



Left Ventricular Noncompaction in Adults: Imaging and Clinical Findings in 63 Patients

성인 좌심실 비치밀화증: 성인 63명의 영상의학적 소견과 임상양상

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Purpose: To describe imaging and clinical findings for a left ventricular noncompaction (LVNC) in the adult.

Materials and Methods: From 2000 to 2014, 63 patients were diagnosed with LVNC by echocardiography, computed tomography, and magnetic resonance imaging at our hospital. Baseline characteristics, clinical manifestations, combined cardiac or systemic anomalies, and imaging findings were reviewed. We made a comparison between the isolated and combined disease groups.

Results: Among 63 patients with LVNC, 32 (51%) patients did not have combined cardiac anomalies (isolated disease group). The mean age at the initial diagnosis was higher in the isolated than in the combined disease group (54.2 years vs. 40.2 years, $p < 0.001$). The combined disease group presented symptoms more frequently at initial diagnosis than the isolated disease group (94% vs. 75%, $p = 0.082$). Heart failure symptoms were the most common adverse events (60.3% in all patients). Thromboembolic events developed in 20 patients, and were more frequent in the combined disease group than in the isolated disease group (39% vs. 26%, $p = 0.279$). The most common cardiac abnormality was dilated cardiomyopathy ($n = 15$, 24%). There was no significant difference in the mean noncompacted/compacted ratios between both of the disease groups.

Conclusion: Isolated and combined LVNC disease groups showed differences in age at diagnosis and clinical manifestations. The clinical and imaging findings may be helpful to better understand LVNC.

Index terms

Cardiomyopathies
Magnetic Resonance
Multidetector Computed Tomography
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INTRODUCTION

Left ventricular noncompaction (LVNC) is a myocardial disorder characterized by numerous prominent trabeculations and increased depth of inter-trabecular recesses that could contain thrombi (1). Congenital developmental arrest during the first trimester leading to the formation of two layers of the myocardium is the most accepted theory for the onset of LVNC (1-3). However, congenital developmental arrest alone is not sufficient to explain the etiology of LVNC, as suggested by some reports of acquired LVNC cases and even of reversible trabeculation in pregnant women (4, 5).

Although several diagnostic criteria for this disorder have been suggested, a global standard diagnosis for LVNC has yet to be established. Variable clinical manifestations and prognoses for LVNC have also been reported because patient population and imaging criteria have differed between previous studies (1, 6-9). We considered that a description of clinical characteristics and imaging features in an LVNC series, even using retrospective data from a single center, would increase our understanding of this rare disease entity. In our current study, we described imaging and clinical findings for a LVNC in a cohort of 63 adult patients.

MATERIALS AND METHODS

Patients

The Institutional Review Board of our hospital approved this retrospective study, and waived the requirement to obtain informed consent (2015-0570). Using the research-dedicated database system (ABLE, Asan Medical Center, Seoul, Korea) of our institution, which contains cardiac CT, cardiac MRI, and transthoracic echocardiography (TTE) reports, patients with LVNC and aged above 18 years were identified using the keywords noncompaction, spongy myocardium, or hypertrabeculation. Among the 72 searched patients, from 2000 to 2014, 66 patients were diagnosed with LVNC at our hospital based on cardiac imaging. Three cases without image data or for whom the image quality was suboptimal were excluded. Finally, 17 CTs, 18 MRIs, and 51 TTEs of 63 LVNC patients were enrolled. Total 43 patients were diagnosed by just one image modality (34 only by TTEs, 6 only by CTs, and 3 only by MRs). We divided the study patients into two groups: LVNC without cardiac anomaly (isolated disease group, $n = 32$) and LVNC with cardiac anomaly (combined disease group, $n = 31$) (Table 1). Combined cardiac anomalies included dilated cardiomyopathy ($n = 15$, 24%) followed by other types of congenital heart disease ($n = 12$, 19%) including coarctation of the aorta (CoA), tetralogy of Fallot (TOF), Ebstein anomaly, and transposition of the great arteries (TGA). Clinical manifestations at diagnosis included symptoms such as neck vein distention, rales, acute pulmonary edema, nocturnal dyspnea, and other manifestations of congestive heart failure (CHF) based on the criteria of the Framingham Heart Study (10). All detectable symptoms in our study patients were described at their initial presentation. Incidental detection was defined as diagnosis of LVNC on imaging that was conducted to evaluate other cardiac diseases without symptoms of CHF or on screening echocardiography (ECG).

