Type A aortic dissection during late pregnancy in a patient with Marfan syndrome

Dissection aortique de type A en fin de grossesse chez une patiente atteinte du syndrome de Marfan

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Abstract

Purpose To report the management of a 38-yr-old patient with known Marfan syndrome who presented with acute Stanford type A dissection of the aorta in the 34th week of pregnancy.

Clinical features A Cesarean delivery was performed under deep general anesthesia with high-dose opioid administration to avoid tachycardia and hypertension during tracheal intubation and obstetric surgery. Delivery took place less than five minutes after induction of anesthesia, and tracheal intubation of the newborn was required due to opioid-induced hypoventilation. Subsequently, aortic arch repair was performed in the mother after connection to extracorporeal bypass. Despite extensive replacement of coagulation factors, severe vaginal bleeding persisted after weaning from extracorporeal bypass, and the bleeding stopped only after a hysterectomy was performed. Postoperatively, after a short period in the

intensive care unit, mother and child were discharged from hospital in excellent condition ten days after surgery.

Conclusion Deep general anesthesia for emergency Cesarean delivery while accepting the risk of respiratory depression in the newborn is a viable option for the anesthetic management of life-threatening events such as Stanford type A dissection.

Résumé

Objectif Décrire la prise en charge d'une patiente de 38 ans atteinte d'un syndrome de Marfan connu qui s'est présentée avec une dissection aiguë de l'aorte de type A selon la classification de Stanford en 34^e semaine de grossesse.

Éléments cliniques Un accouchement par césarienne a été réalisé sous anesthésie générale profonde avec une administration d'opioïdes en doses élevées pour éviter la tachycardie et l'hypertension pendant l'intubation trachéale et la chirurgie obstétricale. L'accouchement a eu lieu moins de cinq minutes après l'induction de l'anesthésie, et il a fallu procéder à une intubation trachéale du nouveau-né en raison d'une hypoventilation induite par les opioïdes. Par la suite, une réparation de la crosse de l'aorte a été réalisée chez la mère après avoir mis en place une circulation extracorporelle. Malgré le remplacement considérable de facteurs de coagulation, des saignements vaginaux graves ont persisté après le sevrage de la circulation extracorporelle, et ces saignements n'ont cessé qu'après une hystérectomie. En période postopératoire, après un court séjour dans l'unité des soins intensifs, la mère et l'enfant ont reçu leur congé de l'hôpital en excellente santé dix jours après la chirurgie.

Conclusion Une anesthésie générale profonde pour un accouchement par césarienne, en tenant compte du risque de dépression respiratoire chez le nouveau-né, est une

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option viable pour la prise en charge anesthésique d'événements potentiellement fatals tels qu'une dissection de type A selon la classification de Stanford.

Stanford type A dissection of the aorta is a life-threatening disease. Without treatment, 50% of these patients die within 48 hr. Causes of death are mainly intrapericardial rupture of the aorta followed by pericardial tamponade and acute severe aortic valve insufficiency.¹ Marfan syndrome is a hereditary autosomal-dominant disease which primarily affects connective tissue, skeletal muscle, the cardiovascular system, and the eyes. Patients with Marfan syndrome also suffer from cardiovascular pathologies, and an aneurysm of the ascending aorta can be found with concomitant aortic valve insufficiency.^{2,3} The risk of developing a dissection of the aorta increases in these patients during pregnancy and reaches a maximum in the third trimester. The 30-day mortality for elective surgical repair of ascending aortic aneurysms in Marfan patients is reported to be only 0–1.5%.^{4,5} Therefore, in patients with Marfan syndrome planning to have children, elective surgical repair of an ascending aortic aneurysm must be seen as a first-line strategy.^{6,7} Herein, we report a case of a 38-yr-old woman with Marfan syndrome with a known aneurysm of the ascending aorta who developed a type A dissection in the 34th week of gestation. In accordance with our institutional guidelines, the patient gave written informed consent for the publication of this report.

Case description

A 38-yr-old female with Marfan syndrome and a known 51-mm ascending fusiform aneurysm of the aorta presented for a periodic echocardiographic follow-up in her 34th week of gestation. After the initial diagnosis of this fusiform aneurysm, the patient refused the recommended elective aortic repair before pregnancy. When she presented for her exam at 34 weeks, the echocardiogram was unchanged compared with previous examinations (Fig. 1). Shortly after this echocardiographic study, the patient experienced a sudden tearing pain in the chest, and the echocardiogram was immediately repeated only 90 min later. The results of this second test strongly suggested acute aortic dissection with evidence of an intima flap (Fig. 2). This suspicion was verified by thoracic computed tomography confirming a Stanford type A dissection of the aorta, which was characterized by evidence of a dissection membrane extending from the aortic bulb to the left subclavian artery.

