中文題目：病因不明性單獨右心室擴張型心肌病變：個案報告

英文題目：Isolated idiopathic right ventricular dilated cardiomyopathy: a case report

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Abstract

Dilated cardiomyopathy is characterized by ventricular enlargement, but such a structural change occurring only in the right ventricle (RV) is rare. We report the case of a 52-year-old man with chronic alcohol consumption who was admitted for dyspnea on exertion and peripheral edema. Echocardiography suggested dilated RV and impaired RV systolic function but preserved left ventricular function. Subsequent magnetic resonance imaging revealed neither significant delayed enhancement nor fatty change of the RV myocardium. Endomyocardial biopsy of the RV revealed no fibrofatty tissue replacement. This rare case is considered to be an isolated idiopathic RV dilated cardiomyopathy.

Introduction

Dilated cardiomyopathy (DCM) is characterized by ventricular enlargement with left ventricle (LV) or biventricular involvement [1]. DCM leads to progressive heart failure and a decline in LV contractile function, arrhythmias, conduction system abnormalities, thromboembolism, and sudden or heart failure–related death [2]. Right ventricular (RV)

dilated cardiomyopathy is not a well-defined entity in current classifications of the cardiomyopathies [2, 3]. There are few reports about isolated (RV) dilated cardiomyopathy with preserved (LV) function [4, 5]. Herein, we report a rare case of isolated (RV) cardiomyopathy.

Case report

A 52-year-old male patient presented at our institution with dyspnea and lower-limb edema that had persisted for several weeks followed by oliguria and orthopnea for 1 day. Dyspnea was aggravated by mild activity. He had neither chest pain nor any history of diabetes mellitus or hypertension and no family history of heart disease. The patient was a chronic heavy smoker and drinker.

On admission, his vital signs were relatively stable. Pitting edema of the bilateral lower limbs was noticed. Physical examination was unremarkable. Electrocardiography showed sinus arrhythmia with a prolonged QT interval (491 milliseconds, normal range = 360–440 milliseconds). Chest radiography revealed cardiomegaly and pulmonary congestion. Biochemical analysis indicated elevated *N*-terminal prohormone of brain natriuretic peptide (8429 pg/mL, normal range <900 pg/mL). Echocardiography revealed a dilated right atrium (RA) and RV and hypokinesis of the RV (*Figure 1*), although the LV was normal in size and

contractility (ejection fraction = 76%). We found mild tricuspid valve regurgitation and mild pulmonary hypertension, along with an estimated peak systolic pulmonary artery pressure of 39 mmHg. The patient also presented a small amount of pericardial effusion and mild liver congestion.

After admission, the patient's dyspnea was relieved gradually by diuretic therapy, and he was discharged in stable condition but was lost to follow-up. Exacerbated dyspnea recurred 2 months later, and the patient presented with dyspnea on exertion and swelling of the neck and right upper limbs. Computed tomography from the neck to the chest revealed dilatation of the right internal jugular vein with thrombus within. He refused catheter-directed thrombolytic therapy, so oral warfarin was used. Follow-up echocardiography showed that the RV factional area change (FAC) was 25.7% (normal range >35%), the tricuspid annular plane systolic excursion was 1.02 cm (normal range >1.6 cm), and the pulsed tissue Doppler velocity at the RV free wall (RV S′) was 7.37 cm/s (normal range >10 cm/s), all of which indicated impaired RV systolic function (*Figure 1*). Cardiac magnetic resonance imaging (MRI) revealed mild enlargement of the right atrium and reduced ejection fraction of the RV (26%). There was no significant delayed enhancement or fatty change of the myocardium (*Figure 2*), which may exclude arrhythmogenic RV cardiomyopathy/dysplasia (ARVC/D), acute myocarditis, or infiltrative diseases. Coronary angiography showed patent coronary arteries, which exclude chronic ischemic cardiomyopathy. Right heart catheterization revealed mild pulmonary

hypertension (mean pulmonary artery pressure = 31 mmHg) and no significant step-up of oxygen. Endomyocardial biopsy of the RV was also performed to investigate any histologic changes that could indicate specific infiltrative disease. The pathology revealed no intervening fibrous or adipose tissue between muscle fibers, and hematoxylin and eosin staining revealed the absence of iron or amyloid deposition (*Figure 3*).

