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CASE REPORT REGIONAL ANESTHESIA

Massive thoracic aortic dissection in the subacute postpartum period in a patient with Marfan syndrome

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ABSTRACT

Aortic dissection is a life-threatening condition that can result in rupture, massive hemorrhage, and death. Parturients with Marfan syndrome are at increased risk of aortic dissection due to connective tissue dysfunction and physiologic changes secondary to pregnancy. Aortic dissection typically manifests during the intrapartum period, rather than the postpartum course. This article discusses a case of a parturient with Marfan syndrome who suffered a massive thoracic aortic dissection in the subacute postpartum period after an uncomplicated vaginal delivery.

Abbreviations: CT - computerized tomography; AD - Aortic dissection; MFS - Marfan syndrome; PPD - postpartum day; TEVAR - thoracic endovascular aortic repair; TTE - transthoracic echocardiogram; VAVD - vacuum assisted vaginal delivery; VD - vaginal delivery

Keywords: Marfan Syndrome; Pregnancy; Aortic Dissection; Neuraxial Anesthesia

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1. INTRODUCTION

Almost one half of all aortic dissections in women younger than 40 y old manifest during pregnancy.[1] Aortic dissection (AD) presents at an average of 30 y of age and 32 weeks' gestation.[1] The majority of these cases occur in patients with Marfan syndrome (MFS) or other genetic syndromes.[1] Though less frequent, MFS patients continue to be at risk for AD in the postpartum period.[2,3] Most postpartum cases of AD generally occur within the first 8 weeks, but instances have occurred as late as 18 months after delivery.[4] We present management of a parturient with MFS, who suffered a massive thoracic aortic dissection (extending from the left subclavian artery to the left common iliac artery) on the postpartum day (PPD) 5 following an uncomplicated vacuum assisted vaginal delivery (VAVD). Written consent from the patient was obtained according to our institutional policy.

2. CASE REPORT

A 23-year-old G2P1001 at 37w2d with maternal MFS presented for a scheduled induction of labor (IOL) due to known aortic root dilation and favorable cervical exam. Two years prior, the patient underwent an uncomplicated VAVD and lumbar epidural placement. At that time, the transthoracic echocardiogram (TTE) noted an ejection fraction of 58%, normal left ventricular diastolic function, dilated sinuses of Valsalva measuring 31 mm, and an aortic root diameter of 38.7 mm.

During this pregnancy, the patient had serial echocardiographic evaluation approximately every 8 weeks. TTE at 20 weeks' gestation noted an ejection fraction of 58%, dilated sinuses of Valsalva measuring 31 mm, and an aortic root diameter of 39 mm. TTE at 27 weeks' gestation noted no changes to the sinuses of Valsalva or aortic root diameter.

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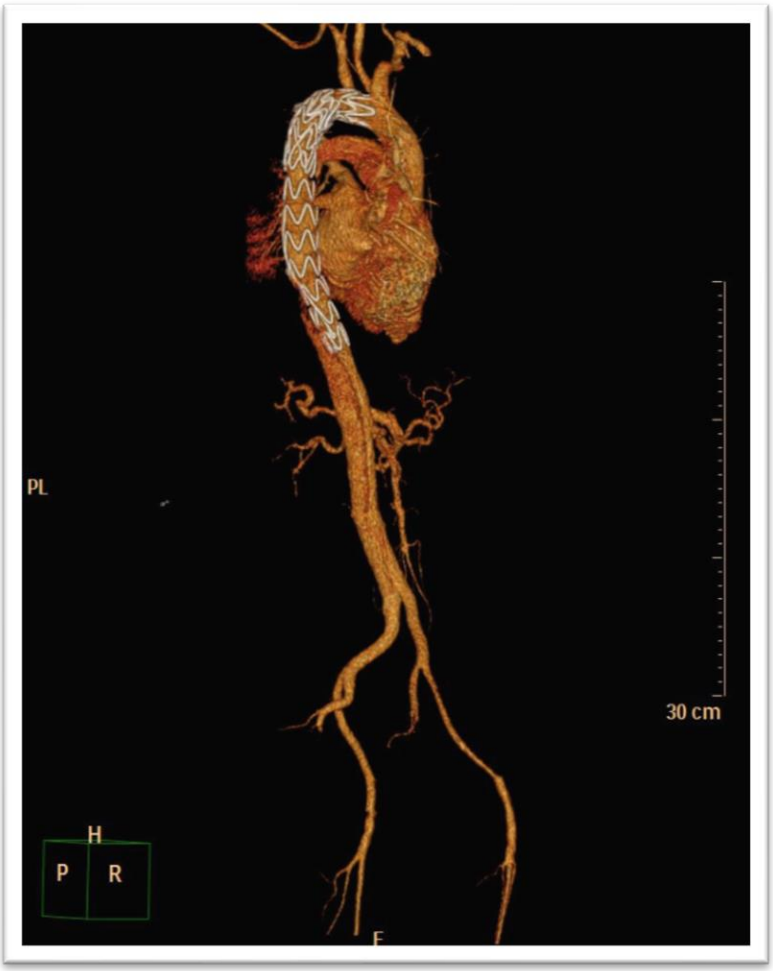

