Case study

Isolated Double-chambered Right Ventricle: a rare congenital heart disease.

ABSTRACT

Double-chambered right ventricle (DCRV) is an uncommon congenital heart condition characterized by a progressive obstruction in the right ventricular outflow tract. This obstruction is caused by anomalous muscles or fibrous tissues that divide the right ventricle into two cavities: a proximal high-pressure chamber (anatomically lower) and a distal low-pressure chamber (anatomically higher). We present a case of a middle-aged man with a medical history of recurrent symptomatic dyspnea. Upon presentation, there were no signs of congestive cardiac failure. The diagnosis was confirmed using transthoracic two-dimensional (2-D) echocardiography and Transesophageal echocardiography (TEE) for more precise characterization. The primary objective of this case report is to highlight the rarity of this congenital heart disease, particularly in adults.

Keywords: Double-chambered right ventricle; Adult congenital heart disease ; Right ventricular outflow tract obstruction ; echocardiography

1. INTRODUCTION :

Double-chambered right ventricle (DCRV) is an uncommon congenital heart disease marked by the division of the right ventricular cavity into two chambers by anomalous muscle bundles. Typically, DCRV is diagnosed during childhood ages or adolescent ages, and most DCRV patients have associated congenital anomalies, such as ventricular septal defect (VSD), pulmonary stenosis, and subaortic stenosis. (1) Here, we report a case of an isolated DCRV in an adult patient

2. PRESENTATION OF CASE

We describe the case of a 53-year-old man, with no known personal history of disease or previous diagnosis of a congenital heart disease, who was admitted due to a progressive shortness of breath over the preceding 4 months. Upon examination, his heart rate was 90beats/minute, the blood pressure was 107/57 mm Hg and his respiratory rate was 20 breaths per minute. On physical examination his extremities were well perfused with strong and symmetric pulses with no sign of left or right heart failure. Upon auscultation of his heart, it was noted that he had a systolic ejection murmur at the left parasternal border. He had ECG which displayed evidence of right ventricular hypertrophy

with right axis deviation, indicative of right ventricular overload. He had a chest x-ray which confirmed cardiomegaly with a 0.7 cardiothoracic ratio. Two-dimensional (2D) transthoracic echocardiography was perfomed which revealed a turbulent Doppler color flow jet indicative of a stenotic mid-right ventricle (Figure 1). Continuous-wave Doppler analysis across this turbulent jet exhibited flow acceleration measuring 4.53 m/s, corresponding to a pressure gradient of approximately 82 mmHg, as calculated using the simplified Bernoulli equation. It is important to note that the obstructive gradient might have been underestimated due to the presence of severe tricuspid regurgitation. Additionally, the examination identified right atrial enlargement, right ventricular dilation, moderate dilatation of the pulmonary artery with normal infundibular contraction, and severe tricuspid regurgitation with a dual primitive and hemodynamic component exacerbated by right ventricular mid-ventricular stenosis

Transesophageal echocardiography (TEE) was performed for a better analysis which demonstrated an anomalous muscle bundle dividing the RV into two parts, with turbulent flow jet on color Doppler images

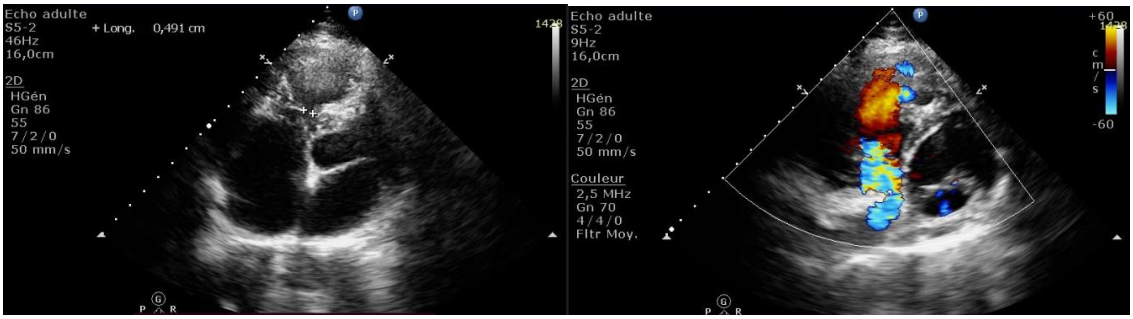

Figure 1: Transthoracic echocardiogram on the parasternal short axis view showed mid-ventricular

