Isolated noncompaction of left ventricular myocardium with fetal sustained bradycardia due to sick sinus syndrome

Süheyla Özkutlu[1], Lütfü Önderoğlu[2], Tevfik Karagöz[1], Alpay Çeliker[1]
Ümit Murat Şahiner[1]

[1]Section of Cardiology, Department of Pediatrics and [2]Department of Obstetrics and Gynecology, Hacettepe University Faculty of Medicine, Ankara, Turkey

SUMMARY: Özkutlu S, Önderoğlu L, Karagöz T, Çeliker A, Şahiner ÜM. Isolated noncompaction of left ventricular myocardium with fetal sustained bradycardia due to sick sinus syndrome. Turk J Pediatr 2006; 48: 383-386.

Noncompaction of the ventricular myocardium is a rare congenital cardiomyopathy resulting from an arrest in normal endomyocardial embryogenesis. This disease is characterized by numerous and prominent trabeculations and deep intertrabecular recesses. It may be isolated or associated with other congenital heart diseases. The disorder is clinically accompanied by depressed ventricular function, systemic embolization and arrhythmias. Sustained bradycardia is infrequent in prenatal life and associated with maternal collagen vascular diseases, structural heart diseases or long QT syndrome. Herein we report a case of isolated noncompaction of left ventricular myocardium diagnosed in the first day of life and followed previously by serial fetal echocardiograms for the sustained sinus bradycardia. On postnatal electrocardiography, there was junctional escape rhythm due to profound sinus bradycardia, leading us to consider sick sinus syndrome. To our knowledge, this is the first case in the literature of isolated noncompaction of left ventricular myocardium with sustained bradycardia due to sicksinus syndrome.

Key words: fetal echocardiography, isolated noncompaction of ventricular myocardium, cardiomyopathy, sustained bradycardia.

Noncompaction of the ventricular myocardium is a rare disorder characterized by numerous, excessively prominent trabeculations and deep intertrabecular recesses in one or more ventricular wall segments. The disorder may be familial and associated with distinctive facial dysmorphism[1,2]. Diagnosis can be made by echocardiography from its characteristic morphological features. The disorder is clinically accompanied by depressed ventricular function, systemic embolization and arrhythmias.

Persistent fetal bradycardia is infrequent in prenatal life. The mechanisms of bradycardia include sinus bradycardia, blocked atrial extrasystoles and complete heart block[3-6]. Sustained bradycardia is usually associated with maternal collagen vascular diseases like systemic lupus erythematosus, structural heart diseases and long QT syndrome[5,6].

Herein we report a case of isolated noncompaction of ventricular myocardium (INCVM) diagnosed on the first day of life and followed previously by serial fetal echocardiograms for the sinus bradycardia. To our knowledge, this is the first case in the literature of INCVM with sustained bradycardia.

Case Report

A 37-year-old woman was referred to the hospital at 30 weeks of gestation for sustained fetal bradycardia. She was her husband's cousin. Her obstetric history was remarkable for the delivery of a male fetus who died in the postpartum first hour in the first pregnancy and intrauterine exitus of a male fetus at term in the third pregnancy. Autopsy was not available in either of the cases. Her second and fourth pregnancies ended with one male and one female healthy infant, respectively. Her fifth pregnancy ended with a missed abortus.

The current pregnancy was referred for sustained fetal bradycardia. Cordocentesis was performed and showed no viral infection [Toxoplasma, cytomegalovirus, rubella, Parvovirus B19,

hepatitis, human immunodeficiency virus (HIV)] and a normal female karyotype was found. Fetal ultrasonography was within normal limits and there was no intrauterine growth retardation. On fetal echocardiography, valvular and great vessel diameters were normal for gestational age. No valvular regurgitation was noted and venous flows (inferior vena cava and umbilical vein) were normal. The arterial duct was patent as well as the foramen ovale. Cardiac functions were normal. The ratio of the early to late filling peaks showed no inversion at tricuspid and mitral valvular orifices and the morphology was found to be normal with sustained bradycardia. Ventricles and atria were found to be synchronous and fetal heart rate varied between 84-94 beats/minute (Fig. 1). All these findings indicated sinus bradycardia. The fetus was followed with serial fetal echocardiograms. During the follow-up period cardiomegaly developed. The sinus bradycardia persisted. She was hospitalized at 34 weeks of gestation and betamethasone was given for fetal lung maturation. Cesarian section was performed at 35 weeks of gestation and a 2600 g female baby was delivered with 2/6/6 Apgar scores at first, fifth and tenth minutes respectively. The baby was intubated and taken to the neonatal intensive care unit. On physical examination the liver was palpable 4 cm in the midclavicular line below the right costal margin. The chest X-ray showed cardiomegaly with a cardiothoracic ratio of 65%. On electrocardiography there was a junctional escape rhythm, corrected QT interval was 0.37 seconds and the heart rate was approximately 60 beats/minute (Fig. 2).

