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Case report

Double-chambered right ventricle in a patient with hypertrophic cardiomyopathy. A case report

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ABSTRACT

We present the case of a 42-year-old male patient with a history of bilateral lung transplantation and chronic graft dysfunction. The patient presented to the adult emergency department due to acute heart failure. During his stay in the emergency room and subsequent outpatient follow-up, multimodality cardiac imaging led to the diagnosis of a double-chambered right ventricle with associated hypertrophic cardiomyopathy. Given the presence of advanced lung disease and poor adherence to both immunosuppressive therapy and clinical follow-up, the patient was deemed unsuitable for repeat lung transplantation. The optimization of his immunosuppressive medication was prioritized, and beta-blockers were added as part of the treatment for dynamic right ventricular outflow tract obstruction. He was referred to pulmonary rehabilitation and is currently showing a partially favorable clinical course, with improvement to New York Heart Association functional class II.

Keywords: Cardiomyopathy Hypertrophic; Heart Ventricles; Diagnostic Imaging (Source: MeSH-NLM).

RESUMEN

Ventrículo derecho de doble cámara en un paciente con miocardiopatía hipertrófica no obstructiva. Reporte de caso

Se presenta el caso de un paciente masculino de 42 años de edad con antecedente de trasplante bipulmonar con disfunción crónica del injerto. El paciente acudió a la central de emergencias de adultos por cuadro de insuficiencia cardíaca aguda. Durante su estancia en guardia y en seguimiento ambulatorio, mediante las multiimágenes cardíacas se diagnosticó ventrículo derecho de doble cámara con miocardiopatía hipertrófica asociada. Debido a su enfermedad pulmonar avanzada y escasa adherencia al seguimiento médico, se decidió no realizar un re-trasplante pulmonar. Se optimizó su medicación inmunosupresora y se añadieron betabloqueantes como parte del tratamiento de la obstrucción dinámica intraventricular derecha. Además, se derivó a rehabilitación pulmonar, mostrando actualmente una evolución parcialmente favorable en clase funcional II.

Palabras clave: Cardiomiopatía Hipertrófica; Ventrículos Cardíacos; Diagnóstico por Imagen (Fuente: DeCS-Bireme).

Introduction

Double-chambered right ventricle (DCRV) is an uncommon cardiac defect. Its aetiology is congenital in most cases, accounting for approximately 0.5% to 2% of all congenital heart diseases, although there are a few reports of acquired forms [1]. It may result from anomalous muscular bundles, trabecular hypertrophy, or an aberrant moderator band [1,2].

Several studies have reported its association with other cardiac anomalies (particularly ventricular septal defects) as well as extracardiac abnormalities [3]. However, only a few case reports have described the coexistence of DCRV and hypertrophic cardiomyopathy (HCM) [4,5]. We describe a patient diagnosed in adulthood with both HCM and DCRV.

Case report

We report the case of a 42-year-old male patient with no cardiovascular risk factors. His relevant family history includes a first-degree relative (father) with HCM. Regarding his personal medical history, he underwent bilateral lung transplantation in 2020 due to acquired bronchiectasis. Post-transplant, he developed obliterative bronchiolitis and graft rejection at three months, attributed to poor adherence to immunosuppressive therapy, ultimately resulting in chronic graft dysfunction.

The patient presented to the adult emergency department with dyspnoea classified as New York Heart Association (NYHA) functional class III, without orthopnoea or paroxysmal nocturnal dyspnoea. He also reported symptoms of sleep apnoea, excessive daytime sleepiness, and dynamic tracheal obstruction as a postoperative sequela.

On physical examination, the patient was normotensive, tachycardic, and eupnoeic, with adequate ventilatory mechanics. Bibasilar fine "velcro-like" crackles were auscultated, along with a 2/6 systolic murmur at the pulmonary focus. Bilateral lower limb oedema (grade 2/4) was also noted. An initial electrocardiogram revealed sinus rhythm with signs of biventricular hypertrophy and marked repolarisation abnormalities **(Figure 1)**. Based on these findings, a transthoracic echocardiogram was performed, which showed severe left ventricular (LV) hypertrophy. Right ventricular (RV) hypertrophy with apical obliteration and an intraventricular pressure gradient of 24 mmHg were also noted **(Figure 2)**. In addition, mild tricuspid regurgitation, right atrial (RA) dilatation with a RV–RA pressure gradient of 41 mmHg and mildly reduced biventricular ejection fraction due to global hypokinesis were observed.

