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Case Report

Anaesthetic management for a patient with Eisenmengers syndrome undergoing ovarian cystectomy

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ABSTRACT

Background: Eisenmenger's syndrome is a cyanotic congenital heart disease, where septal defects or patent ductus arteriosus advances to pulmonary hypertension with reversed/ bidirectional shunt. Any condition that lowers systemic vascular resistance, increases the degree of right to left shunt and the risk for the patient. Case Presentation: We report anaesthetic management of a 25-year-old P2L2 Female weighing 50kg with Eisenmenger's syndrome presented with an 8.3cm×7.5cm×8.4cm right ovarian cyst posted for ovarian cystectomy.

Materials and Methods: The procedure was done under epidural anaesthesia, & post-operatively epidural analgesia was administered for 48 hours. After removal of epidural catheter, multimodal analgesia and anticoagulant therapy with Low Molecular Weight Heparin (LMWH)5000 IU by subcutaneous route was initiated.

Discussion: The mainstay of anaesthetic management in Eisenmenger\'s syndrome revolves around providing analgesia without decreasing systemic vascular resistance, or increasing pulmonary vascular resistance. Other concerns are hypercoagulable state, global hypoxia, Volume overload, hypercarbia, acidosis, hypovolemia.

Conclusion: Systemic hypotension and increased pulmonary resistance associated with General Anaesthesia, & marked sympathectomy associated with spinal anaesthesia make both these modes of anaesthesia relatively risky compared to the plain epidural. Other concerns addressed during perioperative management were Oxygen support, early thromboprophylaxis, infective endocarditis prophylaxis, fluid restriction, and correction of acidosis.

1. Introduction

The term Eisenmenger complex was coined by Victor Eisenmenger in 1897, to define large ventricular septal defects (VSD) with pulmonary hypertension.[1,2] In 1958, Wood redefined Eisenmenger complex as pulmonary hypertension with reversed or bidirectional shunt associated with patent ductus arteriosus or septal defects.[3–6] Prognosis is grim in these patients, with life expectancy mostly being 20 to 30 years.[7] High pulmonary artery pressure with a fixed vascular resistance is not a surgically reversible condition.[8] Degree of right-to-left shunt depends on three factors: (a) Severity of pulmonary hypertension and size of the communication, (b) the relation between Pulmonary Vascular Resistance (PVR) and Systemic Vascular Resistance (SVR) and (c) the contractile state of the right ventricle.[9] Early and sudden death in the postoperative period is often associated with Eisenmenger syndrome. Non-cardiac surgeries carry a mortality risk of as high as 30% in these patients.[10,11] We report successful anaesthetic management of exploratory laparotomy in a patient with Eisenmenger's Syndrome.

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2. Case Presentation

A 25-year-old P2L2 female weighing 50 kg, of ASA physical status III, tubectomised, had complained of difficulty in passing urine for ten days. She gave a history of New York Heart Association (NYHA) grade II breathlessness since last year, grade II clubbing since childhood. On auscultation, a loud P2, ejection systolic murmur (prominent over the pulmonary area), and pansystolic murmur (in the tricuspid area) were heard. Bilateral Jugular Venous Pressure (JVP) waves were above clavicles at 45° inclination. Oxygen saturation was 92% on room air. Routine biochemical investigations revealed: haemoglobin 16.7, erythrocyte count: 561000/cubic mm, (indicating erythrocytosis) Haematocrit: 48.9, normal values of Serum Electrolytes, Proteins, renal & liver function tests. Covid rapid antigen test was negative. High-Resolution Computerised Tomography scan of thorax portrayed: Coronavirus disease Reporting and Data System (CORADS) score of 1, prominent pulmonary arteries and right ventricular hypertrophy. Twelve-lead electrocardiogram (ECG) illustrated ST elevation in aVR, aVL, T inversion in V4, V5, V6, RV strain pattern in Lead V1. Two-Dimensional Echocardiography (2D echo) revealed severe pulmonary arterial hypertension with raised Right Ventricular Systolic Pressure (RVSP) 115 mm Hg, a 16 mm peri-membranous ventricular septal defect allowing bidirectional flow, mild Mitral Regurgitation (MR) and Tricuspid Regurgitation (TR). Transvaginal USG demonstrated an 8.3 cm x 7.5 cm x 8.4 cm complex multiloculated single right ovarian cystic lesion with low-level internal echoes and septations, mild free fluid in the Pouch of Douglas and a few Nabothian cysts (largest: 4 mm) in the vaginal cervix. We informed the patient of her cardiac risks of anaesthesia before posting for a right ovarian cystectomy. Perioperative Intensive Care with continuous intensive cardiac monitoring (invasive and non-invasive) was arranged. Preparation for a central line, radial arterial line and mechanical ventilation were kept ready. Preoperative Arterial Blood Gas (ABG) depicted a hypoxemic picture with pH 7.36, pCO2 27.3, pO2 50.6, sO2 90.2 and HCO3- 23. Two peripheral large-bore intravenous cannulas were inserted. Dopamine infusion was kept ready at a rate of 10 μg/kg/min. The plan of anaesthesia was epidural. In the operation theatre, Ringer Lactate infusion was started at 50 mL/hour, urine output was monitored hourly till post-operative 24 hours. Monitors were attached for non-invasive blood pressure, ECG and pulse oximetry monitoring. The patient was premedicated with intravenous injection pantoprazole 40 mg on the night before surgery and intravenous injection ondansetron 4 mg on the morning of surgery. After sensitivity test, intravenous Inj. Amoxycillin 1g and Inj. Gentamycin 80 mg were given at induction of anaesthesia as a prophylaxis for infective endocarditis. A 16-gauge epidural catheter was inserted through the 3rd and 4th lumber vertebral interspace using the Loss of Resistance (LOR) technique, fixed at 8 cm in situ after positive meniscus sign. 3 mL of 2% lignocaine hydrochloride - adrenaline solution (1:10,000) was given as a test dose. Toxic doses of local anaesthetics in this patient were calculated for lignocaine & bupivacaine, although the use of bupivacaine was restricted due to its potent systemic vasodilatory effect. The patient was made supine after securing an epidural catheter and 100% oxygen was supplied with Bain's Circuit. Oxygen saturation improved to 98%, the patient remained hemodynamically stable throughout the procedure. Epidurally given anaesthetic doses are mentioned below: (Table 1)

