Case Report

Anesthetic management of a parturient with mirror syndrome: a case report

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Abstract: Mirror syndrome is a rare clinical entity consisting of fetal and placental hydrops with maternal edema. It is associated with an increase in fetal mortality and maternal morbidity. We describe the anesthetic management of a parturient with Mirror syndrome complicated by HELLP syndrome and massive postpartum hemorrhage, who required general anesthesia for cesarean delivery.

Keywords: Anesthesia, mirror syndrome

Introduction

Mirror syndrome is a rare obstetric entity that occurs in pregnant women and is secondary to fetal and placental hydrops. The name was derived from the maternal signs and symptoms that “mirror” those of the hydropic fetus and placenta [1, 2]. Patients often also have hemodilutional anemia, hypertension, hypoproteinemia and pulmonary edema [1]. Mirror syndrome is not yet well recognized in clinical practice because of its rare incidence. The imperative treatment for mirror syndrome is to terminate pregnancy immediately by cesarean section or induced labor if the cause of fetal hydrops cannot be treated [3, 4]. However, the anesthetic management of maternal mirror syndrome has been rarely reported. The considerations and precautions of anesthetic management were discussed in the current report. In the report, a patient with mirror syndrome who was treated with cesarean section under anesthesia and followed by a postpartum hemorrhage 5 hours postoperatively was successfully treated by secondary surgery under general anesthesia.

Case report

A 28-year-old woman gravida 2 para 0 (G2P0) with severe bilateral lower extremity edema and severe edema of vulva for 1 week was admitted to our hospital at 31 weeks and four days’ gestation. The patient’s body weight was 54.5 kg and height was 158 cm. She had no significant medical history and was healthy before the pregnancy. The baby was conceived naturally and the patient underwent regular prenatal examinations. Ultrasonography performed in the second semester revealed a singleton with normal fetal umbilical blood flow but thickening of the right ventricular myocardium. Ultrasonography at 31 weeks’ gestation indicated moderate fetal growth restriction (9th percentile) with oligohydramnios, fetal pleural effusion, and fetal hydrops. Magnetic resonance imaging of the fetus indicated subcutaneous edema of the entire body, massive abdominal effusion and pericardial effusion.

The placenta was significantly thickened without signs of placental abruption or implantation. Physical examination showed that the patient was of clear consciousness and a fair general condition. Physical examination revealed normal blood pressure (110/60 mmHg) with a heart rate of 80 beats/min and respiratory rate of 15 breaths/min. Chest auscultation revealed clear breath sounds without rales. Bilateral lower extremity pitting edema was observed. Maternal laboratory tests showed: hemoglobin, 90.0 g/L; hematocrit, 29.6%;
Anesthesia of mirror syndrome

platelets, $124.4 \times 10^9$/L; aspartate aminotransferase (AST), 76 U/L; alanine aminotransferase (ALT), 71 U/L; albumin, 25 g/L; uric acid, 806 µmol/L; total bile acid, 42 µmol/L; urea nitrogen, 6.7 mmol/L; and creatinine, 72 µmol/L. The diagnosis of mirror syndrome was made.

On the third day after admission, the patient presented increased dyspnea with a respiratory rate of 22 breaths/min, tachycardia and abdominal pain. The chest X-ray findings were suggestive of pulmonary congestion. Laboratory tests showed the following: hemoglobin, 80 g/L; hematocrit, 26%; platelets, $90 \times 10^9$/L; ALT, 110 U/L; AST, 124 U/L; and 24-h urinary protein, 1.7 g/L. The features of HELLP (Hemolysis, Elevated Liver enzymes, and Low Platelet counts) syndrome were noted, and the decision was made to perform an emergency cesarean section to terminate pregnancy.

