Primary Pulmonary Hypertension - A Case Report

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Abstract: Pregnancy in women with pulmonary hypertension stresses the already compromised cardiovascular system and is associated with very high peripartum mortality. Though the mode of delivery is decided by obstetric factors the optimal anaesthetic management is either controversial or inconclusive.

We present a case of a parturient with primary pulmonary hypertension who underwent caesarean section under general anaesthesia with good maternal and neonatal outcome.

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A patient with primary pulmonary hypertension for caesarean section

A case report

Introduction

Primary pulmonary hypertension (PPH) is a rare disease characterized by elevated pulmonary artery pressure with no apparent cause. PPH is also termed precapillary pulmonary hypertension or, more recently, idiopathic pulmonary arterial hypertension (IPAH). The diagnosis is usually made after excluding other known causes of pulmonary hypertension. Dresdale and colleagues first reported a haemodynamic account of IPAH in 1951.

Incidence

- In the US: IPAH is responsible for approximately 125-150 deaths per year and has an incidence rate of approximately 1-3 cases per million population per year. The incidence and prevalence of IPAH are considerably higher than those for pure IPAH.
- Internationally: The worldwide incidence of IPAH approximates that observed in the United States.

Pregnancy in women with pulmonary hypertension stresses the already compromised cardiovascular system and is associated with very high peripartum mortality.

Though the mode of delivery is decided by obstetric factors the optimal anaesthetic management is either controversial or inconclusive.

We present a case of a parturient with primary pulmonary hypertension who underwent caesarean section under general anaesthesia with good maternal and neonatal outcome.

History

A 25 year old primary gravida at 29 wks of gestation was referred to our hospital as a case of

pregnancy with primary pulmonary hypertension. She was diagnosed as a case of primary pulmonary hypertension during regular follow up in a private hospital.

She presented to us with complaints of dyspnea on exertion – grade 1-2 and pedal edema since a few days. She had no other significant history.

On examination:-

P= 110/min, BP= 150/90mmhg in right arm in supine position, There was no pallor, icterus, cyanosis, clubbing and pedal edema present, JVP was normal

Cardiovascular system revealed a loud P_2, S_3 gallop and an ejection systolic murmur in the pulmonary area. Respiratory system examination was normal with no added sounds

Per abdominal examination revealed uterus at approximately 28 to 29 wks of pregnancy

Central nervous examination was normal.

Patient was diagnosed as Primigravida at 29wks with pulmonary hypertension. It was decided to monitor the patient till the termination of pregnancy as she was a case of high risk pregnancy.

Patient was on the following treatment during her stay before delivery

T. digoxin 0.25mg OD, T. lasix 40 mg OD, T. Luminal 60mg TDS, T. alpha methyl dopa 250mg TDS, T. iron and folic acid

Patient was also started on Inj Enoxaparin 5000 IU OD

PATIENT COMPLAINED of decreased fetal movements after about 11/2 months of admission i.e. around 35 wks of gestation. Same complain was repeated on the subsequent day .Hence taking this into consideration it was decided to terminate the pregnancy by caesarean section. Prior to surgery, Inj. Enoxaparine was discontinued for 24 hrs.

Preoperative investigations:-

Hemoglobin-12.7gm%, TC-8000, BT-2 min, CT-8 min, APTT-WNL, RFT WNL, ECG WNL, ABG WNL

Echocardiography revealed a dilated right ventricle with mild right ventricular hypertrophy. IVS/IAS was bulging to the left. Ejection fraction was 70% & there was diastolic dysfunction .Pulmonary art systolic pressure was 50mmhg ; there was no other valvular abnormality or associated defects.

Anaesthetic management

Patient refused consent for epidural anaesthesia. .After obtaining high risk informed consent, the patient was taken to the theatre. Base line pulse-90/min BP-110/70mmhg SaO_2-98%.ECG, Pulse oximeter, NIBP were attached to the patient

Premedication: - IV Metoclopramide 10 mg, IV Ranitidine 50 mg

Preoxygenation with 100%O_2 for 5min was done

Induction: - IV Thiopentone sodium (1.25%) 325mg, IV Succinylcholine 80mg along with IV Xylocard 80mg was given prior to intubation to attenuate pressor response.

Patient was intubated with cuffed oral endotracheal tube no 7 (polyvinyl chloride)

Maintenance: - 66%O_2 + 33%N_2O throughout surgery.

After delivery of baby, the patient was given IV oxytocin 5units bolus + 15units in IV fluid, IV 20mg Pethidine +10mg Phenargan, IV Vecuronium 3mg.when patient starts breathing. Surgery lasted for about 50 min. Throughout the surgery vitals were stable SaO_2 remained 99-100%. The patient reversed with IV Neostigmine 2.5 mg + 0.5mg glycopyrrolate. Post extubation patient was conscious, cooperative, vitals stable, maintaining SaO_2. RS & CVS Examination -NAD

Post operatively the patient was monitored in CCU. Her course in hospital was uneventful and she was discharged after 7days in good physical health.

