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Iatrogenic aorta-to-right atrial fistula: A rare cause of pulmonary hypertension

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Background: Aorta-to-right atrial fistula is an extremely rare late complication of prosthetic aortic valve replacement. Increased survival and improved patient outcomes following complex aortic valve surgeries poses paramount significance to the recognition of this unusual complication.

Case: An 80-year-old female presented with two month history of exertional dyspnea and reduced exercise tolerance. She underwent 10 months post aortic valve replacement with a pericardial aortic bio-prosthesis and a left internal mammary artery graft to left anterior descending artery, on a background of severe aortic stenosis and ischemic heart disease. Clinical examination demonstrated raised jugular venous pressure and a faint systolic murmur.

Decision-making: Transthoracic echocardiogram revealed a dilated right atrium (RA) and right ventricle (RV), severe tricuspid regurgitation (TR) and a D-shaped septum in diastole suggesting raised RV end-diastolic pressure, possibly pulmonary hypertension. These findings were absent prior to cardiac surgery. Cardiac magnetic resonance imaging (CMR) was performed to evaluate the unexplained TR. CMR raised the possibility of an aortic root to RA shunt, but aortic valve artefact limited complete evaluation. The RV to LV stroke volume ratio of 2:1 was more compatible with a shunt rather than isolated TR. Subsequent trans esophageal echocardiogram (TEE) color Doppler demonstrated a systolic shunt through an echo-free space from the aortic root into the RA, converging with the severe TR jet, with evidence of RA and RV volume overload. The patient was referred back to the cardiothoracic surgeon in view of revision surgery. The prosthetic aortic valve was excised with placement of a new aortic bio-prosthesis and suturing of the fistula, followed by insertion of a tricuspid annuloplasty band. The patient made a good recovery without any complications and reported significant improvement in her symptoms in clinic follow-up at eight weeks.

Conclusion: Denovo cardiac symptoms and evidence of pulmonary hypertension on transthoracic echocardiogram in a patient post aortic valve surgery warrants further investigation with CMR and TEE, considering the possibility of aorta-to-RA fistula.

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