ISSN: 1607-8322, e-ISSN: 2220-5799 Anaesthesia, Pain & Intensive Care
Vol 27(4); August 2023 DOI: 10.35975/apic.v27i4.2208

CASE REPORT ANESTHESIA & CONCURRENT DISEASE

Anesthetic management of a patient with pheochromocytoma and peripartum cardiomyopathy for cesarean section

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ABSTRACT

Pheochromocytoma and peripartum cardiomyopathy is associated with significant maternal mortality. Cause of death can be heart failure, myocardial infarction, arrhythmias and thromboembolism. Pheochromocytoma can be one of the causes of dilated cardiomyopathy. Anesthesia in an obstetric patient with pheochromocytoma and peripartum cardiomyopathy is challenging. Early diagnosis and a multidisciplinary management is required to improve outcome. Elective cesarean section must be planned and epidural anesthesia should be the preferred technique. We present a case of 34 y old female patient with pheochromocytoma and peripartum cardiomyopathy who underwent cesarean section under epidural anesthesia and had uneventful recovery.

Key words: Pheochromocytoma; Peripartum Cardiomyopathy; Cesarean Section; Epidural Anesthesia

Citation: Ayesha K, Khan J, Sikander RI, Fatima N. Anesthetic management of a patient with pheochromocytoma and peripartum cardiomyopathy for cesarean section. Anaesth. pain intensive care 2023;27(4):596−599.

DOI: 10.35975/apic.v27i4.2208

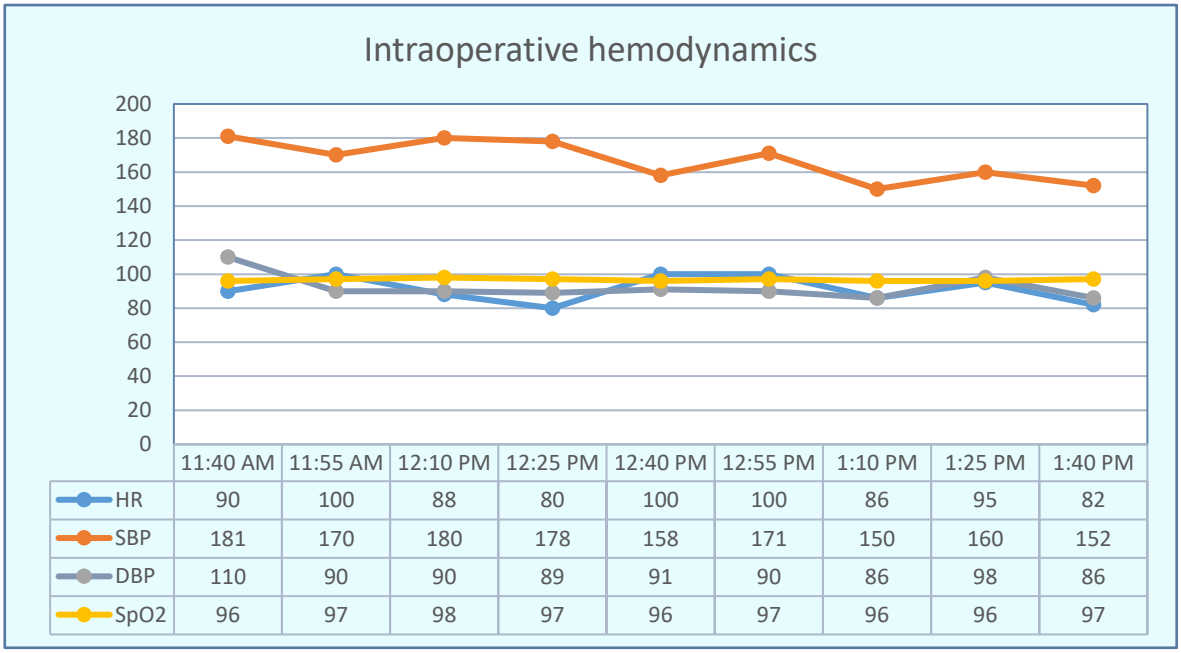
Received: April 16, 2023; **Reviewed:** May 09, 2023; **Accepted:** June 06, 2023

1. INTRODUCTION

Pheochromocytoma is a catecholamine secreting tumor that arises from chromaffin cells, it is extremely rare in pregnancy, with an incidence of 0.0002%. It can mimic severe preeclampsia and undiagnosed cases can have mortality up to 50%. With timely management, mortality can be reduced to less than 15%.[1] Peripartum cardiomyopathy is a rare cause of heart failure that occurs towards the end of pregnancy and in the early postpartum period.[2] Peripartum cardiomyopathy is also associated with up to 50% mortality.[3] When pheochromocytoma and peripartum cardiomyopathy, both occur in a patient, the situation becomes a really big challenge for all of the physicians involved in the management, including the anesthetist. Early recognition and prompt aggressive management can improve outcome in such high-risk patients. W present a case of cesarean section (CS) in a patient with pheochromocytoma and peripartum cardiomyopathy which was operated under epidural anesthesia and the patient had uneventful recovery.

