Short Communications

Pregnancy-Induced Severe Left Ventricular Systolic Dysfunction in a Patient with Hypertrophic Cardiomyopathy

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Summary: This paper reports the first case of hypertrophic cardiomyopathy (HCM) that developed postpartum congestive heart failure (CHF) and severe left ventricular (LV) systolic dysfunction. Review of the literature and clinical implications are discussed.

Key words: hypertrophic cardiomyopathy, congestive heart failure, peripartum cardiomyopathy

Introduction

Despite frequent morbidities, pregnancy in patients with hypertrophic cardiomyopathy (HCM) was considered safe.^{1–3} Morbidity usually improves or resolves after delivery.^{2,3} This sense of security became less certain after reports of maternal deaths among pregnant patients with HCM.^{4,5} This report presents another pregnancy-induced complication in a patient with HCM.

Case Report

A 30-year-old Jamaican woman with a history of HCM, diagnosed 14 years previously, presented to our institution 2

years earlier, complaining of atypical chest pain. She denied any history of dyspnea on exertion, orthopnea, paroxysmal nocturnal dyspnea, pedal edema, palpitations, or syncope. An electrocardiogram (ECG) was significant for normal sinus rhythm and Q waves anterolaterally consistent with HCM. The echocardiogram (Fig. 1) revealed normal left ventricular (LV) size and systolic function, asymmetric septal hypertrophy (septal thickness 2.2 cm, free wall thickness 1.4 cm), without significant LV outflow tract pressure gradient. Verapamil 240 mg daily was started and her symptoms were subsequently controlled. Ten months later, the patient became pregnant; however, she denied any new symptoms. Verapamil was replaced by propranolol. Repeat echocardiogram during the second trimester was unchanged from before. Subsequently, the patient had an uneventful vaginal delivery at term. Three weeks postpartum, she developed severe and progressive symptoms of dyspnea, orthopnea, paroxysmal nocturnal dyspnea, and pedal edema. On physical examination, the patient was moderately distressed, blood pressure was 130/90 mmHg, pulse was regular at 88 beats/min, and respiratory rate was 20/min. Her neck veins were moderately distended. The lung examination revealed bilateral rales. Her heart examination was significant for displaced apical impulse to the sixth intercostal space and anterior axillary line, the first and second heart sounds were normal, and an S₃ gallop was audible. There were no murmurs or pericardial rub. The liver was not enlarged and there was +2 pedal edema. The ECG showed no new changes. A chest x-ray was consistent with cardiomegaly and congestive heart failure (CHF). The echocardiogram (Fig. 2) revealed four dilated cardiac chambers, resolution of asymmetric septal hypertrophy with diffuse hypokinesis of the left ventricle, and severe LV systolic dysfunction (calculated ejection fraction = 20%). There was mild mitral and moderate tricuspid regurgitation. The patient was hospitalized with a diagnosis of new onset CHF. She was treated with angiotensin-converting enzyme inhibitors, diuretics, and digoxin. Subsequently, she improved and was discharged home with minimal symptoms. Two months later, the patient was readmitted with decompensated CHF and atypical chest pain. Cardiac catheterization revealed normal coronary arteries.

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FIG. 1 Echocardiogram showing asymmetric septal hypertrophy (septum thickness = 2.2 cm, free wall thickness = 1.4 cm). (A) Long-axis view, (B) short-axis view. S = septum.

Discussion

To our knowledge, this is the first reported case of a patient with HCM developing severe postpartum LV systolic dysfunction. Progression of HCM to LV dilatation and systolic dysfunction, that is, dilated cardiomyopathy occurs in 10–15% of patients.^{6,7} It appears to result from fibrosis and scar formation as a consequence of myocardial ischemia caused by small vessel coronary artery disease.^{6,8} However, this process usually is slow. The rapid clinical and echocardiographic deterioration in this patient after delivery suggests that pregnancy was the precipitating factor.

The outcome of patients with HCM when they become pregnant is controversial. Pregnancy is associated with distinct and competing physiologic alterations. Increased blood volume increases LV end-diastolic volume; thus, outflow obstruction should diminish. On the other hand, a fall in systemic arterial resistance and uterine obstruction of the inferior vena cava results in worsening of LV outflow obstruction. In addition, increased adrenergic activity related to pain and

emotional stress could aggravate outflow obstruction. Earlier reports suggested that despite symptomatic worsening, pregnancy in patients with HCM is generally safe with excellent maternal outcomes.^{1–3} Symptoms usually improve or disappear after delivery.^{2,3} Recently, there have been reports of maternal deaths in pregnant patients with HCM.^{4,5} These deaths were sudden and were felt to be related to arrhythmias.

An alternative explanation of CHF in this case is a peripartum cardiomyopathy occurring in a patient with preexisting cardiac disease. The relation of HCM to the usual peripartum cardiomyopathy is not clear at this time. Similarly, the incidence of peripartum LV systolic dysfunction in patients with HCM has not been routinely investigated. Thus, a milder form of LV dilation and/or systolic dysfunction may be much more common than the rapid and severe presentation of this patient. Moreover, mild LV dilation and systolic dysfunction can deteriorate slowly into CHF. Therefore, in pregnant patients with HCM, serial echocardiography may be necessary during pregnancy and after delivery even in the absence of symptoms.

FIG. 2 Postpartum echocardiogram showing dilated LV and resolution of asymmetric septal hypertrophy. (A) Long-axis view, (B) short-axis view. S = septum, LV = left ventricle.

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