Fig. 1 Transthoracic echocardiogram of the left ventricle (LV) and ascending aorta (Ao)

Fig. 2 Transthoracic echocardiogram of the left ventricle (LV) and ascending aorta (Ao) showing intima flap (arrow)

Immediately after confirmation of this diagnosis and through close interdisciplinary collaboration with obstetricians, neonatologists, cardiac surgeons and anesthesiologists, the decision was made to deliver the baby via Cesarean delivery under general anesthesia in the cardiac operating room. The anesthesiologists informed the combined team of their plan to administer a high opioid dose for general anesthesia, and they advised the neonatologists to be prepared for a newborn with respiratory depression once the fetus was delivered. Furthermore, the cardiac surgeons were instructed to be prepared to connect the patient immediately to extra-corporal bypass in case of aortic rupture.

In the operating room, the patient was positioned supine with left lateral uterine displacement to avoid aortocaval compression. Baseline monitoring (electrocardiography, pulse oximetry, and noninvasive blood pressure) was established. In the meantime, the obstetricians monitored

the fetal heart rate by intermittent ultrasound examination using a portable device. The patient was sedated lightly with an injection of midazolam 3 mg *iv*, and the right radial artery was cannulated for invasive blood pressure measurement after local infiltration of 1% lidocaine 0.5 mL. Additionally, a large-gauge peripheral venous line was installed and connected to a blood warmer with compression chambers for immediate high-flow volume and blood administration in the case of acute severe bleeding. At this time, the patient's heart rate was 95 beats·min⁻¹, her blood pressure was low at 85/45 mmHg, and she was on no medication affecting blood pressure.

After hemodynamic monitoring was established and all preparations for induction of general anesthesia were completed, the patient's chest and abdomen were prepped and draped. Induction of general anesthesia was then performed by means of modified "rapid sequence induction". After two minutes of preoxygenation with 100% oxygen, sufentanil 60 µg, propofol 200 mg, and rocuronium 100 mg were injected, cricoid pressure was performed, and tracheal intubation was accomplished without any difficulty. General anesthesia was maintained using sevoflurane (2.0% end-tidal in 100% oxygen) and sufentanil 0.7 µg·kg⁻¹·min⁻¹ by continuous infusion. After induction of anesthesia, mean arterial pressure decreased progressively to values near 45 mmHg, and her heart rate decreased to 80 beats·min⁻¹. Tracheal intubation did not increase her blood pressure or heart rate. Norepinephrine (initially at 0.03 µg·kg⁻¹·min⁻¹ and later increased to a maximum of 0.1 µg·kg⁻¹·min⁻¹) was administered to prevent a further decrease in blood pressure. In this situation, a minimum mean arterial pressure of 45 mmHg was tolerated, and this could be achieved by norepinephrine administration.

In this situation of hypotension, the obstetricians performed an expeditious delivery of the baby. During the Cesarean delivery, a central venous catheter and a 7.5 F introducer sheath were placed in the right jugular vein for central venous pressure monitoring and administration of drugs and fluids. Time from tracheal intubation of the mother until delivery was less than five minutes. During this interval, her heart rate was 80–85 beats·min⁻¹, and a mean arterial pressure varied from 45 to 55 mmHg. Oxygen saturation was 99% at all times.

After delivery, the newborn presented with an Apgar score of 3, and tracheal intubation was performed approximately two minutes later after external stimulation of the newborn failed to trigger sufficient spontaneous breathing. The newborn was transferred to the neonatal intensive care unit and mechanically ventilated under light morphine and midazolam sedation. No further pharmacological interventions were required, especially no administration of catecholamines. Sedation was discontinued six hours later,

and tracheal extubation was possible 12 hr after delivery without complications. Following delivery of the fetus, the obstetricians excised a broadly pedunculated myoma of the posterior wall of the uterus which was yet to be diagnosed. The myoma had a diameter of about 8 cm and an irregular surface and consistency.

After extirpation of the myoma, maternal aortic repair was started. After 25,000 units of heparin were administered to the mother, extracorporeal bypass was established by cannulation of the subclavian artery for arterial pump flow and cannulation of the right atrium for venous drainage. Her aorta was clamped proximal to the brachiocephalic trunk, and her heart was arrested using blood cardioplegia. The cardiac surgeons performed an aortic valve-preserving replacement of the ascending aorta with reimplantation of the coronary arteries under moderate hypothermia (28°C). By clamping the proximal brachiocephalic trunk and the left internal carotid artery, circulatory arrest was achieved, and selective cerebral perfusion via the right subclavian artery was performed for 22 min for aortic arch reconstruction. The period of time on cardiopulmonary bypass was 372 minutes, and 55,000 units of heparin were administered. After rewarming the patient, weaning her from extracorporeal bypass was uneventful with stable hemodynamics.

