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Anesthetic management of a pregnant patient with Eisenmenger's syndrome: A case report

Anesthetic management in pregnancy with ES

Abstract

BACKGROUND

Eisenmenger's syndrome (ES) is a rare complication of congenital heart disease that includes pulmonary artery hypertension and reversed or bidirectional shunts. The mortality rate of pregnant women with ES is 30%-70% due to pathophysiological deterioration. Successful perioperative management of a pregnant patient with ES is a challenge for anesthesiologists.

CASE SUMMARY

A 38-year-old pregnant woman was admitted to the cardiology department of our hospital at 22 wk of gestation with complaints of chest tightness and shortness of breath for 3 wk. Transthoracic echocardiography revealed a bidirectional shunt between the descending aorta and pulmonary artery after interventional closure of the patent ductus arteriosus and severe pulmonary hypertension. ES in pregnancy was our primary suspicion. The patient elected to terminate the pregnancy under adequate preoperative preparation, rigorous intraoperative monitoring, and perfect epidural anesthesia. She was discharged successfully on postoperative day 16.

CONCLUSION

Our experience in this case suggests that successful outcomes are possible in pregnant patients with ES for termination of pregnancy under epidural anesthesia and intensive monitoring.

Key Words: Eisenmenger's syndrome; Pulmonary artery hypertension; Pregnancy; Monitoring; Epidural anesthesia; Case report

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Core Tip: Eisenmenger's syndrome (ES) is a rare complication of congenital heart disease, with a high maternal mortality of 30%-70%. We present a rare case of a pregnant woman with ES after cardiac surgery. The patient elected to terminate the pregnancy under adequate preoperative preparation, rigorous intraoperative monitoring, and perfect epidural anesthesia. Our experience in this case suggests that successful outcomes are possible in pregnant patients with ES for termination of pregnancy under epidural anesthesia and intensive monitoring.

INTRODUCTION

Eisenmenger's syndrome (ES) is a rare complication of congenital heart disease during pregnancy[1]. It manifests as pulmonary artery hypertension and a right-to-left or bidirectional shunt secondary to septal defects or patent ductus arteriosus (PDA)[2]. As the mortality rate is as high as 30%-70%, pregnant women with ES are advised to terminate their pregnancy[3,4]. In recent years, advanced medical technology has allowed congenital heart disease patients to undergo corrective surgery, and some patients have decided to conceive. Although there have been a few successful births, it is still recommended to avoid pregnancy after cardiac correction due to the high mortality and maternal complication rates[5]. This case report describes a pregnant patient with ES after

cardiac surgery and discusses the safety and efficacy of epidural anesthesia and intensive monitoring for termination of pregnancy.

CASE PRESENTATION

Chief complaints

A 38-year-old 62-kg pregnant woman (gravida 4, para 0) was admitted to the cardiology department of our hospital at 22 wk of gestation with complaints of chest tightness and shortness of breath for 3 wk.

History of present illness

The patient's symptoms started 3 wk prior with chest tightness and shortness of breath. The degree of heart failure was New York Heart Association grade II.

History of past illness

At 25 years of age, the patient had a history of transcatheter closure of a PDA in a local hospital. The patient stated that the PDA was completely closed by the transcatheter interventional procedure, and she attended irregular follow-up in the local hospital after the operation.

Personal and family history

Between 2010 and 2020, the patient experienced three spontaneous abortions in the first trimester without any pregnancy discomfort. No similar family history was noted.

Physical examination

After admission to our hospital, the patient's temperature was 36.2°C, heart rate was 75 bpm, respiratory rate was 19 breaths per minute, blood pressure was 92/53 mmHg, and oxygen saturation in an oxygen concentration of 33% was 97%. The clinical examination revealed mild cyanosis of the lips and a loud P2 systolic murmur in the second to fourth intercostal spaces of the left margin of the sternum.

Laboratory examinations

Blood analysis revealed a mildly elevated hemoglobin level of 15.3 g/dL, with a normal hematocrit of 44.4% and a platelet count of 1.22×10^{11}/L. The prothrombin time was normal, and the partial thromboplastin time was slightly decreased, at 27.6 s. Electrocardiography (ECG) showed an abnormal Q wave in the high lateral border, slight ST-segment change in the lower border, right axis deviation, and clockwise transposition. Arterial blood gas analysis revealed pH: 7.47, PCO_2: 28.2 mmHg, PO_2: 85.3 mmHg, and SaO_2: 97.4% at 21% oxygen concentration.

