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Double Trouble: Surgical Management and Characterization of Two-Chambered Right Ventricle - Poster Presentation

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Abstract

Introduction

Double-Chambered Right Ventricle (DCRV) is a rare congenital anomaly in which hypertrophied muscular bundles divide the right ventricle into two chambers, producing subvalvular obstruction of the right ventricular outflow tract (RVOT). The resulting pressure gradient between the proximal right ventricle and pulmonary artery can progressively worsen as muscular hypertrophy increases. Although most cases are diagnosed in childhood, delayed adult presentation is uncommon and may mimic other causes of right-sided obstruction such as pulmonic valve stenosis. We report an adult presentation of DCRV highlighting diagnostic evaluation, surgical management, and postoperative hemodynamic improvement.

Methods

Retrospective chart review was performed for the entirety of this report. Informed consent was obtained to present the case. This case report was reviewed by the Mount Sinai Hospital Institutional Review Board, IRB#: MSH-IRB-26-12; Principal investigator Dr. Jonathan Bell, MD.

Case Presentation

A middle-aged woman with a longstanding systolic murmur presented with progressive exertional dyspnea. Transthoracic echocardiography demonstrated a structurally normal pulmonic valve but severe turbulent flow within the RVOT, raising suspicion for muscular obstruction. Right heart catheterization confirmed markedly elevated right ventricular systolic pressure (111 mmHg) with a large RV–pulmonary artery gradient (83 mmHg) and normal pulmonary artery pressures. The patient underwent surgical resection of an obstructing mid-right ventricular muscular band via right ventriculotomy with right ventricular myectomy and bovine pericardial patch augmentation. The resected band measured 3.5 × 2.5 × 0.8 cm and pathology showed hypertrophic myocardial tissue with fibrosis. Postoperatively, right ventricular systolic pressure improved to 39 mmHg and the RVOT gradient decreased to 28 mmHg. The patient recovered without complications and was discharged on postoperative day eight.

Discussion

Adult DCRV is rare and may represent delayed recognition or progressive muscular hypertrophy over time. The diagnostics obtained in this patient case matched and reinforced the proposed pathophysiology of the condition especially well. Multimodal imaging and invasive hemodynamic testing are critical for distinguishing DCRV from other RVOT obstructive conditions. Surgical myectomy remains definitive therapy, with significant symptomatic and hemodynamic improvement when obstruction is relieved.

Conclusions

This case illustrates an uncommon adult presentation of DCRV successfully treated with surgical myectomy and patch augmentation. Early recognition and definitive surgical management can produce substantial hemodynamic improvement and favorable clinical outcomes.