

Type A Aortic Dissection in a Postpartum Woman with Undiagnosed Marfan Syndrome

Naruenart Lomarat, M.D.

Department of Anesthesiology, Faculty of Medicine Siriraj Hospital, Mahidol University, Bangkok 10700, Thailand.

ABSTRACT

Objective: To describe perioperative management for type A aortic dissection in a postpartum woman with undiagnosed Marfan syndrome.

Case presentation: A 30-year-old G2P1 Thai female at 39 weeks of gestation with undiagnosed Marfan syndrome gave birth at Siriraj Hospital, Bangkok, Thailand. After delivery, she developed episodes of hypotension and right hemiparesis. She was diagnosed with Marfan syndrome by clinical features of severe myopia, aortic root diameter Z-score ≥ 2 and bonafide FBN1 mutation detected by genetic study. The imaging studies showed strong evidences of type A aortic dissection and she underwent uneventful ascending aortic replacement under general anesthesia with extracorporeal bypass. The perioperative management was described.

Conclusion: Hemodynamic instability and pregnancy-related physiologic changes are the key factors to be concerned for perioperative management. The collaboration among cardiac surgeon, cardiac anesthesiologist and perfusionist is of great importance.

Keywords: Aortic dissection; Marfan syndrome; pregnancy (Siriraj Med J 2018;70: 359-362)

INTRODUCTION

Marfan syndrome is an autosomal dominant disorder which primarily affects connective tissue of the body, and clinical manifestations result in the cardiovascular, skeletal, and ocular systems.^{1,2} The diagnosis of this particular syndrome relies on revised Ghent nosology criteria includes aortic root aneurysm, ectopia lentis (dislocated lens), and FBN1 mutation. In the absence of any family history, the presence of the two features is sufficient for unequivocal diagnosis and in case of the absence of one of these two cardinal features, the presence of an FBN1 mutation of positive systemic score is required. Stanford type A aortic dissection is a severely life-threatening condition in Marfan syndrome and is considered definitely for surgical treatment. It has been documented that the mortality is extremely high during the first 48 hours.³ The risk of aortic dissection or dilatation dramatically increases during and after pregnancy in patients with Marfan syndrome due to imposition of the

hyperdynamic and hypervolemic states.² However, there are several disorders that can cause acute aortic dissection in postpartum woman such as familial thoracic aortic aneurysm, Ehlers-Danlos syndrome-vascular type or arterial tortuosity syndrome. The following case profiles the uncommon case of a 30-year-old undiagnosed-Marfan-syndrome woman with Stanford type A aortic dissection in a postpartum period. In accordance with our institutional guidelines, the patient gave written informed consent for the publication of this report.

CASE PRESENTATION

A 30-year-old Thai female, G2P1, in the 39th week of pregnancy had an uneventful vaginal delivery. She developed an episode of hypotension after undergoing tubal sterilization under local anesthesia and she underwent emergently exploratory laparotomy under general anesthesia due to suspicious of intraperitoneal bleeding. Unfortunately, it was negative finding and

Correspondence to: Naruenart Lomarat

E-mail: nong075@gmail.com

Received 1 August 2017 Revised 4 October 2017 Accepted 17 October 2017

doi:10.14456/smj.2018.58

explained causes of hypotension were not detected. Six hours later, she developed motor aphasia and right hemiparesis and her physical examination showed diastolic murmur grade III of VI at right parasternal border. Transthoracic echocardiogram revealed aortic root dilatation (4.3 cm in diameter) with moderate aortic insufficiency and intimal flap in ascending aorta. CT of her brain strongly suggested left middle cerebral artery territory infarction and CTA of thoracic aorta showed Stanford A aortic dissection involving brachiocephalic trunk, along left common carotid artery, proximal left subclavian artery, proximal part of celiac trunk and superior mesenteric artery without evidence of rupture of aortic dissection. She was scheduled for emergently ascending aortic replacement.

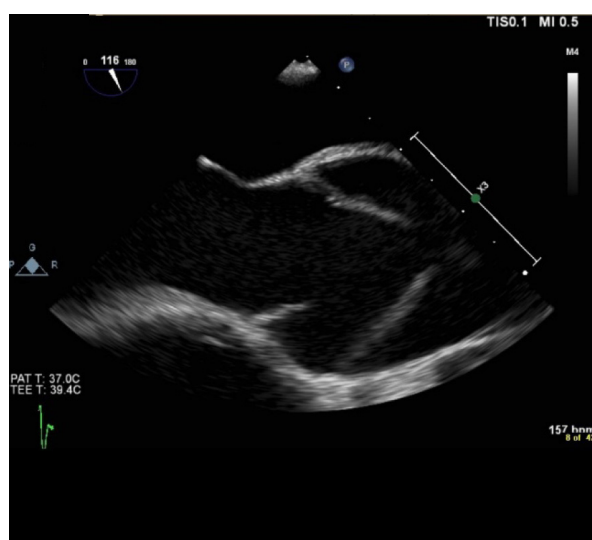


Fig 1. Transesophageal echocardiography showed the intimal flap in the ascending aorta.