Imaging examinations

Transthoracic echocardiography (TTE) revealed the occlusion (manifesting as a stronger echo) between the descending aorta and pulmonary artery (Figure 1A). Color Doppler echocardiography revealed a bidirectional shunt signal on both sides of the occlusion (Figure 1B). Spectral Doppler echocardiography revealed the pressure gradient (PG) and blood velocity of the bidirectional shunt (Figure 1C). The pulmonary artery systolic pressure (PASP) was estimated at 117 mmHg, and the brachial systolic pressure at 124 mmHg.

FINAL DIAGNOSIS

The final diagnosis of this case was pregnancy complicated with ES.

TREATMENT

On day 2 after her admission, the patient underwent surgery to terminate the pregnancy. In the operating room, the patient was supplemented with oxygen *via* a nasal tube. Ringer's lactate (500 mL) was slowly administered during the procedure. Standard monitoring, including SpO_2, noninvasive blood pressure, and ECG, was established. Under ultrasonographic guidance and local anesthesia, an arterial line, central venous catheter, and Swan-Ganz catheter were placed for pressure monitoring and venous

access. In the right lateral position, an 18-G epidural needle was inserted in the L1-L2 intervertebral space, and a 22-G epidural catheter was introduced 4 cm toward the cephalad direction in the epidural space. A 3 mL test dose of 2% lignocaine was administered, and the patient was placed in the supine position. Over a period of 15 min, a total of 9 mL of 0.75% ropivacaine was added gradually to achieve the anesthetic effect. After anesthesia, norepinephrine was given to achieve a systemic blood pressure equal to the pulmonary pressure. Over a period of 15 min, a block height of T4 was obtained in the patient, as assessed using cold stimulation and touch. There were no significant changes in her hemodynamics. The central venous pressure (CVP) was kept at 10-12 mmHg throughout the surgery. The pulmonary artery pressure was always kept lower than the systemic circulation pressure to avoid right-to-left shunt aggravation. The systemic artery pressure was kept at 110-130/60-70 mmHg, the pulmonary pressure at 100-110/50-55 mmHg, and SpO_2 at 98%-100% during the procedure. The procedure proceeded uneventfully, without pain or discomfort. The blood loss was estimated to be 300 mL. At the end of the operation, 1.5 mg morphine was administered epidurally for postoperative analgesia. Since we planned postoperative heparin anticoagulation, the epidural catheter was removed before the patient left the operating room to avoid epidural hematoma and other complications. After surgery, the patient was transferred to the intensive care unit for further monitoring. She was treated with alprostadil for pulmonary hypertension and heparin anticoagulation.

OUTCOME AND FOLLOW-UP

The patient had stable hemodynamics and was transferred to the ward 6 d after surgery. The patient was discharged on the 16th day after surgery. At the follow-up visit one month after hospital discharge, the patient was asymptomatic, and a new TTE showed moderate pulmonary hypertension with a residual bidirectional shunt, which was mainly from left to right (Figure 2). The patient was advised to avoid pregnancy and to attend regular follow-up for her cardiac disease.

DISCUSSION

In the progression of congenital heart disease, the pulmonary artery pressure exceeds the systemic circulation pressure due to the gradual increase in pulmonary vascular resistance (PVR), resulting in a bidirectional or reverse shunt, which is defined as ES. Pregnant women with ES have a mortality rate as high as 30%-70%[3]. The main cause of death in such patients is pregnancy-induced decreased systemic resistance and increased cardiac afterload that exacerbates the right-to-left shunting, leading to pulmonary hypertension, right heart failure, and hypoxemia[6]. Therefore, experts unanimously recommend contraception for women with ES and, once pregnant, early termination of pregnancy.

Guidelines for the management of cardiovascular disease in pregnancy indicate that women with uncomplicated congenital heart disease have no risk or a slightly increased risk of maternal mortality after pregnancy. However, women with congenital heart disease combined with ES have a high mortality due to unbearable hemodynamic changes[7]. The factors related to maternal mortality are severe pulmonary hypertension, ES, and reduced right ventricular systolic function. A previous study showed significantly higher mortality in patients with severe pulmonary hypertension and ES[8]. Therefore, patients with congenital heart disease combined with ES are advised to avoid pregnancy and terminate any pregnancy quickly. In this case, the patient had a history of transcatheter closure of a PDA in a local hospital. She was advised to attend regular follow-up to determine her eligibility for pregnancy. At the last follow-up, she was not diagnosed with pulmonary hypertension or ES. Until the symptoms of chest tightness, she did not undergo cardiac echocardiography, and she did not have any uncomfortable symptoms during early pregnancy. Considering the high mortality rate of continued pregnancy, timely termination of pregnancy may be the best choice for pregnant women with ES.

