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Left ventricular noncompaction mimicking peripartum cardiomyopathy.

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Left Ventricular Noncompaction Mimicking Peripartum Cardiomyopathy

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Left ventricular (LV) noncompaction (NC) is a morphogenetic disorder involving an arrest of compaction of the loose myocardial meshwork during fetal development. The clinical presentation of this disorder is varied and patients could be asymptomatic or present in childhood or adulthood with heart failure, atrial or ventricular arrhythmias, or thromboembolic phenomena.[1] To our knowledge, LVNC presenting as heart failure during pregnancy has not been previously reported.

CASE REPORTS

Case 1

A 26-year-old woman with a history of gestational hypertension treated with calcium channel blockers presented to the emergency department at 31 weeks of gestation with new-onset dyspnea and dizziness. She had a history of pre-eclampsia resulting in induction of labor at 35 weeks with her prior pregnancy. Her medical history included a repaired tracheoesophageal fistula and recurrent hospitalizations for "pneumonias" during childhood. Her mother had died at age 40 years of congestive heart failure. She denied alcohol or recreational drug use. Physical examination was significant for a heart rate of 100/min, blood pressure of 120/80 mm Hg, jugular venous distension, clear lung fields, a systolic outflow murmur in the aortic area, and a gravid abdomen without lower extremity edema. Laboratory tests revealed mild iron deficiency anemia, normal thyroid stimulating hormone, and negative HIV testing. An electrocardiogram showed sinus tachycardia with nonspecific ST-T abnormalities. An echocardiogram revealed an ejection fraction (EF) less than 20%, mild mitral regurgitation, and significant LV trabeculations at the apex and apical lateral walls with associated wall-motion abnormalities (Figures 1 and 2). The right ventricle was normal in size and function. The LV posterior wall and interventricular septal end-diastolic thickness was 1 cm each, respectively. The LV end-diastolic and end-systolic diameter was 5.7 and 5.5 cm, respectively. The end-systolic ratio of NC to compacted layers of endomyocardium was measured to be 3.2. These findings were diagnostic of LVNC. She was medically treated with metoprolol, hydralazine, digoxin, nitrates, and iron supplementation. She had induction of labor at 37 weeks gestation and delivered vaginally without complications. She was subsequently treated with lisinopril in addition to metoprolol and digoxin. Because of symptoms of palpitations, a 24-hour Holter monitor was placed and demonstrated frequent premature atrial complexes, premature ventricular complexes, and runs of atrial flutter. Two months after delivery of her child, radiofrequency ablation was carried out to treat atrial flutter. Electrophysiologic testing at that time demonstrated in addition bundle branch re-entry ventricular tachycardia that was also ablated. Three months postpartum, echocardiography demonstrated improved LVEF of 30% with an end-systolic ratio of NC to compacted layers of 2.5. The patient has subsequently been lost to follow-up.

Case 2

A 14-year-old girl at 36 weeks of pregnancy with a history of "asthma" treated with albuterol presented to the emergency department with bilateral lower extremity edema and headaches. Her pregnancy was complicated by a positive group B streptococcal culture, sickle cell trait, poor weight gain, and gonorrhea. Her medical history included human papillomavirus condyloma and a mature teratoma at age 9 years. She denied any tobacco, alcohol, or illicit drug use. Physical examination was significant for a heart rate of 115/min; blood pressure of 132/99 mm Hg; tachypnea clear lungs; no murmurs, gallops, or rubs; gravid abdomen; and dependent edema. Laboratory tests revealed mild anemia and abnormal liver function. Chest radiograph showed cardiomegaly and pulmonary edema. She was intubated for respiratory distress and then taken to the operating department for a caesarean section. After delivery, the patient was taken to the pediatric intensive care department and her heart failure was treated with nesiritide, milrinone, and dobutamine. She was extubated and, during the next 2 weeks, weaned off the intravenous vasodilators and inotropes. Her initial echocardiograms revealed a LVEF 17%, mild mitral regurgitation, and significant LV trabeculations at the apex and apical lateral walls