Image 1: History of type B aortic dissection with interval new patent endovascular aortic stent graft of thoracic aorta(TEVAR) extending to the true lumen of supraceliac proximal abdominal aorta with exclusion of distal aortic arch -proximal descending aorta aneurysm without evidence of perigraft contrast extravasation. Interval slight decrease in size of para-aortic hematoma around the distal aortic arch.

-Residual dissection flap seen at the distal end of the stent graft extending to the abdominal aorta and distal ends at the proximal left common iliac artery. Symmetric perfusion of both true and false lumen.

-Stable aneurysm of aortic root measuring 4.7 cm in maximum diameter at the level of sinus of Valsalva.

At admission, the patient was continued on her home metoprolol regimen. All initial laboratory work-up was unremarkable. Her baseline vitals were as follows: T 36 °C, BP 100-120 mmHg/60-80 mmHg, HR 70-80 bpm, RR 12-20 rpm, and BMI 24 kg/m^2. The patient had an early placement of a combined spinal epidural and the time to delivery from hospital admission was 16 h. All recorded vital signs throughout labor and delivery were within 20% of the baseline. The patient was discharged home on PPD-2 after an uneventful VAVD.

However, on PPD-5 the patient reported to an outside facility with complaints of generalized back pain which was inaccurately attributed to the epidural placement. In addition, her urinalysis was suggestive of new onset UTI. Hence, she was prescribed oral antibiotics and was discharged home by the outside facility. However, her generalized back pain continued to worsen in severity and on PPD-9 the patient was readmitted to our institution. An emergent computerized tomography (CT) demonstrated an acute thoracic Stanford type B aortic dissection. The dissection flap originated 1.5 cm distal to the left subclavian artery takeoff and extended through the abdominal aorta, terminating distal to the proximal left common iliac artery. There was also an interval increase in the diameter of the aortic aneurysm (now 47 mm from earlier 39 mm) along with active contrast extravasation from the lumbar and intercostal arteries (Image 1).

The patient was medically optimized and stabilized on infusions of nicardipine and esmolol prior to surgery. On PPD-11, she underwent thoracic endovascular aortic repair (TEVAR) with coverage of the left subclavian artery and a lumbar drain placement (at L3-L4). CT imaging following the TEVAR showed a small focal dissecting aneurysm in the proximal right subclavian artery. At time of discharge (postop day 10/ PPD-21), it was discussed that the patient would need hemiarch replacement in the future. Two months after discharge, CT follow up showed no interval changes in aortic measurements: the aortic root remained stable at 47 mm (Image 2).

3. DISCUSSION

Marfan syndrome is a hereditary disorder with an incidence of 1 in 5,000 people.[1,5,6] Parturients with MFS have higher rates of preterm deliveries, premature membrane rupture, cervical incompetence, postpartum bleeding, neonatal mortality, and aortic dissection.[3,7] MFS patients exhibit a mutation in fibrillin-1 resulting in an increase in Transforming Growth Factor (TGF)-beta signaling. This leads to medial deterioration and an elevated risk of aneurysm formation and dissection.[1,8] In

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addition, since the aortic tissue contains significant estrogen receptors, the risk of dissection may be magnified due to rising levels of estrogen during pregnancy.[1] Mouse models on MFS indicate that oxytocin, a hormone involved with uterine contraction and milk letdown, may play a role in aortic dissection as well.[12]