Fig 2. Thoracic CTA showed dilated ascending aorta with intimal flap.

In the operating room, the patient was placed supine and standard monitoring (electrocardiography, noninvasive blood pressure, and pulse oximetry) was established. The right radial artery was cannulated for continuous invasive blood pressure monitoring under local infiltration of 2% lidocaine 0.5 mL and the large-bored peripheral lines were placed. After three minutes of denitrogenation with 100% oxygen, midazolam 8 mg, fentanyl 200 µg, and rocuronium 50 mg were injected, tracheal intubation was uneventfully accomplished and general anesthesia was maintained with isoflurane (1% end-tidal in 60% oxygen). After induction of anesthesia, a 7 F double-lumen catheter and a 8 F introducer sheath were placed in the right internal jugular vein for administration of medication and fluids and central venous pressure monitoring. Transesophageal echocardiographic probe was inserted for evaluating severity and the entry site of aortic dissection and cardiac morphology and function were assessed.

The patient's chest, abdomen, and groins were prepped and draped under aseptic fashion, median sternotomy and the right groin incision were performed and 15,000 units of heparin were administered via central venous line. The extracorporeal system was established by cannulation of the right femoral artery for arterial pump flow and cannulation of the right atrium for venous drainage. Distal ascending aorta was cross-clamped, ascending aorta was opened and blood cardioplegia was directly given via both coronary artery ostiums. The cardiac surgeons performed an ascending aortic replacement with Gelweave graft no.24 and aortic valve commissural resuspension under moderate hypothermia (28 °C). During extracorporeal bypass, mean arterial pressure (MAP) averaged 40 mmHg with 3-5 L/min of cardiac output and norepinephrine was continuously administered to maintain MAP over 60 mmHg. The period of time on cardiopulmonary bypass was 121 minutes and aortic cross-clamped time was 87 minutes. After rewarming the patient, discontinuation from extracorporeal bypass was uneventful with stable hemodynamics.

Reversal of heparin effect was by administering 15,000 units of protamine and the first activated clotting time was prolonged (167 seconds) after protamine administration. The patient was transfused 2 units of packed red blood cells, 5 units of fresh frozen plasma, 5 units of platelet concentration and 5 units of cryoprecipitate for optimizing coagulation, the patient was transferred to the cardiothoracic intensive care unit in the hemodynamically stable condition. After 10 hours of postoperative mechanical ventilation, the patient was successfully extubated. Neurological symptom and sign were evaluated and motor power of

the right-side was slightly increased with grade III of V. She was transferred to the general postpartum ward and was treated in rehabilitation program. Genetics medicine was consulted for evaluation and assessment of Marfan syndrome, and direct DNA sequencing of both DNA strands was performed for mutation detection in the entire coding sequences (exon 2-66; which consists of 66 fragments) for FBN1 gene, and FBN1 NM_000138.4:C.6658C>T[p.Arg2220Term] was identified in this patient. She was given β -blocker. She was discharged on the 14th postoperative day.

DISCUSSION

Aortic aneurysm or aortic dissection uncommonly occurs in young women, but it can occur. It is usually associated with pregnancy with the most incidence of aortic dissection in the third trimester caused by hyperdynamic and hypervolemic state of pregnancy and inhibition of elastin and collagen deposition by estrogen and relaxin.⁴ In this case, the onset of aortic dissection was in the early postpartum period. Aggravating factors of aortic dissection in this case possibly were multifactorial such as the effects of pregnancy, pathophysiologic changes of Marfan syndrome, and hemodynamic changes in heart rate, systemic blood pressure, and systemic vascular resistance (vaginal delivery, tubal sterilization, exploratory laparotomy). Based on the revised Ghent nosology criteria, Marfan syndrome can be diagnosed by just cardinal cardiovascular manifestation plus Bonafide FBN1 mutation. Therefore, the patient might have been underdiagnosed due to subclinical physical appearance (myopia, high-arched palate) and the absence of familial history or this was due to unawareness of her clinical features. If this case was diagnosed before or during pregnancy, antenatal care strategy to prevent aortic dissection such as frequent echocardiography monitoring every 3 months and taking β -blocker would be necessary. Vaginal delivery with epidural analgesia can be performed safely and cesarean section also be performed under general anesthesia and regional anesthesia. Marfan syndrome is an autosomal dominant disorder, and her children might be affected, so genetic study should be done during childhood period. β -blocker is a beneficial medication to prevent cardiovascular sequelae from Marfan.