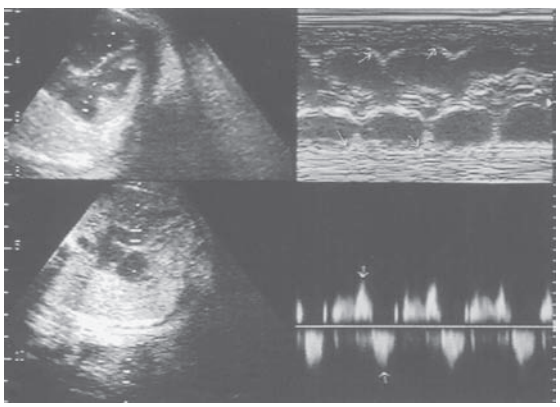

Fig. 1. Two-D and M-mode tracing of the fetal heart (upper figures): The beam passes through the right ventricle and the left atrium. Ventricular (upward arrows) and atrial (downward arrows) contractions are synchronized and fetal heart rate is 94 beat/min. The pulsed Doppler interrogation (lower figures) in the left ventricular outflow tract demonstrates synchronized atrial (upward arrow indicates A wave) and ventricular (downward arrow) contractions.

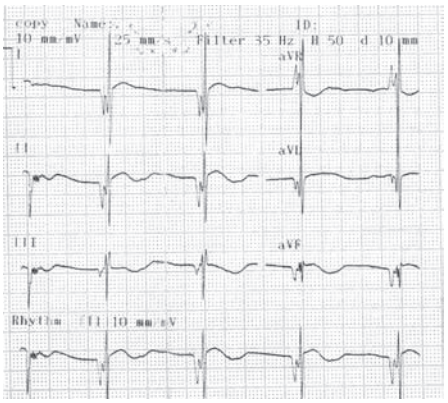

Fig. 2. Postnatal ECG demonstrating junctional escape rhythm with a heart rate of 60 beats/min.

It was thought that the junctional rhythm was due to profound sinus bradycardia as a result of sick sinus syndrome. On echocardiographic examination, there were prominent deep recesses communicating with the ventricular cavity at the left ventricular apex and lateral wall (Fig. 3). Ejection fraction and shortening

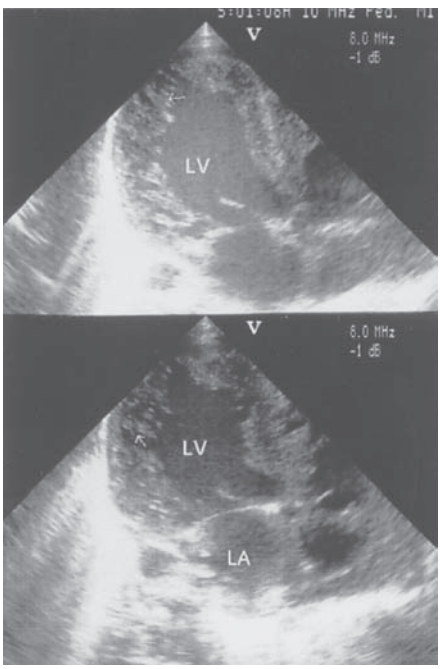

Fig. 3. Postnatal echocardiographic examination showing prominent deep recesses communicating with the ventricular cavity at left ventricular apex and lateral wall. LV: Left ventricle. LA: Left atrium.

fraction were 63% and 32%, respectively. Isoproterenol infusion was started and the patient was taken to the catheter room for transient pacemaker implantation. During the pacemaker implantation the patient developed ventricular fibrillation and defibrillation was performed. The heart rate returned but at the third hour of life the patient again developed cardiac arrest and did not respond to resuscitation.

Discussion

Isolated noncompaction of ventricular myocardium is a rare congenital disorder of endomyocardial morphogenesis, characterized by the presence of numerous prominent trabeculations and deep intertrabecular recesses communicating with the left and right ventricular cavity. The disease uniformly affects the left ventricle, and the right ventricle is also affected on occasion[1,2,7-9]. It may be isolated or associated with other congenital heart diseases, especially with left or right heart obstructive malformations[2,9]. The disease can readily be identified by cross-sectional echocardiography from its characteristic morphological features. Although the etiology of NCVM is not fully elucidated, the disease is thought to be a morphogenetic abnormality that involves arrest of compaction of the loose myocardial meshwork during fetal development. Therefore, it should be present at birth in all patients, whereas ventricular dysfunction might become clinically overt during infancy, childhood or adolescence. In our case, diagnosis was made in the first day of life and this finding supports the theory that INCVM is a disorder of fetal development.

