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Double chambered right ventricle in adulthood: a case report

Erişkinlikte çift odacıklı sağ ventrikül: Olgu sunumu

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Double chambered right ventricle (DCRV) is a rare disorder observed in adulthood and is usually misdiagnosed. The DCRV is a form of right ventricular out flow tract obstruction. In this rare pathology, the right ventricle is divided into two chambers by a muscular band, including high-pressure proximal and low-pressure distal chamber. It develops over time and mostly in patients with an abnormally short distance between the moderator band and pulmonary valve. Most of DCRV patients are diagnosed before 20-years of age. However, despite rarely seen, DCRV may be diagnosed in adults. As it complicates diagnosis, it should be seriously taken into consideration.

Key words: Adulthood; double chambered right ventricle; pulmonary hypertension.

Çift odacıklı sağ ventrikül (DCRV) erişkin dönemde nadiren saptanır ve sıklıkla yanlış tanı konulur. Çift odacıklı sağ ventrikül sağ ventrikül çıkış yolu obstrüksiyonunun bir türüdür. Bu nadir görülen bozuklukta sağ ventrikül musküler bir bant vasıtasıyla yüksek basınçlı proksimal ve düşük basınçlı distal kompartman olmak üzere ikiye ayrılmıştır. Çoğunlukla moderatör bant ve pulmoner kapak arasındaki mesafenin az olduğu hastalarda zaman içinde gelişir. Sıklıkla DCRV hastalarına 20 yaşından önce tanı konulur. Ancak, nadiren görülse de erişkinlerde de DCRV tanısı konulabilir. Bu durumda tanı koymak güçtür ve mutlaka akılda tutulması gerekir.

Anahtar sözcükler: Erişkin çağ; çift odacıklı sağ ventrikül; pulmoner hipertansiyon.

Double-chambered right ventricle (DCRV) is a rare congenital disorder that is usually not observed in adulthood. It makes up 0.5-2% of all congenital heart diseases and is often misdiagnosed. Forster and Humphries[1] reported only one case in 36,000 general autopsies, whereas there was a 1-2.6% incidence of other congenital diseases in their study. The prevalence of DCRV in adults has not yet been studied, and the literature only contains a few cases of this disease that were diagnosed in adulthood.[2] In DCRV, the right ventricle is divided into the high-pressure proximal and low-pressure distal chambers along with two pressure zones by a muscular band, It is considered to be a form of right ventricular (RV) outflow tract obstruction[3] that develops over time, mostly in patients with an abnormally short distance between the moderator band and pulmonary valve. Since most diagnoses occur in patients under 20 years old, diagnosing DCRV in adults can be challenging. Nevertheless, the possibility of its occurrence must be taken seriously if the patient has symptoms which warrant its consideration.

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CASE REPORT

A 45-year-old female patient was admitted to the hospital for progressive dyspnea, fatigue, lower extremity edema, and chest pain. A year earlier she had been diagnosed with primary pulmonary hypertension via echocardiographic evaluation at another cardiology institution and had been on diuretic treatment. She had been suffering from dyspnea for the previous last 10 years, but the severity of the dyspnea and other symptoms had increased in the few months leading up to her admission. On her physical examination, there was no cyanosis, the lungs were clear with auscultation, the heart rate was 76 bpm, and the arterial blood pressure was 120/70 mmHg. At cardiac auscultation, a grade 4/6 harsh systolic murmur was heard at the left sternal border, and peripheral edema was present. The hemoglobin was 12 g/dl, the blood gas analysis was normal, and the blood glucose and electrolyte levels were also within normal ranges.

Her electrocardiography (ECG) findings revealed sinus rhythm, right axis deviation, RV overload, and incomplete right bundle-branch block. Transthoracic echocardiography was repeated to confirm the diagnosis of primary pulmonary hypertension, and it showed RV hypertrophy and a muscular septation inside the RV causing obstruction. The peak gradient was 150 mmHg, and the RV outflow tract and pulmonary valve were normal. However, mild-to-moderate tricuspid regurgitation was found (Figure 1). Right ventriculography was performed, and intracavitary obstruction was observed as well as RV outflow obstruction with a 70 mmHg pressure gradient in the RV (Figure 2). Furthermore, an intraventricular gradient of 120 mmHg was detected. Her pulmonary artery and pulmonary valve were normal upon angiography, and left ventriculography also showed nothing abnormal. Additionally, the results of her coronary angiography were normal except for a small fistula from the right coronary artery (RCA) to the RV.