Approximately one half of aortic dissections, in women younger than 40 y, occur during pregnancy.[1] AD presents at an average of 30 y of age and at 32 weeks of gestation.[1] The highest risk for AD occurs in the third trimester and postpartum period. In particular, the third trimester poses the greatest hemodynamic stress. In the postpartum setting, AD has been reported immediately following delivery and up to 18 months postpartum; with the most common period being within the first 8 weeks postpartum. Parturients continue to have an elevated risk as cardiovascular changes can take weeks after pregnancy to normalize.[1] Rapid increase in blood volume, heart rate, and stroke volume, associated with normal pregnancy induced physiology, intensify shearing forces on the aorta, and the risk for cardiovascular complications.[9] Moreover, compensatory increases in ventricular ejection forces occur as the gravid uterus progressively expands and compresses the aorta and iliac arteries, magnifying the risk intimal tears.[10,11] The proximal aorta is the most common site of pregnancy associated dissections.[1] Though not causally proven, it is the current view that hormonal and physiologic changes are key factors leading to an elevated risk of aortic dissection in pregnancy.

Management is individualized based on the parturient and fetal wellbeing, the gestational age, and the type of aortic dissection.[1] Stanford type B dissections are typically managed medically, while type A dissections require emergency surgery. If dissection occurs before 28 weeks' gestation, aortic repair with the fetus kept in-utero is recommended.[1] If dissection occurs after 32 weeks, primary cesarean delivery followed aortic repair at the same operation is recommended. Between 28 to 32 weeks' gestation, the strategy of delivery is determined by maternal and fetal condition.[1] However, delivery may be considered in patients as early as 24 weeks' gestation (the age of fetal viability) if surgical intervention is recommended for the parturient. If surgery is advised, the obstetricians and neonatologists facilitate informed decision making based on fetal survival and long-term neuro-developmental data.

In patients with MFS and connective tissue disease, open repair is cited as the preferred surgical approach.[13] Currently, TEVAR has limited safety and efficacy data, while open repair offers a longer track record and better outcomes. However, TEVAR may be desired in emergent settings or when the risk of an open repair is too great. In time, TEVAR may become a more frequent alternative to open repair.[13]

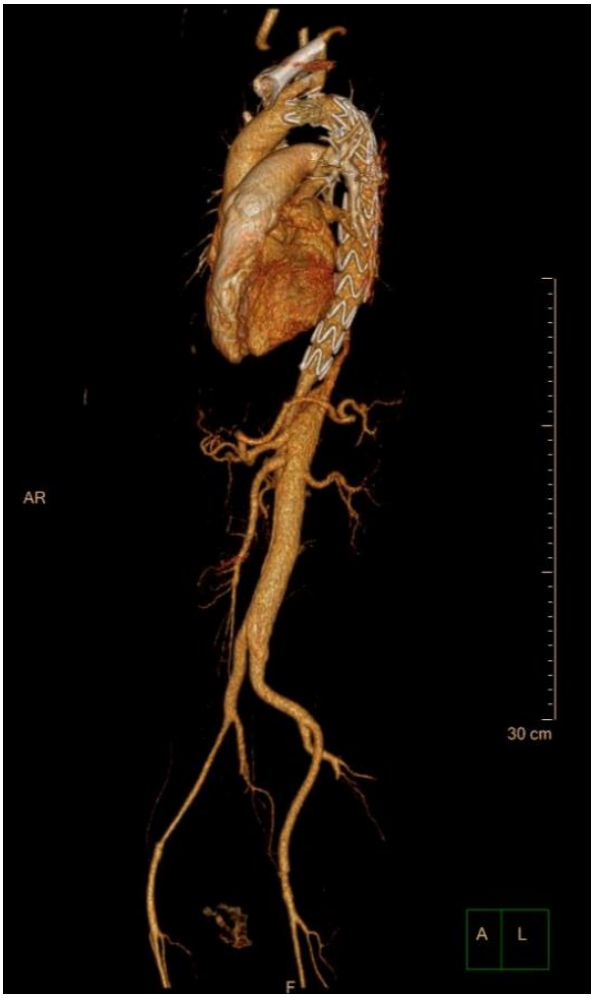

Image 2: History of type B aortic dissection with stable patent endovascular aortic stent graft of thoracic aorta (TEVAR) extending to supraceliac proximal abdominal aorta. Residual dissection flap seen at the distal end of the stent graft extending to the abdominal aorta and distal ends at the proximal left common iliac artery. Symmetrical perfusion of both true and false lumen via multiple fenestrations. -Stable aneurysm of aortic root measuring 4.7 cm in maximum diameter at the level of sinus of Valsalva.