Diagnosis of the NCVM is a difficult aspect of fetal echocardiography. There are few reports about the fetal or neonatal diagnosis of INCVM[10-13]. Kohl et al.[10] reported a case of INCVM at the age of five years which was retrospectively evaluated, and they concluded that this diagnosis could have been made at 23 weeks of gestation. In another study, Bleyl et al.[11] reported three cases of fetal INCVM which were not recognized during fetal echocardiographic examinations, but diagnosed postnatally on autopsy. Winer et al.[12] reported a case diagnosed prenatally as cardiomyopathy, and fetopathological examination showed persisting spongy myocardium. Halbertsma et al.[13] reported a case presenting with cardiogenic shock and diagnosed as NCVM in the neonatal period. A review of 37,555 transthoracic echocardiographic examinations showed that the correct diagnosis was missed in the initial study in 11 of 15 patients. The correct diagnosis was made with a mean time of 3.5 years from the onset of symptoms[10]. In the presented case, the diagnosis was possible at 30 weeks of gestation. With current ultrasound, prenatal detection is possible and if an index case is identified, prenatal examination of further siblings seems to be indicated since familial occurrence has been reported[1,11].

Arrhythmias are one of the major components of INCVM. Usual clinical presentations are ventricular arrhythmias, left bundle branch block or Wolff-Parkinson-White syndrome. Chin et al.[2] reported eight cases of INCVM and described association of ventricular tachycardia and fibrillation in three of these cases. In the largest multicenter study, Ichida et al.[1] reported that left bundle branch block and ventricular tachycardia were rarer than that described in adults, whereas Wolff-Parkinson-White syndrome has relatively high incidence. In a study from our institution, 5 out of 12 noncompaction patients showed rhythm abnormalities (2 patients with ventricular extrasystoles, 1 Wolff-Parkinson-White syndrome, 1 left bundle branch block and 1 complete atrioventricular block)[9]. In the present case, there was fetal sustained sinus bradycardia. During fetal life persistent fetal bradycardia is infrequent. In a fetal ultrasound and electrocardiography study, 3 out of 113 (2.7%) fetal cardiac arrhythmias were classified as sinus bradycardia[4]. Fetal sinus bradycardia is usually associated with maternal collagen vascular disease like systemic lupus erythematosus, structural heart disease or long QT syndrome[5-6]. In our case, there was no sign of lupus in the mother and the corrected QT interval of the baby was within normal limits. As a result of profound sinus bradycardia, junctional escape rhythm was observed, and the patient was diagnosed as having sick sinus syndrome. To our knowledge, our case is the first one diagnosed as INCVM with fetal sustained bradycardia probably due to sick sinus syndrome. Since the fetal diagnosis of INCVM is difficult, the examiner needs to be aware of this rare anomaly; in the case of fetal bradycardia, INCVM should be considered in the differential diagnosis.

REFERENCES

1. Ichida F, Hamamichi Y, Miyawaki T, et al. Clinical features of isolated noncompaction of the ventricular myocardium. J Am Coll Cardiol 1999; 34: 233-240.
2. 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.
3. Miller MS, Shannon KM, Wetzel GT. Neonatal bradycardia. Prog Pediatr Cardiol 2000; 11: 19-24.
4. Lingman G, Lundstrom NR, Marsal K. Clinical outcome and circulatory effects of fetal cardiac arrhythmia. Acta Paediatr Scand Suppl 1986; 329: 120-126.
5. Beinder E, Grancay T, Menendez T, Singer H, Hofbeck M. Fetal sinus bradycardia and the long QT syndrome. Am J Obs Gynecol 2001; 185: 743-747.
6. Meijboom EJ, van Engelen AD, van de Beek EW, Weijtens O, Lautenschutz JM. Fetal arrhythmias. Curr Opin Cardiol 1994; 9: 97-102.
7. Hook S, Ratliff NB, Rosenkranz E, Sterbe R. Isolated noncompaction of the ventricular myocardium. Pediatr Cardiol 1996; 17: 43-45.
8. Oechlin 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.
9. Özkutlu S, Ayabakan C, Çeliker A, Elshershari H. Noncompaction of ventricular myocardium: a study of twelve patients. J Am Soc Echocardiogr 2002; 15: 1523-1528.
10. Kohl T, Villegas M, Silverman N. Isolated noncompaction of ventricular myocardium-detection during fetal life. Cardiol Young 1995; 5: 187-189.
11. Bleyl SB, Mumford BR, Brown–Harrison MC, et al. Xq28-linked noncompaction of the left ventricular myocardium: prenatal diagnosis and pathologic analysis of affected individuals. Am J Med Genet 1997; 72: 257-265.
12. Winer N, Lefevre M, Nomballais MF, et al. Persisting spongy myocardium. A case indicating the difficulty of antenatal diagnosis. Fetal Diagn Ther 1998; 13: 227-232.
13. Halbertsma FJ, van't Hek LG, Daniels O. Spongy cardiomyopathy in a neonate. Cardiol Young 2001; 11: 458-460.