[summarize the trans-thoracic echocardiogram findings related to figure 1 as a legend with an arrow pointing out what you want to illustrate]

3. DISCUSSION :

A double-chambered right ventricle (DCRV) is a congenital cardiac anomaly characterized by the division of the right ventricle into two chambers. Anomalous muscles or fibrous tissues within the right ventricular cavity create a proximal high-pressure (anatomically lower) chamber and a distal low-pressure (anatomically higher) chamber. The incidence of DCRV accounts for 0.5%-2% of all cases of congenital heart disease. [1] [2]

The finding of a DCRV is exceptionally rare as an isolated anomaly. It is more commonly associated with a membranous type ventricular septal defect (VSD). Other reported associated lesions include: subaortic stenosis, pulmonary valve stenosis, double outlet right ventricle, tetralogy of Fallot,

anomalous pulmonary venous drainage, complete or corrected transposition of the great arteries, pulmonary atresia with intact ventricular septum and Ebstein's anomaly of the tricuspid valve (1) (3)

The anomalous muscle bundles, vary in number from 1 to many, typically originating from the body of the septal band (septomarginal trabeculation) and traverse the right ventricular chamber to reach the anterior free wall. (4) (5)

A simple classification of the pathology was proposed by Folger, who described two positions of the abnormal muscle bundle: high (or horizontal) and low (or oblique)

Most cases of DCRV are diagnosed and treated during childhood, where as an initial presentation during adulthood has been reported in rare cases [1] [6] [7]

A similar type of case report has been published by Park et al., in which they reported a case of isolated DCRV presenting in an asymptomatic person , (2) One other case report was published by Pansari et al., in which they reported a case of isolated DCRV presenting with congestive heart failure. (8) , another case report was published by Ashok Garg,et al in which they reported a case of isolated DCRV in a 18-year-old male (9) , a rare case report of isolated DCRV with dextrocardia. was published (10)

Presenting symptoms are usually due to severe RVOT obstruction leading to a low cardiac output state or due to associated right heart failure.

Although transthoracic echocardiography (TTE) is widely accepted for demonstrating double-chambered right ventricle (DCRV) in pediatric patients, challenges may arise in studying adults. The implementation of transesophageal echocardiography (TOE) provides better lesion definition in adults. In certain cases, alternative modalities such as magnetic resonance imaging scan or cardiac catheterization may be necessary . (11)

Surgical correction consists of resection of the AMB and closure of the VSD through a transatrial approach to achieve adequate exposure of the AMB. Surgery offers complete relief of the obstruction, and a substantial improvement in symptoms and functional status. The long-term results of surgical treatment are excellent (12)

The timing of surgical repair depends on the presence of associated cardiac anomalies. If there are no other significant coexisting defects,, observation is possible as long as the intracavitary pressure gradient is not more than 40 mmHg and the obstruction is not progressive . (13)

4. CONCLUSION :

Double-chambered right ventricle (DCRV), which leads to obstruction in the right ventricular outflow tract, is an uncommon congenital heart disease, particularly when initially diagnosed in adults. The

diagnosis can be challenging, especially when it manifests with atypical symptoms. The majority of DCRV cases are associated with congenital anomalies, with ventricular septal defect (VSD) or fixed subaortic stenosis being the most common. An isolated DCRV, without associated anomalies, is exceptionally rare.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

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