Case Report

Anaesthesia for a patient with Eisenmenger's syndrome undergoing caesarean section

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ABSTRACT

Eisenmenger's syndrome is a cyanotic congenital heart disease that includes pulmonary hypertension with reversed or bidirectional shunt associated with septal defects or patent ductus arteriosus. The decreased systemic vascular resistance associated with pregnancy increases the degree of right to left shunting, thereby carrying substantial risk to both the mother and the foetus. The maternal mortality rate of pregnancy in the presence of Eisenmenger's syndrome is reported to be as high as 30–70%. We present a case of a 22-year-old primigravida with Eisenmenger's syndrome who gave birth at 37 weeks of gestation via caesarean section to a live female baby under general anaesthesia. On the third post-operative day, the patient developed tachycardia, tachypnoea, hypotension and decrease in oxygen saturation despite supplemental oxygen, clinically suspected pulmonary thromboembolism. We describe the anaesthetic management for caesarean section and its complications in a patient with Eisenmenger's syndrome. Although pregnancy should be discouraged in women with Eisenmenger's syndrome, it can be successful.

Key words: Anaesthesia, caesarean section, Eisenmenger's syndrome, pulmonary thromboembolism

INTRODUCTION

Victor Eisenmenger in 1897 coined the term Eisenmenger complex, which included large ventricular septal defect (VSD) and pulmonary hypertension.[1,2] Wood redefined this in 1958 as pulmonary hypertension with reversed or bidirectional shunt, associated with septal defects or patent ductus arteriosus.[3-6] The maternal mortality rate is high, with a cumulative risk of 30–70%.[7,8] Death can occur anytime during pregnancy or puerperium.[8] We describe the successful anaesthetic management for caesarean section in a patient with Eisenmenger's syndrome.

CASE REPORT

A 22-year-old primigravida weighing 46 kg, known case of Eisenmenger's syndrome at 37 weeks of gestation, was scheduled for an elective caesarean section. At the age of 15 years, a cardiac catheterization was performed, which revealed a large mid-muscular VSD with severe pulmonary arterial hypertension (120/50 mmHg). She was explained about the existing cardiac condition and advised to avoid strenuous work.

Pre-operatively, apart from exertional dyspnoea (grade II), she was asymptomatic. Physical examination revealed grade II clubbing, pulse rate of 92/min, blood pressure of 130/90 mmHg and room air oxygen saturation (SpO_2) of 90–92% with no rise in jugular venous pressure (JVP). No peripheral oedema or cyanosis was observed. Her lungs were clear to auscultate and cardiac examination showed regular rate and rhythm with loud P_2 and pansystolic murmur over the lower left sternal border. Electrocardiography showed right axis deviation with right ventricular hypertrophy. Two-dimensional echocardiography (2D Echo) with colour Doppler revealed a large muscular VSD (19 mm × 20 mm) with a bidirectional shunt and a good left ventricular function with ejection fraction

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of 58% with severe pulmonary hypertension. Her haemoglobin was 12.6 g%, with a haematocrit of 40% and platelet count of 1,50,000/mm^3. Arterial blood gas (ABG) on room air was pH-7.32, $PaCO_2$-35 mmHg, PaO_2-59 mmHg and SaO_2-91%. Cardiologist opinion was taken and she was advised to undergo elective caesarean section.

She was pre-medicated with Ranitidine 150 mg orally the night before surgery. Infective endocarditis prophylaxis was given. In the operation theatre, standard monitors, an arterial line and a central venous catheter were placed. Prophylactic dopamine infusion 10 μg/kg/min was started to maintain the systemic vascular resistance (SVR). After 3 min of pre-oxygenation, SpO_2 increased to 100%. The pre-induction blood pressure (BP) and central venous pressure (CVP) were 130/90 mmHg and 10 mmHg, respectively. Intravenous induction was carried out with a low dose of thiopentone sodium 150 mg and fentanyl 50 μg titrated to anaesthetic and haemodynamic effects. The patient was intubated endotracheally with suxamethonium 50 mg. Anaesthesia was maintained with 50% oxygen and 0.5% halothane in air and neuromuscular blockade was achieved with vecuronium 5 mg. The end tidal carbon dioxide ($EtCO_2$) was maintained between 32 and 35 mmHg.