A chest CT angiography was performed to rule out pulmonary embolism, which returned negative; however, it revealed marked biventricular hypertrophy **(Figure 3).** With a working diagnosis of congestive heart failure, the patient was treated with 20 mg of intravenous furosemide. Following a favourable clinical response, he was discharged for further outpatient evaluation of his structural heart disease.

As part of the anatomical assessment, a cardiac magnetic resonance imaging (CMR) scan was performed **(Figure 4)**, which demonstrated features of non-obstructive left-sided HCM with RV

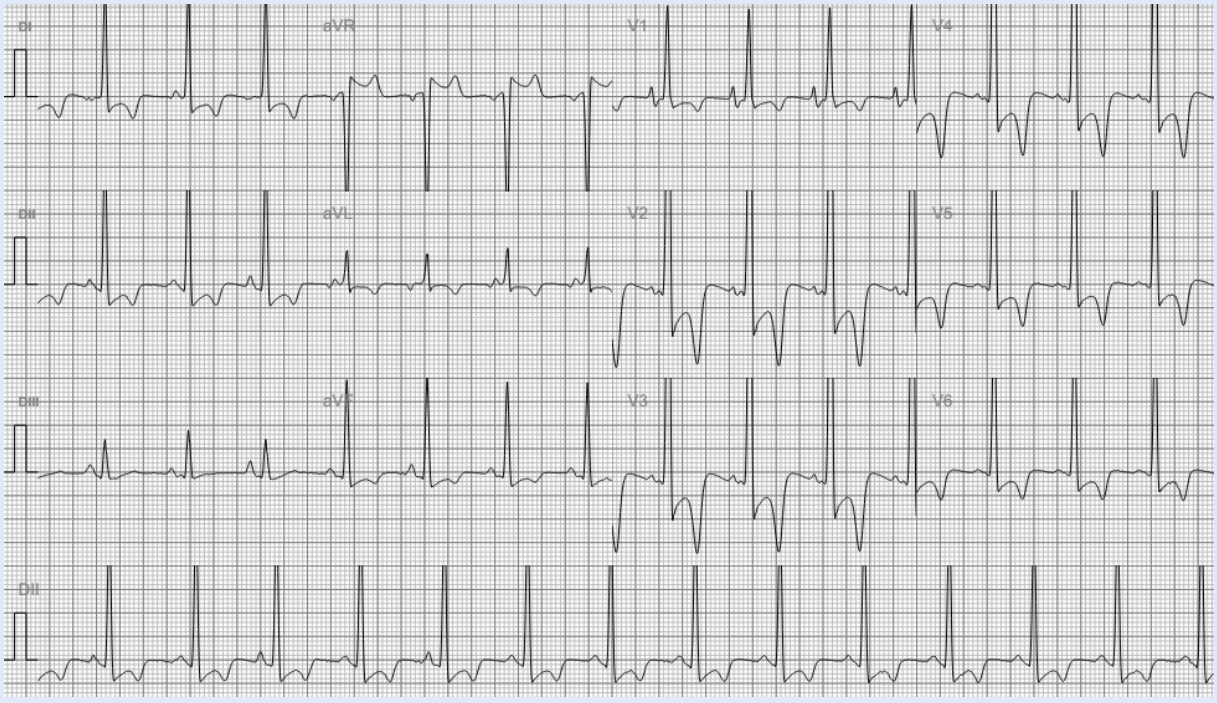

Figure 1. Electrocardiogram showing sinus rhythm at 90 beats per minute. Pulmonary P wave, narrow QRS complex with clockwise rotation, and marked diffuse repolarisation abnormalities.