Imaging Techniques and Analysis

Electrocardiography-gated cardiac CT was performed using either 16-slice CT, first-generation dual-source CT, or second-generation dual-source CT (Sensation 16, Definition, and Definition FLASH respectively, Siemens Healthcare, Erlangen, Germany). Images were obtained after the injection of 60–80 mL of iomeprol-400 (Iomeron; Bracco Imaging, Milan, Italy) followed

by 40 mL of a saline chaser. Body size-adaptive adjustment of tube potential and tube current was performed to reduce the radiation dose. Cardiac MRI was performed using a 1.5-T machine (Intera, Philips, Amsterdam, Netherlands; or Avanto, Siemens Healthcare, Erlangen, Germany). The balanced steady-state free precession sequence applied for cine-MRI data slice thickness of 5–8 mm and imaging matrix 256×256 on the short axis, 4-chamber, 2-chamber, and 3-chamber slice position. Delayed enhancement of MRI was performed using a 2D segmented inversion recovery gradient echo sequence 20 minutes after the intravenous administration of gadoterate meglumine (Dotarem®, Guerbet, Villepinte, France) (0.4 cc/kg) (11). Seventeen patients with cardiac CT and 18 patients with cardiac MR were also reviewed by two radiologists in consensus based on the criterion of Peterson et al. (8) for cardiac MRI and CT at end diastolic phase (6, 8, 9). The noncompacted layer thickness over the compacted layer thickness (NC/C) ratio was obtained from the most severe portion of trabeculation in the cardiac wall on the sagittal view.

TTE, which included two-dimensional and Doppler imaging, was performed using commercially available ultrasonographic equipment (Sonos 7500, Philips Medical Systems, Andover, MA, USA; or Vivid 7, GE Healthcare, Waukesha, WI, USA) with a 35 MHz transducer. The echocardiographic diagnosis of LVNC was based on the criterion of Jenni et al. (6). It included an end-systolic NC/C ratio higher than 2, and evidence of inter-trabecular recesses on color Doppler (6). TTE images were reviewed based on the described criterion to define absence or existence of LVNC. NC/C ratio measured on 2 chamber view, left ventricular ejection fraction (LVEF), indexed LV end diastolic/systolic volume, and indexed LV mass were measured.

Statistical Analysis

Continuous variables are expressed as mean \pm standard deviation, and nominal variables as numbers and percentages. Patient demographics, hemodynamic parameters, and imaging measurements were compared between patients with and those without a combined cardiac anomaly. Continuous variables were compared using the *t*-test, and categorical variables using the chi-square test or the Fisher exact test. Additional Bland-Altman Analysis was done for the inter-modality difference comparison. We performed statistical analysis using SPSS version

21.0 software (IBM Corp., Armonk, NY, USA). A *p*-value < 0.05 was considered statistically significant.

RESULTS

Among the 63 patients with LVNC analyzed in this study, 32

(51%) did not have a combined cardiac anomaly (isolated disease group) and 31 (49%) had combined cardiac abnormalities (combined disease group). The mean age at initial diagnosis of the isolated disease group was higher than that of the combined disease group (54.3 vs. 40.3 years, *p* < 0.001) (Table 1). The combined disease group presented with symptoms at initial diagno-