Anesthetic management

The patient was transferred to the operating room and positioned supine with left uterine displacement and 30°C head elevation. A pre-anesthetic evaluation showed a class II Mallampati airway with mild oral mucosa swelling. Auscultation revealed diffuse crepitation at the base of both lungs. Standard monitoring was attached (electrocardiography, pulse oximetry, and non-invasive blood pressure) and a right cervical cannula was inserted to measure the central venous pressure (CVP). A non-invasive cardiac output (NICOM) system (Cheetah Medical, Wilmington, DE, USA) was also used to measure the peripheral vascular resistance. Respiratory frequency was 20 breath/minute, and pulse oxygen saturation ($\text{SpO}_2$) was 95% on room air. Blood pressure was 123/75 mmHg with a heart rate of 116 beats/min. CVP was 9 cmH$_2$O, NICOM monitoring indicated cardiac output (CO) of 6 L/min, total peripheral vascular resistance (TPR) of 1030 dynes-sec/cm$^5$ (normal range, 800-1200 dynes-sec/cm$^5$) and a total peripheral vascular resistance index (TPRI) of 1589 dynes-sec/cm$^5$/m$^2$ (normal range, 1970-2390 dynes-sec/cm$^5$/m$^2$).

Ringers lactate solution was administered at a rate of 200 mL/h. Cimetidine was injected at dose of 250 mg once the patient was transferred to the operating room. The patient was pre-oxygenated for 5 minutes, after which 60 µg of remifentanil was injected over 30 seconds. Rapid induction was then performed with an injection of propofol 110 mg and succinylcholine 100 mg, and cricoid pressure was applied. Endotracheal intubation with Glide scope video laryngoscope (Saturn Biomedical Systems Inc., Burnaby, BC, Canada) was easily achieved using a 7.0-mm cuffed tube. Anesthesia was maintained with an infusion of propofol at a rate of 300 mg/h with the addition of sufentanil after the umbilical cord was clamped. The lungs were ventilated with positive end-expiratory pressure (PEEP) of 5 cmH$_2$O.

Obstetric surgery

After verification of the correct tube position by capnography, the obstetricians started the operation. Three minutes after the incision, the fetus was delivered. The fetus had severe total body edema and the accumulation of a large amount of exudate in the abdominal cavity (Figure 1). The placenta and umbilical cord were also severe edema. The fetus was immediately intubated with Apgar scores of 2 and 3 at 1 and 5 minutes, respectively. Resuscitation was initiated by the neonatal team. Abdominal paracentesis was performed and approximately 40 mL of yellow liquid was drawn out. Resuscitation attempts ultimately failed.

The patient received oxytocin 20 IU by intravenous infusion followed by intravenous carbetocin 100 µg after umbilical cord clamping. However, the uterus did not sufficiently con-
Anesthesia of mirror syndrome

tract despite carboprost tromethamine administration. To prevent atonic postpartum hemorrhage, bilateral ascending uterine artery ligation was performed and B-lynch sutures were used. During surgery, the patient was hemodynamically stable. Estimated blood loss was 1200 mL and two units of packed red blood cells (PRBC) were transfused. Albumin (10 g) and furosemide (20 mg) were administered. A total of 500 mL lactated Ringers solution was given during the entire procedure. The patient was awakened and extubated once fully awake. The early postoperative period in the postanesthesia care unit was uneventful. The patient was then transferred to the ward 1 hour later.

At 5 hours postoperative, the patient presented with tachycardia and hypotension, and the wound drainage increased to 500 mL without vaginal bleeding. Laboratory values demonstrated hemoglobin of 55 g/L and hematocrit of 20%. Intraperitoneal hemorrhage may have occurred. Accordingly, the patient was transferred to the operating room immediately for emergency surgery to locate the source of the bleeding. Her blood pressure was 70/50 mmHg and she had tachycardia of 130 beats/min. The invasive arterial blood pressure was monitored by radial artery cannulation. A phenylephrine infusion was started to maintain her blood pressure and three units of PRBC was transfused. Urgent surgical exploration was performed under general anesthesia with endotracheal intubation, and the right ovary showed marked edema with ovarian artery bleeding. The bleeding was controlled after ligation of the right ovary. Approximately 3000 mL of blood was suctioned from the peritoneal cavity. A total of 11 units of PRBC, eight units of fresh frozen plasma, and two units of platelets were transfused. After the surgery, the patient was transferred to the intensive care unit and extubated 1 hour later. The edema gradually vanished 3 days after operation and the patient was discharged 6 days later uneventfully.

