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A case of double-chambered right ventricle diagnosed by cardiac magnetic resonance imaging and catheterization

Przypadek dwujamowej prawej komory rozpoznanej na podstawie badania metodą rezonansu magnetycznego i cewnikowania

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Abstract

A double-chambered right ventricle (DCRV) is a heart defect, typically congenital, in which the right ventricle is separated into a proximal high-pressure (anatomically lower) chamber and distal low-pressure (anatomically higher) chamber. Commonly ventricular septal defect is found concomitantly in patients with DRCV. In this case report, we present a 20-year-old female patient who was found to have a DRCV without any concomitant congenital heart disorder.

Key words: double-chambered right ventricle, cardiac MRI, cardiac catheterization, echocardiography

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Introduction

A double-chambered right ventricle (DCRV) is a heart defect, typically congenital, in which the right ventricle (RV) is separated into a proximal high-pressure (anatomically lower) chamber and distal low-pressure (anatomically higher) chamber. It can be caused by the presence of anomalous muscle bands, by hypertrophy of endogenous trabecular tissue, or occasionally by an aberrant moderator band.

Case report

A 20-year-old female patient was admitted to our clinic with the complaint of the increasing dyspnea on minimal exertion. It was learned from her history that warfarin therapy was begun due to the suspicion of pulmonary embolism in another hospital previously. However, computerized tomography (CT) of the chest revealed no evidence of pulmonary embolism. Cardiac auscultation revealed a grade 3/6 systolic murmur over the left sternal border. ECG showed evidence of right atrial enlargement with nonspecific ST-T wave changes across the precordial leads.

Transthoracic (TTE) and transesophageal echocardiography (TEE) revealed evidence of a double-chambered right ventricle (RV) across a prominent moderator band (Figures 1, 2). There was no evidence of obstruction noted across the pulmonary outflow tract. The left ventricular systolic function was within normal limits, with no valvular abnormalities. Cardiac magnetic resonance imaging (MRI) showed a hypertrophied muscle bundle dividing the RV into two chambers (Figure 3). Left and right heart cardiac catheterization was done to confirm the diagnosis. It was detected that high-pressure proximal and low-pressure distal right heart chambers were communicated to each other with a narrow duct. Maximal systolic pressure gradient was measured as 160 mm Hg between two chambers of right ventricle. There was no left-to right shunt according to the blood oximetry values taken in different parts of cardiac chambers, pulmonary artery, superior and inferior vena cava. Left ventriculography seen from left anterior oblique

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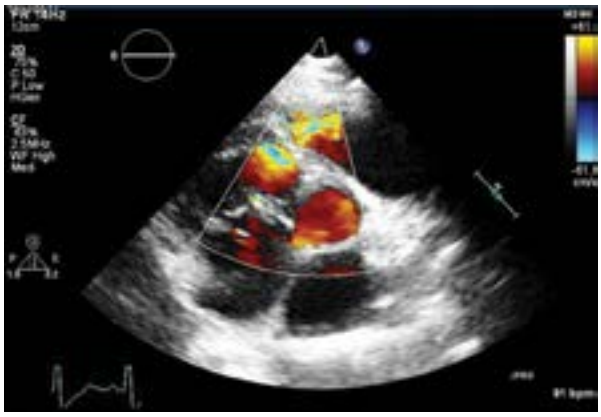

Figure 1. Color flow Doppler image showing the connection between two chambers of right ventricle (short axis view)

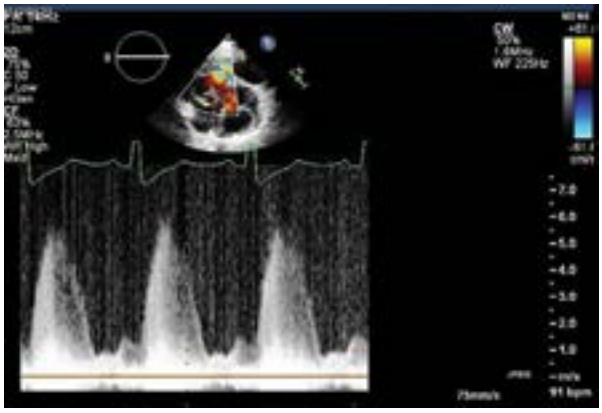

Figure 2. The pressure gradient measured with transthoracic echocardiography in short axis view between two chambers of the right ventricle (6 m/s and 144 mm Hg)

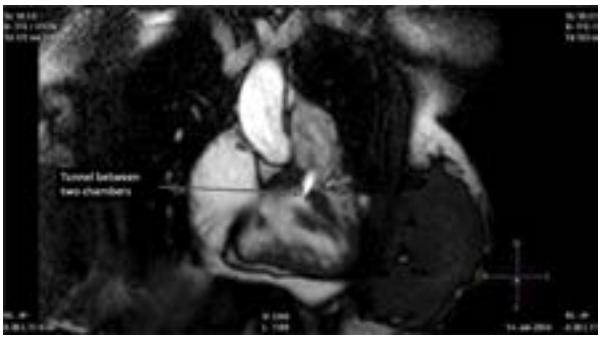

Figure 3. Magnetic resonance imaging of the heart shows anomalous muscle band (arrowheads) dividing the right ventricle