Table 1. Baseline Characteristics of Left Ventricular Noncompaction Patients

	Total (n = 63)	Isolated (n = 32)	Combined (n = 31)	<i>p</i> -Value
Age	47.4 ± 16.0	54.3 ± 12.2	40.3 ± 16.6	0
Sex				
Male, n (%)	43 (68)	24 (75)	19 (61)	0.243
Female	20 (32)	8 (25)	12 (39)	0.243
Chief complaint at diagnosis				
CHF symptom	38 (60)	15 (47)	23 (74)	0.027
Incidental	10 (16)	8 (25)	2 (7)	0.082
Chest pain	8 (13)	8 (25)	0 (0)	0.005
Known heart disease	2 (3)	0 (0)	2 (7)	0.238
Systolic BP	117.7 ± 20.0	122.4 ± 21.0	112.8 ± 18.0	0.055
Diastolic BP	72.6 ± 12.1	75.7 ± 14.2	69.4 ± 8.5	0.037
Hypertension	14 (22)	11 (36)	3 (11)	0.031
Heart rate	77.7 ± 19.4	74.4 ± 19.2	81.2 ± 19.2	0.170
Chest pain	35 (56)	16 (52)	19 (69)	0.440
Shock	17 (27)	3 (10)	10 (32)	0.059
DM	6 (10)	6 (24)	0 (0)	0.024
Smoke	16 (25)	12 (40)	5 (17)	0.084
Combined NMD	3 (5)	2 (6)	1 (3)	
Thromboembolic event	20 (32)	8 (26)	12 (39)	0.279
Significant CAD	11 (18)	8 (26)	3 (11)	0.182
Ischemic heart disease	8 (13)	7 (23)	1 (3)	0.053
Cardiac intervention or OP	15 (24)	1 (3)	14 (45)	
Implantation for HR control	11 (18)	5 (16)	6 (19)	1
Heart transplantation	4 (6)	1 (3)	3 (11)	0.355
ECG				
LBBB	13 (21)	9 (28)	4 (13)	0.222
Normal sinus rhythm	11 (18)	8 (25)	3 (11)	0.062
Atrial fibrillation	10 (16)	2 (6)	8 (26)	0.082
Paced rhythm	5 (8)	3 (9)	2 (7)	0.668
RBBB	4 (6)	1 (3)	3 (11)	0.355
Ventricular arrhythmia	4 (6)	2 (6)	2 (7)	0.238
Sinus tachycardia	3 (5)	0 (0)	3 (11)	0.113
WPW syndrome	1 (2)	0 (0)	1 (3)	0.492
Others	12 (19)	7 (23)	5 (17)	
Cardiac function				
LVEF < 50%	44 (72)	23 (71)	21 (72)	
Initial LVEF, %	36.9 ± 17.5	37.8 ± 16.4	35.7 ± 18.8	0.642
Initial LV ESV index, mL/m ²	74.5 ± 49.8	68.1 ± 42.7	81.5 ± 56.6	0.299
Initial LV EDV index, mL/m ²	110.1 ± 53.5	104.8 ± 47.5	115.9 ± 59.7	0.426
Initial LN mass index	188.1 ± 277.8	148.0 ± 50.5	234.4 ± 403.4	0.250

Table 1. Baseline Characteristics of Left Ventricular Noncompaction Patients (continued)

	Total (n = 63)	Isolated (n = 32)	Combined (n = 31)	p-Value
Wall motion				
Normal	12 (19)	8 (26)	4 (15)	0.672
Hypokinetic	40 (64)	20 (65)	20 (74)	0.721
Akinetic	5 (8)	3 (10)	2 (27)	1
Dyskinetic	0 (0)	0 (0)	0 (0)	
Paradoxical	1 (2)	0 (0)	1 (4)	0.344
Death	4 (6)	4 (13)	0 (0)	
Combined heart abnormality			31 (49)	
Dilated cardiomyopathy			15 (24)	
Kawasaki disease			2 (3)	
Hypertrophic cardiomyopathy			1 (2)	
ARVD			1 (2)	
Congenital heart disease			12 (19)	
Image modality				
Only TTE	34 (54)	22 (69)	12 (39)	
Only CT	6 (10)	1 (3)	5 (16)	
Only MR	3 (10)	3 (9)	0 (0)	
TTE and CT	4 (13)	1 (3)	3 (5)	
TTE and MRI	8 (26)	4 (13)	4 (6)	
CT and MRI	3 (10)	3 (9)	0 (0)	
ALL	5 (16)	3 (9)	2 (3)	

Continuous variables are presented as mean \pm standard deviation. Categorical variables are expressed number (percentage).