The primary anesthetic goal in a patient with ES is to maintain cardiac output and systemic vascular resistance (SVR) and avoid any hemodynamic changes that might increase PVR and exacerbate the right-to-left shunt[9,10]. General anesthesia has clear risks

and disadvantages. First, catecholamine release may increase the PVR during laryngoscopy and intubation[11]. Second, positive-pressure ventilation during general anesthesia reduces venous return and cardiac output. Third, general anesthesia increases the risk of reflux aspiration in pregnant women. In addition, induction drugs reduce myocardial contractility and SVR. Epidural anesthesia has been successfully used in pregnant patients with ES[11,12]. Epidural anesthesia reduces catecholamine levels and PVR by sympathetic blockade. Epidural anesthesia is preferable to spinal anesthesia because of its slow onset, which avoids abrupt changes in hemodynamics that increase the right-to-left shunting. Furthermore, the epidural catheter provides assurance for further anesthesia and postoperative analgesia. We consider epidural anesthesia while keeping the hemodynamics as close to normal as possible to be the best choice for these patients.

Rapid hemodynamic changes in pregnant women with ES can lead to a pulmonary hypertension crisis, right heart failure, hypoxemia, and even sudden death. The purpose of monitoring is to detect these changes early and provide timely intervention and treatment. However, the placement of invasive monitoring equipment increases the risk of infection and thromboembolism. Considering the risks and benefits, we placed an arterial catheter, central venous catheter, and pulmonary arterial catheter in this patient. Invasive blood pressure monitoring facilitates real-time supervision of hemodynamic changes. CVP monitoring helps with fluid guidance and medication use. We found that the right-to-left shunt was minimized when the CVP was maintained with an appropriate preload (CVP 10−12 mmHg). The pulmonary artery pressure was monitored to ensure a systemic resistance equal to the pulmonary artery pressure and to minimize the right-to-left shunt. The patient had severe pulmonary artery hypertension before surgery (PASP 117 mmHg). Keeping her SVR higher than her PVR was the key to reducing the right-to-left shunt. Transesophageal echocardiography is uncomfortable for awake patients. TTE is widely used for the diagnosis of ES because it is noninvasive and readily available. Intraoperative echocardiography aids in assessing shunts, pulmonary artery pressure, and right ventricular function[13]. TTE showed that the PASP decreased from 117 mmHg

preoperatively (R-L: V: 2.02 m/s; PG: 16 mmHg) to 78 mmHg postoperatively (R-L: V: 1.18 m/s; PG: 6 mmHg). Therefore, early termination of pregnancy is a correct and effective decision for patients with ES.

Epidural anesthesia reduces the SVR by blocking the sympathetic nerves. Norepinephrine decreases the SVR during anesthesia because β-agonism increases myocardial contractility and cardiac output[14]. In this case, epidural anesthesia combined with norepinephrine successfully maintained the hemodynamic stability as close to normal as possible.

We chose not to administer oxytocin because it causes direct vasodilation and reduces the SVR, with a compensatory increase in the heart rate and cardiac output. Considering that the patient's preoperative coagulation function and platelets were normal, an epidural catheter was placed during the operation. Therefore, heparin was used for anticoagulation after surgery.

CONCLUSION

Here, we present a rare case of a pregnant woman with ES after cardiac surgery. ES is a rare complication of congenital heart disease with high maternal mortality. Our experience in this case suggests that successful outcomes are possible in pregnant patients with ES under epidural anesthesia and intensive monitoring.

ACKNOWLEDGEMENTS

The authors thank Ji-Hong Zhu for assistance with figure preparation.

Figure Legends

Figure 1 Preoperative echocardiography. A: Image showing the occlusion (manifesting as a stronger echo) between the descending aorta and pulmonary artery (yellow arrow); B: Color Doppler echocardiography revealed a bidirectional shunt signal on both sides of the occlusion (yellow arrow); C: Spectral Doppler echocardiography revealed the

pressure gradient and blood velocity of the bidirectional shunt (yellow arrow). Vel: Velocity; PG: Pressure gradient.

Figure 2 Follow-up echocardiography one month after hospital discharge. A: Color Doppler echocardiography revealed only a minor bidirectional shunt signal on both sides of the occlusion (yellow arrow); B: Spectral Doppler echocardiography revealed a residual bidirectional shunt, which mainly was from left to right (yellow arrow).

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