Discussion

As an uncommon obstetric medical entity, mirror syndrome is not well recognized and clinicians still lack experience, especially regarding anesthetic management. Mirror syndrome shares many features with preeclampsia, including hypertension, edema and proteinuria, which makes it difficult to distinguish between the two syndromes [1, 4]. Mirror syndrome and preeclampsia may occur simultaneously [5]. Hemodilution has been a distinct pathophysiological feature of mirror syndrome compared to hemoconcentration in preeclampsia [6, 7]. Other important clinical features of mirror syndrome are progressively elevated uric acid, pruritus and dyspnea [1]. In the current case, the patient also had a significantly decreased platelet count, hypoproteinemia and slightly elevated ALT and AST, and she might have had HELLP syndrome. Her blood circulation had been monitored by NICOM, which revealed normal measurements of CO, TPR, and TPRI, suggesting that peripheral vascular resistance was not elevated, while preeclampsia is usually associated with increased peripheral vascular resistance. Our finding indicated that TPR and TPRI could be used as parameters to differentiate between the two syndromes.

Mirror syndrome threatens both mother and fetus. Braun et al [1] reported that pulmonary edema occurred in 21.4% of 56 patients with mirror syndrome and the average rate of intrauterine death and stillbirth was 35.7%. Delivery of the fetus remains the only way to reverse maternal complications if attempts to resolve the hydrops fail [8].

Studies on the anesthetic management of patients with mirror syndrome are rare. McCann et al [7] reported the successful use of epidural analgesia in a patient with mirror syndrome at 21 weeks’ gestation. Zlotnik et al [6] reported a case of mirror syndrome at 25 weeks’ gestation in which the pregnancy was terminated by the induction of labor. At the third stage of delivery, the patient required manual revision under anesthesia of the undelivered placenta that was retained by the uterus. Since she had no epidural catheter in situ and her case was complicated by HELLP syndrome and a difficult airway, awake fiberoptic intubation with subsequent general anesthesia was performed. Tayler et al [2] recently reported a cesarean section under epidural anesthesia for a patient with mirror syndrome at 34 weeks’ gestation. The anesthetic management was relatively simple because the patient had edema only and no HELLP syndrome, thrombocytopenia, anemia, pulmonary edema or dyspnea.
Anesthesia of mirror syndrome

The patient in the current case presented with severe overall edema that might complicate epidural placement. Epidural hematoma formation could be induced as a result of the progressively decreasing platelets. Also, ventilation support might be needed due to the pulmonary edema. Taking all of the above concerns into consideration, epidural anesthesia was not the optimal anesthetic procedure for this patient. Therefore, general anesthesia with endotracheal intubation was chosen. However, the airway edema was suspected in the patient due to the edema of oral mucosa. As such, a smaller tracheal tube was utilized and preparation for difficult airway management was made accordingly. Fortunately, the endotracheal incubation was accomplished smoothly using a video laryngoscope. Volume control in combination with 5-cm H₂O PEEP ventilation was used to improve pulmonary function.

Fluid administration was restricted because there was evidence of lung edema on the chest roentgenogram as well as hemodilutional anemia. Albumin and furosemide were used to improve serum albumin level and reduce volume overload. Volume status was evaluated by central venous pressure and stroke volume variation was provided by NICOM.

Uterine atony was found in the patient after labor, and blood loss was estimated to be approximately 1200 mL. Bilateral uterine artery ligation in combination with Blynch suturing was performed, and various medicines were administered to reinforce uterine contraction and prevent hemorrhaging at that time. However, an acute hemorrhage occurred 5 hours after surgery, and a secondary surgical exploration indicated active bleeding at one ovary. It remains to be further elucidated whether the latter complication of hemorrhage is connected with mirror syndrome; however, it is worthwhile for clinicians to be cautious that postpartum hemorrhage might occur in a patient with mirror syndrome [9].

Mirror syndrome is associated with an increase in perinatal mortality [9, 10]. The pediatricians in this case were well prepared for neonatal resuscitation and the emergent medical treatments were carried out, including tracheal intubation, umbilical vein catheter placement and exudation extraction from the thoracic and abdominal cavities. However, the resuscitation attempts failed due to the neonate’s poor physiological condition.

In summary, here general anesthesia was administered for a patient with mirror syndrome conducting cesarean section and the peripheral vascular resistance was measured by NICOM to differentiate it from preeclampsia. These features have not been reported before. Our findings suggest that after careful planning is made for possible complications such as difficult airway, pulmonary edema and hemorrhage, general anesthesia may be safely administered to a patient with mirror syndrome.

Disclosure of conflict of interest

None.

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Anesthesia of mirror syndrome