ARVD = arrhythmogenic right ventricular dysplasia, BP = blood pressure, CAD = coronary artery disease, CHF = congestive heart failure, DM = diabetes mellitus, ECG = echocardiography, EDV = end diastolic volume, EF = ejection fraction, ESV = end systolic volume, HR = heart rate, LBBB = left bundle branch block, LV = left ventricle, LVEF = left ventricular ejection fraction, NMD = neuromuscular disease, OP = operation, RBBB = right bundle branch block, TTE = transthoracic echocardiography, WPW syndrome = Wolff-Parkinson-White syndrome

sis more frequently than the isolated disease group (94% vs. 75%, $p = 0.082$). The most frequent chief complaint at initial diagnosis was CHF in both groups ($n = 38$, 60%), which was more frequent in the combined than in the isolated disease group (74% vs. 47%, $p = 0.027$). Thromboembolic events were more common in the combined than in the isolated disease group, without statistical significance (39% vs. 26%, $p = 0.279$). ECG findings were heterogeneous, varying from normal sinus rhythm to Wolff-Parkinson-White (WPW) syndrome. Left bundle branch block was the most common finding in the isolated disease group ($n = 9$, 28%), whereas atrial fibrillation was the most frequent in the combined disease group ($n = 8$, 26%). The number of patients who underwent cardiac intervention or operation was much higher in the combined than in the isolated disease group (14 vs. 1, respectively). Four patients underwent heart transplantation, 1 in the isolated and 3 in the combined disease group. In the combined disease group, the most common comorbid cardiac abnormality was dilated cardiomyopathy ($n = 15$, 24%), followed by other congenital heart diseases ($n = 12$, 19%) in-

cluding CoA, TOF, Ebstein anomaly, and TGA.

From the retrospective review of the data from each image modality, patients with LVNC showed extensive trabeculation, increased NC/C ratio, and inter-trabecular recess (Fig. 1). The mean NC/C ratio was 2.8 ± 0.6 on TTEs in 51 patients, 2.8 ± 0.7 on CTs in 17 patients, and 2.9 ± 0.8 on MRIs in 18 patients. There was no significant difference in the NC/C ratios between the isolated and the combined disease group (NC/C ratio on TTE: 2.8 ± 1.7 vs. 2.8 ± 0.3 ; $p = 0.227$) either on TTE or MRI (Table 2). In the 17 patients with CT data, the combined disease group showed a higher NC/C ratio than the isolated disease group ($p = 0.010$). In the patients who underwent both TTE and CT ($n = 9$), TTE and MRI ($n = 13$), and CT and MRI ($n = 8$), there were no significant inter-modality differences in the NC/C ratios (Table 3). And, the limits of agreement for inter-modality differences in the NC/C ratios on TTE and CT, TTE and MRI, and CT and MRI are 0.11 ± 1.23 , 0.12 ± 1.24 , and 0.01 ± 0.72 , respectively by Bland-Altman Analysis. Among the 18 patients who received MRI, delayed myocardial enhancement was observed in 6 pa-

