization** demonstrated only minimal coronary artery disease and revealed a tortuous Vieussens' arterial ring connecting the conus branch of the right coronary artery to the left anterior descending artery, with drainage into the left pulmonary artery — confirming a Dogan Type 1B VAR with coronary-pulmonary artery fistula.

• **Cardiac MRI** showed no late gadolinium enhancement and no stress perfusion defects, thereby confirming nonischemic cardiomyopathy secondary to coronary steal.

DISCUSSION

- The fistulous connection in this Type 1B Vieussens' arterial ring allows blood to flow abnormally from the higher-pressure coronary system into the low-pressure pulmonary trunk, resulting in chronically reduced perfusion to the LAD territory — a classic coronary steal phenomenon.
- Over time, this persistent steal led to myocardial dysfunction, adverse remodeling, and a progressive decline in left ventricular ejection fraction despite widely patent epicardial coronary arteries.
- During the patient's sepsis, the induced tachycardia markedly shortened diastolic filling time, further worsened the coronary steal, and produced dynamic subendocardial ischemia that manifested as inferolateral ST depression on ECG.
- These ischemic changes were fully reversible once heart rate control was achieved and sepsis resolved.
- The patient was successfully treated for sepsis and restarted on guideline-directed medical therapy consisting of sacubitril-valsartan 24/26 mg twice daily, metoprolol succinate 25 mg daily, dapagliflozin 10 mg daily, and spironolactone, with plans for elective surgical ligation or coil embolization of the fistula.

CONCLUSION

• Vieussens' arterial ring with fistulization (Dogan Type 1B) is a rare but treatable cause of progressive nonischemic cardiomyopathy. It should be considered in the differential diagnosis for patients who present with unexplained left ventricular dysfunction despite minimal coronary artery disease on angiography.

• Early recognition with multimodality imaging (CTA, echocardiography, cardiac catheterization, and MRI) followed by timely intervention can prevent further deterioration and improve long-term cardiac outcomes.