

우심실부전이 동반된 단독우심실비치밀화증

순천향대학교 의과대학 순천향대학교 ¹천안병원 내과, ²구미병원 내과

임상혁¹ · 안지훈² · 오태훈¹ · 이동현¹ · 박민우¹

Isolated Right Ventricular Noncompaction Accompanied by Right Ventricular Failure

Sanghyok Lim¹, Jihun Ahn², Taehun O¹, Donghyun Lee¹, and Minwoo Park¹

*Department of Internal Medicine, ¹Soonchunhyang University Cheonan Hospital,
Soonchunhyang University College of Medicine, Cheonan;*

²Soonchunhyang University Gumi Hospital, Soonchunhyang University College of Medicine, Cheonan, Korea

Noncompaction of the ventricular myocardium is a rare congenital cardiomyopathy caused by arrest of normal endomyocardial embryogenesis. Isolated right ventricular noncompaction (IRNC) is an even rarer form of this disease. We report herein on a 68 year-old male diagnosed with IRNC who presented with right-sided heart failure, without involvement of the left ventricle. Diagnosis was achieved with the aid of echocardiography and ventriculography. Medical treatment including prescription of diuretics, a calcium channel blocker, and digitalis, improved both the symptoms and right ventricular function. (Korean J Med 2015; 88:69-73)

Keywords: Right ventricular noncompaction; Echocardiography; Right-sided heart failure

INTRODUCTION

Noncompaction of the ventricular myocardium (NCVM) is a disease characterized by an increase in ventricular trabeculation, caused by arrest of normal endomyocardial embryogenesis [1,2]. The usual clinical features of NCVM are heart failure, thromboembolic events, and atrial fibrillation [3,4]. NCVM usually develops in the left ventricle. Thus, patients with NCVM com-

monly present with left-sided heart failure. Recently, some authors have reported cases of right ventricular (RV) noncompaction with or without involvement of the left ventricle [2,5]. Herein, we describe a rare case of a patient with isolated right ventricular noncompaction (IRNC) who presented with right-sided heart failure. Our diagnosis was facilitated by transthoracic echocardiography (TTE), transesophageal echocardiography (TEE), and right ventriculography. Both the symptoms and RV function

Received: 2014. 1. 23

Revised: 2014. 5. 20

Accepted: 2014. 7. 29

Correspondence to Jihun Ahn, M.D.

Department of Internal Medicine, Soonchunhyang University Gumi Hospital, 179 1gongdan-ro, Gumi 730-706, Korea
Tel: +82-54-468-9062, Fax: +82-54-468-3638, E-mail: jichoona@gmail.com

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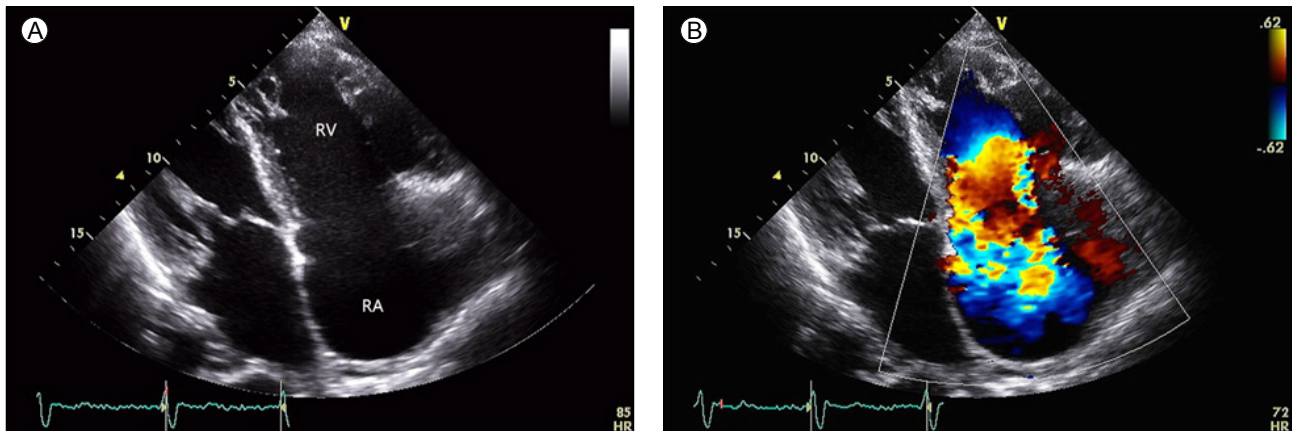


Figure 1. Initial transthoracic echocardiography (TTE). (A) A four-chamber view showing an extremely dilated right atrium and right ventricle. (B) Color flow Doppler data evidencing severe tricuspid regurgitation. RA, right atrium; RV, right ventricle.

improved upon prescription of a calcium channel blocker, digitalis, and diuretics.