Table 1: Intermittent epidural bolus dosage of anaesthetics given to the patient against time (in minutes) from test dose

Time in minutes (From test dose)	Drug name	Volume	Dose
05:00	Plain Lignocaine (2%)	4 ml	80 mg
15:00	Plain Lignocaine (2%)	2 ml	40 mg
45:00	Bupivacaine (0.5%)	4 ml	20 mg
60:00	Tramadol (50 mg/ml)	1 ml	50 mg

The achieved level and depth of anaesthesia was adequate during the 50-minute-long procedure. Mild sedation was given with Injection Midazolam 1 mg and Injection Fentanyl 0.5 μg/kg by intravenous route during the surgery. The patient was kept on 4 litres per minute of Oxygen by Hudson mask and closely monitored for 3 hours in the Post-Anaesthesia Care Unit before shifting to the Intensive Care Unit. The epidural catheter was kept in situ for postoperative epidural analgesia for two days. Four epidural doses of 0.125% bupivacaine, 6 ml each, were administered 12 hours apart. Multimodal analgesia and anticoagulant therapy with subcutaneous LMWH 5000 IU were initiated on the third day. One episode of shivering and fall of oxygen saturation occurred 8 hours after surgery with no breathlessness, tachycardia or tachypnoea. The oxygen flow rate was increased to 8 litres per minute and saturation improved to 96%, with ABGA showing similar improvement in pO2 (80.3 mm Hg), sO2 (97.2%). Oxygen was tapered on the 3rd postoperative day. Discharged on the 5th postoperative day, the patient was advised routine follow up with a cardiologist regarding her cardiac conditions.

3. Conclusion

Eisenmenger's Syndrome is a result of pulmonary vascular disease arising in a patient with an aberrant connection between pulmonary and systemic circulation, where blood pressure in the pulmonary circulation is similar to or more than that of the systemic

circulation, resulting in a right to left/ bidirectional shunt through the aberrant connection.[12,13] The degree of shunt present depends mostly on the ratio between SVR and PVR. Any condition that raises PVR, or lowers SVR, results in an increased right-to-left shunt, and a global hypoxic picture. Cyanotic episodes, Polycythaemia due to chronic hypoxia and thromboembolic complications are prone to occur. Active Right-to-Left/ bidirectional shunt promotes chronic hypoxic and hyper carbic changes or acute episodes intraoperatively.[5] The chosen anaesthesia technique should avoid such changes in SVR/PVR ratio, hypoxia, hypercarbia, acidosis, systemic hypotension, hypovolemia, pulmonary hypertension. Although several papers portray general anaesthesia as a safer choice, it poses clear risks and disadvantages avoidable with epidural anaesthesia. SVR should be properly maintained, which dictates avoidance of loss of peripheral resistance, prohibiting the use of intravenous Thiopentone or Propofol as an induction agent. Catecholamines released during or after laryngoscopy or during Extubation can increase PVR. Intermittent Positive Pressure Ventilation (IPPV) increases intrathoracic pressure, decreases venous return, rising pulmonary arterial pressure and right to left shunt. Spontaneous ventilation can exacerbate hypercarbia, and nitrous oxide, hypoxia.[12] Inhalational agents can alter SVR/PVR ratio. Sevoflurane and Desflurane, although decrease SVR, do not affect Cardiac Output (CO) profusely.[14]) Systemic opioids may fail to provide adequate analgesia, and sympathetic pain response, i.e., tachycardia and increased cardiac output in the presence of VSD and pulmonary hypertension depict increased right to left shunt.[15]

