Left Ventricular Non-Compaction Associated With Patent Ductus Arteriosus

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Isolated left ventricular non-compaction (LVNC) is a rare congenital anomaly. It is characterized by numerous prominent ventricular trabeculations and deep intertrabecular recesses. The incidences of associated cardiovascular complication and congenital heart disease are high. It is associated with congestive heart failure, ventricular arrhythmia, embolic events, and distinctive facial dysmorphism. Familial tendency has also been reported. It is believed to represent an arrest in endomyocardial morphogenesis. The prognosis for patients is grim. The most common cause of death in these patients was sudden cardiac collapse. Many patients suffered from transient ischemic attacks, pulmonary embolisms, heart failure events, pulmonary edema episodes, cardiogenic shock and sustained ventricular tachycardia. Even asymptomatic patients were still at risk of unpredictable ventricular arrhythmia and sudden cardiac collapse. We report a 36-year-old female with patent ductus arteriosus (PDA) and an incidental finding of left ventricular non-compaction. As far as we know, this is the first report of left ventricular non-compaction associated with PDA.

Key Word: Left ventricular non-compaction

INTRODUCTION

Isolated left ventricular non-compaction (LVNC) is a rare congenital anomaly. It is characterized by numerous prominent ventricular trabeculations and deep intertrabecular recesses.

The incidence of cardiovascular complication is high. It is associated with congestive heart failure, ventricular arrhythmia, embolic events, and distinctive facial dysmorphism. Familial tendency has also been reported. It is believed to represent an arrest in endomyocardial morphogenesis. Previous investigators have viewed it as either persisting sinusoids or spongy myocardium. A study of 8 children published by Chin et al.1 demonstrated the presence of a continuous layer of endothelium from the ventricular cavity into the recesses without coronary communication to the ventricular cavity.

LVNC has also been associated with various congenital heart diseases. The prognosis for patients is grim: according to a Mayo clinic report,2 59% of patients with isolated ventricular noncompaction had died or had a heart transplant within 6 years of diagnosis. The most common cause of death in these patients was sudden cardiac collapse. Many patients suffered from transient ischemic attacks, pulmonary embolisms, heart failure events, pulmonary edema episodes, cardiogenic shock and sustained ventricular tachycardia. Even asymptomatic patients were still at risk of unpredictable ventricular arrhythmia and sudden cardiac collapse.3-9

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tricular non-compaction. As far as we know, this is the first report of left ventricular non-compaction associated with PDA.

CASE REPORT

A thirty-year-old female came to cardiology services for further evaluation of an incidentally found cardiac murmur during a routine physical check-up. Her vital signs were unremarkable. We found no preceding episodes of syncope, embolic, or heart failure events in her medical history review.

A typical continuous murmur was found in the precordial pulmonic area. We carried out a transthoracic echocardiography (TTE) study to search for the presence of PDA. Transthoracic echocardiographic study showed an abnormal continuous shunt flow from the aorta into the main pulmonary artery. These findings suggested the presence of PDA. At the same time, we noted prominent left ventricular trabeculae and deep intertrabecular recesses. Color Doppler echocardiography showed the color flow traversed within the left ventricle between these abnormally hypertrophic trabeculae (Figure 1). We found no left ventricular (LV) or right ventricular (RV) outflow tract obstruction. Transesophageal echocardiography (TEE) showed abnormally hypertrophic muscle bundles occupying nearly 2-thirds of the left ventricle cavity space, with flow in and out between inter-trabecular areas.

She reported no episodes of sudden death, facial dysmorphism or thromboembolic events among her family members or herself. She was physically well developed, and her mental condition was normal. The results of an electrocardiogram (ECG) were unremarkable. Borderline cardiac shadow size, left atrial enlargement, and pulmonary vessel prominence appeared in the chest film.

Through a review of the relevant medical literature, we found that lethal arrhythmia and occult myocardial ischemia were usually reported. We arranged the various studies referred to below for the patient.

An ambulatory Holter ECG showed basically normal sinus rhythm with rare ventricular premature beat (VPC) and rare atrial premature beat (APC). No lethal ventricular arrhythmia was recorded. Thallium-201 myocardial scan after a persantin injection showed no definite evidence suggestive of perfusion defects or focal myocardial ischemia.

The patient also received a magnetic resonance im-

Figure 1. Transthoracic 2-dimensional echocardiography: (A) Parasternal short-axis view end-diastolic phase showed plentiful hypertrophic trabecular muscles traversing within the left ventricular cavity. (B) Modified apical view end-diastolic phase showed plentiful hypertrophic trabecular muscles traversing within the left ventricular cavity, especially toward apex. (C) Transthoracic color Doppler echocardiography short-axis view showed intracardiac flow traversing in and out of the left ventricle between the hypertrophic trabecular muscles.
age (MRI) study that showed numerous prominent trabeculae and deep intertrabecular recesses (Figure 2). The angiographic left ventriculography is shown in Figure 3.

She received right ventricular endocardial biopsy. Light microscopic examination of the endocardial biopsy specimen revealed focal widening of the perivascular space and focal organizing of the mural thrombus. Subendocardial fibroelastosis or fibrosis was absent according to Verhoeff van Gieson and Masson-trichrome stains. Ultrastructural examination by transmission electron microscope (Figure 4) showed perinuclear cytoplasmic clearing due either to a markedly reduced content of myofibrils or an accumulation of glycogen with relatively increased mitochondria in some of the myocytes. Large subsarcolemmal vacuoles were also identified.

All of the above findings supported the clinical diagnosis of left ventricle non-compaction (LVNC) associated with PDA. The narrowest diameter was measured about 5 mm by aortogram. It was a left-to-right continuous shunt from the aorta into pulmonary artery. The estimated QP over QS was about 1.46. The patient received Cook coil (MWCE-38-12-10, 10 mm in length) embolization for the PDA. The result was good and she was regularly followed up at outpatient department.

The patient has 1 son and 1 daughter. The son was also found to have LVNC and 1 muscular interventricular septal defect (VSD). The daughter was found to be normal.

**DISCUSSION**

LV non-compaction has almost always been associated with other congenital anomalies, including obstructive LV or RV outflow tracts, and an anomalous origin of the left coronary artery from the pulmonary artery. Various forms of outflow tract obstruction during fetal life, such as left ventricular outflow tract obstruction or pulmonary atresia...
with intact interventricular sputum, are responsible for the persistence of deep endomyocardial spaces surrounded by exaggerated hypertrophy of the trabeculae. This non-compaction of the myocardium has been found in ventricles exposed to excessively high pressures during intrauterine development.

In contrast, the reasons for isolated non-compaction are unknown, and it rarely occurs. However, according to many previous reports, patients with isolated ventricular non-compaction have a high risk of progressive left ventricular dysfunction (around 60%), ventricular arrhythmia (around 60%), and embolic events (around 40%). In the Chin et al. series, facial dysmorphism, characterized by prominent foreheads, strabismus, low-set ears, high arched palates and micrognathia, was reported. The incidence of facial dysmorphism in isolated ventricular non-compaction was about 40% (3 in 8 patients). However, in the Mayo clinic report, the figure was lower (2 in 17 patients).

Electron microscopy has been performed in only a few cases and was either normal or showed abnormal mitochondrial. Our case reveals some new information about ultrastructural findings which has not been described before, such as perinuclear cytoplasmic clearing due to a markedly reduced content of myofibrils or accumulation of glycogen and large subsarcolemmal vacuoles. However, the significance of these ultrastructural findings is unknown. Since similar ultrastructural findings have not been described in the literature and this is our first experience, the significance of the ultrastructural findings cannot be more specifically stated. Perhaps, detailed ultrastructural analysis on more cases in the future will give answer to the significance of the ultrastructural findings.