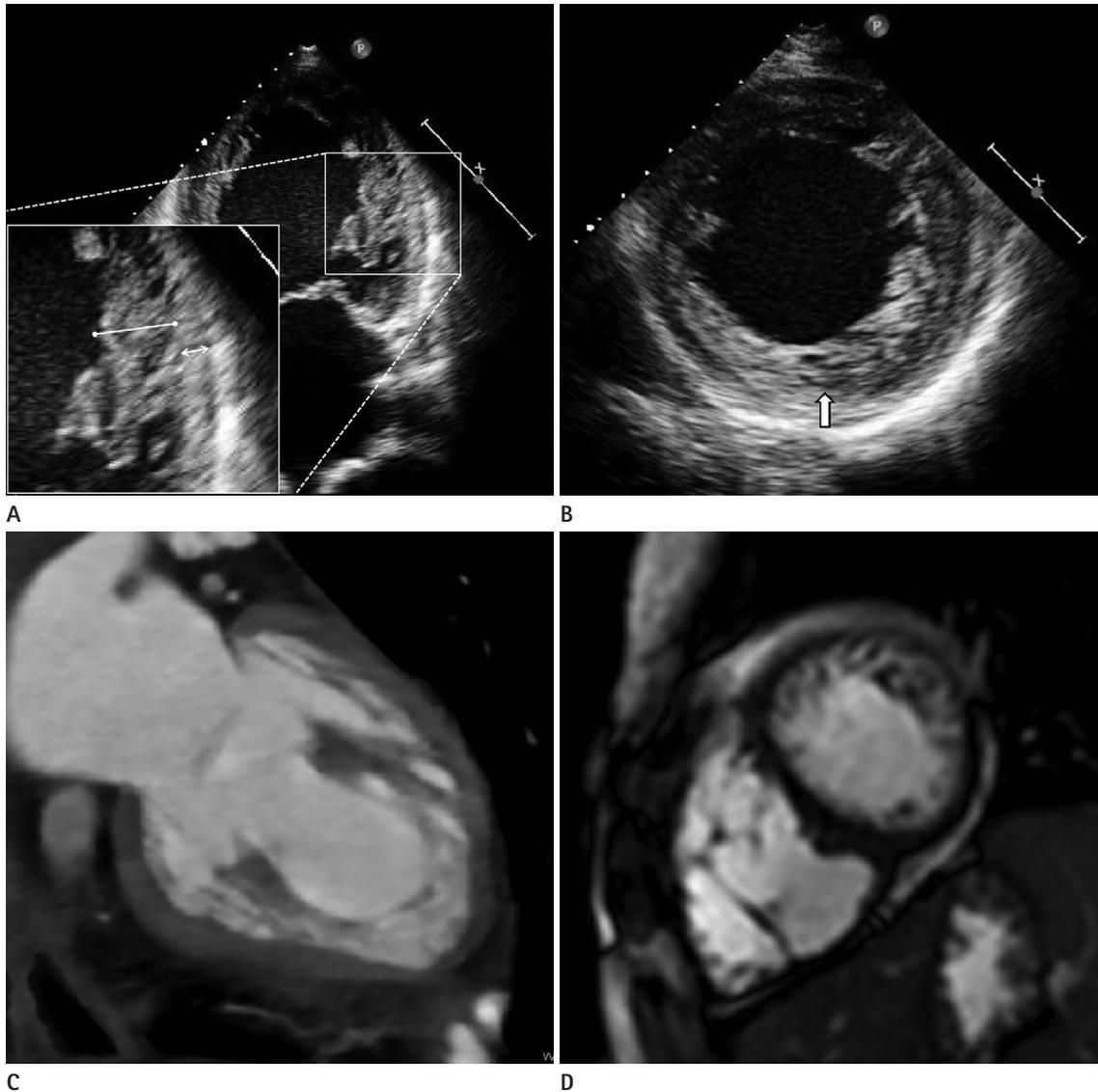


Fig. 1. 63 years old male with typical image findings of left ventricular noncompaction. Long and short axis view on echocardiography at end systolic phase of a 63 years old male hospitalized for dyspnea (**A, B**). Note that extensive trabeculations, two inner compacted (**A**, marking with bidirectional arrow end) and outer noncompact layer (**A**, marking with bidirectional circle end) with different echogenicity, increased the non-compacted layer thickness over the compacted layer thickness ratio, and visible low echogenic inter-trabecular recess (**B**, arrow). The CT (**C**) and MRI (**D**) images at end diastolic phase of a 34 years old female with prominent inner noncompact layer, especially in mid to apical wall of left ventricle.

tients, 4 of whom showed a decreased LV ejection fraction of less than 50% at presentation (Fig. 2).

DISCUSSION

The major findings from our current analysis of clinical and imaging data in LVNC patients collected over 14 years at a single tertiary center were: 1) the combined form of LVNC shows a younger age at diagnosis, more frequent symptoms, and higher frequency of thromboembolic events than the isolated form;

and 2) the mean NC/C ratios of 2.8 ± 0.6 on TTE, 2.7 ± 0.7 on CT, and 2.9 ± 0.8 on MRI indicate no significant inter-modality difference.