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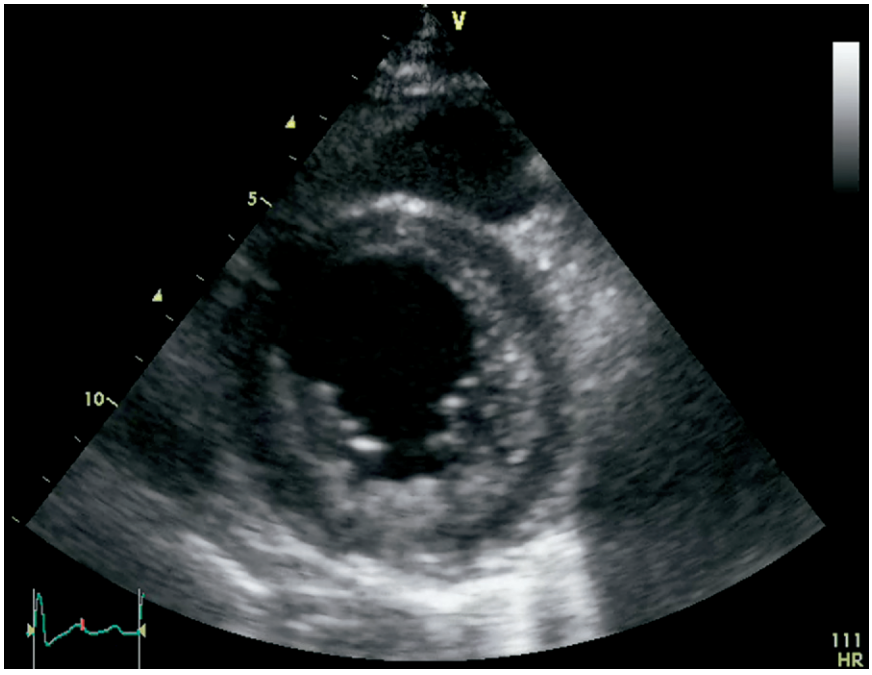

Figure 1 Two-dimensional echocardiogram (short-axis view) showing prominent trabeculations and recesses in noncompacted portion of left ventricle.

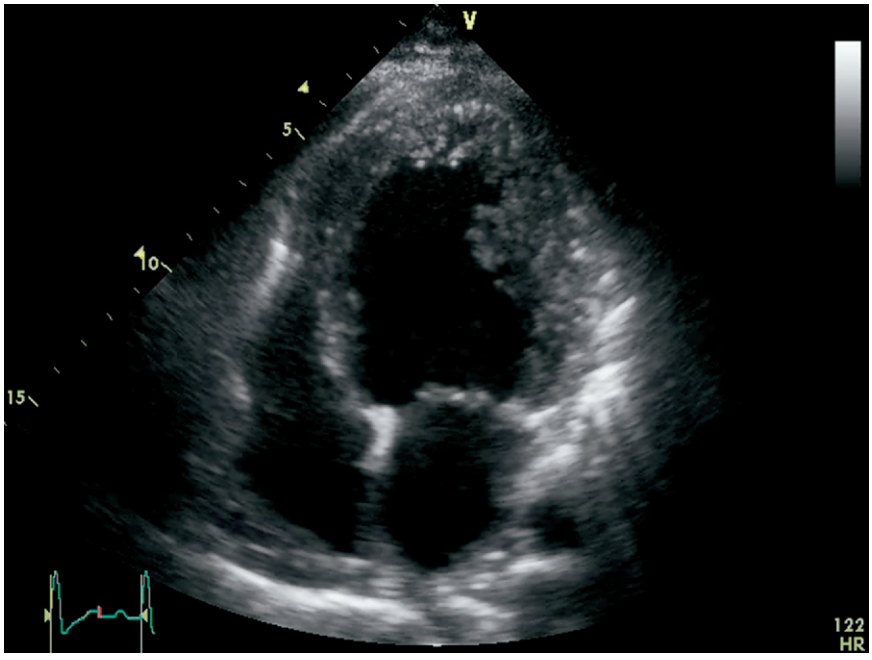

Figure 2 Two-dimensional echocardiogram (apical 4-chamber view) showing prominent trabeculations and recesses in noncompacted left ventricle.

with associated wall-motion abnormalities. The right ventricle was normal in size and function. The LV posterior wall and interventricular septal end-diastolic thickness were .65 and .53 cm, respectively. The LV end-diastolic and end-systolic diameter was 6.3 and 5.5 cm, respectively. The end-systolic ratio of NC to compacted layers of endomyocardium was measured to be 1.63. Approximately 1 month after presentation, she was discharged in stable heart failure on an intensive oral medical regimen. At 17 months follow-up, the patient had persistent LV systolic

dysfunction albeit improved LVEF of 30% with a end-systolic ratio of NC to compacted layers decreasing to 1.4 and remained in marginally compensated heart failure.

