



Type A Aortic Dissection in an Asymptomatic 28-Week Pregnant Patient with Marfan Syndrome

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Abstract

Acute aortic dissection during pregnancy is a rare and potentially fatal disease. Connective tissues disorders, including Marfan syndrome, in combination with the physiological stress of pregnancy and delivery substantially increase the risk of aortic dissection. It is vital to recognize symptoms early, if present, in order to undergo medical and surgical management to increase the chance of survival. We report the case of an asymptomatic 32-year-old pregnant female with Marfan syndrome found on echocardiogram to have a markedly dilated ascending aorta and Stanford type A dissection, undergoing successful Caesarean section and Bentall procedure to rescue both mother and fetus.

Keywords

Aortic dissection, Marfan syndrome, Pregnancy

Introduction

Aortic dissection in pregnancy is a rare complication that occurs most commonly in the 3rd trimester or in the early postpartum period. Conditions that predispose patients to the development of an aortic dissection include hypertension, collagen disorders, advanced age, male sex, preexisting aortic aneurysms, bicuspid aortic valve, pregnancy and delivery. It is a disease characterized by significant morbidity and mortality, particularly in the presence of associated risk factors. In pregnancy, aortic dissection is most commonly associated with connective tissue disorders such as Marfan syndrome. We present a rare case of an asymptomatic Type A aortic dissection in a pregnant woman with Marfan syndrome who underwent successful delivery of fetus and an uneventful postoperative course.

Case Report

A 32-year-old female (gravida 2, para 1) of Indian descent with a past medical history of hypertension and Marfan syndrome was referred to our institution at 28 weeks gestation during her 2nd pregnancy. Two weeks prior to admission, a transthoracic echo was completed, revealing mild to moderate aortic insufficiency along with severe aortic root dilation of 54 mm at the sinotubular junction (Figure 1). This was compared to a previous echo completed in 2014 revealing normal dimensions of the aorta. Magnetic resonance angiography (MRA) of the chest confirmed

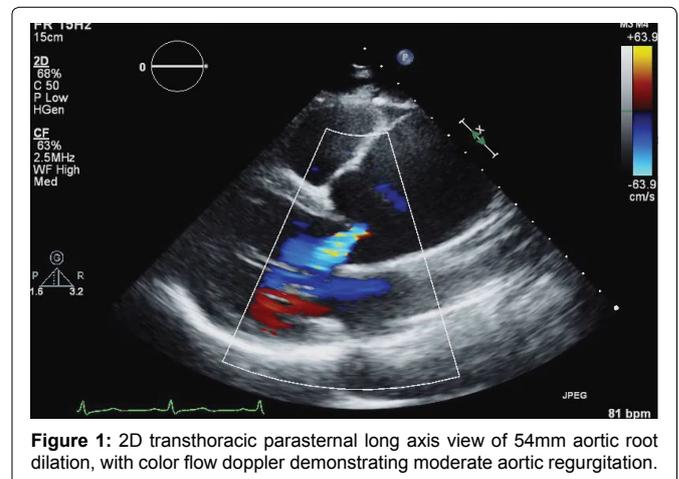


Figure 1: 2D transthoracic parasternal long axis view of 54mm aortic root dilation, with color flow doppler demonstrating moderate aortic regurgitation.

type A aortic dissection, beginning at the sinotubular junction and extending into the proximal aortic arch (Figure 2). A separate type B dissection was also found extending from the distal thoracic aorta to just before the aortoiliac bifurcation. Cardiothoracic surgical and high risk obstetric consultation was obtained and a multidisciplinary approach concluded that immediate repair of the aortic root and type A dissection should be undertaken after scheduled Caesarean section in the cardiac operating suite. The type B dissection was managed medically.

The patient has a strong family history of Marfan syndrome. Her mother, brother, and two-year-old son have Marfan syndrome. Her mother underwent aortic valve replacement, and brother underwent valve-sparing surgery of the aortic root. There is no family history of aortic dissections. The patient's previous pregnancy was an uncomplicated, normal spontaneous vaginal delivery at 37 weeks in early 2014. It was after her first pregnancy, when her brother was diagnosed, that she became aware of her own diagnosis. Since the diagnosis, she had never had routine screening of her aorta.

