Anomalous Pulmonary Artery Membrane: A Rare Membrane Obstructing Right Pulmonary Artery

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I. Introduction:

Several types of anomalous bands in various chambers of heart have been reported and some are clinically significant. [1-5] Here we report a 3 years female child having a murmur and ECHO Revealing presence of moderate sized ostium secundum atrial septal defect (ASD) with rare anomalous right pulmonary artery (RPA) orifice. She underwent surgical resection of membrane and ASD closure.

II. Clinical Presentation:

A 3 years old female child presented with cough, cold and failure to thrive. CVS revealed grade 2/6 ejection systolic murmur at left upper sternal border, conducted to the right mid chest. PA stenosis was clinically impressed. ECG revealed rsR’ pattern in V1 lead. ECHO (Parasternal short axis view) showed a long membranous measuring 9 mm, originating at the PA confluence, extending to RPA orifice (5.3 mm) and obstructing at the origin of right pulmonary flow with a gradient of 66 mmHg. Proximal right and left PA were measured as 7.7 mm and 8.8 mm respectively. Subxiphoid bicaval view of transthoracic ECHO showed high type moderate size ostium secundum ASD measured as 9-10 mm in width with left to right shunt. It was a borderline divisible septal defect. CTVS team advised for surgical intervention in view of ASD in association of RPA stenosis caused by membrane obstructing at the orifice of RPA. Resection of obstructing membrane and plasty had been done at the proximal portion of RPA along with the ASD closure.

III. Discussion:

The filamentous membranous structure can be visualized rarely by ECHO at the confluence or at the region of angle of bifurcation of PA at various lengths, which is usually benign in nature. Rarely it can be longer and may obstruct the orifice of RPA. The need for surgical intervention depends upon the gradient across the RPA which in our case was 66 mmHg producing unilateral PA hypertension and right ventricular dysfunction. Relieving the stenotic gradient at the PA was necessary to prevent other complications.
IV. Conclusion:

Different types of anomalous bands, membranes, tendons and venous valves have been described in the chambers of heart, but not within the pulmonary artery bifurcation level. This membrane can be seen normally at various lengths without any obstruction. But when it becomes longer, may obstruct the orifice of pulmonary artery which can be well demonstrated by ECHO and needs surgical intervention.

References:
