Successful closure of ventricular septal defect in a patient with noncompaction of the left ventricular myocardium

Sol ventrikülde miyokard nonkompaksiyonu olan bir hastada ventriküler septal açıklığın başarılı tamiri

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Left ventricular noncompaction (LVNC) is a rare clinical condition characterized by numerous prominent intertrabecular recesses in the ventricle, and is an unclassified cardiomyopathy. The pathology has been almost invariably associated with other congenital cardiac malformations. We presented a two-year-old patient with LVNC accompanied by ventricular septal defect (VSD). Two-dimensional echocardiography showed a perimembranous VSD, 8 mm in size, and LVNC. The ratio of left ventricular noncompacted/compacted myocardial layers was 4.2. Since there was no evidence for cardiomyopathy, the patient underwent open heart surgery for the closure of VSD. Four months after the operation, echocardiographic findings showed normal myocardial performance.

Key words: Echocardiography; cardiomyopathies; heart defects, congenital/complications/surgery; heart septal defects, ventricular; heart ventricles.

Sol ventrikül nonkompaksiyonu, ventrikülün trabeküllü alanlarında çok sayıda boşlukla karakterize, nadir bir anomalidir; sınıflandırılmamış kardiyomiyopatiler içinde yer alır. Patolojiye çoğu kez başka doğumsal kalp malformasyonları eşlik eder. Bu yazıda, sol ventrikül nonkompaksiyonuna ventriküler septal defektin (VSD) eşlik ettiği iki yaşında bir hasta sunuldu. Hastanın ikiboyutlu ekokardiyografi ile incelenmesinde 8 mm büyüklüğünde perimembranöz bir VSD ve sol ventrikül nonkompaksiyonu saptandı. Sol ventrikülün nonkompakte/kompakte miyokard tabakalarının oranı 4.2 olarak hesaplandı. Hastada herhangi bir kardiyomiyopati bulgusu olmadığından, VSD'nin kapatılması amacıyla açık kalp ameliyatı yapıldı. Ameliyattan dört ay sonraki ekokardiyografik incelemede miyokard performansı normal bulundu.

Anahtar sözcükler: Ekokardiyografi; kardiyomiyopati; kalp defekti, doğuştan/komplikasyon/cerrahi; kalp septal defekti, ventriküler; kalp ventrikülü.

During the early phase of the embryo, ventricular intertrabecular spaces communicate with the ventricular cavity. As the heart develops, the myocardium becomes compacted and the mesh-like morphology disappears. Left ventricular noncompaction (LVNC), which is known as an unclassified cardiomyopathy, is thought to result from an arrest in endomyocardial morphogenesis. It is characterized by excessively prominent trabeculations in the affected ventricle(s), sometimes affecting the right ventricle and interventricular septum, as well.[1,2] This anomaly is generally associated with congenital obstructive lesions of the left or right ventricular outflow tract, such as pulmonary atresia with intact ventricular septum and aortic atresia.[3,4] Noncompaction of ventricular myocardium can be seen as an isolated malformation or associated with other congenital cardiac malformations.

Herein, we present a patient with LVNC concomitant with ventricular septal defect (VSD), who was successfully treated surgically via cardiopulmonary bypass (CPB). To our knowledge, this is the first case of LVNC concomitant with VSD that was surgically corrected by open heart surgery.

CASE REPORT

A two-year-old patient was referred to our institution due to suspicion of a congenital cardiac defect. His heart rate was 110 beats/min in electrocardiography and a chest x-ray showed normal situs and an enlarged heart with a cardiothoracic ratio of 0.55. Two-dimensional echocardiography (2D-ECHO) showed a perimembranous VSD, 8 mm in size, and LVNC (Fig. 1a, b). Fortunately, left ventricular systolic function was not depressed and ejection fraction was 68%; however, there was a prominent trabeculation and recesses of left ventricle. The ratio of left ventricular noncompacted/compacted myocardial layers was 4.2, being diagnostic for noncompac-

