Abstract: 1648

Partial anomalous pulmonary venous connection - a rare diagnosis in an asymptomatic adult

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Introduction: Partial anomalous pulmonary venous connection (PAPVC) is a rare congenital heart defect whose clinical presentation depends on the degree of left-to-right shunting and the presence of other associated cardiac defects. Surgery is recommended for symptomatic patients and those with pulmonary arterial hypertension (PAH). We present a case of an asymptomatic adult incidentally diagnosed with PAPVC.

Case Report: A 64 years-old woman with no relevant past medical history presented to her primary care physician complaining of palpitations. Her outpatient workup included: EKG revealing a dominant R wave in V1 and a right ventricular strain pattern; a normal Holter monitor, a chest X-ray showing right atrial (RA) and ventricular enlargement and a transthoracic echocardiogram that raised the possibility of an ASD and confirmed right heart volume overload. She was subsequently referred to our institution for assessment of ASD possibility. On consultation, the patient reported intermittent self-limited palpitations, denying angina or dyspnea. A transthoracic echocardiogram was performed, followed by transesophageal echocardiogram, which revealed an intact, but floppy, interatrial septum, right pulmonary veins draining into the inferior vena cava, marked RA and RV enlargement, and an enlarged pulmonary trunk. Cardiac Magnetic Resonance Imaging revealed a markedly dilated RV with a preserved systolic function (RV ejection fraction of 48%), partial anomalous right pulmonary venous return to the inferior vena cava and a hemodynamically significant left-to-right shunt (QP:QS of 2.0). On cardiac catheterization, it was possible to determine a pulmonary blood flow (QP) 7.16L/min, a systemic blood flow (QS) of 3.99L/min, a QP:QS of 1.79, pulmonary vascular resistance 2.3UW/m², a systolic pulmonary artery pressure of 25mmHg and a pulmonary artery occlusion pressure of 8mmHg. Despite being asymptomatic and the absence of pulmonary hypertension, with a QP:QS<2, there was a significant right ventricular dilation imposed by the volume overload, so the patient was referred to cardiac surgery.

Conclusion: PAPVR may be incidentally diagnosed in asymptomatic patients undergoing routine diagnostic imaging, however its correct diagnosis requires a high level of suspicion.