Discussion

World Health Organization (WHO) has proposed a classification system for pulmonary

hypertension based on common clinical features using five categories:

1. Pulmonary arterial hypertension (PAH)
2. Pulmonary venous hypertension,
3. Pulmonary hypertension associated with disorders of the respiratory system,
4. Pulmonary hypertension due to chronic thrombotic or embolic disease, and
5. Pulmonary hypertension due to disorders directly affecting the pulmonary vasculature.[1]

Under this system, primary pulmonary hypertension (PPH) is a subset of PAH. Secondary pulmonary arterial hypertension, another subset of PAH, is commonly caused by certain immunologic diseases such as CREST syndrome (calcinosis, Raynaud's disease, esophageal dysmotility, sclerodactyly, and telangiectasia) and drug therapies such as PHEN-FEN.

The following diagnostic criteria for primary pulmonary hypertension were developed on the basis of findings in the National Institutes of Health (3) and Mayo Clinic series (6):

- Mean PA pressure greater than 25 mm Hg at rest or 30 mm Hg with exercise
- No evidence of left-sided myocardial, valvular, or congenital heart disease (with the exception of high-flow and low-pressure shunts,)
- No clinically significant lung disease, pulmonary emboli, or active vasculitis

The classic pathologic presentation is pulmonary arteriolar hypertrophy with obliterating arteriolar plexiform lesions. Presentations that are seen much less often are pulmonary veno-occlusive disease and pulmonary hemangiomatosis.

As seen from the echocardiographic findings of our patient these criteria are fulfilled.

The enhanced activity of thromboxane or diminished activity of prostacyclin or impaired synthesis of nitric oxide has also been associated with PPH. But whether these are the cause or the result of the disease is unclear.

The hormonal changes associated with pregnancy allow large amount of fluid to accumulate

in the interstitial space. Following delivery, this fluid is suddenly shifted to maternal circulation, increasing preload significantly and further increasing pulmonary hypertension. This lethal risk is highest during first 10 days of post partum period.

In healthy pregnant woman PVR is 34% less than non pregnant states, as the prostacyclin production is increased five fold. The withdrawal of this pulmonary vasodilator effect of prostacyclin in immediate post partum period may account for majority of deaths. When a patient with PH must undergo a Caesarean section, the prognosis worsens. The mortality is around 70% in patients with Eisenmenger's syndrome who are delivered by Caesarean section. Roberts et al published the only report of a patient with PPH who survived after Caesarean section. The maternal mortality is increased after Caesarean births in women with normal cardiovascular systems. Women with marginal cardiac reserve and pulmonary hypertension face added risks associated with postoperative fluid shifts, post-surgical pain and a higher incidence of thromboembolism after operative delivery. The increased mortality after Caesarean section may also be related to the pre-surgical status of the patient. Johnson et al. advocated elective delivery near term, when fetal lung maturity is reached, to avoid further cardiovascular stresses, and suggested that Caesarean delivery will avoid the stress of labor. The stress of labor, however, can be minimized with good analgesia and careful management. We think it is prudent to avoid elective Caesarean section because of the increased mortality associated with operative delivery in Eisenmenger's syndrome and PPH. Caesarean section may, however, be unavoidable for obstetric reasons, or because the mother's condition is so precarious that she may not survive long enough to deliver vaginally. In such cases, the anaesthetic management is of critical importance for a favorable outcome.

The anaesthetic management of pregnant patient with PPH remains controversial. However the goals of anaesthetic management include:

- Avoiding marked decrease in venous return

- Avoid marked reduction in SVR
- Avoid myocardial depression

Both the inhalational induction and narcotic based induction are recommended. But the disadvantage includes slow induction, neonatal depression, maternal myocardial depression, effects of positive pressure ventilation among others.

Nitrous oxide may increase PVR in patients with preexisting PH. Ketamine, by releasing catecholamines, can increase PVR. The other IV or inhalation anaesthetics,

neuromuscular blockers and antagonists have little effects on PVR. The use of PA catheters has not reduced the mortality in parturients. The difficulty in insertion of PA catheter, due to low CO, large RV, precipitation of arrhythmias and difficulty in obtaining accurate measurements due to elevated PVR, Avoiding further increase in PVR measurements due to elevated PVR, lead to its questionable usefulness. Echocardiography can be very useful monitor in these patients as it also provides information regarding contractility, EF, wall motion abnormalities, biventricular interaction etc.

If preload and after load are well maintained, regional or peripheral blocks are ideal. The single biggest predictor of outcome in patients with pulmonary hypertension is presence of RV failure. We decided to perform general anaesthesia as this was a first kind of case in our institution and review of literature suggested both kinds of anesthesia being administered to such patients.

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