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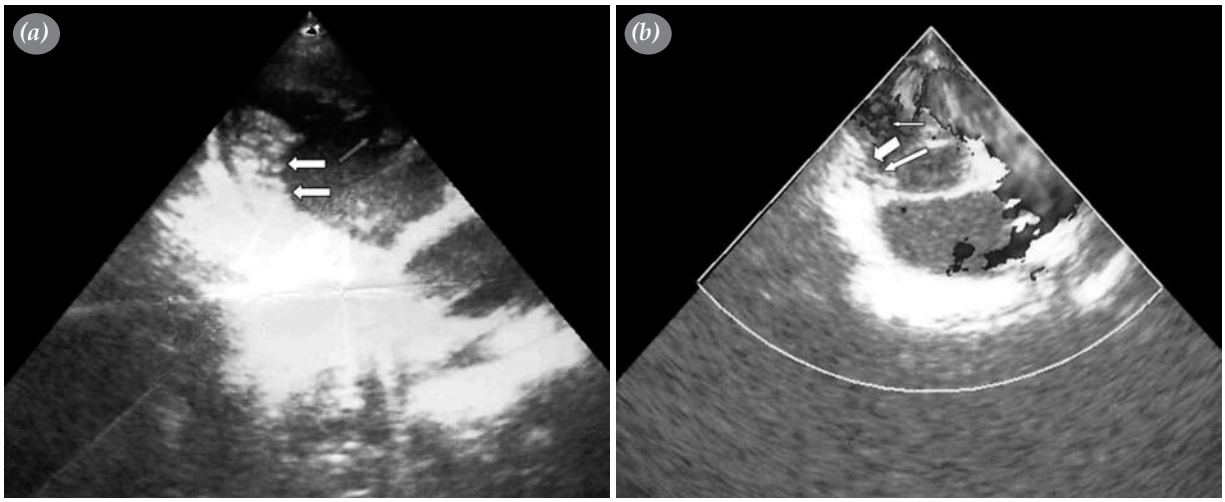

Fig. 1. Two-dimensional echocardiograms showing left ventricular noncompaction. **(a)** Parasternal long-axis view demonstrates a thin epicardial layer and an extremely thickened endocardial layer with prominent left ventricular trabeculations (white arrowheads), deep recesses in the apex and posterior wall of the ventricle and ventricular septal defect (gray arrowhead). **(b)** Modified parasternal long-axis view clearly shows prominent trabeculations and deep recesses (arrows) in the left ventricular free wall.

tion. Data recorded at the catheterization laboratory were as follows: PA 57/23 mmHg (mean 36 mmHg), left ventricle 98 mmHg, right ventricle 14 mmHg, Ao 92/54 mmHg (mean 74 mmHg). There was no mitral or tricuspid valve insufficiency. The perimembranous VSD and left ventricular hypertrabeculation were also confirmed during cardiac catheterization (Fig. 2). Since there was no evidence for cardiomyopathy, the patient underwent open heart surgery for the closure of VSD. The defect was closed with the use of a Dacron patch and the patient was weaned from CPB. Inotropic support with dopamine or dobutamine was not required after the weaning from CPB. The patient's general and clinical condition improved after the operation. Postoperative echocardiography showed normal ejection fraction, and the patient was discharged from the hospital on the tenth postoperative day with a good clinical condition. Four months after the operation, echocardiographic findings showed normal myocardial performance.

DISCUSSION

In the early phase of embryogenesis, the heart consists of a loose network of muscle fibers that normally condense gradually, and it becomes compacted with the disappearance of large intertrabecular spaces. It is thought that ventricular noncompaction represents an arrest in this process. Sengupta et al.[5] demonstrated that the myocardium was essentially 2-layered in the areas of noncompaction, with the noncompacted endocardium-to-epicardium ratio of ≥2. They measured the thickness during end-systole for better visualization of LVNC.[5] In our case, the ratio of left ventricular noncompacted/compacted myocardial layers was 4.2.