As an unknown myocardial disease, LVNC has shown variable clinical findings and its diagnosis is the subject of some controversy. The clinical characteristics of LVNC have been reported in several studies, showing high variability from an asymptomatic state to sudden cardiac death (1, 7, 12-21). The most frequent chief complaint at initial diagnosis was heart failure-related symptoms in both groups of our current study ($n = 38$,

60%), was more frequent in the combined than in the isolated disease group (74% vs. 47%, $p = 0.003$) (3). In our present analysis, the disease was detected incidentally in the absence of symptoms in only 10 (16%) patients, and these cases were more frequent in the isolated than in the combined LVNC group (25% vs. 7%). Thromboembolic events were evident in 20 patients, consistent with the previously reported incidence range from 0 to 38% (1, 3, 12-14). In addition, the incidence of this complication was higher in the combined than in the isolated disease group (39% vs. 26%, $p = 0.279$). Several comorbid ECG abnormal findings have been reported, such as WPW syndrome (3-32%), ventricular tachycardia (15-38%), bundle branch block (5-56%), paroxysmal supraventricular tachycardia and others (1, 3, 12-14, 22). In our current study, a left bundle branch block was the most common ECG finding in the isolated disease group ($n = 9$, 28%). Conversely, atrial fibrillation was the most frequent ECG finding in the combined disease group ($n = 8$, 26%).

Several reports have recently suggested a relatively benign natural course and lower frequency of symptomatic presenta-

tion than the previous notion that NC typically leads to heart systolic dysfunction on follow-up. For example, a recent the MESA-9.5 year follow-up study reported a benign course in the relatively asymptomatic adult population (the so-called "asymptomatic trabeculation"). That report suggested that regular and frequent imaging and clinical follow-up may be unnecessary in subjects with a low pre-test probability of LVNC but with marked trabeculation based on traditional imaging criteria (16, 20, 23). Some reports have also shown that some factors can interfere with the measurement of the NC/C ratio, in particular some mimickers such as a false tendon and aberrant bands (24, 25). On current study population, only 10 among 63 patients (16%) diagnosed as LVNC incidentally. That means the majority of the included population in this study basically focus on the symptomatic patients. We considered that is the main difference about study populations compared to the MESA-9.5 year follow-up study (20).

This study has some limitations of note. First, definite diagnostic and classification criteria for LVNC are still lacking. According to the classification of cardiomyopathy by the American Heart Association, LVNC is considered to be a genetic cardiomyopathy distinguished from other cardiomyopathies. However, some reports have tried to phenotypically subcategorize LVNC as dilated/ hypertrophic/restricted-type LVNC, or categorize according to the involved chamber (23). To that same purpose, we retrospectively included all LVNC patients with/without combined cardiac abnormalities including congenital heart disease, acquired cardiac disease, and other kinds of cardiomyopathy, regardless of disease category. Therefore, some inclusion bias was unavoidable. Furthermore, the difference of the diagnostic criteria among the image modalities take a role as a limitation. Second, since most cases were diagnosed following hospitalization in a large tertiary hospital, the prevalence of LVNC was hard to evaluate and the true incidental findings were probably underestimated. In addition, there was a limitation in the compar-

Table 2. NC/C Ratio on Each Three Image Modality

	Compacted Layer	Noncompacted Layer	NC/C Ratio
TTE			
Total ($n = 51$)	5.9 ± 1.2	16.4 ± 4.2	2.8 ± 0.6
Isolated ($n = 31$)	6.0 ± 1.2	16.7 ± 4.3	2.8 ± 1.7
Combined ($n = 20$)	5.8 ± 1.2	15.9 ± 4.1	2.8 ± 0.3
CT			
Total ($n = 17$)	6.4 ± 2.1	16.6 ± 3.5	2.8 ± 0.7
Isolated ($n = 6$)	8 ± 1.9	16.4 ± 3.3	2.2 ± 0.4
Combined ($n = 12$)	5.6 ± 1.6	17.2 ± 4.3	3 ± 0.7
MRI			
Total ($n = 18$)	5.9 ± 1.6	16.2 ± 3.3	2.9 ± 0.8
Isolated ($n = 6$)	7 ± 1.4	16.5 ± 3.6	2.6 ± 0.6
Combined ($n = 13$)	5.3 ± 1.4	16 ± 3.3	3.1 ± 0.8

Parameters are shown in millimeters, presented as mean ± standard deviation.

