CASE OF THE ISSUE

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Anomalous origin of Right Coronary Artery from Pulmonary Artery (ARCAPA) in an Adult Woman Presenting with Atypical Chest Pain

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CLINICAL PRESENTATION

A 37-year-old woman presented with occasional chest pain on exertion. Physical examination and ECG were normal. Transthoracic echocardiography demonstrated dilated, tortuous coronary arteries, and the right coronary artery (RCA) origin could not be identified. The patient was then referred for coronary CT angiography (CTA).

IMAGING FINDINGS

Retrospective ECG-gated Coronary CTA was performed on a 512-slice scanner (GE HealthCare), which demonstrated the following abnormalities:

Anomalous origin of the RCA from the main pulmonary artery (Figures 1 and 2).

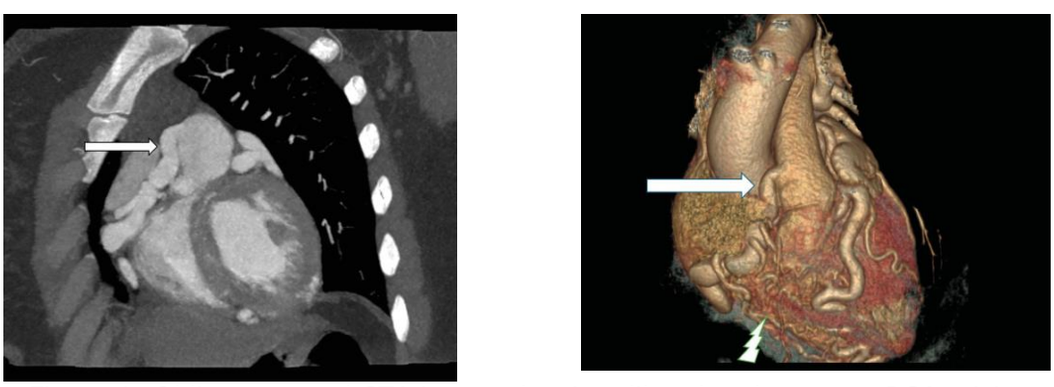

Figure 1: Oblique coronal CTA image showing dilated and tortuous RCA arising from the pulmonary artery

- Dilated and tortuous left coronary artery (LCA), RCA and left anterior descending artery (LAD) (Figure 2)
- Prominent intercoronary collaterals, predominantly between RCA and LAD (Figure 2)

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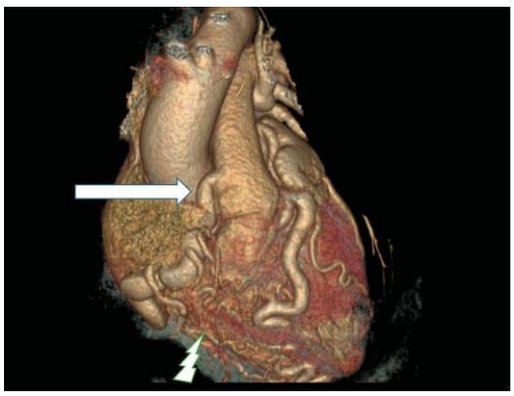

Figure 2: 3D-Volume rendering technique (VRT) image showing anomalous origin of RCA from pulmonary artery (straight arrow) with intercoronary collaterals between RCA and LAD (Zigzag arrow)

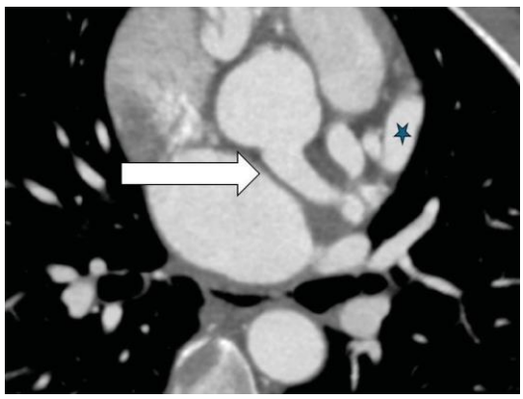

Figure 3: CTA axial image showing normal origin of LCA from left coronary sinus (white arrow). Dilatation and tortuosity of RCA and LAD (shown by stars)

DISCUSSION

ARCAPA is an extremely rare congenital coronary anomaly in which the RCA arises from the pulmonary artery rather than the aorta. The estimated prevalence is less than 0.002% in the general population. Unlike its more common counterpart, i.e., anomalous origin of the left coronary artery from the pulmonary artery (ALCAPA), in which patients are symptomatic from an early age, patients with ARCAPA are often asymptomatic and are detected incidentally or later in life when the abnormality is isolated. This is because of the formation of collaterals between the left and right coronary arteries, which masks the hemodynamic effects. However, it can also result in "coronary steal," with increased flow in the LCA, retrograde flow from collaterals into the RCA and then into the pulmonary artery, and patients may present with features of ischemia and heart failure.[1,2,3]

Noninvasive modalities like transthoracic echocardiography (TTE) and coronary CT angiography (CTA) are useful in demonstrating anatomical as well as physiological abnormalities. TTE is a useful first-line exam; however, coronary CTA is considered the gold standard in the assessment of coronary abnormalities. In our case, TTE showed the presence of tortuous coronaries, and the diagnosis was made following coronary CTA.[2,4]

Surgical reimplantation of the RCA into the aorta is recommended even in asymptomatic cases to prevent the long-term complications of heart failure or sudden cardiac death.[3]

TEACHING POINTS:

- Dilated coronaries with non visualization of RCA origin in TTE should raise suspicion for ARCAPA.
- Normal origin of LCA from left coronary sinus (Figure 3)
- Coronary CTA confirms pulmonary origin of RCA and shows intercoronary collaterals..
- Extensive collaterals allow survival into adulthood.
- Surgical correction is advised even in asymptomatic or minimally symptomatic patients.

CONFLICT OF INTEREST

None

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