Patients with an aortic root diameter of > 40 mm have a 10% risk of dissection during pregnancy.[14] Diagnostic criteria obtained by echocardiography or magnetic resonance imaging (MRI) are critical in assessing risk of potential AD and establishing a plan for delivery.[15] TTE is the screening modality of choice, with a sensitivity and specificity up to 75% and 90% respectively.[11] CT and

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aortography may be utilized, but involve exposure to radiation and intravenous contrast to the fetus.[1] In emergent situations, these modalities are not contraindicated in pregnancy.

Management of MFS parturients involves close maternal hemodynamic control, while minimizing fetal cardiovascular and respiratory depression. To decrease heart rate and reduce shear forces that contribute to dissection, beta-blockers are advised, particularly if the aortic root is larger than 40 mm or progression of aortic root enlargement is observed.[1] Side effects of beta-blockers in pregnancy include increased uterine tone, decreased umbilical flow, fetal bradycardia, fetal growth restriction and hypoglycemia.[1] Other treatment options include use of propranolol, esmolol and nicardipine.

There is no established consensus regarding anesthetic choice for patients with MFS. Patients with an aortic diameter < 40 mm and sinus of Valsalva < 40 mm are typically cited as being safe for vaginal delivery.[9] vaginal delivery (VD) is associated with increased hemodynamic lability.[11] Every effort should be made to minimize acute and wide swings in hemodynamics. Early epidural anesthesia and laboring in a semi-upright position or on the left side is recommended.[1] VAVD or forceps-assisted delivery is advised to expedite the second stage of delivery.[1,9]

Our patient, a parturient with MFS with stable aortic root diameter (< 40 mm) and a history of a successful VD, had less than one percent chance of AD. Management of delivery included routine monitoring (pulse oximetry, noninvasive blood pressure), a combined spinal epidural for improved sacral coverage, and an uncomplicated VAVD with an active second stage of labor spanning only three minutes. Nonetheless, our patient suffered a massive thoracic aortic dissection in the subacute postpartum period. The aortic dissection presented in the postpartum period; however, it is possible that it may have occurred during delivery and only later became appreciable as it progressed. Though it may not have prevented the occurrence of an AD, an aggressive approach at hemodynamic monitoring, control with invasive arterial catheter placement, telemetry during labor and delivery, and monitoring for up 24 h postpartum may have led to an earlier recognition.

In this case, a post-delivery echocardiogram and complete aortic CT or MR angiography may be necessary for earlier detection of dissection or interval aortic root diameter growth. Another area of intervention should be patient education and timely access to healthcare. Patients' and their support group should receive a focused education prior to discharge. It was clear that our patient was not fully aware of what to look out for as warning signs in the postpartum setting. Additionally, her closest hospital facility lacked knowledge of handling complications in patients with Marfan syndrome. Telemedicine appointments may have been advantageous, considering the patient's residence in a remote location.

4. CONCLUSION

Aortic dissection during pregnancy carries a high mortality rate for both mother and fetus. The primary anesthetic goal for patients during the antepartum, intrapartum, and postpartum periods all demand careful hemodynamic management and the reduction of cardiovascular stress. Type B aortic dissections are not well predicted by aortic root size close to 40 mm; and a type B aortic dissection may be missed on echocardiography alone. In these cases, imaging the entire aorta post-delivery with a CT or MR angiogram prior to discharge should be considered.

This case report highlights the importance of patient education regarding signs and symptoms of dissection, as well as provider education for those who may infrequently encounter parturients with the Marfan syndrome.

5. Conflict of interest

The study utilized the hospital resources only, and no external or industry funding was involved.

