Case Report

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Anaesthetic Management of a Case of Severe Pulmonary Artery Hypertension for Excission of A Vulval Mass

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Abstrat

It is known that patients with pulmonary hypertension are at high risk for anaesthesia and surgery. Primary pulmonary hypertension (PPH) has now been replaced byidiopathic pulmonary hypertension or pulmonary artery hypertension (PAH). PAH is arareform of progressive fatal disease. Occasionally these patients may be posted for non cardiac surgeries.Ideal anaesthetic technique involves maintaining of stable pulmonary and hemodynamicparameters for a possible good outcome. Here we present a 31 year old female with severe pulmonary artery hypertension who underwent successful excision of a hugevulvalmass.

Keywords: Pulmonary Hypertension; BiPAP; Regional Anesthesia.

Introduction

Pulmonary hypertension (PH) is a disorder characterized by abnormally high blood pressures in the pulmonary vasculature. PH is a progressive, fataldisease [1] associated with high mortality due to the stress of surgery andanesthesia [2]. A detailed understanding of PAH as well as associated risks must be known for safe and smooth conduct of anesthesia andpost operative care. Patients with pulmonary hypertension are amongst the most challenging for anaesthesiologists to manage. Furthermore, such patients undergoing surgery have a high perioperative morbidity and mortality. Any factor contributing to further increase in pulmonary hypertension will lead to decompensation of a stable disease which can lead to unfavorable results. This case report describes successful management of non-cardiac, non-obstetric surgery in a 31 year old female with severe PH who underwent excision of vulval

Case Report

A 31 year old female was admitted to our hospital with history of fever and cough for three days. She wasa known case of primary pulmonary hypertension and hypothyroidismon regular medications and following up at our hospital clinics. She had no drug allergies. Her medications included bosentan, iloprost and thyroxin. Patient deteriorated during her hospital stay and developed respiratory distress. She was put on oxygen via face mask @ 5l/min. Vital parametersat thistime were, heart rate of 135/min, tachypnoea with respiratory rate of 22-24/min, blood pressure130/70mmhg, SpO_2 87-90%. Chest revealed bilateral equal air entry with scattered crepitations.

A big vulval mass measuring 20x23 cm in size, pedunculated, non-tender with areas of necrosis and foul smelling was noticed on examination. This swelling was having a thick peduncle.A historyof recent increase in the size of swelling was noticed.

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Patient's condition worsened further in the intensive care unit, where she was diagnosed to be suffering from sepsis. The vulval mass was attributed to be the source of infection and hence the gynaecologists advised excision of the vulval mass.Laboratory investigations showed a high neutrophil count. Coagulation profile and other biochemical parameters were within normal limits. EKG showed right ventricular strain with right axis deviation. Transthoracic echocardiogram was carried that showed the following, a good systolic function with EF 58%. Tricuspid valve calcification with moderate tricuspid regurgitation, normal left ventricular dimension and wall thickness, no regional wall motion abnormality, dilated right ventricle (RV) with impaired right ventricular function, no pericardial effusion, no vegetation's and pulmonary arterial systolic pressure recorded as 100mmhg Before the surgery and during the ICU stay the patient deteriorated further. She became further tachypneic. She was placed on, BiPAP of 15/5cm H_2O. SpO2 was maintained at 90-92% with BiPAP support. As the vulval swelling was assumed to be the source of infection, it was decided to excise the mass . Patient was accepted as ASA 4E in view of sepsis and severe pulmonary artery pressures of100mmhg. Patient was wheeled into the operation room with BiPAP, where she was connected to the standard ASA monitors. She was anxious looking, with HR112 -114/min, SpO_2 88-90%. She had right radial artery canula. Invasive blood pressure could be monitored BP 112/ 67 mmhg. Blood gas carried out, showed features of respiratory acidosis.The plan was to institute local anaesthesia into the peduncle of the mass and get it excised. However the peduncle was very vascular and local infiltration could not be given. It was decided to give her subarachnoid block. Under all aseptic precautions 1.5ml of 0.5% heavy bupivacaine was administered intrathecally at lumbar 3/4 levels using 27G pencil point needle in the sitting position. later she was placed 45 degrees head up and in lithotomy position. BiPAP 15/5cmh_2o continued during the whole procedure. Her vital parameters remained stable, HR 115-118/min, blood pressure 100/64-110/68mmhg, SpO_2 90-92%. Her temperature was also kept with in normal limits. All those factors that could have resulted in precipitation of the symptoms of PH were avoided .

Surgery lasted for hour and a half. After the procedure, patient was shifted back to ICU on BiPAP. She was gradually weaned off from BiPAP, the next day and placed on oxygen via face mask. She remained stable in ICU and was shifted to gynaecology ward after 3 days. She was discharged home 8 days after surgery. She is being regularly followed up at our hospital clinics. Her cardiac status remains stable. She is continuing her medications for pulmonary hypertension.

Discussion

Although mild PH does not affect anaesthetic outcome, patients with moderate to severe PH can have adverse outcome [1]. Patients with PH are at high risk to undergo cardiac as well as non cardiacsurgery [2].

Accordingto 4th world symposium[3] Pulmonary hypertension is as mean pulmonary arterial pressure of more than 25 mmhg at rest or more than 30mmhg at exercise.