CASE REPORT

A 68 year-old male was admitted to our hospital with abdominal distension, and edema in both legs, on June 15, 2012. He had been treated for heart failure at a local clinic for the past several years. On physical examination, a regular pulse of 76 beats per minute was noted, and his blood pressure was 100/60 mmHg, with an elevation in jugular venous pressure. Electrocardiography revealed a normal sinus rhythm and an incomplete right bundle branch block. An initial chest X-ray identified cardiomegaly; we thus performed TTE. This technique revealed an extremely dilated right atrium (RA) and RV, associated with severe regurgitant tricuspid flow (Fig. 1A and 1B). On the subcostal view, the inferior vena cava was flat and dilated and the RV fractional area change (FAC) had decreased to 28% (Fig. 2A). We next performed TEE, to rule out shunt development. No shunt was apparent, but prominent trabeculation and deep intertrabecular recesses in the apical and mid-lesion of the RV were apparent (Fig. 3A). No other cardiac abnormality was noted, and the end-systolic thickness ratio (that of the noncompacted compared to the compacted myocardium) was greater than two (Fig. 3D). Subsequently, we performed TTE using contrast echocardiography (agitated saline). The modified

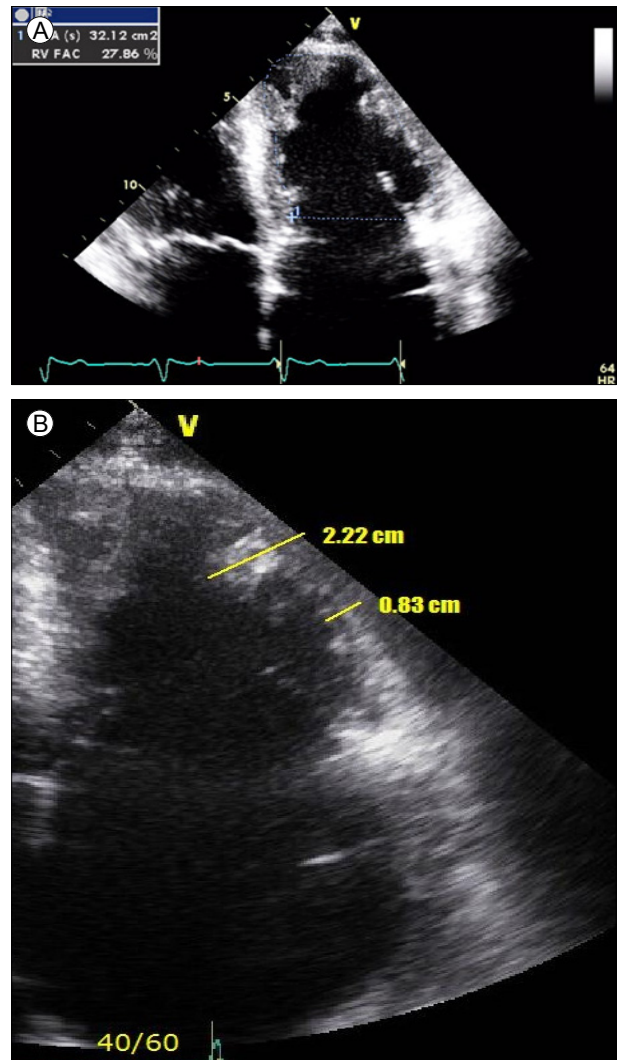


Figure 2. The right ventricular fractional area change (FAC). (A) The initial FAC was 24%. (B) The follow-up FAC improved to 38%.