The cause of left ventricular dysfunction is not clear. Epicardial coronary artery was intact in almost all reported cases, but intramural hypoperfusion was suspected. The increased levels of elastic and fibrous tissues in trabeculae and recesses might be associated with subendocardial ischemia. The underlying mechanism might be failure of the coronary microcirculation to grow with the increase in ventricular mass, or compression of the intramural coronary vascular bed by the hypertrophied myocardium, or a combination of both mechanisms.

Junga et al. performed positron emission tomography using N-13 ammonia as a flow marker and intravenous dipyridamole for stress testing in 5 patients with isolated ventricular non-compaction. The preliminary results demonstrated that subendocardial ischemia was the 1 of the causes of ventricular dysfunction. Their report supports the conclusion that subendocardial ischemia is 1 of the reasons for ventricular dysfunction and ventricular arrhythmia. The incidence of prolonged QT dispersion and the occurrence of late potential were high in patients with isolated ventricular non-compaction (3 in 5 patients). This might explain why ventricular arrhythmia occurs. In the case under consideration here, there was neither sign of myosclerosis nor lethal arrhythmia.

REFERENCES

左心室 Non-compaction 合併開放性動脈管（Patent Ductus Arteriosus）
個案報告及文獻回顧

蕭世宏 1 李道興 1 馬光遠 1 劉俊鵬 1 彭南靖 2
王志生 3 吳銘庭 4 謝凱生 5
高雄市 高雄榮民總醫院 內科部 心臟內科 1 核子醫學部 2 病理科 3 放射線部 4 兒童醫學部 5

單獨的左心室 non-compaction 在先天性心臟疾病是很少見的，其特徵是具有粗大的心室肌
肉束（trabeculation）及心室肌肉束之間有深層的凹陷，心血管系統上的併發症發生率很高，
回顧先前的文獻報告，可以看出其常合併有其他先天性心臟病、心衰竭，發生致病的心室
性心律不整，血栓形成及特殊的臉部畸型，有家族遺傳的傾向。現在報告一位 36 歲女性，
有開放性動脈管及左心室 non-compaction。本病例為臺灣地區第一例有關左心室 non-
compaction 的個案報告。病患為 36 歲女性，於理學檢查發現心雜音，於是進一步檢查。其
生命跡象是正常狀態。沒有暈倒、心衰竭、猝死病史。心電圖及胸部 x 光片檢查並無異常
之處。於聽診時發現於肺動脈有連續性雜音。於是患者接受經胸前心臟超音波，發現有異
常的血流從主動脈到肺動脈，診斷為開放性動脈導管。同時意外發現患者之左心室內有粗
大的肌肉束（trabeculum）形成，其間有深層的凹陷。彩色都卜勒心臟超音波檢查顯示有不
正常的血流穿梭於這些不正常之肥大的肌肉束間，此外並沒有左心室或右心室出口道阻塞
現象。此病人家庭成員中無猝死案例，亦無臉部的畸型或血栓形成阻塞血管等病史，病人的
生理狀況及智能皆正常。24 小時心心電記錄並未發現重大之心律不整。核子醫學心臟灌
流檢查並未發現重大之心肌缺氧病變。病患接受心臟核磁共振及心導管檢查後，確定是左
心室 non-compaction（LVNC）合併開放性動脈導管。此病人後續接受開放性動脈導管的栓
塞手術治療。現況良好。據文獻回顧，左心室 non-compaction 的特徵是粗大的肌肉束形成
佈滿於左心室腔內。一般相信是胚胎演化抑制至異常形態所造成。此病常會合併有心室先
天異常，包括左心室或右心室出口道阻塞現象，從肺動脈到左冠狀動脈有不正常的血流來
源，也常見心衰竭、心室性心律不整、中風等，部分患者會出現特殊的臉部畸型。致死原
因與心衰竭及心室性心律不整有關。本疾病預後不佳。然而在我們的病人，並無心衰竭、
心律不整跡象，至今仍規則於門診追蹤中。現況良好。患者育有一兒一女，兒子亦被發現
有左心室 non-compaction 現象且合併心室中隔缺損。女兒則完全正常。

關鍵詞：左心室 non-compaction。