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Learning Objectives

- Consider undiagnosed congenital heart disease in the differential diagnosis of an adult patient presenting with heart failure

Case

HPI: A 58-year-old woman presented with palpitations & shortness of breath, & reports experiencing progressive dyspnea on exertion, reduced exercise tolerance and mild lower extremity edema in the past year. Patient denies chest pain, dizziness, or syncope.

PMH: Rheumatoid arthritis, well-controlled on methotrexate, von Willebrand disease, & paroxysmal atrial fibrillation, on beta blocker therapy

PE: notable for a grade III/VI continuous murmur along the LSB & trace b/l pitting edema to the ankles

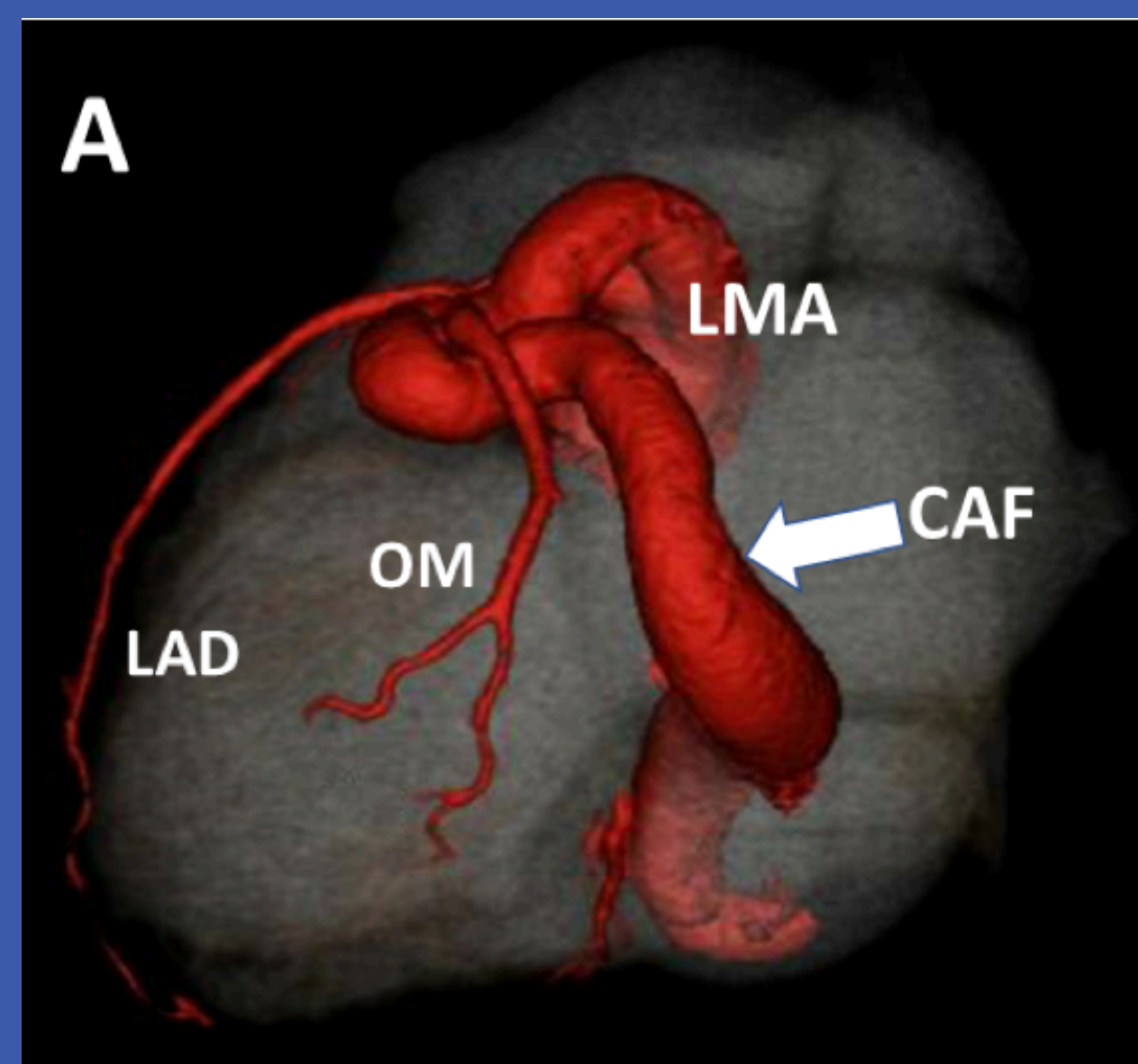


Figure A. Coronary CTA

Epidemiology:

- CAFs occur equally between men and women, irrespective of ethnicity, and account for approximately 0.002% of new cases of coronary anomalies annually in both the pediatric and adult populations.
- Single fistulas account for 90% of all CAFs, with 75% of incidentally found CAFs being clinically silent, especially during childhood.

Diagnostic Work Up

- **Gold standard:** Selective coronary angiography
- Alternative modalities include CCTA & TEE with Doppler
- Newer technologies include multidetector CT & cardiac MRI, which provide great detail on fistula course & drainage
- Evolving tools, such as fractional flow reserve, instant wave-free ratio, & intravascular ultrasound, help to assess vessel morphology, integrity, & the presence of thrombi

Implications

- Data is limited regarding long-term outcome and optimal medical management post-closure, but post-closure myocardial infarction remains a concern.

Discussion

Treatment

- The decision process in managing fistulae depends on site of origin of the fistula (proximal vs distal), size of the fistula, patient's symptoms, presence of any complication caused by the fistula (eg, angina, heart failure, endocarditis, rupture, etc), age of the patient, the anatomy of the fistula, & presence of other indications to undergo an invasive procedure.
- The class 1 recommendations by the AHA/ACC include:
 - Patients with continuous murmur should undergo exact delineation of the origin and termination of the fistula by either echocardiography or computed tomography/magnetic resonance imaging.
 - Patients with large fistulae should undergo closure (surgical or percutaneous) after delineation of the exact anatomy.
 - Patients with small to moderate fistulae with complications (eg, ischemia, arrhythmias, or ventricular dysfunction of unexplained etiology) should undergo fistulae closure.

Post-op Therapy

- After transcatheter closure of a large symptomatic fistula, heparin should be started six hours after the procedure and adjusted to keep the partial thromboplastin time at 1.5 times normal while warfarin is started.
- Patients should be discharged home on warfarin & be adjusted to keep the international normalized ratio (INR) around 2.5 for a period of 6 to 12 months .
- Patients should receive antiplatelets indefinitely.

The patient underwent successful transcatheter device CAF closure, with symptom resolution, normalization of pulmonary artery pressures, & improvement in her four chamber dilatation at 3 months. However, four months post CAF closure, the patient re-presented with acute chest pain & ischemic ECG changes with a peak troponin-I level of 30 ng/mL consistent with a NSTEMI. The ischemic event was thought to be thromboembolic with showering of microemboli into the distal LCX circulation. Clopidogrel & atorvastatin were added to her regimen. At present, she is asymptomatic & has returned to an active lifestyle.