On admission and throughout her entire pregnancy, the patient denied any chest pain, back pain, abdominal pain, dyspnoea with rest or exertion, or light-headedness. Her pregnancy was uneventful other

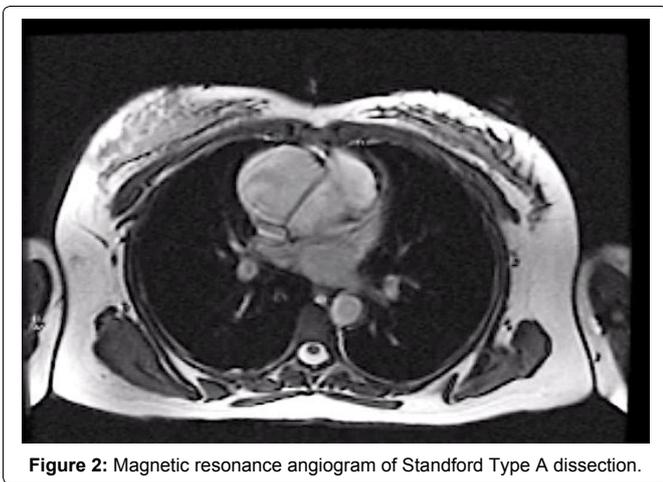


Figure 2: Magnetic resonance angiogram of Stanford Type A dissection.

than some minor weakness and fatigue. Given the lack of symptoms, it was believed that the dissection was most likely sub-acute or chronic, occurring sometime within the past two years, since the prior echo report from 2014 was normal.

On presentation, the patient's blood pressure ranged from 100-120 systolic and 80-90 diastolic with a heart rate ranging from 70s to 80 s. Physical exam of the patient was unremarkable, except for a 2/6 diastolic murmur of aortic insufficiency. Patient had non-bounding palpable pulses bilaterally in the upper and lower extremities. Per guidelines, goal heart rate was strictly maintained around 60 beats per minute with a systolic blood pressure less than 120 mmHg with labetalol [1].

Patient was initiated on steroids for fetal lung maturity, with a plan for Caesarean section under regional anesthesia in one week, after which, the patient would undergo the Bentall procedure. If at any point the patient were to become unstable, emergent surgical intervention would be considered.

The fetus was delivered by Caesarean section without complication 1 week after admission to the hospital. The patient recovered well in the CCU and was hemodynamically stable. The infant was and remained healthy.

Four days later, the patient was stable enough to undergo a Bentall procedure using a St. Jude mechanical valve conduit with a 23 mm mechanical valve, and an ascending aortic aneurysm repair along with hemi-arch replacement.

Patient recuperated well post operatively without any surgical complications and discharged on post-op day 5 with outpatient cardiology and OBGYN follow up.

Discussion

Aortic dissection is a fatal disease that may occur in the 3rd trimester of pregnancy and the early postpartum period. The incidence of acute aortic dissection in pregnant patients with Marfan syndrome is limited. In 1997, 4 out of 36 (11%) pregnant Marfan patients developed acute aortic dissection [2]. Recently in 2016, it was shown that 2 out of 31 (6.5%) pregnancies in a total of 19 Marfan patients, developed aortic dissections [3].

Hemodynamic alterations in the third trimester of pregnancy, including increased total circulatory volume and blood pressure change the aortic wall structure [4]. Increased estrogen and progesterone suppress the synthesis of collagen and elastin, which ultimately weakens vascular walls. This, along with disruption of the integrity of structural proteins in the aortic wall seen in Marfan syndrome, increases the risk of the development of aortic root dilation and dissection. Thus, in patients who are pregnant with Marfan syndrome, clinicians must have a high index of suspicion for aortic dissection, even in the absence of symptoms.

