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Case Report

Anaesthesia Section

Emergency Caesarean Section in a Patient with Undiagnosed Eisenmenger's Syndrome with Severe Pulmonary Hypertension

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ABSTRACT

Pregnancy is associated with major physiologic changes in various organ systems of the body, especially the cardiovascular system. This is especially dangerous in the presence of pre-existing Pulmonary Artery Hypertension (PAH) and maternal mortality is unacceptably high (30-50%) in such patients. We report the successful perioperative management of a parturient with severe PAH due to an undiagnosed Eisenmenger's syndrome, posted for emergency caesarean section.

Keywords: Mortality, Pregnancy, Pulmonary artery hypertension

CASE REPORT

An unbooked, 31-year-old lady (Gravida 3, Para1, No live issue, Abortions 2) with 35 weeks pregnancy, presented to the obstetric casualty in labor. She gave a history of fatigue since childhood (20-22 yrs), occasional haemoptysis, paroxysmal nocturnal dyspnea and breathlessness and palpitations on exertion since last five years. She also complained of cyanosis of nails and lips for two years, which had increased in the last 15 days. She had pulmonary tuberculosis two years back, for which she had taken complete treatment.

On examination the patient was of thin built, lying comfortably in the supine position. She had clubbing and central cyanosis, along with pedal oedema. On auscultation the chest was bilaterally clear with an ejection systolic murmur over the pulmonary area, and fixed S_2. All blood investigations were unremarkable except for a slightly raised haematocrit (44.3%).

Oxygen saturation (SpO_2) on room air was 69% and Arterial Blood Gas analysis (ABG) revealed a PaO_2 of 47 mmHg, $PaCO_2$ of 38 mmHg, pH 7.38. The ECG showed T wave inversion along with Right Axis Deviation (RAD) and Right Ventricular Hypertrophy (RVH). An emergency Echocardiography (ECHO) was performed and it revealed a dilated Right Atrium (RA), Right Ventricle (RV) and Pulmonary Artery (PA), severe Pulmonary Artery Hypertension (PAH) and severe Tricuspid Regurgitation (TR). Ejection Fraction (EF) was 60% and there was no regional wall motion abnormality. A provisional diagnosis of severe primary PAH was made. The cardiologist advised oxygen supplementation and injection furosemide 40 mg Intravenously (IV) twice a day.

In view of severe pulmonary hypertension along with episodes of fall in the fetal heart rate, the obstetricians decided to take up the patient for an emergency caesarean section.

A high-risk consent was taken and the patient was shifted to the operation theatre. Baseline monitoring i.e., Heart Rate (HR), Non-Invasive Blood Pressure (NIBP), Electrocardiography (ECG) and SpO_2 was established. The preoperative pulse rate was 85/min, Blood Pressure (BP) was 130/64 mmHg and respiratory rate was 20/min. SpO_2 which was 69% on room air, increased to 85% and 90% on 50% and 100% oxygen respectively. Intravenous access was achieved through two large bore IV cannulae and the patient was preloaded with 500 mL Ringers Lactate (RL).

Combined spinal epidural anaesthesia was planned. A low dose spinal anaesthesia was given with 1.6 mL of 0.5% hyperbaric bupivacaine and 10 µg fentanyl (total volume 1.8 mL). An epidural catheter was placed in L3-4 inter space in the left lateral position. Level of blockade achieved was T8 (to cold spray) with the subarachnoid block. A further top up of 4 mL of 2% bupivacaine was given through the epidural catheter to achieve a level of T6 (to cold spray) after 6 minutes. A 1.6 kg female child was delivered with APGAR of 7/7/8 at 1, 5 and 10 minutes, respectively. Five U syntocinon was given stat intravenously after birth of the baby and 10 U was added in 500 mL RL, given over 30 minutes. There were two episodes of hypotension which were managed with phenylephrine boluses. Her intraoperative ABG after delivery of baby showed PO_2 of 86.6, PCO_2 38.6, pH 7.40 and SPO_2 96.6%. Total 1200 mL of fluid was given intraoperatively. Postoperative vitals were HR-87/min, BP-132/65 mm hg, SpO_2 88% on 0.5 FiO_2. Chest was bilateral clear. Patient was comfortably shifted to postoperative room with oxygen via face mask.

Patient was kept in recovery room, under observation for four hours and then shifted to high dependency unit with oxygen supplementation via venturi mask. She was started on Enoxaparin, Bosentan, Sildenafil and Acenocoumarol. She was asymptomatic till 24 hours after surgery when suddenly; she developed breathlessness, tachypnoea and fall in SpO_2. Patient was propped up, nebulized with levo-salbutamol; given furosemide injection and oxygen supplementation was continued. A repeat ECHO was performed which revealed suspicion of Atrial Septal Defect (ASD) with very high right ventricular systolic pressures (144+RA pressure). A transesophageal echocardiography was advised after stabilization. Patient continued to have dyspnea for which she was shifted to ICU and put on Non-Invasive Ventilation (NIV). She improved symptomatically within a few hours.

A pulmonary artery catheter was inserted and pressures were measured on day four of surgery which showed Central Venous Pressure (CVP)-14/2(9) mm Hg, Right Ventricular Pressure (RVP)-134/7(58) mm Hg, Pulmonary Artery Pressure (PAP)-135/61(86) mmHg, Pulmonary Capillary Wedge Pressure (PCWP)-20/14(16) mm Hg, Cardiac Output (CO)-3.58 l/min, Cardiac Index (CI)-2.13 l/min/m^2. In ICU, further blood investigations were sent and her NT-Pro BNP and D dimer levels were found to be significantly elevated. RA factor turned out to be negative.