2. CASE REPORT

A 34 y old female patient in her fourth pregnancy presented at 33 weeks of gestation with a history of worsening dyspnea progressed into orthopnea and paroxysmal nocturnal dyspnea. She was hypertensive for one and half years, which remained uncontrolled and she complained of persistent headache. She was taking antihypertensives; tablet labetalol, methyldopa, and amlodipine, along with diuretics, tablet ursodiol and iron supplements. Three of her previous pregnancies were



	11:40 AM	11:55 AM	12:10 PM	12:25 PM	12:40 PM	12:55 PM	1:10 PM	1:25 PM	1:40 PM
HR	90	100	88	80	100	100	86	95	82
SBP	181	170	180	178	158	171	150	160	152
DBP	110	90	90	89	91	90	86	98	86
SpO2	96	97	98	97	96	97	96	96	97

Figure 1: Intraoperative hemodynamic parameters during the operative period

uneventful and she delivered vaginally. Patient underwent laparoscopic cholecystectomy seven years back under general anesthesia, after which she developed postoperative nausea and vomiting which was managed with intravenous antiemetics.

Examination showed the patient was tachypneic, had an oxygen saturation of 96% on room air. Her blood pressure (BP) was 182/119 mmHg and heart rate (HR) 89 beats/min. She was pale and anxious. There was no sign of heart failure. Rest of the examination was unremarkable. Investigations showed patient's hemoglobin was 10.1 g/dL, had slightly raised AST and serum bile acids. Initially there was a suspicion of suffering from severe preeclampsia, but she did not respond to the treatment. So with suspicion of pheochromocytoma, 24 h urinary metanephrines and normetanephrines were advised which were markedly raised. Her electrocardiograph showed sinus rhythm with heart rate 90 beats/min. Transthoracic echocardiography revealed ejection fraction (EF) of 35%, concentric left ventricular hypertrophy, moderate systolic dysfunction and grade 1 diastolic dysfunction, pulmonary artery pressure 12 mmHg and normal valvular function. MRI scan was not done.

The patient was optimized in the preoperative period. Her raised BP was managed with injection hydralazine 5 mg boluses and infusion hydralazine. Alpha blocker doxazosin 2 mg was started twice daily orally. Patients with less functional capacity are unable to tolerate hemodynamic changes of pregnancy especially towards the end of pregnancy. So, after discussion with the obstetrician and cardiologist elective CS was planned at 35 weeks of gestation. But on surgeon's request, case was proceeded earlier at 34 weeks.

Patient was counselled in detail. Informed consent was taken. When she came to operating room, two wide bore intravenous (IV) lines were secured. In addition to standard ASA monitoring, urine output and invasive BP monitoring was initiated. Case was proceeded under epidural anesthesia. Before induction, patient had BP of 180/110 mmHg. Infusion of dexmedetomidine was started @ 0.5 µg/kg/h along with infusion glyceryl trinitrate and both infusions were titrated to effect. Procedure lasted for around 2 h and throughout the surgery, BP was maintained around 160/90 mmHg with HR 80 to 90 beats/min and oxygen saturation 97% with oxygen @ 6 L/min via face mask (Figure 1). Injection oxytocin 5 IU were given slow IV after the delivery of the baby along with diuretic and antiemetics. Intraoperative blood loss was around 1 L for which 500 ml fluid and 1 red cell concentrate were transfused.

Postoperatively, there was risk of rebound hypertension, heart failure, nausea vomiting and thromboembolism. Postoperative pain management was another important concern. We managed the patient accordingly. Patient had uneventful recovery. She was discharged on third postoperative day on medication; doxazosin, labetalol, valsartan, furosemide + spironolactone and aspirin. Her BP was maintained around 160/90 mmHg.

Two weeks after CS, CT scan abdomen and pelvis was performed which confirmed right adrenal mass. Her

echocardiography was repeated 3.5 weeks after CS, which showed EF 40%, left ventricular global hypokinesia, moderate systolic dysfunction, grade 1 diastolic dysfunction, pulmonary artery pressure 17 mmHg and normal valvular function. Patient underwent laparoscopic right adrenalectomy under general anesthesia uneventfully at PIMS hospital

3. DISCUSSION

Pheochromocytoma and peripartum cardiomyopathy, both are associated with significant mortality and morbidity. Cause of death can be myocardial infarction, heart failure, arrhythmias, intracranial bleeding and thromboembolism. 30% patients with peripartum cardiomyopathy die due to a thromboembolic event. Almost 60% patients improve by 6 months in the postpartum period.[2]

