Case Report

Marfan's Syndrome: Successful term pregnancy after repair of a dissecting thoracic aortic aneurysm at 21 weeks gestation

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ABSTRACT

We report a 38 year-old Chinese woman with Marfan' syndrome that had an acute type A aortic dissection and severe aortic regurgitation at 21 weeks of gestation. Emergency aortic valve repair and ascending aortic replacement was successfully performed under circulatory arrest with mild hypothermia. The patient underwent a cesarean section and delivered a healthy baby at 38 weeks.

KEY WORDS: Marfan' syndrome, Aortic Dissection, Bentall procedure, Pregnancy.

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INTRODUCTION

Acute type A aortic dissection during pregnancy can be fatal to both the mother and the fetus. Although rare, an association between pregnancy and aortic dissection has been reported.[1] We report the management for a scheduled cesarean section of a 38-year-old woman with Marfan' syndrome who previously had successful Bentall procedure to repair her aortic aneurysm and replace her aortic valve during the 21th week of her pregnancy.

CASE REPORT

A 38-year-old, gravida 2, para 1, 21-week gestation, Chinese woman was admitted to our institution complaining of crushing chest pain that radiated into her back. According to her family history, there was no one else with Marfan' syndrome. In addition to typical bilateral lens subluxation, arachnodactyly and long and thin extremities were found in the patient.

She was also known to have gestational hypertension. The chest computed tomography (CT) scan revealed the presence of a type A aortic dissection involving the ascending aorta (Fig-1-2). An echocardiogram demonstrated moderately dilated

left ventricle with normal systolic function, dilation of the aortic root with maximum diameter of 69mm, and severe aortic regurgitation (Fig-2A-2B).

The patient was counseled in great detail about the risk of continuing the pregnancy, but she chose repair of the aneurysm with continuation of the pregnancy. We performed an emergency Bentall procedure with mild hypothermic (33°C) with the fetus remaining in situ. Median sternotomy and cardiopulmonary bypass were established via the femoral artery with direct right atrial drainage and left atrial venting in this patient. The aortic root diameter was 65mm. A 23mm ATS Aortic Valve Graft (ATS Medical, Minneapolis, MN) was implanted with re-implantation of the coronary artery and aortic valve replacement were performed in this patient. The total cardiopulmonary bypass time and clamp time were 104 and 62 minutes, respectively. There were no intraoperative complications. The fetus survived the surgery. Uterine and vaginal bleeding did not occur during and after cardiovascular repair. The mother and fetus were discharged in good condition after a total hospital stay of 15 days.

Her labour started spontaneously at 38 weeks of gestation. She was under general anaesthesia caesarean section through a midline abdominal incision was performed. A normal healthy male newborn weighted 2330g was delivered with Apgar score of 8/10. Neurological or mental impairment or Marfan' syndrome-like appearance was not observed in the newborn.

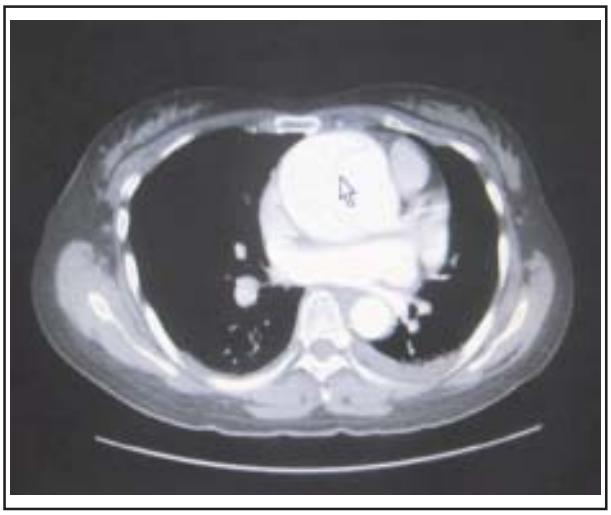

Fig-1: Contrast computed tomography scan showing the type A dissecting aneurysm of the aorta (arrow) with a maximal diameter of 8cm. The arrow points to the intimal flap between true lumen and false lumen.