Epidural was preferred over general anaesthesia due to its superiority in producing perioperative analgesia. A 16-gauge epidural catheter was inserted through the 3rd and 4th lumber vertebral interspace using the LOR technique. Use of Bupivacaine was restricted due to its potent systemic vasodilatory effect. 4 ml of 2% lignocaine (without preservatives) was given 5 minutes after test dose, and another 2 ml after 15 minutes. The sensory level of blockade achieved was T8. Complete motor blockade (modified Bromage score 3) was produced with the abovementioned dosage. 4 ml of 0.5% bupivacaine was given epidurally at 45 minutes to maintain the level of the blockade and, 50 mg tramadol was given epidurally to supplement postoperative analgesia at the end of surgery. The total dose did not produce any hemodynamic alteration.

100% oxygen was supplied using Bain's Circuit. The patient's oxygen saturation was 98%. Intravenous sedation was given with inj. Midazolam 1 mg and Inj. Fentanyl 0.5 μg/kg. The right ovarian cystectomy was performed within 50 minutes with stable vitals and no signs of hypoxia throughout the procedure. Postoperatively the patient was provided 4 litres per minute of oxygen by Hudson mask and monitored closely in post-anaesthesia care unit for 3 hours with no post-operative complications. The patient was shifted to the Intensive Care Unit (ICU) for observation for postoperative 48 hours, during which, epidural analgesia was provided. The patient had one episode of shivering and fall of saturation 8 hours after surgery without accompanying tachycardia, tachypnoea or breathlessness. The oxygen flow rate was increased to 8 litres per minute with subsequent improvement in pulse oximetry (97%) and ABG analysis pO2 (80.3 mm Hg), sO2 (97.2%). Oxygen was tapered on the 3rd day. The patient was discharged five days after surgery, with advice to follow up with cardiologists regarding her cardiac conditions.

The benefit of invasive central venous and arterial catheterisation and monitoring is controversial in Eisenmenger syndrome, as it comes with the risk of thromboembolism and air embolism, and large VSD can lead to paradoxical emboli with catastrophic consequences.[16] Hence Central venous line and Arterial Line were kept as standby measures only. Early thromboprophylaxis with 5000 IU subcutaneous LMWH was started to avoid the risk of thromboembolism during post-operative rest and immobilization.[17] Early ambulation is advocated. In post-surgical patients, the presence of septal defects is often linked with infective endocarditis. Thus, prophylactic intravenous doses of Inj. Amoxicillin 1 g and Inj. Gentamicin 80 mg were indicated.[18] Intravenous Inj. Dopamine at a rate of 10 μg/kg/min has a significant role in maintaining SVR and could be essential in an intraoperative emergency, thus an infusion pump with Dopamine was kept on standby.[17] Spinal route of anaesthesia with intrathecal 0.5% hyperbaric bupivacaine should be avoided as the loss of SVR is much intense with spinal anaesthesia, as are the risks of cardiotoxicity and central nervous system toxicity. Spinal route of anaesthesia with intrathecal isobaric 0.75% ropivacaine or isobaric 0.5% levobupivacaine has a less hemodynamic effect, less cardiotoxicity and central nervous system toxicity.[19] However due to the restriction on intravenous fluids in Eisenmenger syndrome, sympathectomy associated with spinal anaesthesia was unwelcome. Discussion on the use of additives like Tramadol, Dexmedetomidine and Clonidine with intrathecal anaesthesia is beyond the scope of this report, as literature is inadequate on their hemodynamic safety in Eisenmenger patients.

Maintaining good CO and SVR keeps baseline SVR/PVR value and shunt direction preserved. Control over the hemodynamic condition, strong analgesia, thromboprophylaxis and infective endocarditis prophylaxis avoids complications.

4. List of Abbreviations

LMWH = Low Molecular Weight Heparin; VSD = Ventricular Septal Defect; PVR = Pulmonary Vascular

Resistance; SVR = Systemic Vascular Resistance; NYHA = New York Heart Association; JVP = Jugular Venous Pressure; CORADS = Coronavirus disease Reporting and Data System; ECG = Electrocardiogram; 2D Echo = Two-Dimensional Echocardiography; RVSP = Right Ventricular Systolic Pressure; MR = Mitral Regurgitation; TR = Tricuspid Regurgitation; USG = Ultrasonography; ABG = Arterial Blood Gas; LOR = Loss of Resistance; IPPV = Intermittent Positive Pressure Ventilation; CO = Cardiac Output; ICU = Intensive Care Unit

5. Consent

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

6. Availability of Data & Materials

Data sharing is not applicable to this article as no datasets were generated or analysed during the current study.

7. Conflict of Interests

The authors declare that they have no competing interests.

Acknowledgements

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