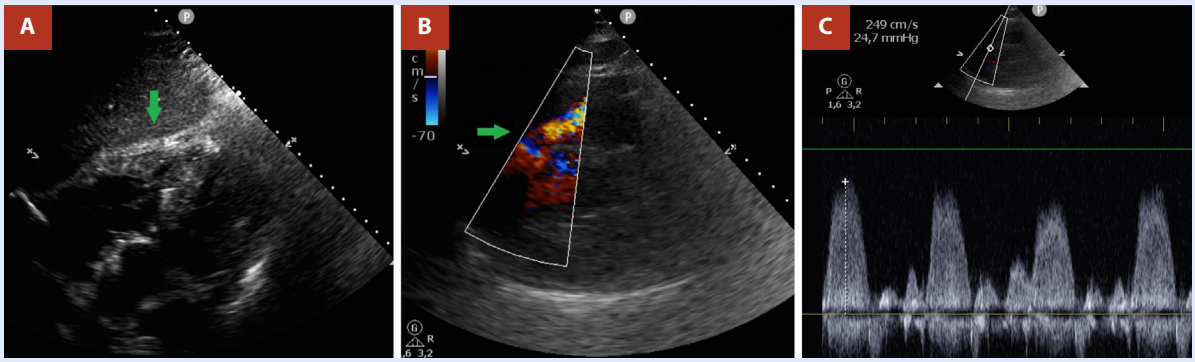

Figure 2. Transthoracic echocardiogram with suboptimal acoustic window from **(A)** a subxiphoid view showing marked mid-apical hypertrophy of the right ventricular (RV) free wall (green arrow) **(B)** short-axis view at the level of the great vessels demonstrating mid-ventricular flow acceleration within the RV (green arrow) and **(C)** intraventricular pressure gradient in the RV.

involvement predominantly affecting the outflow tract and apical segments (maximum thickness of 17 mm in the RV outflow tract), resulting in subpulmonary stenosis and systolic obliteration of the RV cavity, along with mildly reduced RV ejection fraction (51%). LV mass was markedly increased (153 g/m^2), with a maximum wall thickness of 24 mm in the anteromedial region, and a preserved LV ejection fraction. Post-contrast images following intravenous gadolinium injection revealed diffuse, faint intramyocardial late gadolinium enhancement in the mid anteroseptal, mid inferoseptal, anteroapical, apical septal, apical inferior, apical lateral, apical tip, and the RV free wall. The latter finding ruled out subendocardial diseases such as endomyocardial fibrosis. Native T1 mapping showed a value of 1123 milliseconds, consistent with diffuse interstitial fibrosis, while extracellular volume and T2 mapping values were within normal limits.

Cardiopulmonary exercise testing revealed a mixed ventilatory defect with a predominant severe obstructive component and severely reduced forced vital capacity, consistent with chronic lung allograft dysfunction.

Given the presence of advanced pulmonary disease and the patient's poor adherence to immunosuppressive therapy and clinical follow-up, he was deemed unsuitable for lung re-transplantation. Management focused on optimising immunosuppressive medication, and beta-blockers were introduced to address the dynamic right intraventricular obstruction. The patient was referred to pulmonary rehabilitation and currently shows a partially favourable clinical course, with improvement to NYHA functional class II.

Discussion

RV involvement in HCM is uncommon, occurring in approximately 15% of patients, and predominantly or isolated RV disease is even rarer [6]. The underlying mechanism is congenital in most cases, accounting for 0.5% to 2% of all congenital heart diseases, although it may also be acquired, such as in right-sided chamber involvement in HCM [1]. This condition is characterised by a right