The presentation of aortic dissection varies from patient to patient. Typical symptoms include acute onset of severe tearing

chest, back, or abdominal pain [5]. Our patient presented without any symptoms, suggesting that the dissection had been chronic or subacute, developing over a matter of weeks to months. Very few case reports have been found where patients presented asymptotically, as well as reports on how to manage these patients appropriately. Data obtained from the International Registry of Acute Aortic Dissection (IRAD) shows that of 977 IRAD patients, only 63 patients (6.4%) presented with no pain symptoms [6].

Imaging studies should be utilized to diagnose a patient with aortic dissection. A meta-analysis by Shiga, et al. reviewed studies of the diagnosis of aortic dissection by TEE, helical CT and MRI. The data showed that these tests are comparable in terms of diagnostic value: TEE had 99% sensitivity and 95% specificity, helical CT had 100% sensitivity and 98% specificity, and MRI had 98% sensitivity and 98% specificity [7].

When deciding on how to manage a patient with an aortic root aneurysm that could potentially result in a dissection, it is imperative to accurately measure the aortic root diameter. In a prospective study of 21 Marfan patients, two out of four women with aortic root diameter of between 40 and 43 mm at pregnancy developed an aortic complication [8]. In a retrospective study in 36 patients, four Marfan women with aortic root diameters of between 40 and 45 mm developed aortic dissection [5]. In 2003, Immer, et al. showed an increased risk for the development of type A dissection in Marfan women with aortic root diameters greater than 40 mm. The absolute diameter of the aortic root and how rapid it dilates should both be taken into account [9]. It is recommended that surgery be performed to prevent rupture or dissection of the ascending aorta when the ascending aortic diameter reaches 55 mm for non-Marfan patients and 40 to 50 mm in Marfan patients [1]. A study of acute type A aortic dissection in pregnancy showed an incidence of 0.4 cases per 100,000 person-years, and a pre-admission mortality rate of 53% [10].

Studies have shown that women with Marfan syndrome with aortic dilation less than 40 mm tolerated pregnancy well. Women with dilations larger than 40 mm at pregnancy have a higher risk for aortic dilation, rupture, or dissection. Consequently, it is recommended that women with aortic root diameter greater than 40 mm should avoid pregnancy due to risk of aortic dissection [11]. When the aortic root is less than 40 mm, vaginal delivery can be tolerated. Caesarean section is recommended in women whose aortic root diameter is more than 40 mm [9]. It is recommended, though, that before 28 weeks of gestation, aortic repair be performed prior to delivery, and after 32 weeks gestation, emergency Caesarean should be performed prior to aortic repair [8]. Between 28 and 32 weeks, the risks and benefits should be taken into account by the team in order to proceed with either delivery or Caesarean section. Since our patient presented to us just after 28 weeks of gestation in the setting of a 54 mm dilated aortic root with type A aortic dissection, it was decided that our patient undergo Caesarean section prior to aortic repair.

Treatment depends on the site of the dissection, with surgery recommended for acute type A dissections and conservative therapy for type B dissections. Approximately 65% of aortic dissections have been found to occur in the ascending aorta, 30% in the descending aorta, less than 10% in the aortic arch, and about 1% in the abdominal aorta [12]. IV nitroprusside, if presenting with dangerously elevated pressures, and a β -blocker should be started upon initial diagnosis. Blood pressure control is one of the major actions that can be taken to avoid the development of aortic dissections in pregnancy, and to prevent further worsening of them if they have already developed [13].

It is imperative that, even without other risk factors, pregnant patients with Marfan syndrome be evaluated for aortic dissection if deemed necessary. Most importantly, in the presence of an aortic dissection, coordination between OB/GYN, cardiology, and CT/vascular surgery teams must be effective in order to manage and treat these patients appropriately.

Conclusion

Marfan syndrome is the leading cause of aortic dissection in third trimester women, followed by pregnancy itself. It is imperative that

physicians understand the risk factors for developing aortic dissection as patients may present asymptotically. Immediate evaluation along with further necessary medical or surgical interventions is essential to the fetus and patients probability of survival.

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