DISCUSSION

LVNC is an unclassified cause of cardiomyopathy, a result of an abnormality in the morphogenesis of the endomyocardium resulting in separate and distinct compact and NC layers.[2] LVNC has been well described in children and adults, with varying presentations and clinical courses.[3] The timing of the patients' presentation may be related to the hemodynamic alterations that occur during pregnancy such as a dramatic increase in plasma volume by up to 50%, with maximal hemodilution occurring at 30 to 32 weeks of gestation. Within an hour after delivery, cardiac output increases by 50% above baseline because of a shift of blood from the uterus to the vascular space, increased venous return to the heart as a result of the acute release of inferior vena cava compression, and mobilization of extravascular fluid into the intravascular compartment.[4] These hemodynamic perturbations with a transient decrease in EF seen during pregnancy[5] could likely precipitate symptomatic heart failure in patients with otherwise asymptomatic LVNC. Population-based echocardiographic screening provides information on the natural course of the disease in patients with LVNC, suggesting that patients can be asymptomatic with preserved EF initally and subsequently develop LV dysfunction.[3] It can, therefore, be hypothesized that patient 1 had compensated LV function during her first pregnancy, but symptomatic heart failure during her second pregnancy as a result of a later decline in EF.

The diagnosis of LVNC is based on distinct echocardiographic features including hypertrabeculation commonly at the apex and lateral walls, associated hypokineses in the affected areas, and a ratio of the NC to compacted layer being greater than 1.4.[6] Case 1 fulfilled all these criteria and furthermore the absence of significant LV cavity dilation makes it unlikely that heart failure was caused by an idiopathic dilated or peripartum cardiomyopathy.[7] The patient's mother prematurely died of heart failure and because LVNC can have familial recurrence, possibly in an autosomal dominant pattern, echocardiographic screening of family members is usually recommended.[8] Atrial and ventricular arrhythmias are uncommon in patients with LVNC[9] reported in anecdotal case reports,[10] however, bundle branch re-entry ventricular tachycardia as demonstrated in this patient has not been previously described. Implantable cardioverter defibrillators have been placed in these patients for standard indications such as sudden cardiac death and sustained ventricular arrhythmias.[11,12] There is probably no current knowledge base to justify the prophylactic placement of implantable cardioverter defibrillators in all patients with LVNC except in patients with a persistent LVEF less than or equal to 35% based on extrapolation of current literature in patients with chronic heart failure.[13]

Therapy for patients with systolic heart failure and LVNC includes standard medical therapy as for idiopathic dilated cardiomyopathy and includes angiotensin-converting enzyme inhibitors, beta-blockers, and diuretics as needed.[14,15] Improvement in LVEF as seen in both patients, with or without medical therapy, has been described in LVNC.[1,14] Advice regarding subsequent pregnancies for patients with LVNC can only be extrapolated from the peripartum cardiomyopathy literature and is, hence, controversial. Patients with persistent LV dysfunction caused by peripartum cardiomyopathy have a high mortality and women in whom LV function has returned to normal have a decline in EF with subsequent pregnancies.[16] Individualized preconception counseling may be helpful, using general criteria developed by Siu et al.[17]

The asymptomatic presence of LVNC, with the possibility of decompensation during the hemodynamic stress of pregnancy, and subsequent spontaneous improvement of reduced LVEF observed in LVNC, is similar to the natural history of peripartum cardiomyopathy. The lack of awareness of LVNC and limitations in earlier imaging technology to delineate the endocardium especially near the apex may have led to an underappreciation of this condition. Patients with peripartum cardiomyopathy should, therefore, be carefully screened for the possibility of LVNC, a relatively uncommon cause of heart failure.

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