For the anesthetic consideration, the difficult airway can be suspected due to the airway mucosal edema and narrow airway passage (gestation hormonal effects) and Marfan manifestation (high arched palate). Smaller endotracheal tube, stylet, and video laryngoscope should be prepared. This patient was successfully intubated without any complications with McCoy blade laryngoscope

no.4. Hypertension and tachycardia must be avoided, so high dose of opioids, lidocaine and β -blocker can attenuate hemodynamic changes during laryngoscopy and intubation. Ruptured aortic dissection is the lethal complication, so cardiac surgeons, perfusion and extracorporeal circuit should standby in the operating room in case of hemodynamic collapse after induction and intubation. During on cardiopulmonary bypass with moderate hypothermia, adequate organ perfusion should be optimized by adequate pump flow 3-5 L/min maintaining mean arterial pressure 50-60 mmHg. However, this patient's MAP was continuously lower than 50 mmHg (40mmHg) even with full pump flow. The low MAP in this patient might be caused by physiologic anemia and hemodilution (pregnancy physiology) and vasodilatation from progesterone effect on vascular tone. Progesterone might play a role on vascular integrity and permeability promoting decreased systemic vascular resistance and interstitial edema, and progesterone effect might last 6-8 weeks after placental delivery.⁵ Norepinephrine is drug of choice to counter severe vasodilatation phenomenon during extracorporeal bypass, so 0.05-0.1 μ g/kg/min of norepinephrine was continuously infused to maintain MAP 50-60 mmHg in this patient.

Coagulopathy commonly occurs after prolonged extracorporeal circulation, continuous bleeding, residual heparin effect, and hypothermia. A hypercoagulable state is usually present in the third trimester of pregnancy and late postpartum,^{6,7} but there was no significant effect in this case. Therefore, the patient was treated coagulopathy as a non-pregnant patient, adequate heparinization reversal, appropriate quantity of fresh frozen plasma and coagulation factors replacement, for maintaining normothermia and coagulation test are the important keys to correct coagulopathy. Fortunately, there was no massive bleeding from vagina and the previous surgical wound, and coagulation test showed no abnormality.

CONCLUSION

Subclinical features and the absence of familial history are the difficult points for definite diagnosis of Marfan syndrome. Acute aortic dissection Stanford type A in pregnancy is rare and difficulty to diagnose. Transthoracic echocardiography is a first-line useful technique for diagnosis and detection of severity and consequences of aortic dissection and computerized tomography of aorta is definitely a tool to detect origin of dissecting flap and the extension of dissection. Hemodynamic instability, pregnancy-hormonal effect and coagulopathy are the important keys for safe perioperative management during aortic surgery and extracorporeal bypass.

ACKNOWLEDGMENTS

The author gratefully acknowledges this patient for authorizing release of patient case data.

Conflict of interest declaration: The author hereby declares no personal or professional conflicts of interest regarding any aspect of this report.

Funding disclosure: This was an unfunded study.

REFERENCES

1. Keane MG, Pyeritz RE. Medical management of Marfan syndrome. *Circulation*. 2008;117:2802-13.
2. Amnash NM, Sundt TM, Connolly HM. Marfan syndrome: Diagnosis and management. *Curr Probl Cardiol*. 2008;31:7-39.
3. Jayaram A, Carp HM, Davis L, Jacobson SL. Pregnancy complicated by aortic dissection: Caesarean delivery during extradural anesthesia. *Br J Anaesth*. 1995;75:358-60.
4. Williams EJ, Benyon RC, Trim N. Relaxin inhibits effective collagen deposition by cultured hepatic stellate cells and decreases rat liver fibrosis in vivo. *Gut*. 2001;49:577-83.
5. Pomini F, Mercogliano D, Cavalletti C, Caruso A, Pomini P. Cardiopulmonary bypass in pregnancy. *Ann Thorac Surg*. 1996;61:259-68.
6. Anderson JA, Weitz JI. Hypercoagulable states. *Clin Chest Med*. 2010;31:659-73.
7. Arya R. How I manage venous thromboembolism in pregnancy. *Br J Haematol*. 2011;153:698-708.