CASE REPORTS

Non Cardiac Surgery in a Patient with Eisenmenger Syndrome-Anaesthesiologist's Challenge

Sohan Lal Solanki, Vaibhav Vaishnav, A. K. Vijay

CASE REPORT

A 44 year old female scheduled for vaginal hysterectomy was referred for pre anaesthetic evaluation. She had complaints of breathlessness, palpitation, fatigue, headache and visual disturbances. She had pulmonary tuberculosis 5 year back and recurrent chest infection since childhood. She had 2 live healthy issues and her pregnancies were uneventful.

On examination she was 46 kgs in weight, 150 cms in height. She had grade 2 clubbing, central cyanosis and right parasternal heave. On auscultation ejection systolic murmur was heard over 2nd left intercostal space and precordium, with loud and widely split 2nd heart sound. Lungs were bilaterally clear. Blood pressure (NIBP) was 110/70 mmHg and pulse was 84/min. Hemogram, coagulogram, Renal & Liver function tests were within normal limits. Pre-operative chest X-ray revealed changes of pulmonary tuberculosis, dilated central pulmonary arteries and cardiomegaly. Electrocardiogram (ECG) showed right ventricular hypertrophy with right axis deviation. Echocardiograpy showed enlarged right atrium and right ventricle with atrial septal defect (Secundum Type) with bidirectional flow and predominantly right to left shunt and ejection fraction of 50%. Arterial blood gases revealed pH-7.39, pO₂-54, pCO₂-26, cHCO₃-15, BE- -7 and SO₂- 89.0%.

After explaining anaesthetic procedure and risks involved informed consent was taken. In the operating room, standard ECG, NIBP and pulse oximeter were attached. 18 G intravenous cannula secured in left upper limb. Preoperative blood pressure was 110/72 mmHg and pulse rate was 86/min. Oxygen saturation (SpO₂) was 89% on room air. Oxygen was supplemented with venturi mask (FiO₂-0.5). SpO₂ increased from 89% to 95% on oxygen supplementation. A right internal jugular central venous catheter was inserted under local anaesthesia. Her pre induction central venous pressure (CVP) was 7 cm of Normal saline (NS). Under full aseptic precautions and after adequate local infiltration, epidural anaesthesia was given

in left lateral position in L3-L4 space with midline approach by Tuohy 18 gauge epidural needle using loss of resistance technique. A 20 gauge epidural catheter was inserted and test dose was given. 10 ml of 2% lignocaine with 1:200000 epinephrine admixed with 50 µg fentanyl was administered in fractionated 'graded' doses of 5 ml every 10 min; while assessing the pulse, blood pressure, respiration and keeping constant verbal contact. After 20 minutes there was sensory block up to T8 with adequate motor block. Epidural anaesthesia was maintained with 0.5% bupivacaine intermittently depending upon the recession of the sensory block. During surgery one litre of crystalloid (ringer lactate) was given. Intraoperatively her pulse rate ranged between 82-120/min, systolic blood pressure between 120-98 mmHg and SpO_a between 94-97%. CVP was maintained around 10 cm of NS. Blood loss was about 250 ml. After surgery patient was kept in post anaesthesia care unit for 2 hours and then shifted to ward. Arterial blood gases at end of surgery was pH-7.34, pO₂-58, pCO₂-30, HCO₃-15.5, BE-12, SO₂- 92%. Post operatively 4 ml boluses of 0.125% bupivacaine with fentanyl was given every 6 hours for pain relief until 24 hours. She developed one episode of hypotension (BP of 82/60 mmHg) with fall in oxygen saturation in evening on day of surgery. She was managed with intravenous fluid and oxygen. Subsequently her stay in ward was uneventful and she was discharged on 7th post operative day.

DISCUSSION

Eisenmenger syndrome is one of the leading causes of perioperative death (up to 19%)^{1,2} in patients undergoing non cardiac surgery (NCS). Eisenmenger syndrome is an uncommon complication of many congenital heart diseases, simple such as ventricular and atrial septal defect, as well as complex, such as single ventricle.^{3,4} Survival up to fifth and sixth decade is rare.⁴ Perioperative risk and outcome depends on urgency, duration of surgery, anaesthesia used and underlying pathology. Profound hemodynamic variability such as heart rate, blood pressure, volume status,

Drs.Sohan Lal Solanki, Vaibhav Vaishnav, Residents, A. K. Vijay, Associate Professor, Department of Anaesthesiology and Intensive Care, J.L.N. Medical College and Associated Hospitals, Ajmer (Rajasthan), India Correspondence: Dr. Sohan Lal Solanki, E-mail: me sohans@yahoo.co.in

oxygenation, and neurohormonal activation adds an extra stress on an already abnormal cardiopulmonary system. 5,6 Hematocrit >60%, arterial oxygen saturation <80%, right ventricular hypertension, syncopal attack and a fixed pulmonary hypertension not responsive to oxygen carries poor prognosis.7 Maintenance of adequate filling pressure and systemic vascular resistance, optimal analgesia, and timely or earlier management of clinical deterioration are the key points. Pregnancy is not well tolerated by patients with this condition. When pregnancy occurs in women with Eisenmenger's syndrome, medical termination is considered safer than any mode of delivery. Normal pregnancy outcome in our patient is probably because patient's with atrial septal defect develop Eisenmenger syndrome in later age group (90% during adulthood) than do patient with ventricular septal defect and patent ductus arteriosus who develop this condition earlier in life (80% during infancy).2

Both general and regional anaesthesia technique have been used, 1,8 numerous studies have suggested that there is no anaesthetic technique that is superior to other in offering myocardial protection in these patients.⁶ Patients with Eisenmenger's syndrome pose a challenge to anaesthesiologists due to inability to adapt to sudden changes in hemodynamics because their pulmonary vascular bed is fixed.6 Many agents used for induction and maintenance of general anaesthesia depress myocardial function and reduce systemic vascular resistance. So the choice of induction agents for general anaesthesia should be those agents which would cause least hemodynamic disturbance i.e. ketamine or opioid induction with etomidate. Epidural or spinal anaesthesia with higher dosage of local anaesthetic would be extremely hazardous because of inevitable peripheral vasodilatation associated with severe sympathetic blockade. Slowly titrated dose of local anaesthetic combined with epidural opioids is mandatory because it avoids abrupt fall in SVR. Oxygen is a pulmonary vasodilator which resulted in reducing the flow across the right to left shunt, this explains the improvement in SpO2 in our patient, when supplemental oxygen was given and hence implying a reversible shunt. Hypotension and desaturation in our patient in evening may be due to multiple pulmonary thrombosis or emboli. There is no conclusive evidence of thromboembolic phenomenon as no ventilation perfusion scan is available in our setup.

Volume depletion and prolonged fasting should be

avoided before surgery and all intravenous lines should be equipped with a device to filter air bubbles to prevent paradoxical air embolism. Alpha adrenergic agonist should be used if there is systemic arterial hypotension and hypovolemia should be treated with intravenous volume replacement.² Blood loss should be minimized. Some patient's with Eisenmenger syndrome may require higher hematocrit to maintain adequate oxygenation. Prevention of thromboembolism should be encouraged by early ambulation and if prolonged immobilization is anticipated subcutaneous administration of heparin should be given.^{2,9} In conclusion adult patients with Eisenmenger syndrome require special care while undergoing NCS, directed toward ameliorating their preoperative status and avoiding complications related to Eisenmenger syndrome.

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