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Case Report

Pregnancy with severe hypertrophic obstructive cardio myopathy fighting against all odds: a case report

Kalyani Sai Dhandapani*, Anushri Manimaran, Hajira Fathima

Department of Obstetrics and Gynaecology, Southern Railway Headquarters Hospital, Chennai, Tamil Nadu, India

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***Correspondence:**
Dr. Kalyani Sai Dhandapani,
E-mail: drkalyanis@yahoo.co.in

ABSTRACT

Hypertrophic obstructive cardiomyopathy (HOCM) is an inherited condition causing left ventricular outflow tract (LVOT) obstruction, with a prevalence of 1 in 500. Widespread use of echocardiography has led to this disorder being increasingly diagnosed in pregnant women. Knowledge of HOCM is imperative for obstetricians. Pregnancy is generally well tolerated in HOCM but women who are symptomatic pre pregnancy or have significant LVOT obstruction can develop complications such as heart failure, arrhythmias and sudden cardiac death.

Keywords: HOCM, LVOT, Echocardiography

INTRODUCTION

Hypertrophic cardiomyopathy (HCM) is a genetic disorder that has an autosomal dominant mode of transmission characterized by primary left ventricular hypertrophy that is not related to abnormal loading conditions (such as hypertension, valvular heart disease). This disease often caused by mutations within genes encoding cardiac sarcometric proteins. The prevalence of HCM in the general population is 1 in 500; therefore, a similar incidence in pregnant women is probable.

HCM is usually well tolerated in pregnancy. Most patients fall in the class II group of the modified world health organization (WHO) maternal cardiovascular risk scheme. However, a subgroup of women with HCM may be at higher risk of adverse pregnancy outcomes. Women with severe LVOT obstruction (LVOTO), symptomatic arrhythmias, and moderate systolic left ventricular dysfunction are in the higher-risk class III group of the modified WHO scheme-like in our case. In addition, the small minority of pregnant women with severe systolic left ventricular dysfunction or severe symptomatic LVOTO have a very high pregnancy risk. These women would be placed in the modified WHO class IV group in whom pregnancy is contraindicated.[1]

The reported total mortality in women with HOCM is 0.5% but worsening of symptoms such arrhythmias, heart failure, sudden cardiac death, thromboembolic events occurred in 29% of the patients.[2] Hence the MACE (Major adverse cardiovascular events) diagnosis, prevention and management is the main aspect in HOCM.

CASE REPORT

A 33-year-old G4P2L1A1 high risk HOCM patient, who was lost to follow up for the preceding 3 years presented at 20 weeks gestation with worsened dyspnoea (NYHA class III), palpitations and multiple episodes of near-syncope over the past 1 month.

She had a history of sudden cardiac death of her brother and mother at the age of 20 years and 30 years respectfully. Her first pregnancy was well tolerated, which was LSCS done at Madya Pradesh, delivery details not known (patient was not diagnosed HOCM). During second pregnancy patient referred from her home town with

symptoms of breathlessness to our hospital (apex cardiac centre in Indian railways). Emergency LSCS done after consulting cardiologist at 28 weeks of gestation in view of severe pulmonary oedema not responding to medical management (patient was diagnosed HOCM with severe LVOT obstruction). Baby died postnatally after 18 days. Patient recovered after 10 days of intensive cardiac care. Patient was sent home with optimised dose of beta blocker and advised genetic counselling, contraception, biannual review to cardiac OPD. Patient underwent one medical termination of pregnancy using over the counter MTP kit after one year of last LSCS. Did not turn up for cardiac or OBG review.

After 2 years she came to our hospital at 20 weeks of gestation with history of dyspnoea (NYHA class III) with minimal daily activities, palpitations and multiple episodes of near syncope for one month. Multidisciplinary approach initiated. Patient was admitted in cardiac intensive care under continuous monitoring. Echo showed a high resting LVOT gradient of 60 mm Hg, moderate to severe MR and dilated LA. Patient developed atrial fibrillation which was managed with verapamil and dose optimised based on heart rate monitoring.

Despite these indicators of adverse prognosis, she was managed successfully with anti-failure measures and antiarrhythmics, under combined cardio-obstetric care. Patient chose to remain as an inpatient, even after attaining NYHA II functional status. Stage I FGR was diagnosed and a course of antenatal corticosteroids was given at 33 weeks. Elective LSCS planned at 35 weeks 6 days in view of decreased fetal movements. Cardiologist, obstetrician, anaesthetist, paediatrics team were prepared. Though vaginal delivery with regional analgesia is appropriate in HOCM, our patient was delivered by LSCS due to a history of previous 2 LSCS deliveries, including one preterm LSCS. Intraoperative complications encountered were atonic PPH, managed with B lynch sutures and pulmonary oedema, managed with judicious diuretics and CPAP. Delivered an alive female baby of 1.9 kg with APGAR score 7/10, 8/10.

Patient shifted to cardiac ICU after surgery with CPAP. Patient was kept in mechanical ventilator, vitals were normalised. Patient weaned off from ventilator on POD-1. Contraception, cardiac follow up, neonatal follow up, contraception and genetic counselling advised to patient.