The patient underwent surgical correction while using transesophageal echocardiography. Her defect was repaired through a right atriotomy and featured a resection of the hypertrophied and fibrotic muscle at the os infundibulum and at additional hypertrophied septal and parietal muscle bands. There were no surgical complications. Postoperative echocardiography demonstrated no residual gradients. After surgery, all the patient's symptoms disappeared, and she was discharged from hospital.

DISCUSSION

Other cardiac anomalies frequently accompany DCRV; therefore, patients with this disease must be examined carefully for the presence of defects such as ventricular septal defects and pulmonary valve stenosis.[4] However, our patient had isolated DCRV which produced 120 mmHg gradients in the RV.

Patients with DCRV usually have symptoms like chest pain, dyspnea, and syncope, and our patient was no exception.[5] Diagnosis of DCRV is usually problematic, even with angiography, but it must be kept in mind for patients with the aforementioned complaints. Screening for DCRV should especially be considered if there is an unexplained etiology regarding pulmonary hypertension or tricuspid regurgitation. Hachiro et al.[6] defined the criteria for diagnosis of DCRV as the following: the demonstration of a

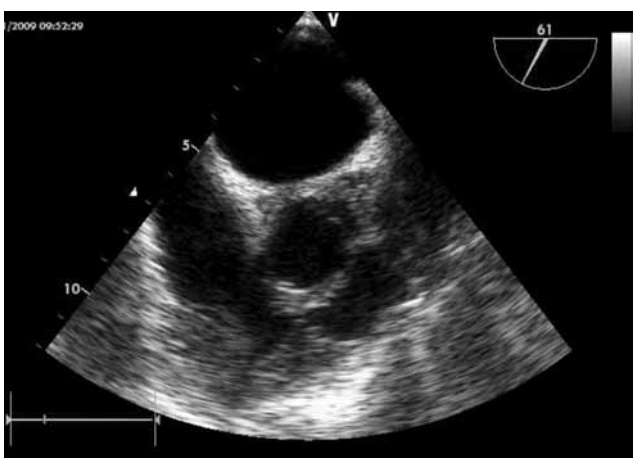

Figure 1. Intracavitary obstruction was showed at transesophageal echocardiography.

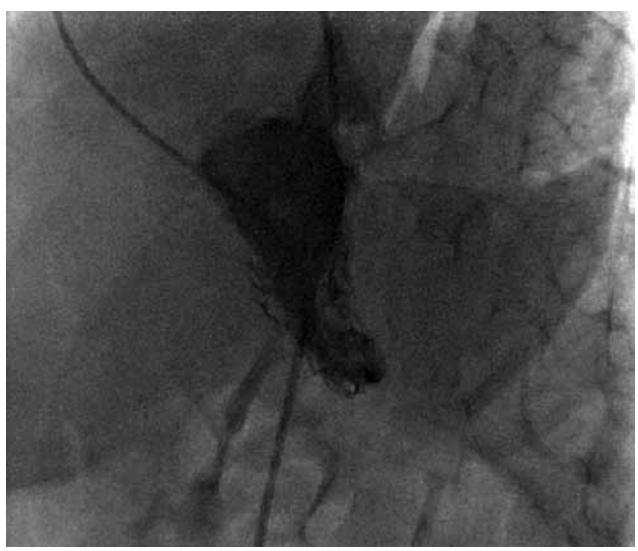

Figure 2. Double chamber was showed at angiography.

systolic pressure gradient in the RV cavity during RV catheterization, the visualization of the obstruction under the RV infindubulum caused by an abnormal muscle band, the absence of infundibular hypoplasia, and the direct visualization of an intracardiac muscle band during surgery.[6] In our case, we were able to demonstrate intraventricular obstruction by transthoracic echocardiography and angiography before surgery. Double-chambered right ventricle is a progressive disease, and patients may have different symptoms with varying levels of severity. Therefore, McElhinney et al.[7] suggested surgical correction in asymptomatic DCRV patients with a high intracavitary pressure gradient.

In conclusion, DCRV is usually seen in childhood and frequently is accompanied by cardiac disorders. However, as in our patient, it may present in adulthood without other cardiac defects. Therefore, when there is an unexplained etiology involving either pulmonary hypertension or tricuspid regurgitation, screening for DCRV should be conducted so that proper treatment options can be explored.

Declaration of conflicting interests

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