Regarding clinical presentation of pheochromocytoma, the most common symptom is headache, and the most common sign is hypertension.[1] Pheochromocytoma during pregnancy can mimic severe preeclampsia. Hypertension that occurs after 20 weeks of gestation along with proteinuria is gestational hypertension; whereas hypertension that occurs before 20 weeks of gestation and accompanied by headache, sweating, palpitations and cardiomyopathy is suggestive of pheochromocytoma.[4] Pheochromocytoma in pregnancy is diagnosed clinically as well as with biochemical tests and non-contrast MRI scan. Plasma free metanephrines level is the most sensitive test. Whereas, 24 h urinary metanephrines is the best urine screening test. Metanephrines have higher sensitivity than plasma or urinary catecholamines as diagnostic tests and consist of fractionated plasma or urinary metanephrine or normetanephrine. Level of urinary metanephrines and nor metanephrines more than twice is suggestive of pheochromocytoma. It is medically treated with alpha blockers which must be taken for at least 2 weeks before surgery. Phenoxybenzamine, prazosin and doxazosin are safe in pregnancy. Beta blocker can be added in order to avoid tachycardia but it must be either cardioselective or alpha + beta blocker. During pregnancy, best time for excision is 2nd trimester of pregnancy. But if diagnosed late in pregnancy, elective CS must be planned as labor and vaginal delivery can precipitate hypertensive crisis.[5]

Peripartum cardiomyopathy is a rare cause of heart failure that occurs toward the end of pregnancy and in the early postpartum period. Diagnosed on the basis of diagnostic criteria: heart failure developing towards the end of pregnancy or up to 5 months postpartum, absence of another identifiable cause of cardiac failure, absence of cardiac symptoms prior to late pregnancy, left ventricular dysfunction defined by an ejection fraction of less than 45%.[6] Risk factors include advanced maternal age, multiparity, multiple pregnancies, pregnancy induced hypertension and essential hypertension. Pheochromocytoma can be one of the causes of dilated cardiomyopathy.[7] Regarding clinical presentation, orthopnea and paroxysmal nocturnal dyspnea is more specific.[8] Electrocardiograph and echocardiography confirms the diagnosis. It is treated with oxygen supplementation, vasodilators, diuretics and beta blockers. Inotrope and anticoagulant can be added.[6]

Literature shows that in 2012, a 35 y old lady in her last trimester of pregnancy developed sudden shortness of breath, had uncontrolled blood pressure, and was rushed to the hospital. Her echocardiograph showed cardiomyopathy. Patient developed ventricular fibrillation, was resuscitated but could not survive. Her autopsy revealed right adrenal mass.[4] Pheochromocytoma along with cardiomyopathy has high maternal mortality if not timely managed. Another case was reported in 2018 about a 24 y primigravida who developed left flank pain at 37 weeks. She had a history of headache for 2 y and had gestational diabetes as well. Her investigations confirmed left pheochromocytoma. She was treated with phenoxybenzamine and atenolol. After 10 days, patient underwent CS and excision of left adrenal mass uneventfully.[1] If pheochromocytoma is diagnosed late in pregnancy, removal of mass must be delayed till the time of CS.

Obstetric patient with pheochromocytoma and peripartum cardiomyopathy is a challenge for the anesthetist. So, preoperative optimization is extremely important. Alpha blocker reduces blood pressure and restores intravascular volume. Cardioselective beta blocker treats tachycardia. Vasodilators reduce preload and afterload and diuretics relieve pulmonary congestion. Goals of management of pheochromocytoma and peripartum cardiomyopathy must be kept in mind while managing such high risk patient. For pheochromocytoma, goals are to maintain blood pressure, restore intravascular volume, avoid factors that cause catecholamine surge and maintain hemodynamic stability.[5] For peripartum cardiomyopathy; in order to maintain cardiac output, avoid increase in afterload, maintain preload and contractility and to reduce myocardial work. Goal is to avoid tachycardia and arrhythmias.[7] In addition to standard ASA monitoring, urine output and invasive blood pressure monitoring is essential.

Regarding mode of anesthesia, it is debatable whether to choose regional anesthesia or general anesthesia. Epidural anesthesia must be the preferred technique because it reduces stress response, provides better analgesia and better control of hemodynamics when compared with general anesthesia. Drugs of general anesthesia are myocardial depressants and tend to reduce

myocardial contractility. However, induction and extubation can lead to hypertensive crisis. Whereas, spinal anesthesia can lead to sudden hypotension and patient with peripartum cardiomyopathy cannot compensate for it.[7] Intraoperatively, blood pressure must be controlled with short acting antihypertensives like phentolamine, nitroglycerine and magnesium sulphate.[5] Uterotonic oxytocin must be used cautiously as it causes vasodilation and tachycardia. Postoperative management includes oxygenation, strict monitoring and control of blood pressure, adequate pain management, fluid balance, thromboprophylaxis and cardiology review.

Timely diagnosis and proper management can improve maternal and fetal outcome. Our patient was not a diagnosed case of pheochromocytoma and her MRI scan was not done which is confirmatory. Patient was not well optimized before CS. At our set up, rapidly acting alpha blocker was not available to control intraoperative hypertension. However, we tried our level best to achieve goals of management and patient had uneventful recovery.

4. CONCLUSION

Pheochromocytoma and peripartum cardiomyopathy during pregnancy have high maternal mortality rate. Early identification, optimization of the patient and multidisciplinary management can improve outcome in such high-risk patient.

5. Conflict of Interest

The authors declare no conflicts of interest.

6. Consent to publish

The consent to publish this report was obtained from the patient.

7. Author Contribution

AK: Conduct of the case and manuscript writing

KJ: Literature review and manuscript editing

SRI, FN: Manuscript editing

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