Abstract: P1488

**Anomalous origin of left coronary artery from right pulmonary artery in association with scimitar syndrome**

**Authors:**
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**Topic(s):**
Imaging: Congenital Heart Disease

**Citation:**
Clinical Presentation: A full-term neonate was referred to our institution because of respiratory distress. CXR was significant for right lung hypoplasia and mild cardiomegaly. ECG showed normal sinus rhythm, right atrial enlargement, and right ventricular hypertrophy with no signs of ischemia.

**Imaging Findings:**
The initial echocardiogram demonstrated PAPVD with the right upper pulmonary vein draining into IVC/RA junction with flow acceleration (mean gradient= 7 mmHg), moderate ASD, small muscular VSD with left-right shunting, moderate PDA with bidirectional shunting. Forward flow was seen in the proximal part of left main coronary artery (LMCA). RV systolic pressure was supra-systemic with a qualitatively moderately reduced RV systolic function.

The patient was taken to the catheterization lab where MPA angiography revealed an antegrade flow from the RPA into LMCA supplying both the anterior descending and the circumflex arteries. A selective injection within the scimitar vein showed drainage of the right lung into a vertical vein connecting with stenosis to IVC.

A follow up echocardiogram to re-examine the coronary origin revealed an anomalous origin of LMCA from proximal RPA; 3 mm distal to branch pulmonary artery bifurcation with mainly antegrade low velocity flow into LMCA and LAD. (Image 1)

**Role of Imaging in Patient Care:**
- Imaging of the coronary origin in patients with ALCAPA can be challenging especially if the LMCA originates from RPA. Also, the presence of pulmonary hypertension might contribute to maintain coronary perfusion and lead to misinterpretation of the antegrade flow in LMCA and its branches.
- In certain situations, cardiac catheterization is essential to make the diagnosis of ALCAPA which prevented a potentially catastrophic outcome. Catheter intervention with a series of balloon dilations of the stenotic scimitar vein was successful in relieving the stenosis.

**Summary/Discussion Points:**
- Extensive review of the available literature revealed only three cases of Scimitar syndrome associated with ALCAPA. In all of these cases, the LMCA originated from the posterior sinus of MPA. Our case is the first to report ALCAPA from RPA in association with Scimitar syndrome. This presentation might have led to the initial misinterpretation of the echocardiography images.
- The presence of pulmonary hypertension in our patient maintained an adequate antegrade flow across the LMCA preventing significant coronary steal and signs of myocardial ischemia.
- The report highlights the challenges in making the diagnosis of ALCAPA with echocardiograms. Moreover, we discuss the role of cross-sectional and invasive imaging to rule out potential coronary arteries anomalies in patients with Scimitar syndrome, as this a rare although a very significant association that may have important implications in their outcomes.
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