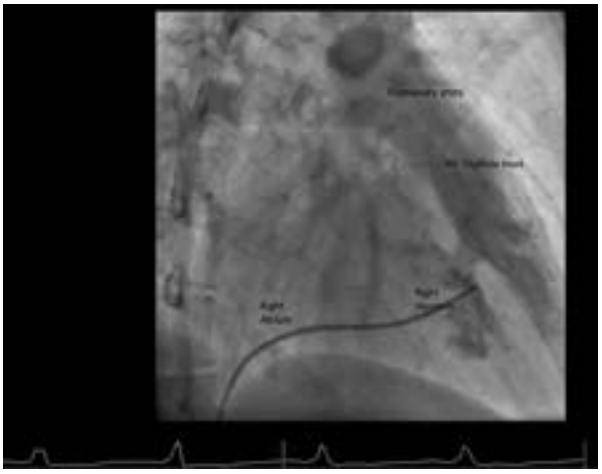

Figure 4. Cardiac catheterization with angiography of double-chambered right ventricle (RV) in RAO position. An abnormal muscle bundle separates the high-pressure inflow chamber and the low-pressure outflow chamber

position did not show any ventricular septal defect. Right ventriculography seen from right anterior oblique position showed that there was a double-chambered right ventricle separated from each other with an abnormal muscle bundle and dilated pulmonary artery (Figure 4). It was decided to operate the patient but she did not accept it.

Discussion

Double-chambered right ventricle is a rare congenital heart disorder involving 2 different RV pressure-compartments that is often associated with ventricular septal defect (VSD) [1, 2]. In our case we could not find VSD. Usually, the obstruction is caused by an anomalous muscle bundle crossing the RV from the interventricular septum to the RV free wall [1]. Because of its evolving nature, its diagnosis is usually made during childhood/adolescence and very rarely during adulthood [3]. It accounts for approximately 0.5–1% of patients with congenital heart disease [4]. Diagnosis can be challenging when it is asymptomatic. Cardiac magnetic resonance imaging and invasive testing can provide further information to confirm the diagnosis and guide therapeutic decisions [5]. Many authors have reported a high incidence of concomitant congenital disease; up to 90% of cases are associated specifically with membranous-type ventricular septal defects [6].

Patients have been reported to present with dyspnea, syncope, both stable and unstable angina and exercise intolerance [3]. Left-sided parasternal systolic murmurs have been auscultated in DCRV, which are typically displaced lower along the sternal border compared to a typical systolic murmur [3]. Electrocardiogram testing on patients may reveal right ventricular hypertrophy. Right axis deviation, prominent R waves in the right precordial leads, and an the

absence of prominent S waves in the left precordial leads have all been reported in cases of DCRV and suggest right ventricular hypertrophy [3].

A diagnosis of DCRV should be considered when right ventricular hypertrophy is apparent with no signs of infundibular hypertrophy or valvular pulmonary stenosis [7]. The most effective form of diagnosis of DCRV is TEE [6]. Use of continuous Doppler also allows for the calculation of intraventricular pressure gradients and obviates the need for additional cardiac catheterization [6, 7]. On echocardiography, fibrotic and hypertrophic tissue is more evident during systole, since the muscle bands are contracted and thickened [8]. Cardiac catheterization and angiocardiography can be used to confirm the diagnosis [7].

It is important to note that tetralogy of Fallot and DCRV with an associated ventricular septal defect may look similar on echocardiography. However, these can be easily distinguished since tetralogy of Fallot is characterized by decreased pulmonary flow whereas DCRV is associated with increased pulmonary flow [9].

Once diagnosed, the most effective treatment of DCRV is the surgical removal of hypertrophic or anomalous tissue using a right atriotomy [7]. However, a DCRV repair using a right ventriculotomy is also described. Indications for surgery may include a pressure gradient over 40 mm Hg between the proximal chamber and the pulmonary artery, the presence of aortic regurgitation and symptoms of heart failure [10]. If a patient has mild symptoms and refuses surgery, beta-blocker therapy may be sufficient to improve their general condition [7, 11].

Since DCRV is generally a progressive pathology, it may still be beneficial to operate before the emergence of symptoms [7]. Patient groups at high risk for DCRV include those with ventricular septal defects, increased pulmonary flow, and rapid onset or primary right heart failure without associated pulmonary hypertension [3, 7, 9].

Conclusion

Double-chambered right ventricle is a rare congenital heart defect in which the right ventricle is separated into a high-pressure proximal and low-pressure distal chambers. It is usually associated with ventricular septal defect unlike in our case. We could not find any case like ours who has very high systolic pressure gradient between the two chambers of right ventricle without ventricular septal defect in the literature.

This defect is considered to be congenital and typically presents in infancy or childhood but has been reported to present rarely in adults. DCRV is typically found concomitantly with congenital cardiac disorders, most notably ventricular septal defect and subaortic stenosis. Due to its rarity and the difficulty of visualization, DCRV continues to be misdiagnosed. The most effective diagnostic studies are transesophageal echocardiogram, continuous flow Doppler, and cardiac catheterization. Surgical excision of the obstructive tissue is the most effective form of treatment.

In conclusion, multimodality cardiac imaging using echocardiography, cardiac CT, cardiac MRI and cardiac catheterization are often required for complete characterization of complex congenital heart anomalies in adults like our case.

Conflict of interest(s)

The authors declare that they have no conflict of interest.

Streszczenie

Dwujamowa prawa komora (DCRV) to wada serca, zwykle wrodzona, w której prawa komora jest podzielona na dwie części — proksymalną (położoną anatomicznie niżej), wysokociśnieniową oraz dystalną (położoną wyżej), w której ciśnienie jest niskie. U pacjentów z DRCV często wykrywa się równocześnie ubytek w przegrodzie międzykomorowej. W niniejszej pracy kazuistycznej przedstawiono przypadek 20-letniej chorej, u której wykryto DRCV bez innych współistniejących wad serca.

Słowa kluczowe: dwujamowa prawa komora, sercowy MRI, cewnikowanie serca, echokardiografia

Folia Cardiologica 2017; 12, 2: 187–190

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