DISCUSSION

Aortic dissection is a life-threatening disease that requires immediate surgical intervention. Marfan' syndrome is a hereditary disease, which is one of the most prevalent connective tissue disorders, presenting a risk of aortic dissection of approximately 1% even without dilatation of the aorta. Because of the increased risk associated with emergency operation for dissection during gestation, a prophylactic elective repair is preferred in women with Marfan's syndrome who contemplate pregnancy.[1]

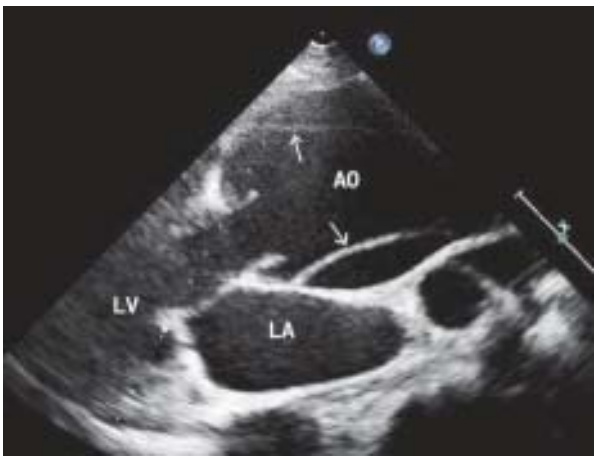

Fig-2A

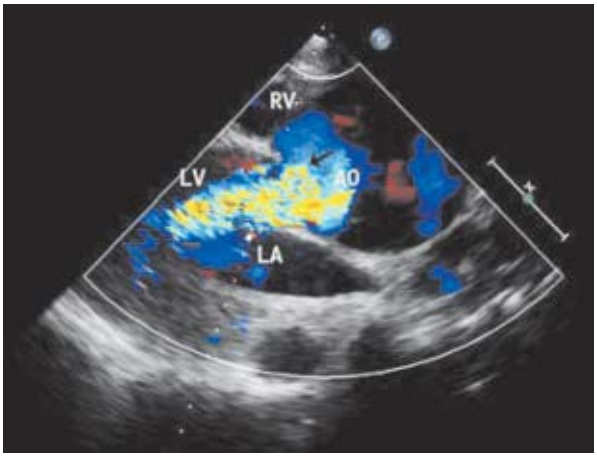

Fig-2B

Fig 2: Transthoracic echocardiography.
(A) Parasternal long-axis view. There is severe dilatation of the aorta and a linear echo is seen just above the aortic valve (white arrow).
(B) Parasternal long-axis view. The dilated aortic root, Severe aortic regurgitation (black arrows) is seen with colour flow mapping. LV, left ventricle; LA, left atrium; Ao, aorta; RV, right ventricle.

Surgery for acute aortic dissection during pregnancy has been described in previous reports in the literature, and favorable outcomes for pregnant women with acute type A dissection occurring late in pregnancy have been reported.[2,3] However, in most cases, the fetal outcome was relatively poor[4,5], likely secondary to variables associated with delivery of the fetus, fetus maturity and the condition of the fetus in situ. Cardiovascular operation using deep hypothermia with total circulatory arrest for aortic repair may be associated with an even higher risk of fetal mortality.

Zeebregts and colleagues[6] proposed a guideline for management according to the gestational age aiming to save both the mother and the fetus. Before 28 weeks gestation, aortic surgery with the fetus kept in the uterus is recommended. Other strategies, including minimization of deep hypothermic circulatory arresting time and institution of concomitant femoral artery cannulation for cardiopulmonary bypass may be of benefit in cases in which the fetus remains in situ. After 32 weeks gestation, primary cesarean section followed by aortic repair at the same setting is the management of choice as in the report of Shihata.[2] But, a difficult decision to be made is when aortic dissection occurs between 28 and 32 weeks of gestation. During this period, delivery must be decided based on the fetus' conditions, and caesarean section should be indicated in case of fetal distress, or pregnancy may be continued if the fetus can tolerate maternal surgery.

In our case, the patient strongly wanted to have a child, even if the fetus were to die during the operation. We did not use deep hypothermia, which would have prolonged the length of the operation and have been dangerous to the fetus. A high flow, high pressure, mild hypothermic or normo thermic flow on cardiopulmonary bypass is probably the safest choice for fetal preservation.

A pregnant Marfan's patient with a dissecting aortic presents a considerable challenge for anesthesiologists, obstetricians and cardiovascular surgeons. Proper management required consultations among the above departments, including cardiology and perinatology physicians. With appropriate care and surgical correction of the dissecting aneurysm timely in pregnancy, a successful outcome for the pregnancy was possible as in our case, where the dissecting aneurysm was repaired during the 21th week, with successful continuation of the pregnancy to 38 weeks.

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