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Challenges of an emergency cesarean section in a patient with hypertrophic cardiomyopathy: Lessons learned from a case

Desafíos de una cesárea de urgencia en una paciente con miocardiopatía hipertrófica. Lecciones aprendidas de un caso

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ABSTRACT

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant genetic disorder[1], characterized by left ventricular hypertrophy unexplained by secondary causes[2]. Its prevalence in the general adult population has been estimated at 0.16% to 0.29%[2] however many patients may not be diagnosed due to absence of signs or symptoms[3]. This cardiac abnormality is characterized by increased left ventricular wall thickness, in the absence of cardiac, systemic, or metabolic disease[1] capable of generating that magnitude of hypertrophy[2], with preserved or increased ejection fraction[3]. HOCM is associated with arrhythmias, heart failure and sudden death, all of the above are exacerbated with pregnancy[4]. During delivery various hemodynamic parameters must be controlled to prevent exacerbation of the obstruction of the left ventricle outflow tract and other complications. These include maintenance of sinus rhythm, reduction of sympathetic activity in order to reduce chronotropy and inotropy, maintenance of left ventricular filling and systemic vascular resistance[5]. We present a case of a patient with HOCM with a 24.4 week twin pregnancy that arrived at the operation room due to a placental abruption who required an emergency cesarean section.

Keywords: Hypertrophic cardiomyopathy, genetic disorder, cardiac abnormality, ejection fraction, pregnancy cardiomyopathy.

RESUMEN

La miocardiopatía hipertrófica (MCH) es un trastorno genético autosómico dominante[1], caracterizado por hipertrofia ventricular izquierda sin causa secundaria[2]. Su prevalencia en la población adulta general se ha estimado entre el 0,16% y el 0,29%[2]; sin embargo, muchos pacientes pueden no ser diagnosticados debido a la ausencia de signos o síntomas[3]. Esta anomalía cardíaca se caracteriza por un aumento del grosor de la pared ventricular izquierda, en ausencia de enfermedad cardíaca, sistémica o metabólica[1] capaz de generar dicha magnitud de hipertrofia[2], con fracción de eyección conservada o aumentada[3]. La MCH se asocia con arritmias, insuficiencia cardíaca y muerte súbita; todas estas afecciones se exacerban con el embarazo[4]. Durante el parto, es necesario controlar diversos parámetros hemodinámicos para prevenir la exacerbación de la obstrucción del tracto de salida del ventrículo izquierdo y otras complicaciones. Estos incluyen el mantenimiento del ritmo sinusal, la reducción de la actividad simpática para reducir la cronotropía y la inotropía, el mantenimiento del llenado ventricular izquierdo y la resistencia vascular sistémica[5]. Presentamos el caso de una paciente con miocardiopatía hipertrófica con un embarazo gemelar de 24,4 semanas que llegó a quirófano debido a un desprendimiento de placenta y requirió una cesárea de urgencia.

Palabras clave: Miocardiopatía hipertrófica, trastorno genético, anomalía cardíaca, fracción de eyección, miocardiopatía gestacional.

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Introduction

Hypertrophic cardiomyopathy (HCM) is an autosomal dominant genetic disorder[1], characterized by left ventricular hypertrophy unexplained by secondary causes[2]. Its prevalence in the general adult population has been estimated at 0.16% to 0.29%[2] however many patients may not be diagnosed due to absence of signs or symptoms[3]. This cardiac abnormality is characterized by increased left ventricular wall thickness, in the absence of cardiac, systemic, or metabolic disease[1] capable of generating that magnitude of hypertrophy[2] with preserved or increased ejection fraction[3].

HOCM is associated with arrhythmias, heart failure and sudden death, all the above are exacerbated with pregnancy[4]. During delivery various hemodynamic parameters must be controlled to prevent exacerbation of the obstruction of the left ventricle outflow tract and other complications. These include maintenance of sinus rhythm, reduction of sympathetic activity to reduce chronotropy and inotropy, maintenance of left ventricular filling and systemic vascular resistance[5].

We present a case of a patient with HOCM with a 24.4 week twin pregnancy that arrived at the operation room due to a placental abruption who required an emergency cesarean section.

Case report

During a night shift at the maternity ward, the surgical team was informed that a 26 years patient with a 24.4 week twin pregnancy was bleeding due to a placental abruption and probably going to need a cesarean section. Clinical history was reviewed, the patient was coursing her second pregnancy, she was hospitalized for a preterm prelabor rupture of membranes associated with retrosternal pain and palpitations. Medical history included obesity, a hypertrophic cardiomyopathy without left ventricle outflow tract obstruction and history of stroke in 2020 of cardiogenic origin with right hemianopsia as a sequelae. Her NYHA classification was II/IV and denied having syncopal episodes. She was taking aspirin, was being treated for depression with sertraline and was anticoagulated with enoxaparin (last dose administered at 8 pm). Beta blocker therapy was initiated.