NC/C ratio = the ratio of noncompacted layer thickness over the compacted layer thickness, TTE = transthoracic echocardiography

Table 3. Inter-Modality Difference of NC/C between Three Different Image Modality in All Groups

	TTE vs. CT ($n = 9$)			TTE vs. MRI ($n = 13$)			CT vs. MRI ($n = 8$)		
	TTE	CT	p -Value	TTE	MRI	p -Value	CT	MRI	p -Value
Compacted layer	5.6 ± 1.2	6.9 ± 2.3	0.147	5.6 ± 1.3	6.1 ± 1.7	0.370	6.5 ± 2.5	5.6 ± 1.8	0.433
Noncompacted layer	14.3 ± 3.0	17.7 ± 4.5	0.083	15.8 ± 4.6	16.3 ± 2.9	0.440	16.8 ± 3.5	15 ± 2.2	0.247
NC/C ratio	2.6 ± 0.4	2.7 ± 0.9	0.737	2.7 ± 0.3	2.9 ± 0.7	0.592	2.8 ± 0.9	2.8 ± 1.9	0.980

Parameters are shown in millimeters, presented as mean ± standard deviation. For determining the p -value, Independent t -test was used.

NC/C ratio = the ratio of noncompacted layer thickness over the compacted layer thickness, TTE = transthoracic echocardiography

