

Type A Aortic Dissection in Pregnancy

Two Operations Yielding Five Healthy Patients

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Abstract

Type A aortic dissection in pregnancy is a rare, life-threatening condition with a higher incidence in patients with connective tissue diseases. Mortality is high, reflecting the challenges of protecting both maternal and fetal well-being. We discuss two pregnancies complicated by aortic dissection, including one twin pregnancy, and describe the successful aortic repair immediately following Caesarean section. A total of three healthy neonates were delivered. The challenging management and implications of this precarious condition are explored in the context of these cases.

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Key Words

Aortic dissection · Surgery · Pregnancy · Aortic aneurysm

Introduction

Type A aortic dissection is a rare and life-threatening complication of pregnancy that most commonly occurs in the context of connective tissue disease (CTD) [1]. We present two cases of successful delivery and surgical repair of aortic dissection.

Case Presentation

Case One

A 32-year-old parturient presented at 32 weeks with anterior chest pain, radiating to her back. Before this, she had been in good health with an

uncomplicated pregnancy. Echocardiography revealed a Type A aortic dissection with torrential aortic regurgitation.

Intravenous antihypertensive agents were administered, followed by urgent transfer to our center. Under general anesthesia (GA), a Caesarean section was performed and the healthy, preterm baby girl was admitted to the neonatal unit.

Subsequently, a midline sternotomy incision was made, confirming Type A aortic dissection with the intimal tear across the sinotubular junction. Cardiopulmonary bypass was established using the right axillary artery for arterial return and the right atrium for venous drainage, and the aorta was clamped. Myocardial protection was by antegrade and retrograde multidose cold-blood cardioplegia. The aorta was opened and the valve excised. The coronary ostia were mobilized on aortic buttons. A bileaflet mechanical ATS valved conduit (ATS Medical, Inc., Minneapolis, MN, USA) was sutured in place and the ostia were reimplanted within the graft. The distal anastomosis was completed with the clamp on, and air was evacuated from the heart. The heart took over the circulation without support. The patient made an uneventful recovery and was discharged on day 5. Histology of the aorta showed only mild myxoid degeneration, with no evidence of CTD, and related genetic tests were all negative. Her Beighton score (which quantifies joint laxity) was 1/9.



Case Two

A 38-year-old woman presented in the 38th week of her twin pregnancy with severe breathlessness and a 2-week history of anterior chest pain radiating to the neck and back with syncopal episodes. There were clinical signs of hypoxia and frank pulmonary edema. The patient had preeclampsia and was being treated with methyldopa. Echocardiography revealed acute Type A aortic dissection involving the entire thoracic aorta with severe aortic regurgitation. She was treated initially with glyceryl trinitrate and transferred to our center.

Under GA, a Caesarean section was performed, delivering a baby boy and then a baby girl, both healthy. A midline sternotomy incision was made, confirming Type A aortic dissection. The intimal tear was in the noncoronary sinus. Cardiopulmonary bypass was established using the right axillary artery for arterial return and the right atrium for venous drainage, and the aorta was clamped. Myocardial protection was by antegrade and retrograde multidose cold-blood cardioplegia. The ascending aorta was resected, including the region of the root containing the tear. The valve was resuspended and the aortic layers were reapproximated with Bioglu. A 30 mm graft was implanted at the sinotubular junction. The distal anastomosis was completed with the clamp on, and air was evacuated from the heart. The heart took over the circulation without support. The patient made an uneventful recovery and was discharged on day 9.

Genetic testing for common CTD was all negative, but the geneticist believed features suggestive of Ehlers-Danlos syndrome type IV were present. Furthermore, there was a family history of aortic dissection and of death of unknown cause during childbirth.

Comment

The challenges of managing an acute aortic dissection in pregnancy begin at diagnosis, where delay increases mortality, which rises by 1 to 3% every hour, with a 25% mortality by 24 hours [2]. The classical presentation of anterior chest pain radiating to the back always merits prompt investigation. The diagnosis of aortic dissection is more challenging in preeclampsia, which presents with hypertension and epigastric pain, potentially masking the presenting features of an acute aortic dissection. Indeed, attrib-

uting epigastric pain and hypertension to preeclampsia alone may delay the diagnosis [3]. Distinguishing between these two conditions is further confounded by the fact that dissection, like preeclampsia, is more likely in the third trimester, when the hyperdynamic circulation and hormonal effects on the vasculature may predispose to dissection [4]. Echocardiography is a noninvasive, highly sensitive, and specific aid to diagnosis [5].

Anesthesia in these circumstances is challenging, requiring experienced input and consideration of the well-being of both mother and baby. Regional anesthesia is generally the method of choice for Caesarean section because GA carries a risk of aspiration, failed intubation, and hypoxemia associated with physiological changes in pregnancy [4]. In the setting of acute Type A aortic dissection, however, the choice of anesthetic type is altered. Regional anesthesia for Caesarean section has been used in the context of acute dissection [6], but the need for full systemic heparinization increases the risk of epidural hematoma. Blood pressure control, needed to reduce the risk of aortic rupture during delivery, is easier in GA. A rapid sequence induction will minimize the risk of aspiration; patients should be preoxygenated, and equipment for difficult airways must be available. A high-dose opioid technique is often used to ensure hemodynamic stability and diminish the hypertensive response to intubation, which could cause extension or rupture of the dissection, but this is associated with respiratory depression, so that the neonate may require intubation and ventilation after delivery. The mother should be placed in a 15° left lateral tilt until delivery to avoid compression of inferior vena cava and aorta by the gravid uterus. Strict blood pressure control is of paramount importance: glyceryl trinitrate, labetalol, hydralazine, and nifedipine can be used in pregnancy without any adverse fetal effects. Esmolol may be used, but propranolol can cause fetal hypoglycemia and bradycardia. Sodium nitroprusside should be avoided, as accumulation of cyanide ions can be fetotoxic.

Cardiac surgical outcomes are poorer in pregnant women, and neonatal survival is poor, especially if dissection happens well before term [7]. Multiple pregnancy poses an even greater risk of dissection complication, due to an elevated maternal cardiac output [8]. This is the first reported mother of twins in the United King-

dom, and one of very few worldwide, to survive aortic dissection during pregnancy without fetal loss.

Genetic investigation is indicated in aortic dissection. A diagnosis of CTD has implications for future pregnancies: 50% of aortic dissections in pregnancy occur in patients with Marfan syndrome [1]. The CTD-caused changes in the aorta, combined with the hemodynamic and vascular changes in pregnancy, elevate the risk of a dissection. Neither of our patients had mutations associated with the most common CTDs, although one may have clinical features of Ehlers-Danlos syndrome.

The detection and management of aortic dissection in pregnancy is challenging. The successful outcomes in these cases highlight two important points. First, due to the life-threatening nature of aortic dissection,

thorough investigation of anterior chest or epigastric pain is essential in pregnancy, even against a background of preeclampsia, and ultrasound offers a quick, safe, and accurate means of diagnosis. Second, the management of aortic dissection in pregnancy requires a multidisciplinary approach in which the anesthesiologist, obstetrician, neonatologist, and cardiac surgeon must urgently collaborate to ensure the protection of both mother and baby.

Conflict of Interest

The authors have no conflict of interest relevant to this publication.

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