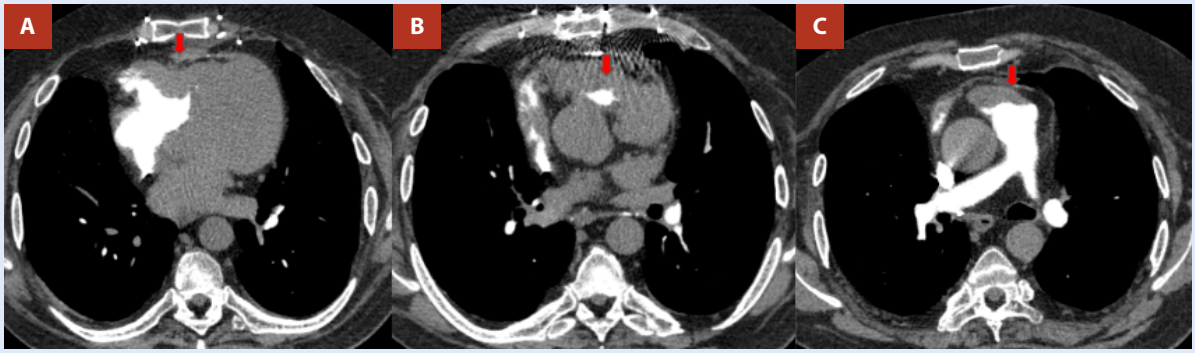

Figure 3. Chest CT angiography showing absence of contrast opacification suggestive of marked hypertrophy of the right ventricular free wall **(A, arrow),** with narrowing of the outflow tract in more cranial slices, progressing to a nearly occlusive lumen **(B, arrow)** approaching the pulmonary artery **(C, arrow)**.