Blood count evidenced hemoglobin in 15.3 g/dL and hematocrit 44%, platelet count was within normal range and prenatal laboratories were normal. EKG showed sinus rhythm, left auricular dilation, bifascicular block, Poor R wave progression, lateral repolarization abnormality and Left ventricular hypertrophy signs. Echocardiogram reported severe eccentric asymmetric hypertrophy pattern predominantly in the basal septum, preserved systolic function with LVEF in 55% (Simpson method). No apparent segmental disorders and indirect signs of type 2 diastolic dysfunction. Left and right atrium with severe dilation. Mild anterior systolic movement of the mitral valve (SAM). Dilation of right cavities, free wall hypertrophy, preserved RV systolic function, TAPSE 31 mm and moderate tricuspid regurgitation, TRV 321 cm/s, allowing estimation of sPAP of 50 mmHg. No signs or symptoms of acute heart failure were documented.

After an hour, the patient continued bleeding and was scheduled for an emergency cesarean section. She didn't have fasting and the last dose of enoxaparin was administered 4 hours before. Plasma, platelets, and packed red blood cells were reserved and available. Protamine was available in the operating room.

She arrived at the operating room, initial vital signs were blood pressure 120/78 mmHg, heart rate 68 b.p.m, respiratory rate 20 b.p.m and SpO_2 98%. She was premedicated with 1mg of midazolam and 50 mcg of fentanyl i.v. to prevent stress response and sympathetic stimulation. Basic ASA monitoring was initiated, a noradrenaline infusion at 0.05 mcg/kg/min was started to maintain blood pressure as phenylephrine was not available. Preoxygenation with oxygen at 6 l/min was started, rapid sequence intravenous induction was performed with remifentanil 0.5 mcg/kg/min, lidocaine 80 mg, succinylcholine 200 mg, propofol 150 mg and dexamethasone 4 mg. A 7.5 tube and arterial line were placed and 30 mg of rocuronium were administered.

After induction and during surgery hemodynamic parameters were strictly controlled with titrated noradrenaline infusion. Balanced anesthesia was maintained with sevoflurane and remifentanil. To prevent uncontrolled hemorrhage 1 g of tranexamic acid was administered before incision. Procedure started with little bleeding and no reversal of the anticoagulation was administered. 2,000 cc of ringer's lactate were infused during the procedure. After the babies were born 100 mcg of carbetocin were administered with adequate uterine tone and no further significant bleeding.

Procedure ended without eventualities. Postoperative multimodal analgesia included 75 mg of diclofenac, 0.7 mg of hydromorphone, 1 g of paracetamol and a TAP block with bupivacaine 0.25% with adrenaline. Neuromuscular blockade was monitored and reversed with neostigmine 1.5 mg, no atropine was administered to prevent tachycardia.

Patient awoke and was extubated, noradrenaline infusion was suspended, no hemodynamic changes or arrhythmias were present. She had no pain and was transported to the UCI without complications.

Discussion

Hypertrophic cardiomyopathy (HCM) is one of the most prevalent genetic cardiac conditions[3] and can present at any age[6]. Approximately 60% of the patients with HOCM have an autosomal dominant inheritance and 60%-40% of them have genetic mutations of the sarcomere unit that compromise contractility[6]. The present discussion is based on the review of literature provided, which addresses relevant aspects related to the management and outcomes of pregnancy in women with HCM.

Risk stratification is essential to identify HCM patients who may be at a higher risk of adverse events during pregnancy[7]. Factors such as a high New York Heart Association (NYHA) functional class before pregnancy, signs of congestive heart failure, and an elevated gradient in the left ventricular outflow tract have been identified as significant predictors of adverse cardiac events during pregnancy[7].

In the perioperative setting it is important to differentiate Hypertrophic cardiomyopathy from Hypertrophic cardiomyopathy with left ventricular outflow tract obstruction (LVOTO)

defined as a systolic pressure gradient in the left ventricular outflow tract of 30 mmHg or greater at rest or while exercising[6]. It's important to note that the LVOTO may be absent at rest but present during different activities, this is known as dynamic LVOTO[8], one of the causes is the systolic anterior motion (SAM) of the anterior mitral leaflet into the LVOT during systole[8].

Pregnancy represents a state of high physiological stress on the cardiovascular system. Therefore, pregnant women with hypertrophic cardiomyopathy face the potential for increased morbidity, and as a result, managing their condition can become a significant challenge when complications arise[9],[10].

The physiological changes that occur during pregnancy, such as decreased vascular resistance, increased blood volume and cardiac output due to increase in stroke volume and heart rate, can lead to a worsening of heart failure in women with HCM[9],[10],[11].

The choice of anesthesia can significantly impact these hemodynamic changes during labor and delivery in the patient with HCM. Adverse effects during labor and delivery include increases in heart rate and contractility from endogenous catecholamines due to pain and anxiety.