TEE was done 16 days after surgery as patient stabilized. It showed a 25 mm ASD with severe TR and enlarged RA and RV. Patient was gradually weaned off and oxygen support was reduced. She was discharged on 17th day of surgery after complete stabilization on tab Acenocoumarol 2 mg once a day, Tab. Bosentan 125 mg twice a day, Tab. Sildenafil 20 mg thrice a day and antibiotics.

DISCUSSION

Pregnancy is associated with dramatic physiologic changes in various organ systems of the body, especially the cardiovascular

system. It poses an immense risk to females with PAH and maternal mortality remains to be unacceptably high (30-50%) in these patients [1]. We hereby reported the successful perioperative management of a parturient with severe PAH due to an undiagnosed Eisenmenger's syndrome, posted for emergency caesarean section.

Pulmonary hypertension per se is an important risk factor for increased perioperative morbidity and mortality. Stress, pain, ventilation, and surgical related inflammation can further increase pressure and resistance within the pulmonary arteries leading to right heart failure. The highest risk for perioperative mortality can be attributed to pregnancy and patients undergoing emergency surgeries [2]. Maternal mortality in patients undergoing caesarean section can be as high as 65% [3].

Common cause of pulmonary hypertension in women of child bearing age is a large intracardiac or aortopulmonary shunt due to CHD [4]. The risk of maternal death in pregnant parturients for unoperated CHD with severe pulmonary hypertension is very high. The progressive increase in plasma volume which peaks in the third trimester, compromises the already overburdened right ventricle [5]. There are various markers for poor prognosis in such patients which include maternal haematocrit >60%, arterial oxygen saturation <80%, right ventricular hypertension and syncopal attacks [3]. Various factors such as hypoxia, hypercapnia, acidosis etc., can lead to an increased pulmonary vascular resistance, increased pulmonary arterial pressures and ultimately reversal of the shunt in ASD, known as Eisenmenger's syndrome.

Pregnancy induced systemic vasodilation is detrimental in patients with Eisenmenger's syndrome. Decreased Systemic Vascular Resistance (SVR) in pregnancy can increase right to left shunting leading to a further worsening of hypoxia with significant risk to both the mother and the fetus. At the time of labor and delivery severe haemodynamic compromise may occur due to labor pain which increases Pulmonary Vascular Resistance (PVR) and mobilisation of blood volume during uterine contractions. Anaesthetic management of a pregnant patient for caesarean section with pulmonary hypertension due to Eisenmenger's syndrome is a challenge even to a seasoned anaesthesiologist.

The anaesthetic goals in a pregnant patient, with Eisenmengers's syndrome, for a caesarean section, is to maintain a stable cardiac output without worsening of intracardiac shunt. Though both central neuraxial block and general anaesthesia have been used, none out of the two are completely safe and both can increase the PVR. Central neuraxial block can increase PVR by decreasing SVR and general anaesthesia due to catecholamine release during laryngoscopy, intubation and during Positive Pressure Ventilation (PPV). Regional anaesthesia has been reported safe in pregnant patients with Eisenmenger's syndrome for caesarean section, though most advocate an epidural block [6]. Epidural anaesthesia, being slow in onset is presumed to have minimal precipitous haemodynamic changes. A meta-analysis has shown that both general anaesthesia and central neuraxial blockade have significant morbidity and mortality, and there was no significant difference in perioperative mortality between the two techniques.

Our patient presented with central cyanosis, clubbing, exercise intolerance and ECHO suggested pulmonary hypertension. All these findings were pointing towards a cardiac cause of pulmonary hypertension but unfortunately sinus venosus ASD is frequently missed on transthoracic echocardiography. Transesophageal echocardiography, which is more sensitive, could not be done in emergency settings in our hospital [7].

Our patient had respiratory distress on day two, which could have been due to pulmonary embolism, right heart failure, septicemia or a sudden rise in PVR. Since she responded to measures to decrease PVR (bosantan, sildenafil, IPPV, lasix), we presume there was an increase in PVR postoperatively due to pain or fluid overload, which is most common.

We wanted to avoid general anaesthesia so as to avoid increases in PVR due to PPV. Though epidural anaesthesia is safe, it always has the risk of failure, which would have necessitated the administration of general anaesthesia. Spinal anaesthesia can lead to a disastrous fall in SVR. So we decided to go ahead with combined spinal epidural anaesthesia using a small dose of bupivacaine in spinal block, with the aim of preventing any precipitous fall in SVR or a dangerous rise in PVR [Table/Fig-1].

	Central neuraxial blockade	General anaesthesia
Advantages	PVR maintained/slight decrease	Airway secured SVR maintained
Disadvantages	Risk of failure (epidural) Disastrous fall in SVR (spinal)	Increases PVR

[Table/Fig-1]: Comparison between central neuraxial blockade verses general anesthesia for Caesarean section.

Though general anaesthesia is more commonly used in Eisenmenger's syndrome, a combined spinal epidural anaesthesia has also been safely used [8]. Spinal anaesthesia has the advantage of ensuring early onset and a better quality of anaesthesia. Epidural anaesthesia provides postop pain relief.

CONCLUSION

Thus, a combined spinal epidural anaesthesia, if used along with strict maintenance of SVR and PVR, is a safe bet in pregnant patients with Eisenmengers syndrome for caesarean section.

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