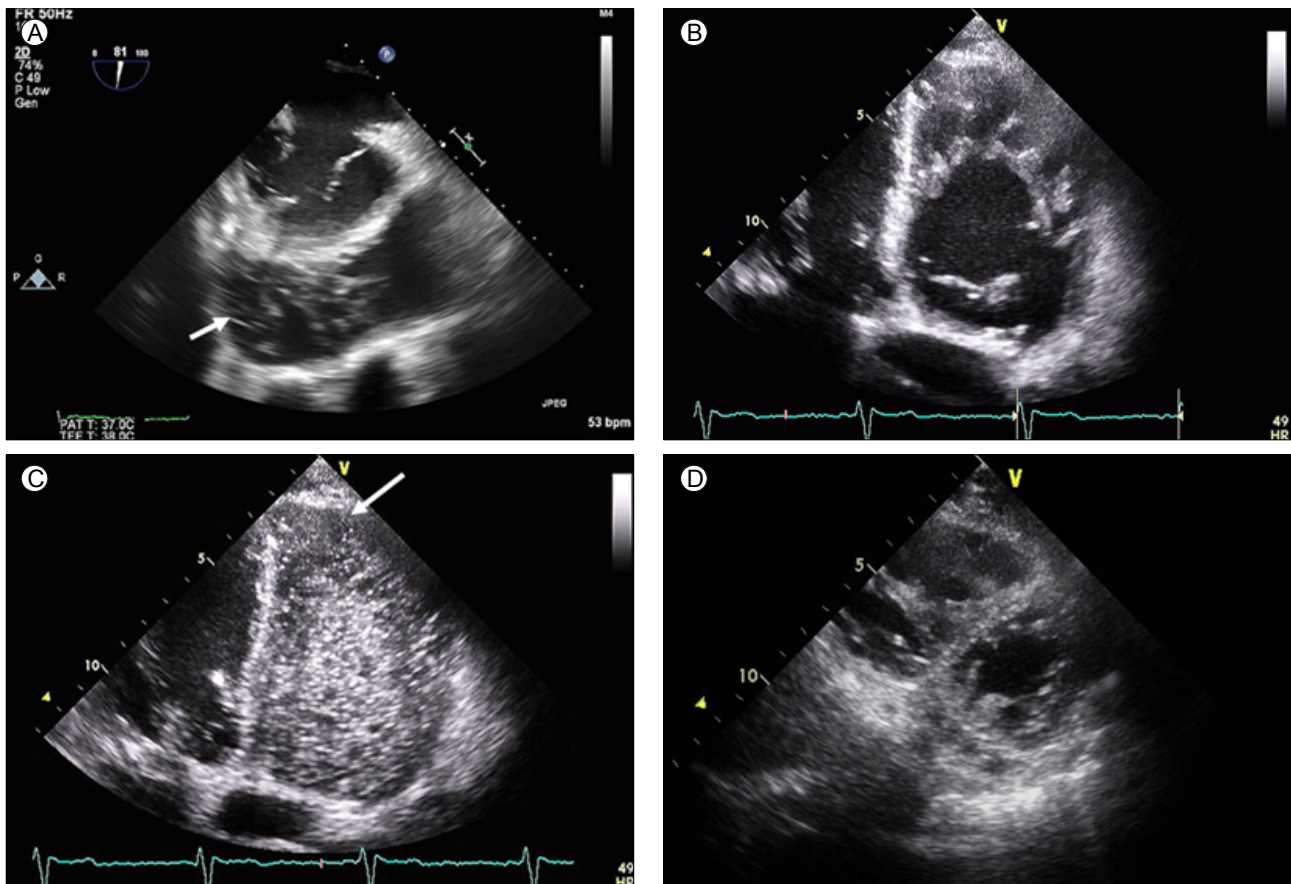


Figure 3. Transesophageal echocardiography (TEE) and TTE with contrast echocardiography. (A) TEE reveals prominent trabeculation and recesses (arrow) in the right ventricle (RV). (B) The modified four-chamber view shows a dilated (RV and prominent trabeculation and recesses. (C) Contrast echocardiography using agitated saline reveals bubble entry into the recesses (arrow). (D) A parasternal short-axis view illustrates the non-compaction morphological features of the RV.

four-chamber view revealed prominent trabeculation and, on contrast echocardiography, the contrast material entered the trabecular recess (Fig. 3B and 3C). Right-side catheterization did not yield evidence of primary pulmonary hypertension, and right ventriculography showed that the non-compacted apical RV myocardium was sponge-like in appearance (Fig. 4A). Also, magnetic resonance imaging (MRI) revealed a spongy noncompacted myocardium at the RV apex (Fig. 4B). We treated our patient conservatively; we prescribed diuretics, digitalis, and a calcium channel blocker. His symptoms and RV function gradually improved (Fig. 2B); the RV size became reduced and the extent of tricuspid regurgitation (TR) severity fell. The patient was discharged in a stable condition.