Discussion

Dilated cardiomyopathy is characterized by LV chamber enlargement and systolic dysfunction in the absence of abnormal loading conditions (e.g., hypertension or valve diseases) or coronary artery disease sufficient to cause global systolic impairment. RV dilation and dysfunction may be present but are not necessary for the diagnosis [3]. ARVC/D, a inheritable heart disease, involves predominantly the RV and is characterized histologically by the presence of adipose and fibrous tissue replacement in the RV myocardium [6]. The structural change associated with ARVC/D may be confined to a localized region of the RV (the triangle of dysplasia) during the early stage, and more diffuse RV disease and LV involvement are common as disease progresses. In this patient, the cardiac MRI revealed no fibrofatty tissue replacement within the RV myocardium, which excludes the diagnosis of ARVC/D.

The patient had a history of chronic alcohol consumption. Long-term heavy alcohol consumption can result in cardiac dysfunction, herein referred to as alcoholic cardiomyopathy (ACM) [7]. ACM is classified among the DCM [2], and, like other DCM, is characterized by a dilated LV, normal or reduced LV wall thickness, and increased LV mass [8]. Kajander et al. assessed the effect of alcohol on the LV and RV by increments in daily alcohol consumption and found that LV size showed a U-shaped reduction associated with increased daily alcohol use, accompanied by an increase in RV size associated with very heavy drinking [9]. Histological examination of ACM revealed various degrees of fibrosis, patchy areas of endocardial fibroelastosis, intramural blood clots, and focal collections of swollen cells in both the epicardium and endocardium [7]. Teragaki et al. reported that myocytic hypertrophy, fibrosis, and nuclear change were less significant in ACM than in DCM [10], suggesting that histologic changes alone do not allow us to differentiate the 2 diseases. The histologic change in the present case revealed no fibrosis, but we cannot absolutely exclude alcohol as an etiological factor in the patient's RV DCM. Following an extensive literature search, we have found no reports to date of ACM with isolated RV involvement.

The nonspecific pathologic change in the present case may also exclude some extremely rare congenital cardiomyopathies limited to the RV, such as Uhl's anomaly of the RV [11] or noncompaction of the myocardium [3].

We found 1 case report of RV ACM in patents with severe sepsis [12], and another 2 case

reports of isolated right ventricular Takotsubo cardiomyopathy occurred in patients after surgery [13, 14]. These 2 kinds of cardiomyopathy appear to be fully reversible. No known acute stressful event precipitated the symptom onset in the present case, and the patient's RV structural change was not transient.

In conclusion, we report a rare case of isolated RV cardiomyopathy in a middle-aged man who presented with dyspnea on exertion and peripheral edema. No known etiology could be identified. Although such a clinical entity is not currently included in classification systems, isolated idiopathic RV cardiomyopathy seems to be the most appropriate diagnosis in the present case.

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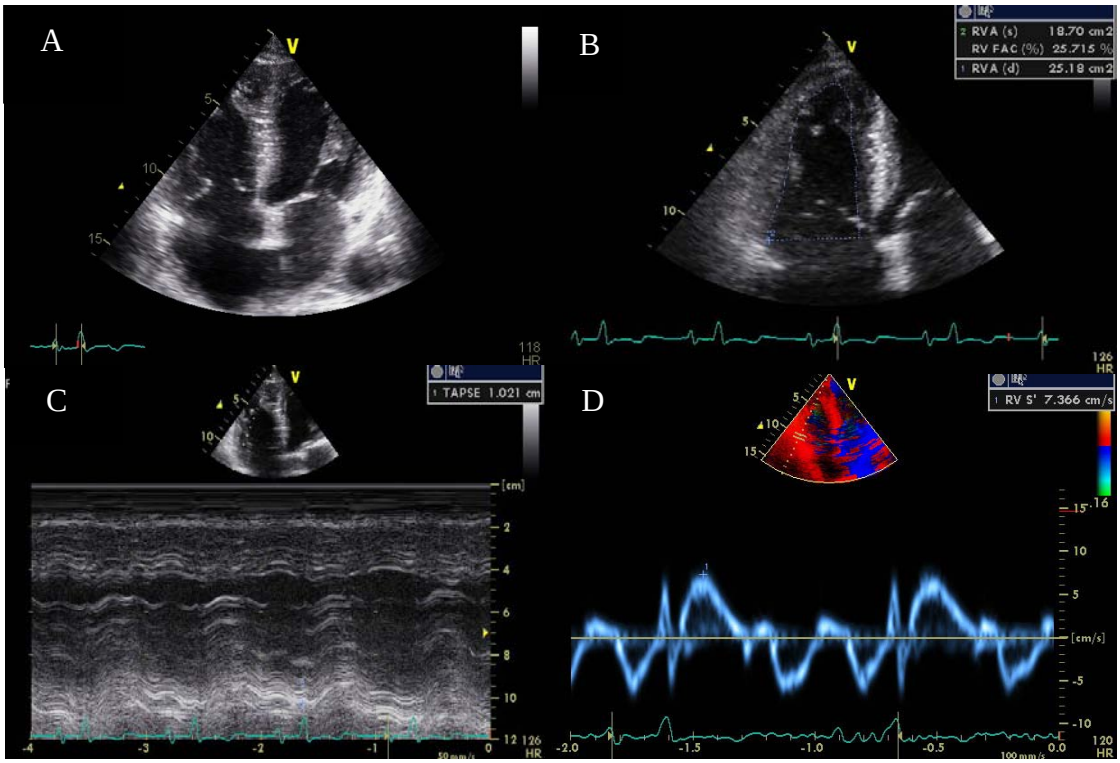