DISCUSSION

HOCM is increasingly diagnosed in women of childbearing age due to more widespread use of echocardiography and familial screening programs. The majority of young women with heart disease, including HOCM, wish to consider pregnancy and therefore obstetric admission is a common cause for hospitalisation in this patient population. However, to date pregnancy outcome data for these patients are scarce.[3]

HCM is an archetypical single gene disorder with an autosomal dominant pattern of inheritance. Autosomal recessive and X-linked modes of inheritance have been described but are rare.[4] The clinical course can range from a lifelong asymptomatic status to one where individuals are highly symptomatic and functionally limited with heart failure and malignant arrhythmias.

Diastolic left ventricular dysfunction is invariably present and some patients also develop systolic dysfunction later in the disease course. The LVOT obstruction is often associated with mitral regurgitation, mostly due to systolic anterior motion of the mitral valve, although intrinsic abnormalities of the mitral valve apparatus also contribute. Dyspnoea and chest pain are the most frequent symptoms and relate to the pathophysiological impact of the diastolic dysfunction, LVOT obstruction, mitral regurgitation, and myocardial ischaemia. Atrial fibrillation is the most common arrhythmia with an associated high risk of thromboembolism. There is an increased risk of sudden death, particularly in those with a family history of sudden death, symptoms of syncope, ventricular tachycardia, blunted blood pressure response on exercise and severe hypertrophy. In these patients, an implantable cardiovert can be used.

Pathophysiology in pregnancy

During pregnancy, plasma volume and cardiac output increase. The increase in cardiac output in the first and second trimesters is achieved by a larger stroke volume, while later in pregnancy there is an increase in heart rate. The additional volume load of pregnancy causes enlargement of the ventricular cavity, which theoretically might reduce the LVOT obstruction; however, the increased cardiac output tends to counteract this effect and the LVOT gradient will increase with advancing gestation. The same volume loading increases distension of the left atrium and thereby risk of atrial fibrillation. In the context of diastolic disease, the volume changes and increased heart rate are not well tolerated, aggravating symptoms of dyspnoea and lowering the threshold for developing left heart failure.

At the time of delivery, cardiac output increases further secondary to auto-transfusion of blood from the contracting uterus and increased catecholamine levels. There is also an increase in heart rate secondary to blood loss, pain and stress, while the expulsive efforts during delivery tend to diminish venous return. All of these physiological changes lead to an increase in LVOT gradient and shorten the diastolic filling period, therefore increasing the risk of pulmonary oedema.

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Medications

Beta blockers are considered safe during pregnancy and should be continued if already used before pregnancy and should be considered in those patients with more than mild LVOTO and/or maximal wall thickness 15 mm to prevent sudden pulmonary congestion during exertion or emotional stress. Beta blockers can be used for rate control in AF and to suppress ventricular arrhythmias. Verapamil can be used as a second choice when b-blockers are not tolerated in general, antiarrhythmic medications should be avoided if possible, during the first trimester. Disopyramide, despite being a category C drug from the former FDA classification, should only be used when potential benefits outweigh risks because it is related to some relevant adverse effects, namely, uterine contraction and placental abruption.[6]

Peri operative considerations

Aim is to maintain good preload to avoid worsening of LVOT obstruction. Central venous pressure monitoring Arterial line for BP monitoring. Lateral decubitus or slight left tilt to avoid supine hypotension. Oxytocin must be given in carefully because of its vasodilation (and compensatory tachycardia) and the abrupt inflow of a large amount of blood into the systemic circulation (central blood volume increase of 10-25%) as a consequence of uterine contraction that can adversely affect cardiac performance. Regional anaesthesia: may be dangerous since vasodilation associated with sympathetic blockade of the lower extremities may lead to a critical reduction of preload and afterload. For treating hypotension with spinal anaesthesia, vasopressors with short onset, short duration of action, and predictable dose-dependent responses are ideal. Phenylephrine is primarily an alpha-1 adrenergic agonist with minimal to no beta-adrenergic activity. It elevates MAP by causing venous and arterial vasoconstriction and increasing cardiac preload without any significant direct effect on cardiac myocytes, thus avoiding tachyarrhythmias. Therefore, phenylephrine has become the preferred vasopressor in this setting. Epidural anaesthesia (±spinal) has been used safely for vaginal delivery in patients with HCM. LSCS can be safely managed with carefully titrated epidural anaesthesia, using CVP monitoring and maintaining euvolemia or slight hypervolemia. (Graded epidural).

Low blood pressure should be promptly evaluated with echocardiography to assess for LVOT obstruction.[7] Continue beta-blockade or diltiazem /verapamil through delivery and postpartum. Careful monitoring is recommended particularly in the immediate peripartum period when large fluid shifts can lead to acute pulmonary edema as occurred in this case. In the setting of acute heart failure, therapeutic aims are similar to those in nonpregnant women, and both intravenous diuretics and vasodilator

CONCLUSION

This case emphasizes close monitoring and multidisciplinary team management in high risk HOCM mothers. Pre-pregnancy evaluation, risk assessment, multi-disciplinary approach, and genetic counseling are stressed. Although pregnancy is well tolerated by HCM patients, related hemodynamic burdens may lead to unfavourable events requiring close monitoring and adequate treatment.

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