Ventricular noncompaction may occur in isolation (isolated ventricular noncompaction) or may be associated with other congenital cardiac malformations.[6,7] Echocardiography, magnetic resonance imaging, and angiography are the main imaging tools to diagnose

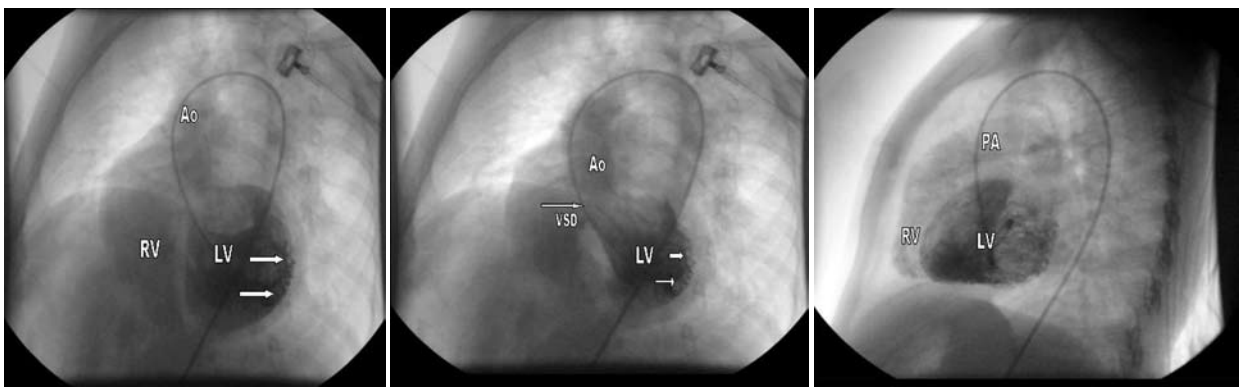

Fig. 2. In the diastolic and systolic phases, anteroposterior and lateral views of cardiac catheterization clearly demonstrate hypertrabeculation and deep intertrabecular recesses in the left ventricle (arrowheads). VSD: Ventricular septal defect; Ao: Aorta; RV: Right ventricle; LV: Left ventricle.

ventricular noncompaction, but two-dimensional color Doppler echocardiography is the standard and first-line diagnostic tool. Despite an increasing awareness, there is still little knowledge regarding the diagnosis of this rare cardiomyopathy. The diagnosis of LVNC may easily be overlooked or delayed in particular cases presenting with heart failure due to depressed ventricular function. Delay in diagnosis is due to similarities between LVNC and other cardiomyopathies, and to the lack of familiarity of the referring physicians with its specific diagnostic pattern.

This rare abnormality is reported to be accompanied by three major conditions, namely, depressed ventricular function, systemic embolization, and ventricular arrhythmias that may sometimes be fatal. Because depressed ventricular function is diagnosed in the late period, patients are sometimes referred to clinics with the complaint of heart failure. Noncompaction primarily affects the left ventricle, but the right ventricle may also be involved.[4,8,9] Asymptomatic cases have normal systolic function and increased left ventricular end-diastolic pressure, mimicking restrictive cardiomyopathy. Most patients are initially asymptomatic,[10] though there are symptomatic cases.[1]

Similarities between LVNC and other cardiomyopathies sometimes make the differential diagnosis difficult. Familiarity of cardiologists with the pathology ensure early and correct diagnosis. Prominent multiple myocardial trabeculations and intertrabecular spaces within this trabecular structure which continue to the ventricular cavity are typical echocardiographic and angiocardiographic findings of LVNC.

The treatment of myocardial noncompaction is not different from that of other cardiomyopathies. Medical treatment is preferred at the beginning of congestive heart failure. Diuretics, angiotensin-converting enzyme inhibitors, and digitalis are preferred for medical treatment in patients with noncompaction of ventricular myocardium. In our case, medical treatment was not started because the patient had no symptoms of the myocardial failure during the preoperative and postoperative period. Transthoracic echocardiography, which shows clear and valuable data about systolic and diastolic functions, was performed periodically to monitor the status of myocardial performance.

To the best of our knowledge, no surgically treated case of LVNC combined with VSD has been reported previously. There is only one case report from our institution, in which congenitally corrected transposition of the great arteries accompanied by noncompaction of both ventricles was treated surgically under CPB.[2]

In conclusion, patients with VSD may have ventricular noncompaction without any manifestation of cardiomyopathy. Surgical correction should be performed in a single session in the early period of life. In these cases, progressive myocardial insufficiency may complicate the clinical condition, and may cause accelerated clinical deterioration. In addition, noncompaction should be suspected and investigated whenever ventricular function is depressed irrelevant to the accompanying heart defect, and if detected, appropriate medical management should be initiated to improve the myocardial performance in the early period of myocardial failure.

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