A live female baby with Apgar score of 5 at 1 min and 8 at 5 min, weighing 1.9 kg, was extracted. Intravenous infusion of oxytocin 15 units was administered slowly over 30 min. Intraoperative analgesia was obtained with morphine 6 mg i.v. Blood loss was estimated to be about 500 mL. The patient remained haemodynamically stable throughout the procedure, which lasted for 45 min. Low-molecular weight heparin (LMWH) 5000 IU was given subcutaneously towards the end of the surgery. Neuromuscular blockade was reversed with neostigmine 2.5 mg and glycopyrrolate 0.5 mg and the patient was extubated and shifted to the post-operative ward with oxygen by mask for observation. Post-operatively, the patient was haemodynamically stable and dopamine was tapered off. Anticoagulation with heparin and analgesia with morphine 4 mg i.v. 8^{th} hourly were continued for the next 1 week. Perioperatively, CVP was used as a guide to administer i.v. fluids and was maintained around 10 cm of H_2O. Volume overload was avoided as it could easily precipitate right ventricular failure in these cases.[1]

On the third post-operative day, she was found to have cyanosis with tachycardia, tachypnoea and hypotension (90/60 mmHg) with SpO_2 of 75% despite supplemental oxygen. Her ABG revealed pH-7.42, $PaCO_2$-32 mmHg, PaO_2-45 mmHg and SaO_2-72%. Dopamine was restarted at 10 μg/kg/min to maintain the systolic BP above 100 mmHg. SpO_2 remained between 75% and 80% despite oxygen therapy. She improved gradually over a period of 3 days and dopamine was tapered off. She was discharged on the 15^{th} post-operative day.

DISCUSSION

In a review of pulmonary vascular disease and pregnancy, it was noted that in patients classified as having Eisenmenger's syndrome, the risk of maternal mortality was in the range of 30–50%. It was suggested that the degree of pulmonary hypertension determined the maternal mortality,[6] while in another quoted a range of 30–70%.[7] The two major problems facing a pregnant patient with Eisenmenger's syndrome are, firstly, a fall in the SVR (which could allow a right to left intracardiac shunt)[3,4] and, secondly, thromboembolism (which could fatally interfere with an already embarrassed pulmonary circulation).[9] The presence of fixed pulmonary hypertension not responding to oxygen therapy may perhaps be an absolute indication for termination of pregnancy. As advised by the cardiologist, to avoid the stress of labour and the late pregnancy, programmed caesarean section was planned. There are reports regarding elective caesarean section to optimize foetal development and to minimize the maternal risks at term in these types of patients.[2,4] These patients, however, are high-risk candidates for anaesthesia.[8,10]

The main anaesthetic goal is to avoid a fall in the arterial blood pressure by maintaining both the cardiac output and the SVR.[3,7,11] The goal of monitoring is to detect sudden changes in the haemodynamics early so as to initiate appropriate treatment and prevent further complications.[11]

The place of invasive monitoring in Eisenmenger's syndrome is controversial and, as with any other form of monitoring, the risk of complications must be weighed against the value of information obtained. These patients are polycythemic and intraarterial catheterization may be associated with a higher incidence of post-canulation thrombus formation. Insertion of central venous catheter has a potential

risk of infection and paradoxical air embolus.[11] The complications of pulmonary catheterization are pulmonary arterial rupture in the presence of pulmonary hypertension apart from arrhythmias and systemic embolisation.[8,12] By considering risks and benefits, we obtained continuous measurement of blood pressure via an intraarterial line but did not insert a pulmonary arterial catheter. Instead, we inserted a central venous catheter to detect right heart failure as the right heart is ejecting against high pulmonary vascular resistance and to optimize the pre-load.