Vaginal bleeding occurred after the extracorporeal bypass was terminated, and the bleeding could not be controlled even by administering 35,000 units of protamine and maintaining normothermia (36.6°C). The first activated clotting time was prolonged (141 sec) after protamine administration. The following were transfused to compensate for blood loss and to optimize coagulation: 13 units of packed red blood cells, 2,500 mL of autologous washed blood (cell saver), 14 units of fresh frozen plasma, 3 platelet concentrates, 500 units of antithrombin III, 3,000 units of prothrombin complex concentrate, and 4 g of fibrinogen. Also, an additional intravenous injection of oxytocin, 5 units as a bolus followed by a continuous infusion of 8 units·hr⁻¹, did not stop the vaginal bleeding; therefore, it was essential to perform an open hysterectomy. The hysterectomy stopped the vaginal bleeding, and after almost 14 hr of surgery, the patient was transferred to the cardiothoracic intensive care unit in a hemodynamically stable condition. After 12 hr of postoperative mechanical ventilation, the patient's trachea was extubated without complication.

Discussion

This case shows that intentional acceptance of respiratory depression in the newborn, which is induced by high-dose opioid administration, can be a successful option for

anesthetic management in an emergency situation such as Cesarean delivery in a patient suffering from an acute aortic dissection.

Case reports have been published in the literature describing the anesthetic management of Cesarean delivery complicated by aortic dissection.^{8,9} In these cases, the surgical incision line was infiltrated with lidocaine and a beta blocker was injected prior to induction of anesthesia to avoid tachycardia and hypertension. Subsequently, anesthesia was induced using thiopental and succinylcholine. In both cases, the newborn was delivered with adequate spontaneous breathing.

In our case, we opted for a different management. After dissection of the ascending aorta, the risk for aortic rupture is extremely high. Our strategy was based on the avoidance of tachycardia and hypertension to minimize the risk of aortic rupture. Therefore, we did not use the classical induction medications for Cesarean delivery — thiopental and succinylcholine — as reported in the two previous cases. In the authors' opinion, this approach can still carry a high risk for the development of hypertension and tachycardia even if a beta blocker is administered and there is prior infiltration of the surgical incision line with local anesthetics. We therefore decided to induce general anesthesia and intubation via a modified rapid sequence induction — which is indicated in this emergency situation — with high-dose opioids and propofol. We chose propofol rather than thiopental because the effect of thiopental on the cardiovascular system may be more unpredictable. However, the key point was to administer high doses of opioids to ensure deep anesthesia and analgesia with a minimized risk of tachycardia and hypertension.

We tried to strike a balance between the requirements for managing an aortic dissection and the needs of the fetus. We accepted a mean arterial pressure of 45 mmHg to minimize the risk of aortic rupture and to keep catecholamine doses low. Aggressive blood pressure therapy with high-dose catecholamines can be dangerous due to unexpected and uncontrollable peaks of blood pressure with potential aortic rupture. Furthermore, high doses of catecholamines are expected to reduce fetal oxygen supply by vasoconstriction in the placenta. In particular, bolus administration should be avoided due to unpredictable effects. In our case, the impending decrease in mean arterial pressure below 45 mmHg was treated with a continuous infusion of nor-epinephrine using a syringe pump. However, because hypotension and catecholamines might result in placental hypoperfusion with potential fetal hypoxemia, the fetus must be delivered as quickly as possible.

We intentionally accepted the possibility of opioid-induced respiratory depression in the neonate and thus the requirement for endotracheal intubation and ventilation. This situation must be handled by an experienced team of

neonatologists. Administration of naloxone to the newborn might be an alternative at this point, but we preferred to secure immediate optimal oxygenation by endotracheal intubation. We potentially had to face additional problems in the newborn, such as hemodynamic instability, in which case, at least oxygenation would have been secured by endotracheal intubation.

After delivery, deep general anesthesia was maintained in the mother using sufentanil and sevoflurane. The main challenges after aortic repair and weaning from extracorporeal bypass were severe vaginal bleeding and coagulopathy. Coagulopathy had to be expected after more than six hours of extracorporeal circulation, hypothermia, and continuous blood loss, and its effect was clinically obvious by the absence of blood clots in the surgical field. Usually a state of relative hypercoagulability is present at the end of pregnancy,^{10,11} but in the authors' opinion, this effect played no significant role in this case. Therefore, we treated coagulopathy in the same manner as we would in a non-pregnant patient. Although the effect of heparinization was reversed by protamine and a large quantity of coagulation factors and fresh frozen plasma was substituted, abdominal-vaginal bleeding did not stop and a hysterectomy had to be performed as therapy of last resort. It is possible that excision of the myoma also contributed to the bleeding, but the initial clinical suspicion of dysregulatory circulation in the myoma (irregular surface and consistency) was confirmed by histopathologic examination showing areas of intramyomal hemorrhage. The obstetricians decided to remove the myoma primarily as the minor procedure.

This case report demonstrates that even very complex clinical cases can be handled successfully when close interdisciplinary collaboration is performed. Both mother and child were discharged from hospital in excellent condition ten days after surgery.

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