DISCUSSION

NCVM is a disorder of the heart muscle that triggers development of multiple trabeculations in the left ventricular myocardium, and is thought to arrest myocardial development. The disease is principally evident in the left ventricle (LV) apex and papillary muscle [1,3]. Heart failure is the prime clinical feature of NCVM, caused by depression of the systolic function of the noncompacted LV. Thromboembolic events and arrhythmia are also common clinical features of NCVM [1,3,6]. Treatment focuses on prevention of complications and control of symptoms [1,4]. Arrhythmias are treated with beta-blockers, calcium channel blockers, and other agents (depending on the features of individual cases). It is important to control NCVM symptoms, to

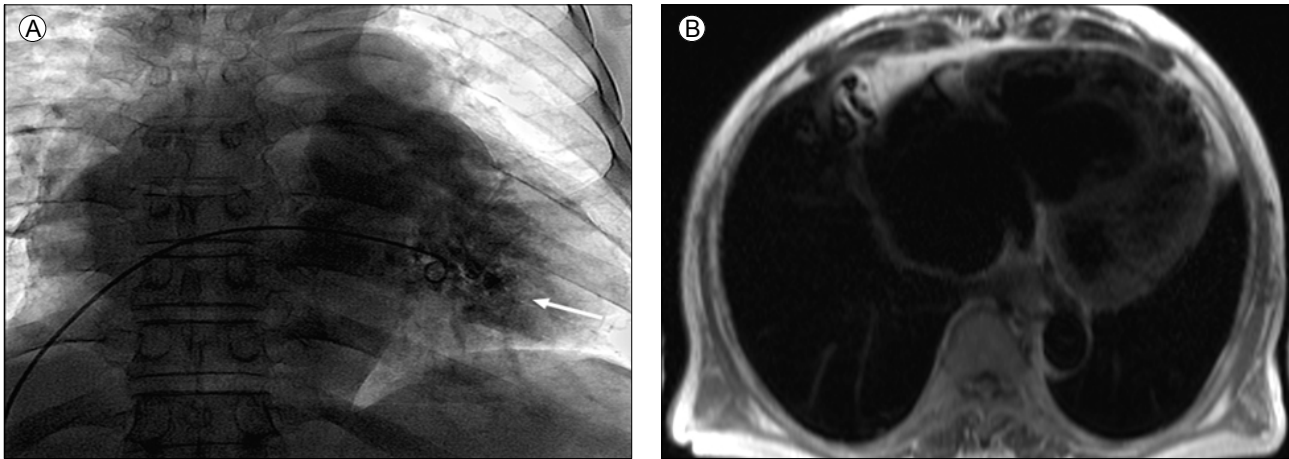


Figure 4. Right-sided ventriculogram and heart magnetic resonance imaging. (A) A right-sided ventriculogram reveals an abnormally prominent trabecular zone and deep intertrabecular recesses in the non-compacted RV myocardium (arrow). (B) Magnetic resonance imaging: a four-chamber view reveals spongy noncompacted myocardium at the RV apex. RV, right ventricular.

prevent heart failure. One report claimed that anti-failure medication (carvedilol) was associated with major improvement in a four-month-old infant with left ventricular noncompaction (LVNC) complicated by congestive heart failure [7]. Thromboembolic events can occur in patients with the disease, but any role for aspirin in terms of primary prevention of NCVM and cardioembolic events remains rather controversial [1,4].

Our knowledge and understanding of the etiology, genetic background, diagnosis, treatment, and outcomes of NCVM have steadily improved. One rare case of IRNC, with or without LV involvement, has been reported. However, the clinical manifestations of IRNC are not well-described, and no definitive consensus on treatment of the condition (including IRNC) has been formed. Patients are usually prescribed anti-failure or anti-congestive medications [6,7]. In our case, the patient presented with severe right-sided heart failure, and the RA and RV were extremely dilated. RV function also decreased upon hypokinetic motion (FAC = 28%). We did not consider IRNC prior to performance of TEE (during initial examination) because our focus was on the volume overload of the right-sided heart chambers. Also, RV enlargement and deviation render it difficult to diagnose IRNC during initial TTE. We diagnosed IRNC with the aid of multiple imaging modalities including TEE, angiography, and MRI. We prescribed a calcium channel blocker and digitalis

to improve RV contractile function, and diuretics to reduce blood volume. Symptoms and RV function improved, and the patient was discharged in a stable condition.

Left-sided heart failure is the most frequent clinical manifestation of NCVM, because the LV apex and mid-region are the principal sites of disease [8]. Treatment of NCVM remains challenging, and no definitive consensus has been reached, apart from heart transplantation in extreme cases. IRNC rarely occurs in the absence of LV involvement [9]. Clinical manifestations vary by lesional site and disease severity. Patients should be treated by reference to symptoms and clinical manifestations. Diuretics, digitalis, and a calcium channel blocker help to control IRNC symptoms and the disease course.

중심 단어: 우심실비치밀화증; 심초음파; 우심실부전

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