

Coexistence of left ventricular noncompaction and double-orifice mitral valve in a patient with congestive heart failure

Fikri Demir, Semra Atalay, Cem Karadeniz, Ercan Tutar

Division of Pediatric Cardiology, Department of Pediatrics, Ankara University Faculty of Medicine, Ankara, Turkey.
E-mail: fikridemir@yahoo.com

SUMMARY: Demir F, Atalay S, Karadeniz C, Tutar E. Coexistence of left ventricular noncompaction and double-orifice mitral valve in a patient with congestive heart failure. Turk J Pediatr 2012; 54: 444-446.

Left ventricular noncompaction (LVNC) is a rare cardiomyopathy. It usually presents with ventricular dysfunction, thromboembolic events and arrhythmias. An asymptomatic clinical course is also possible. LVNC is frequently associated with other congenital heart diseases including heart valve abnormalities. The coexistence of LVNC with double-orifice mitral valve was observed rarely. The presence of such coexistence in a patient presented with heart failure in the newborn period is reported herein because of its rarity.

Key words: noncompaction cardiomyopathy, double-orifice mitral valve, heart failure.

Left ventricular noncompaction (LVNC) is a rare cardiomyopathy in which the ventricular myocardium has a spongy appearance due to prominent trabeculations and deep intertrabecular recesses. It may be asymptomatic or present with ventricular dysfunction, thromboembolic events and arrhythmias^{1,2}. LVNC is frequently associated with other congenital heart diseases including heart valve abnormalities^{3,4}. The coexistence of LVNC with double-orifice mitral valve (DOMV) in a patient is reported herein because of its rarity.

Case Report

A male newborn was admitted to a local hospital with poor sucking, cyanosis, and respiratory distress on the second day of life. He had been diagnosed with atrial septal defect (ASD), patent ductus arteriosus, third-degree mitral regurgitation, and heart failure. He had been treated with digoxin, dopamine, and furosemide, and mechanically ventilated for four days. He had no history of perinatal asphyxia. The investigations for metabolic and infectious causes and cardiac catheterization performed to rule out any coronary anomaly had all been inconclusive.

He was referred to our institution for further investigations on the 32nd day of life. His heart and respiratory rates were 130 and 60 per minute, respectively. An apical 3/6

degree holosystolic murmur was heard. The chest X-ray revealed cardiomegaly, and the electrocardiography showed left ventricular hypertrophy. Echocardiographic examination revealed enlarged left heart chambers, trabeculations and deep intertrabecular recesses in the middle portion of the left ventricular posterior wall and apex (LVNC) (Figs. 1, 2), decreased systolic function (ejection fraction: 30%), and DOMV (Fig. 3) with moderate mitral and aortic regurgitation. The myocardial noncompacted to compacted layer thickness ratio was measured as 2.55. Serum brain

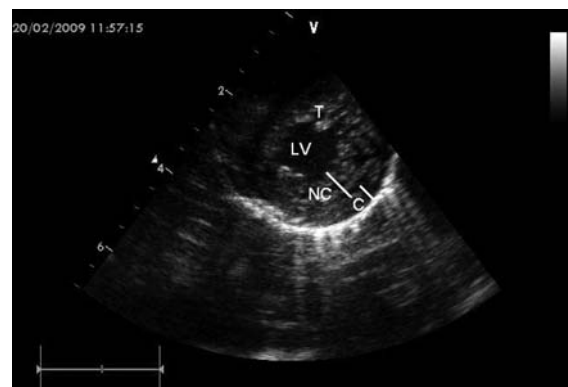


Figure 1. The two-dimensional echocardiographic appearance of left ventricular noncompaction on the parasternal short-axis view. C: Compacted layer. LV: Left ventricular cavity. NC: Noncompacted layer. T: Trabeculations.



Figure 2. The color Doppler echocardiographic appearance of left ventricular noncompaction on the parasternal short-axis view. C: Compacted layer. LV: Left ventricular cavity. NC: Noncompacted layer. T: Trabeculations. The arrow indicates the blood flow from the ventricular cavity into the intertrabecular recesses.

natriuretic peptide (BNP) level was 22110 picograms per milliliter. We did not observe any arrhythmia. He was discharged in a good clinical condition after 11 days of hospitalization under the treatment of digoxin, furosemide, enalapril, and acetylsalicylic acid. LVNC was not determined in any family member by echocardiographic screening.

After two years of follow-up, he was symptom-free with anticongestive treatment. Serum BNP level was normal. Aortic regurgitation disappeared, and he had mild mitral regurgitation and near-normal systolic functions.

Discussion

Left ventricular noncompaction (LVNC) is a genetic cardiomyopathy characterized by prominent trabeculations and deep intertrabecular recesses within the ventricular myocardium. Although it may be asymptomatic, heart failure is a frequent presenting feature, as in our patient^{1,2}.