If the patient requires an operation, either regional or general anaesthesia can be suitable.[9,10,13] Several authors have recently suggested that it is safe to administer epidural anaesthesia to a patient with Eisenmenger's syndrome.[11,12] However, in each of these reports, it appeared that the patient's pulmonary vasculature dilated in response to oxygen. In Eisenmenger's syndrome, the amount of right-to-left shunt depends in part on the ratio of SVR to pulmonary vascular resistance (PVR). Epidural anaesthesia causes sympathetic blockade that reduces SVR. If SVR decreases without a concomitant decrease in PVR, the amount of right-to-left shunt increases.[12,14] Therefore, in our case, we considered it undesirable to induce a sympathetic blockade that might have resulted in an increased right-to-left shunt. Epidural analgesia was not considered because our obstetric plan was to begin heparin perioperatively. Weiss and Hess[6] reviewed the literature and found that the strategies and concepts for providing regional anaesthesia during labour and delivery in anticoagulated patients are controversial, and also mentioned that lumbar anaesthesia contributes to the risk of spinal haematoma in patients receiving antithrombotic drugs. To avoid this, we opted for general anaesthesia. This may be achieved by combining a short-acting i.v. narcotic such as fentanyl in addition to a low dose of induction agent such as thiopentone sodium or ketamine or inhalational agents.[11,14] Although thiopentone causes a decrease in SVR, the effect is dose-dependent.[1] Hence, we used a low dose of thiopentone. Although ketamine has theoretical advantages over barbiturates as an induction agent, in that it does not reduce SVR but causes increase in the heart rate,[15] which is undesirable. Traditional and contemporary teaching considers all obstetric patients to be at increased risk for pulmonary aspiration compared with patients scheduled for non-obstetric elective surgical procedures, mandating antacid prophylaxis as well as rapid sequence induction with cricoid pressure. However, the requirements of a patient at risk for aspiration are difficult to reconcile with a judicious, titrated induction of anaesthesia that is ideal for a patient with severely compromised cardiac function.[16] Thus, rapid sequence induction was avoided in our case. The problems of general anaesthesia and positive-pressure ventilation are decrease in venous return and cardiac output. Hence, we decided to maintain the SVR with prophylactic dopamine infusion titrating to the effect. We avoided nitrous oxide because it is a potent pulmonary vasoconstrictor.[8] Although both halothane and isoflurane have been incriminated to cause systemic hypotension because of varying combination of myocardial depression and vasodilation,[1,3] in these circumstances, it is better to use halothane in low concentration to ensure lack of awareness because the decrease in SVR is less compared with that of isoflurane.[1] After the extraction of the baby, we chose to administer oxytocin as slow infusion. Cole and colleagues[7] stated that the policy of using uterine massage followed by a slow oxytocin infusion proved safe and effective in these patients. Oxytocin as a bolus causes direct vasodilation and reduces SVR with compensatory increase in the heart rate and cardiac output.[7,10]

Because of the risk of pulmonary thromboembolism and paradoxical systemic embolisation, heparin was given in a low dose.[14] Hypoxia unresponsive to O_2 therapy, which was observed on the third post-operative day, probably could be due to the development of multiple pulmonary thrombosis or emboli. This could have lead to an increase in the PVR with worsening of shunt. Although ventilation-perfusion scan was not done, refractory hypoxia could point out toward a thromboembolic phenomenon in our patient, which probably could not be prevented despite anticoagulation.

CONCLUSION

The experience presented in this paper suggests that although pregnancy must be discouraged in women with Eisenmenger's syndrome, it can be successful. Safe anaesthetic management of these patients requires meticulous preparation and familiarity with all the anaesthetic agents to maintain the cardiovascular stability. Then, early extubation should be avoided in such patients because, invariably, they may go for worsening of shunt and

thromboembolic phenomena as these complications can occur as late as the third post-operative day, as seen in our patient and other reports. Thus, we recommend a general anaesthetic technique with maintenance of haemodynamics as close to normal as possible, with adequate control of pain and early initiation of thromboprophylaxis for successful management of similar cases.

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