Pregnancy in individuals with Hypertrophic Cardiomyopathy (HCM) is generally well-tolerated, but it does carry an increased risk of cardiac complications. The mortality rate among pregnant women is 2.4%, compared to 0.007% in the general population. In patients with hypertrophic cardiomyopathy (HCM), pregnancy and childbirth are often associated with a low risk of complications[12]. Cardiac complications include acute heart failure, left ventricular outflow tract obstruction (LVOTO), arrhythmias, and ischemic events[13]. The risk of experiencing these events seems to correlate with the severity of the disease before pregnancy and the presence of certain risk factors[13].

The aim of perioperative care in these patients include: adequate preload and afterload, maintenance of sinus rhythm and reduction of sympathetic activity to reduce chronotropy and inotropy[14],[15].

An analysis of various anesthetic techniques should consider their specific conditions. Spinal and epidural anesthesia, for instance, may lower systemic vascular resistance (SVR) and promote a sympathetic blockade potentially worsening outflow obstruction, rendering them relatively contraindicated[14]. Volatile anesthetic agents, which can decrease myocardial contractility, should also be administered with caution to avoid a significant SVR reduction[11],[14]. Achieving a positive outcome hinges on carefully fine-tuning anesthetic agents, ensuring adequate volume replacement, and promptly replacing blood loss while being guided by invasive hemodynamic monitoring.

Regarding vasopressor support, ephedrine is not advised for managing hypotension in these patients due to the potential for tachycardia and increased inotropy. Indeed, the favored approach for addressing hypotension after sympathetic blockade is the titration of phenylephrine in 50 µg increments[14]. For initial treatment of hypotension in patients with left ventricular outflow tract obstruction (LVOTO), the recommended approach includes an intravenous fluid bolus and the use of an alpha-adrenergic agonist, such as phenylephrine as mentioned before[14].

In the intraoperative period, ECG abnormalities are common, occurring in 75%-90% of cases, with findings such as high-voltage complexes, ST abnormalities, T wave inversion, and signs of left atrial enlargement[15]. However, these ECG changes do not reliably indicate the severity of the condition.

Echocardiography has the capability to show the degree of hypertrophy and the potential for systolic anterior movement of the mitral valve. However, it›s important to note that this phenomenon is dynamic, and its absence in pre-surgery assessments does not guarantee that it won›t occur during the surgical procedure[15],[16].

Premedication can help in lowering sympathetic activity, thereby reducing the workload on the heart. Proper hydration is crucial to maintain ventricular preload, especially when it comes to left-sided pressures[15]. Placing invasive arterial monitoring before the induction of anesthesia enables a swift response to fluctuations in arterial pressure[10].

The choice of agents for both inducing and maintaining anesthesia should be made with the goal of minimizing decreases in systemic vascular resistance and preventing tachycardia or sympathetic surges[14]. When it comes to ventilation, the objective should be to minimize any reduction in venous return that could occur[15]. This can be achieved by using small tidal volumes and rapid ventilatory frequencies. Incorporating transesophageal echocardiography during surgery serves as an ideal continuous monitor to assess the adequacy of left ventricular filling and the potential development of LVOT obstruction[16].

Pulmonary artery wedge pressure measurements can sometimes be misleading, as they may give an exaggerated estimate of left ventricular filling, but they are still valuable for tracking trends over time[17]. When addressing hypotension, it›s important to consider judicious volume resuscitation and the use of alpha-agonists[15]. Vasopressors such as noradrenaline can be used with caution as they have some inotropic and chronotropic effects, and can increase myocardial oxygen demand as well as LVOT obstruction. If dealing with hypertension, it›s advisable to administer beta-blockers after ensuring adequate analgesia and anesthesia. Avoid the use of vasodilator agents like nitroglycerin[16].

In postoperative management, the goal is to prevent sympathetic stimulation by providing effective pain control and avoiding hypothermia[15]. While regional anesthesia is generally discouraged because it can reduce systemic vascular resistance and potentially lead to outflow obstruction, there are some case reports, especially in obstetric anesthesia, demonstrating its safe use[18],[19], Peripheral nerve blocks can help in the multimodal pain management strategies and are safe[19].

Cardiac arrest in hypertrophic cardiomyopathy (HCM) is a unique and critical situation. If the arrest is believed to be caused by left ventricular outflow tract (LVOT) obstruction, the use of inotropic agents is contraindicated, as they can exacerbate the obstruction[15]. Instead, it is more appropriate to consider alpha-agonists, intravenous fluids, and the rapid correction of arrhythmias[15],[16]. Additionally, it›s advisable to apply external defibrillator pads before the induction of anesthesia to be prepared for potential cardiac emergencies[15],[20].

Conclusion

Hypertrophic cardiomyopathy is prevalent in pregnant women. The anesthesiologist must be familiar with echocar-

diography changes, EKG alterations and hemodynamic goals, as well as potential complications and contraindicated medications in these patients to ensure successful maternal and fetal outcomes. No type of anesthesia is contraindicated, the anesthesia with which the anesthesiologist is most comfortable can be administered, however, the specific objectives of these patients must be taken into account in order to have adequate perioperative care and avoid complications.

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