During early embryogenesis, the ventricular wall is composed of loosely arranged myocardial fibers, which condense and later form a normal compact myocardium. It is supposed that an arrest in this process results in noncompaction of the ventricular myocardium, which has an appearance resembling initial developmental stages^{1,5,6}. The presentation of a two-day-old patient with heart failure is consistent with a clinical course that begins antenatally and manifests itself in the early neonatal period.

Mitral leaflets are derived from endomyocardial cushions, which form the papillary muscle and chordae together with primitive trabeculations. Pathomorphologic examinations of the hearts with DOMV have shown various subvalvar apparatus anomalies. Thus, subvalvar developmental abnormality may have a pivotal role in the pathogenesis of DOMV. Abnormal endomyocardial differentiation also results in persistence of primitive trabeculations and LVNC^{5,7,8}. The frequent association of LVNC with other cardiac malformations including DOMV necessitates searching for all those anomalies.

The coexistence of DOMV and LVNC is extremely rare. Sugiyama et al⁵ reported two children: one was admitted with heart failure at three years of age and the other had ASD, ventricular septal defect (VSD) and first-degree atrioventricular block. The other presented cases were of adult age^{5,9,10}. Unlike the previously reported cases, our patient presented with heart failure at an earlier age and had a higher degree of mitral regurgitation. As the mitral regurgitation decreased with clinical improvement, the principal cause of regurgitation was thought to be the mitral annular dilatation rather than the DOMV itself.

Although it was speculated that early presentation in infancy in patients with LVNC was associated with increased risk of mortality, our patient has been doing well for two years. Neither arrhythmias nor thromboembolic events were determined^{2,3}.

In conclusion, since developmental processes involving formation of the mitral valve and

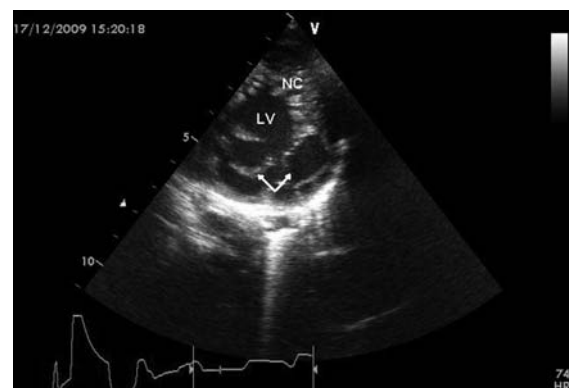


Figure 3. Parasternal short-axis view shows double-orifice mitral valve and noncompacted myocardium. LV: Left ventricle. NC: Noncompacted layer. The arrows indicate the two orifices of the mitral valve.

compaction of the left ventricular myocardium may be pathogenetically related, patients with LVNC may have mitral valve abnormalities, and this should be kept in mind during the echocardiographic examination.

REFERENCES

1. Chin TK, Perloff JK, Williams RG, Jue K, Mohrmann R. Isolated noncompaction of left ventricular myocardium. A study of eight cases. *Circulation* 1990; 82: 507-513.
2. Oechslin EN, Attenhofer Jost CH, Rojas JR, Kaufmann PA, Jenni R. Long-term follow-up of 34 adults with isolated left ventricular noncompaction: a distinct cardiomyopathy with poor prognosis. *J Am Coll Cardiol* 2000; 36: 493-500.
3. Tsai SF, Ebenroth ES, Hurwitz RA, Cordes TM, Schamberger MS, Batra AS. Is left ventricular noncompaction in children truly an isolated lesion? *Pediatr Cardiol* 2009; 30: 597-602.
4. Hughes ML, Carstensen B, Wilkinson JL, Weintraub RG. Angiographic diagnosis, prevalence and outcomes for left ventricular noncompaction in children with congenital cardiac disease. *Cardiol Young* 2007; 17: 56-63.
5. Sugiyama H, Hoshiai M, Toda T, Nakazawa S. Double-orifice mitral valve associated with noncompaction of left ventricular myocardium. *Pediatr Cardiol* 2006; 27: 746-749.
6. Breckenridge RA, Anderson RH, Elliott PM. Isolated left ventricular non-compaction: the case for abnormal myocardial development. *Cardiol Young* 2007; 17: 124-129.
7. High FA, Epstein JA. The multifaceted role of Notch in cardiac development and disease. *Nat Rev Genet* 2008; 9: 49-61. Review.
8. Bano-Rodrigo A, Van Praagh S, Trowitzsch E, Van Praagh R. Double-orifice mitral valve: a study of 27 postmortem cases with developmental, diagnostic and surgical considerations. *Am J Cardiol* 1988; 61: 152-160.
9. Gorgulu S, Celik S, Eksik A, Tezel T. Double-orifice mitral valve associated with nonisolated left ventricular noncompaction--a case report. *Angiology* 2004; 55: 707-710.
10. Wang XX, Song ZZ. A combination of left ventricular noncompaction and double-orifice mitral valve. *Cardiovasc Ultrasound* 2009; 7: 11.