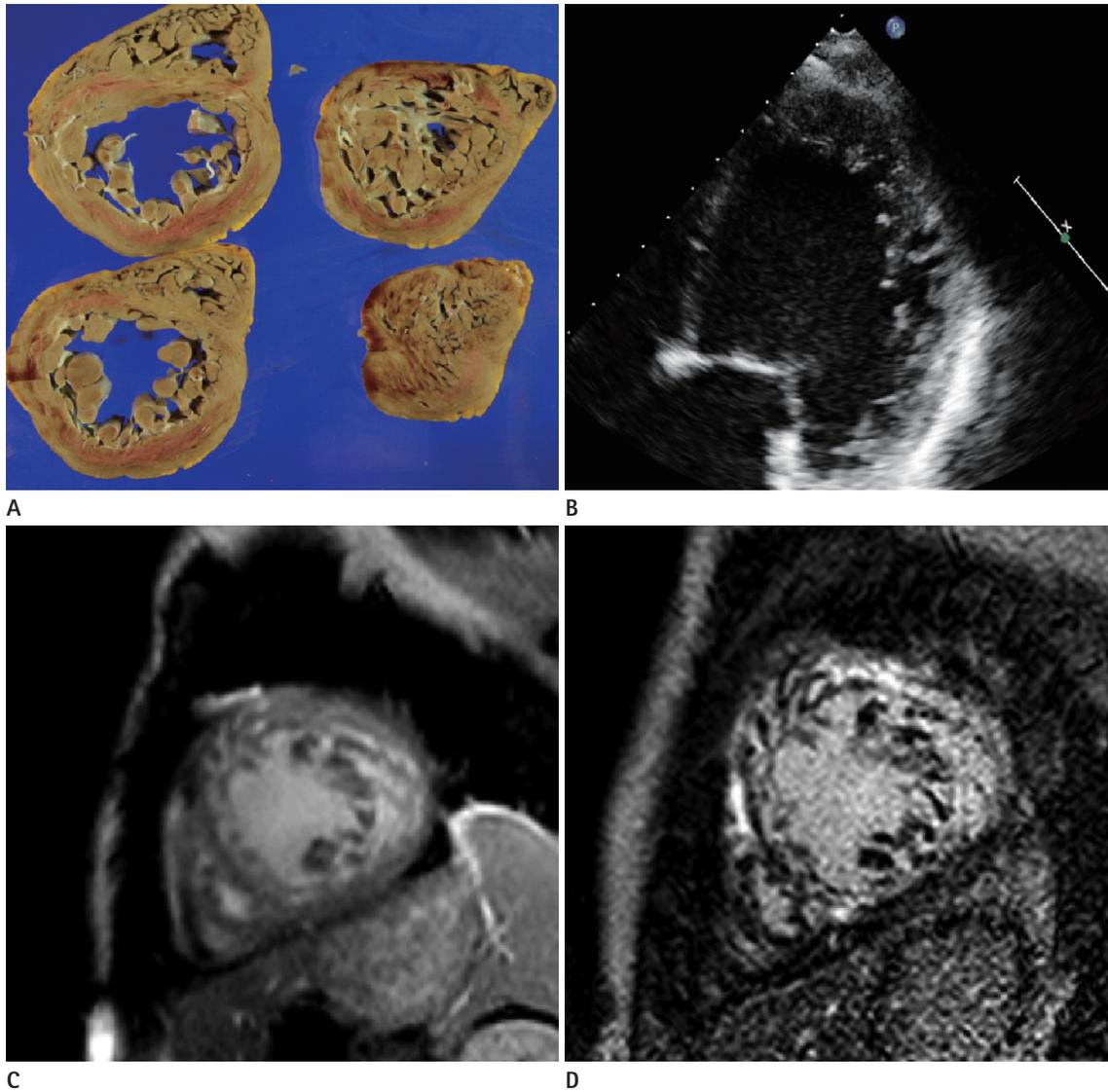


Fig. 2. 31 years old female with LVNC: correlation between the autopsy specimen and image findings. The gross morphology of autopsy specimen **(A)**, TTE finding **(B)**, and MRI findings **(C, D)** of a 31years old female LVNC patient who underwent heart transplantation. There was delayed myocardial enhancement at mid anterior, mid septal, and mid inferior wall on MRI examination **(D)**.
LVNC = left ventricular noncompaction

ative analysis of imaging findings, since only five patients received all image modality simultaneously. Finally, current study was done with retrospective manners.

In conclusion, isolated and combined groups of LVNC showed differences in the age at diagnosis and in the clinical manifestations. The clinical and imaging findings of LVNC presented in this study may assist in the better understanding of LVNC.

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성인 좌심실 비치밀화증: 성인 63명의 영상의학적 소견과 임상양상

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목적: 성인 심실 비치밀화증 환자의 영상의학적 소견과 임상양상을 기술하고자 하였다.

대상과 방법: 2000~2014년까지 본원에서 심 초음파, CT, 그리고 MRI 검사를 통해 63명의 환자가 심실 비치밀화증으로 진단되었다. 해당 심실 비치밀화증 환자를 대상으로 기본 질환 특징, 임상 양상, 동반 심 질환, 영상의학적 소견을 분석하였다. 그리고 동반 심 질환의 유무에 따라 단독 질환군과 복합 질환군을 구분하여 비교하였다.

결과: 63명의 환자 중 동반 심 질환이 없는 단독 질환군 환자는 32명(51%)이었다. 단독 질환군에서 복합 질환군에 비해 처음 진단될 당시의 나이가 많았다(54.2세 vs. 40.2세, $p < 0.001$). 복합 질환군에서 단독 질환군에 비해 진단 당시 증상이 더 많았다(94% vs. 75%, $p = 0.082$). 두 그룹 모두에서 심부전증과 연관된 증상이 가장 많았다(60.3%). 전체 환자에서 혈전색전증이 발생한 경우는 20명이었는데, 복합 질환군에서 단독 질환군보다 많았다(39% vs. 26%, $p = 0.279$). 동반 심 질환 중 가장 흔한 것은 확장성 심근병증이었다($n = 15$, 24%). 영상의학적 소견상 두 질환군의 비치밀화/치밀화 비는 유의한 차이가 없었다.

결론: 성인 심실 비치밀화증은 단독 질환군과 복합 질환군, 두 그룹 간에 진단 당시의 나이와 임상양상에 차이가 있다. 임상양상과 영상의학적 소견은 심실 비치밀화증에 대한 이해에 도움이 될 것이다.

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