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CLINICAL IMAGE

Congenital Aortopulmonary Fistula Presenting with Chest Pain in an Adult: Diagnostic and Interventional Approach

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Congenital aortopulmonary fistula (APF) is a rare abnormal communication between the ascending aorta and the pulmonary artery.[1,2] Its presentation in adulthood is uncommon and may mimic ischemic syndromes, although true myocardial ischemia is typically absent.[1,3] We present the case of an adult patient with recurrent chest pain due to congenital APF that was successfully treated with percutaneous coil embolization.

A 53-year-old man presented with a 1-year history of exertional dyspnea and several weeks of typical angina at rest. His medical history included type 2 diabetes mellitus, hyperlipidemia, and transcatheter coil embolization performed in 2020 for a coronary–pulmonary artery fistula originating from a diagonal branch of the left anterior descending artery.

On admission, electrocardiography (ECG) revealed sinus rhythm without ischemic changes, and echocardiography showed preserved left ventricular function with an ejection fraction of 60% and no valvular abnormalities. The estimated pulmonary artery systolic pressure was within normal limits, with no echocardiographic evidence of pulmonary hypertension. Coronary angiography demonstrated no obstructive stenosis but revealed opacification of the main pulmonary artery from the ascending aorta, which was diagnostic of an APF (Figure 1A). Modified computed tomography (CT) angiography demonstrated the previously implanted coil and the apparent communication between the aorta and the pulmonary artery (Figures 1B and C).

Given the presence of typical angina, coronary fractional flow reserve and myocardial perfusion scintigraphy were performed, both of which were negative for ischemia. After a multidisciplinary heart team discussion, percutaneous coil closure of the fistula was undertaken. Complete angiographic occlusion was achieved without complications (Figure 1D). The patient experienced prompt and sustained relief of chest pain and exertional dyspnea at follow-up.

Congenital APFs are exceptionally rare and often remain undetected until adulthood.[2,3] Most cases are asymptomatic or present with dyspnea or cardiac murmurs; presentation with typical angina is unusual. Given the recent onset of angina at rest, alternative causes of chest pain were systematically investigated. Coronary angiography demonstrated normal epicardial coronary arteries without obstructive disease. Acute coronary syndrome, myocarditis, and valvular pathology were excluded using ECG, cardiac biomarkers, echocardiography, and functional ischemia testing. Therefore, the patient's symptoms were attributed to the hemodynamic and perfusion-related consequences of the APF.

In this patient, the mechanism of pain was likely multifactorial and may have included:

1. Preload-driven myocardial oxygen supply–demand imbalance resulting from a chronic left-to-right shunt that increased ventricular wall stress;
2. Diastolic run-off from the aorta into the pulmonary artery, transiently reducing coronary perfusion pressure;
3. Perivascular nociceptor stimulation secondary to vessel wall stretch;
4. Microvascular ischemia not detectable by conventional fractional flow reserve or perfusion studies.

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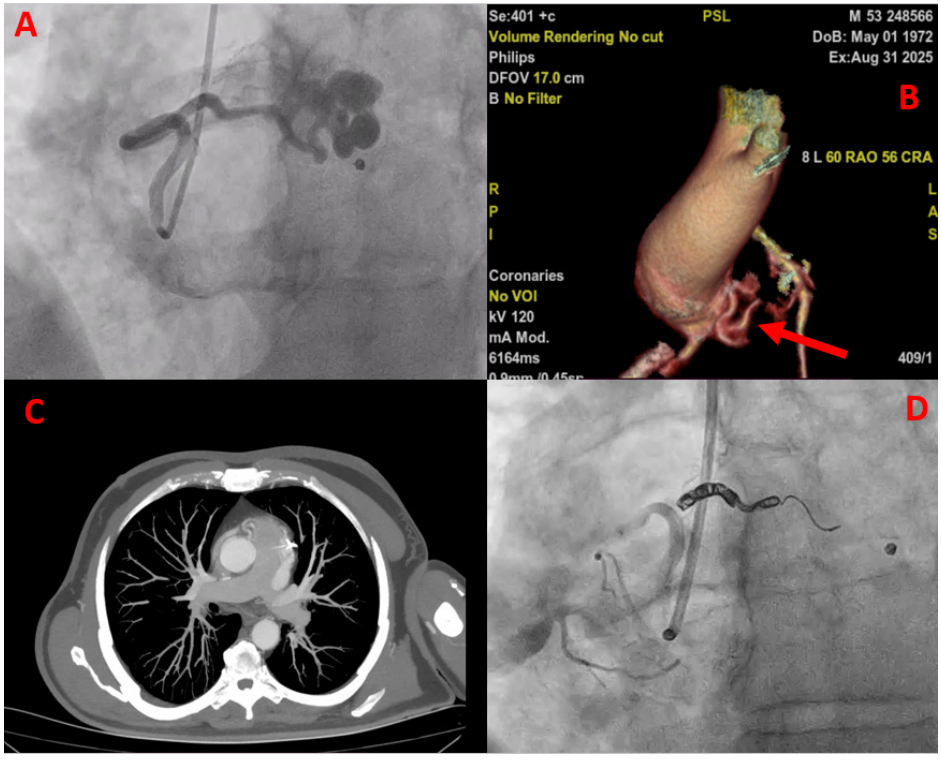

Figure 1. A) Aortopulmonary fistula detected by coronary angiography. B) Three-dimensional computed tomography (CT) angiography demonstrating the origin and course of the aortopulmonary fistula (arrows). C) Two-dimensional CT angiography showing the connection of the fistula to the main pulmonary artery (arrow). D) Angiographic image after percutaneous coil embolization demonstrating complete occlusion of the fistula

These mechanisms may explain the paradox of ischemic-type chest pain despite normal coronary arteries and negative functional ischemia testing.[1,2] Non-invasive assessment suggested the absence of a hemodynamically significant left-to-right shunt; therefore, Qp/Qs measurement by right heart catheterization was not considered necessary.

Multimodality imaging is essential for diagnosis and procedural planning. CT angiography accurately delineates the aortopulmonary connection, defines the fistulous tract, and guides device sizing. Invasive angiography confirms the anatomical features, allows shunt quantification, and enables closure during the same procedure. Functional testing (fractional flow reserve and perfusion imaging) remains useful for excluding concomitant coronary artery disease.[2]

For anatomically suitable fistulas, transcatheter closure is the preferred treatment strategy, offering high procedural success rates, rapid recovery, and durable symptom relief.[2,4,5] Surgical correction is reserved for complex, large, or anatomically unsuitable fistulas. In our case, coil embolization resulted in complete angiographic closure and full symptomatic improvement, highlighting the safety and efficacy of this approach in carefully selected patients.

APF is a rare but important cause of angina in adults with normal coronary arteries. A systematic diagnostic approach integrating multimodality imaging and selective functional testing is essential for identifying the underlying mechanism. Percutaneous transcatheter coil occlusion provides effective and minimally invasive treatment with excellent outcomes in appropriately selected patients.[2,4]

Informed Consent: Written informed consent was obtained from the patient for the publication of this case report and the accompanying images.

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