WHO classifiesPH into five groups based on the mechanism causing the disease:

1. Pulmonary artery hypertension (PAH), idiopathic and develops over the years
2. Pulmonary hypertension owing to left heart disease
3. Pulmonary hypertension secondary to lung disease
4. Pulmonary hypertension associated with chronic embolic/thromboembolic disease.
5. Pulmonary hypertension secondary to disorders affecting pulmonary vasculature

PH Is a progressive fatal disease [4]. The median life expectancy for non-treated patients is approximately 2-3 years from the time of its discovery. Price et al in their study have shown that there is 15% mortality at 1 year after diagnosis. PH is due to a combination of pulmonary arterial vasoconstriction, and in situ thrombosis. Eventually in PH pulmonary artery pressures and pulmonary venous pressures are raised. These high pressures lead to load on left ventricle that in turn leads to right ventricular hypertrophy and dilatation and eventually right ventricular failure. Patients with PH usually have non-specific symptoms like dysponea, fatigue, dizziness, syncope, chest pain, palpitations, cough, peripheraledema, ascites, hepatomegaly, tachycardia, tachypnea and raised jugular veinous pressure. Factors like hypoxemia, hypothermia, hypotension, acidosis, hypervolemia, pain and, increased intrathoracic pressures aggravate PH. In severe PH, surgery should only be considered if it is lifesaving.

A proper pre-operative evaluation should be carried out and involves multidisciplinary approach and includes clinical examination, ECG, Chest X-ray,

arterial blood gas analysis, echocardiography and metabolic profile. Echocardiography provides useful tool for diagnosis of PH. However, Right heart catheterization is gold standard for measuring the hemodynamic pressures in the pulmonary circulation and getting information about the right heart functionality.

Various treatment options are available and they include, Calcium channel blockers: Nifedipine, diltiazem and amlodipi Prosteinoids, they are potent vasodilators, delivered by continuous intravenous infusion. Endothelin receptor antagonist, bosentan [5] is taken orally Phosphodie sterase inhibitors, Sildenafil and milrinone are pulmonary and cardiac inodilators. Inhaled vasodilators includeiloprost [6] and Nitric oxide.

In elective cardiac as well as non-cardiac surgeryall undiagnosed causes of PH should be identified and optimized. On the other hand, in emergency situations we may not have adequate time to correct and optimize the patient condition and surgery may be done with explained risks.Our case was also an emergency surgery where there was not much time for properly optimizing the patient condition.

A proper preoperative evaluation should be carried out and includes clinical examination, ECG, Chest X-ray, arterial blood gas analysis, echocardiography and metabolic profile. Right heart catheterization is gold standard for measuring the hemodynamic pressures in the pulmonary circulation and getting information about the right heart functionality. All undiagnosed causes of PH should be identified and optimized. On the other hand in emergency situations we may not have adequate time to correct all the underlying causes and surgery may be done with risks explained.

Allanesthetic techniques can in principle beapplied to patients with pulmonary hypertension. Regional anaesthesia offers an advantage over general anaesthesia in terms of notimpairings pontaneous breathing and avoidingelevated pulmonary pressures induced by laryngoscopy and mechanical ventilation [14].

Martin et alshowed that operative mortality in patients with Eisenmengers syndrome was 18% with general anaesthesia Vs 5% with regional anaesthesia [7]. Price et al. also suggest in their study that the use of general anesthesia in PH was linked to worse patient outcome than regional anesthesia [8]. Most of these patients are anticoagulants and they need to be stopped before planning regional anesthesia [9] .

Local anesthesia applied alone in a field around surgical site can be the safestapproach. Continuous techniques should be preferred over bolus administration of local anaesthetics to avoid uncontrolled drops in blood pressure, decrease in myocardial perfusion and precipitation of right heart failure. For limb surgery ultrasoundguided plexus or peripheral nerve blocks are the best choices. Spinal anaesthesia when given as bolus can cause sharp fall in peripheral vascular resistance and decrease myocardial perfusion. However saddle block performedwith low dose of local anaesthetic can prevent sudden changes in hemodynamic parameters. In obstetrics, successful application of lumbar epidural anaesthesia has been repeatidly described [15], even though recent literature describes higher morbidity of pregnant women with pulmonary hypertension. Antanasoff P. et al. used epidural anaesthesia for caesarian section [10] in a case of severe pulmonary artery hypertension General anaesthesia on the other hand provides the uncompromised airway and safe oxygenation. Moreover, selective pulmonary vasodilators like iloprost [11] can be administered through the breathing circuits. For abdomen and thoracic surgeries general anaesthesia can be combined with epidural anaesthesia for better haemodynamic control [12]. If general anaesthesia is employed, transesophageal echocardiography (TEE) may provide the best real time monitor of cardiac preload and status of right to left shunting. Patientneeds tobe observed in the ICU for first few dayspost operatively as there is a high risk of sudden death [13]. Finally theyshould be put back totheir usual oral anticoagulants post-operatively.

Conclusion

Perioperative management of patients with PH presents an interdisciplinary challenge that requires adequate involvement of anaesthesiologist, surgeons, cardiologist and intensivist. Although anaesthetic management of PH patients continues to be challenge, athorough assessment of the patient andameticulous attention to details minimizes the possibility of complications and allow best possible outcome.

Risk Disclosure

Patients with PH have increased morbidity and mortality. A pre-operative assessment of risks and possible benefits of surgical intervention plays a vital role. All these risks should be addressed andexplained well to the patient.

Conflict of Interest None of the authors involved in this study have any conflicts of interest to disclose.

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