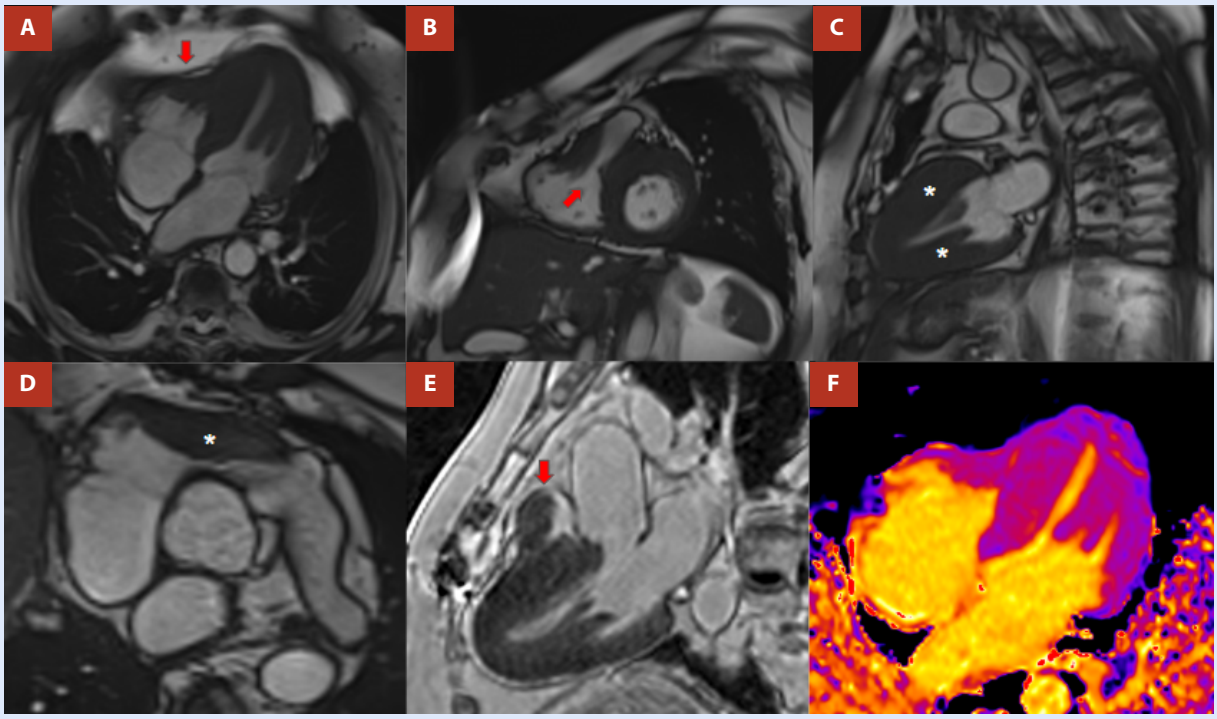

Figure 4. Cardiac magnetic resonance imaging. **A:** Four-chamber view showing obliteration of the apical segments of the right ventricle (RV) (arrow) and biventricular hypertrophy. **B:** Sagittal section through the subvalvular narrowing at the level of the RV outflow tract (arrow), caused by hypertrophy of the interventricular septum and RV free wall. **C:** Two-chamber view of the left ventricle highlighting prominent septal and posterior wall hypertrophy (asterisks). **D:** Short-axis view at the level of the great vessels showing RV outflow tract hypertrophy (asterisk) with luminal obliteration. **E:** Late gadolinium enhancement image demonstrating a faint diffuse intramyocardial pattern and marked RV hypertrophy (arrow). **F:** Native T1 mapping sequence with an elevated myocardial value of 1123.0 ms, consistent with diffuse fibrosis.

intraventricular pressure gradient greater than 20 mmHg between a high-pressure proximal chamber near the tricuspid valve and a low-pressure distal chamber adjacent to the pulmonary valve. The defect may be caused by anomalous muscular bundles, trabecular hypertrophy, or an aberrant moderator band [1,2]. Histological findings appear similar to those observed in LV involvement, suggesting a shared pathogenesis; however, double-chambered RV outflow tract obstruction is less common and may lead to more severe and treatment-resistant symptoms [7]. Although the pathophysiology is not yet fully understood, RV outflow tract (RVOT) obstruction and reduced cardiac output due to impaired RV filling are thought to contribute to symptom progression. In our patient, the association with left-sided HCM and the absence of other congenital cardiac anomalies suggest an acquired aetiology, placing this case within a small subset of reports in the existing literature.

The symptoms associated with this condition are similar to those of other cardiovascular diseases, with dyspnoea and exercise intolerance being the most frequently reported [8]. These symptoms may progress to minimal exertional tolerance or even dyspnoea at rest as the RV obstructive gradient increases. In our case, the patient had chronic lung allograft dysfunction, which contributed to the exacerbation of respiratory symptoms. Nevertheless, the late presentation in adulthood remains a noteworthy and uncommon finding.

Echocardiography and cardiac catheterisation are useful tools for assessing ventricular hypertrophy, diastolic pressures, and RV outflow tract gradients, respectively. CMR is currently the gold standard for diagnosis, as it provides high-resolution images that allow detailed evaluation of the biventricular distribution of hypertrophy and assists in excluding other causes of ventricular abnormalities [9]. Moreover, CMR contributes to the differential diagnosis by identifying alternative causes of apical RV obliteration, such as endomyocardial fibrosis and cardiac tumours.

Although the optimal treatment remains undefined, symptomatic patients have been managed with beta-blockers and calcium channel blockers, which have variably reduced symptoms and intraventricular gradients [10]. Risk stratification for sudden cardiac death is essential in patients with HCM; however, current risk scores do not consider RV involvement as a significant variable, and it remains unknown whether its presence may carry greater prognostic weight than its absence [11,12]. Surgical resolution of double-chambered right ventricle through myectomy, with or without associated HCM, has been described in the literature [13]. In our patient, this option was not pursued due to high surgical risk and the presence of multiple comorbidities, although it

would have represented the most appropriate approach given the severity of the dynamic obstructive component.

In conclusion, the association between double-chambered right ventricle and hypertrophic cardiomyopathy is rare. A thorough cardiovascular assessment using multimodal imaging is essential to establish the diagnosis. This report presents a clinically significant case of an uncommon association with adult-onset presentation, highlighting its relevance. It underscores the importance for adult cardiologists to recognise this condition, despite the limited number of cases reported beyond the second decade of life.

Ethical aspects

Written informed consent was obtained from the patient prior to the preparation and publication of this case report, with clear explanation regarding data confidentiality and privacy protection.

Authors' contributions

JMI, JF, RL, ALM, MNP: Conceptualization, Investigation and Writing - Original Draft. **RB, DP, RP:** Supervision and Writing - Review & Editing.

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