6. Authors' contribution

TD, NAC, RR - Concept, conduction of the study work and manuscript editing

GRR - Concept, and manuscript editing

7. REFERENCES

1. Suresh M, Segal BS, Preston R, Fernando R. Shnider and Levinson's Anesthesia for Obstetrics. 5th ed. Philadelphia, PA: Lippincott Williams & Wilkins Health; 2013.
2. Ryan SL, Bhattacharyya S. Connective tissue disorders in pregnancy. Neurol Clin. 2019;37(1):121-9. [PubMed] DOI: 10.1016/j.ncl.2018.09.006
3. Meijboom LJ, Drenthen W, Pieper PG, Groenink M, van der Post JA, Timmermans J, et al. Obstetric complications in Marfan syndrome. Int J Cardiol. 2006;110(1):53-9. [PubMed] DOI: 10.1016/j.ijcard.2005.07.017
4. Chang CY, Yang JM, Lam CW, Chen PH. Successful management of aortic dissection in a patient with Marfan syndrome during pregnancy. Am J Obstet Gynecol. 2013;208(2). [PubMed] DOI: 10.1016/j.ajog.2012.11.034
5. Bhushan R, Altinbas L, Jäger M, Zaradzki M, Lehmann D, Timmermann B, et al. An integrative systems approach identifies novel candidates in Marfan syndrome-related pathophysiology. J Cell Mol Med. 2019;23(4):2526-35. [PubMed] DOI: 10.1111/jcmm.14137
6. Curry RA, Gelson E, Swan L, Dob D, Babu-Narayan SV, Gatzoulis MA, et al. Marfan syndrome and pregnancy: maternal

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and neonatal outcomes. BJOG. 2014;121(5):610-7. [PubMed] DOI: 10.1111/1471-0528.12515

7. Rossiter JP, Repke JT, Morales AJ, Murphy EA, Pyeritz RE. A prospective longitudinal evaluation of pregnancy in the Marfan syndrome. Am J Obstet Gynecol. 1995;173(5):1599-606. [PubMed] DOI: 10.1016/0002-9378(95)90655-x
8. Delgado García DR, Latorre Andreu P, Fernández Tomás B, Tébar Cuesta MI. Marfan syndrome in a term-pregnant woman with aortic root dilatation between 40 and 45 mm. Rev Esp Anestesiol Reanim. 2019;66(1):49-52. [PubMed] DOI: 10.1016/j.redar.2018.08.007
9. Pyeritz RE. Marfan syndrome: improved clinical history results in expanded natural history. Genet Med. 2019;21(8):1683-90. [PubMed] DOI: 10.1038/s41436-018-0399-4
10. Harris CG, Croce B, Tian DH. Marfan syndrome. Ann Cardiothorac Surg. 2014;3(4):437. [PubMed] DOI: 10.3978/j.issn.2225-319X.2014.07.09
11. Ágg B, Szilveszter B, Daradics N, Benke K, Stengl R, Kolossváry M, et al. Increased visceral arterial tortuosity in Marfan syndrome. Orphanet J Rare Dis. 2020;15(1):91. [PubMed] DOI: 10.1186/s13023-020-01369-w
12. Habashi JP, MacFarlane EG, Bagirzadeh R, Bowen C, Huso N, Chen Y, et al. Oxytocin antagonism prevents pregnancy-associated aortic dissection in a mouse model of Marfan syndrome. Sci Transl Med. 2019;11(490):eaat4822. [PubMed] DOI: 10.1126/scitranslmed.aat4822
13. Harky A, Hussain SMA, MacCarthy-Ofosu B, Ahmad MU. The role of thoracic endovascular aortic repair (TEVAR) of thoracic aortic diseases in patients with connective tissue disorders: a literature review. Braz J Cardiovasc Surg. 2020;35(6):977-85. [PubMed] DOI: 10.21470/1678-9741-2019-0367
14. Minsart AF, Mongeon FP, Laberge AM, Morin F, Dore A, Leduc L. Obstetric and cardiac outcomes in women with Marfan syndrome and an aortic root diameter ≤45 mm. Eur J Obstet Gynecol Reprod Biol. 2018;230:68-72. [PubMed] DOI: 10.1016/j.ejogrb.2018.09.012
15. Frankel WC, Trautner BW, Spiegelman A, Grigoryan L, LeMaire SA. Patients at risk for aortic rupture often exposed to fluoroquinolones during hospitalization. Antimicrob Agents Chemother. 2019;63(2). [PubMed] DOI: 10.1128/AAC.01712-18
16. Goland S, Elkayam U. Pregnancy and Marfan syndrome. Ann Cardiothorac Surg. 2017;6(6):642-53. [PubMed] DOI: 10.21037/acs.2017.10.07

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