Figure 1 Evaluation of right heart systolic function by echocardiography. A. Dilated right atrium (RA) and right ventricle (RV), B. RV two-dimensional fractional area change (FAC) is 25.7cm/s, C. Tricuspid annular plane systolic excursion (TAPSE) is 1.02cm, D. Pulsed tissue Doppler velocities of RV free wall (RV S´) is 7.37cm/s.

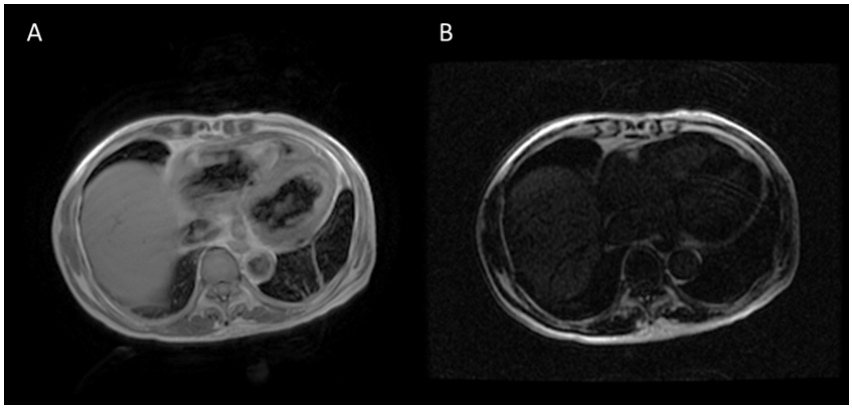

Figure 2. Cardiac magnetic resonance imaging (MRI). T1 weighted image (panel A) revealed no increased signal intensity of RV myocardium as subcutaneous fat. Delayed myocardial enhancement image (panel B) revealed no delayed enhancement of RV myocardium.

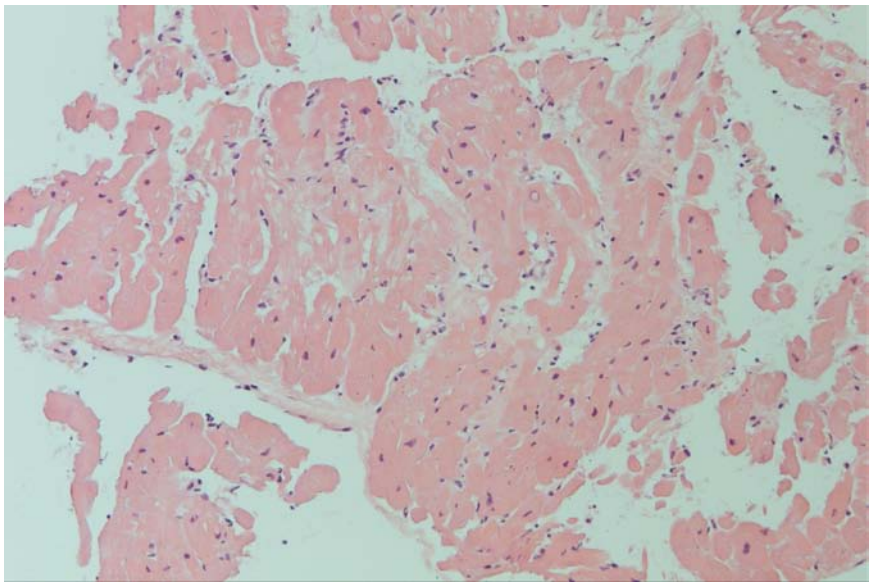

Figure 3. Microscopic images of RV. A hematoxylin and eosin staining of endomyocardial biopsy
revealed no myocyte necrosis, sparse infiltration of lymphocytes, and no intervening fibrous or adipose tissue.