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PERIPARTUM CARDIOVASCULAR DISEASE MINI-FOCUS ISSUE

INTERMEDIATE

CASE REPORT: CLINICAL CASE

A Postpartum Type A Dissection

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ABSTRACT

A 28-year-old woman with familial thoracic aortic aneurysm and dissection syndrome and a mildly dilated aorta presented 3 days postpartum with a type A aortic dissection. This case illustrates the unpredictability of this disease and the challenges with risk stratification of women with underlying aortopathy contemplating pregnancy. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:150–3) Crown Copyright © 2020 Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

We describe a case of a 28-year-old woman who presented to the emergency department 3 days after the birth of her first child. She presented with pleuritic chest pain radiating down both arms, into her neck, and through to her back. Blood pressure was 116/69 mm Hg, and heart rate was 80 beats/min. Her clinical examination was remarkable for a new soft diastolic murmur at the right upper sternal edge. She had normal-quality bilateral upper limb and lower limb pulses.

LEARNING OBJECTIVES

- To recognize acute aortic dissection as a potential diagnosis in pregnancy and in the immediate postpartum period.
- To understand that aortic dimensions alone are not enough as a predictor for aortic dissection in pregnancy.
- To recognize that our current risk stratification tools for determining risk of aortic dissection in pregnancy are suboptimal.

PAST MEDICAL HISTORY

The patient had a background of familial thoracic aortic aneurysm and dissection syndrome, with a pathogenic variant in the *MYH11* gene. Her father experienced an aortic dissection at the age of 20 requiring 2 cardiac surgeries, and was the first to undergo genetic testing, which demonstrated a pathogenic variant in the *MYH11* gene. Her paternal grandfather succumbed to an aortic dissection at the age of 42. The patient was referred for genetic testing after her father's genetic results and was in the first trimester of her pregnancy when she received the result that she carried the same pathogenic gene variant. She was referred to the cardiac obstetrics (COB) clinic for counseling and ongoing care during pregnancy. It was unclear whether she had received prepregnancy counseling. At her first visit to the COB clinic at 18 weeks' gestation, her height was 167 cm and her weight was 61.7 kg. The proximal ascending aorta measured 38 mm on transthoracic echocardiogram (TTE). The indexed aortic dimension was normal at 22 mm/m^2, although this is underestimated using her pregnancy weight. She was counseled as high risk during pregnancy, and she opted to

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Informed consent was obtained for this case.

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continue with the pregnancy. She was placed on metoprolol during the pregnancy and her aortic dimensions were closely monitored by both TTE and cardiac magnetic resonance imaging. The aortic dimensions remained stable and the pregnancy was event free. A high-risk care plan was initiated for her labor and delivery. She went into spontaneous labor at 39 weeks. She had an early epidural and subsequently underwent a vaginal delivery with passive descent in the second stage as planned. The latter part of labor was complicated by early chorioamnionitis and she was started on intravenous antibiotics. She was fully dilated by this stage and breast pumps were used to expediate the latter stage when contractions intermittently slowed down. She had postpartum hemorrhage secondary to retained products, which responded promptly to manual evacuation and bimanual massage. As per the established high-risk care plan, no oxytocin, Hemabate, or ergotamine was administered. She remained normotensive throughout the admission. The patient wished to be discharged on day 2 postpartum, and was clearly advised to return should she have any symptoms of chest pain. She remained on the same dosage of metoprolol on discharge.

DIFFERENTIAL DIAGNOSIS

The differential diagnoses of this presentation in this early postpartum young female patient includes acute aortic dissection, acute pulmonary embolism, and spontaneous coronary artery dissection.

INVESTIGATIONS

Electrocardiogram demonstrated normal sinus rhythm with no ischemic changes. Troponin T was normal at 9 (N <14 ng/l). Hemoglobin was stable at 95 g/l. Renal function and electrolytes were normal. She was referred urgently for cardiac computed tomography, which demonstrated a type A aortic dissection extending from the sinotubular junction to just proximal to the brachiocephalic artery (**Figure 1**). No significant pericardial effusion was noted, and an urgent focused TTE demonstrated new mild aortic regurgitation.

MANAGEMENT

The patient was referred emergently for cardiac surgery. An intraoperative transesophageal echocardiogram confirmed the findings noted on cardiac computed tomography and TTE (**Figures 2A to 2C**). Intraoperatively, the aortic tissue was noted to be thin and friable. She underwent an ascending aorta repair, with a 24-mm tube graft. Her postoperative course was uncomplicated, and she was discharged home in a stable condition.

ABBREVIATIONS AND ACRONYMS

COB = cardiac obstetrics

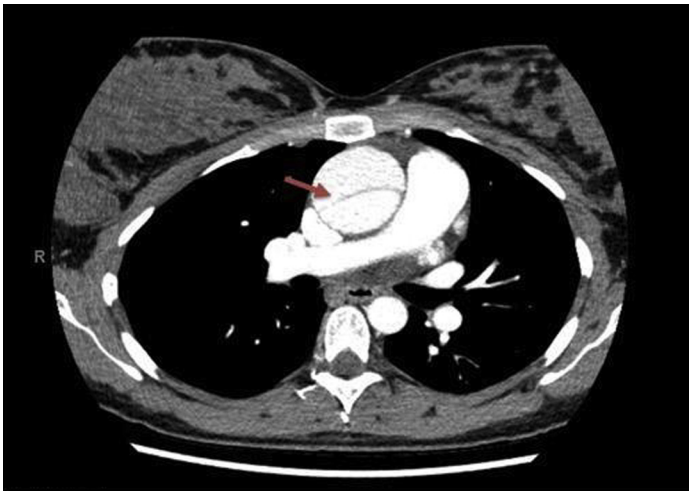
TTE = transthoracic echocardiogram

DISCUSSION

Familial thoracic aortic aneurysm and dissection syndrome refers to the familial inheritance of thoracic aortic aneurysms and dissections in the absence of a genetic syndrome such as Marfan or Loeys Dietz syndromes. They are generally inherited in an autosomal dominant pattern; however, demonstrate variable expression and decreased penetrance (1). Mutations in the *MYH11* gene affect the smooth muscle myosin heavy chain, which is a specific contractile protein of smooth muscle cells. This mutation causes very low smooth muscle cell content in association with large areas of medial degeneration in aortic tissue leading to aortic disease (2).

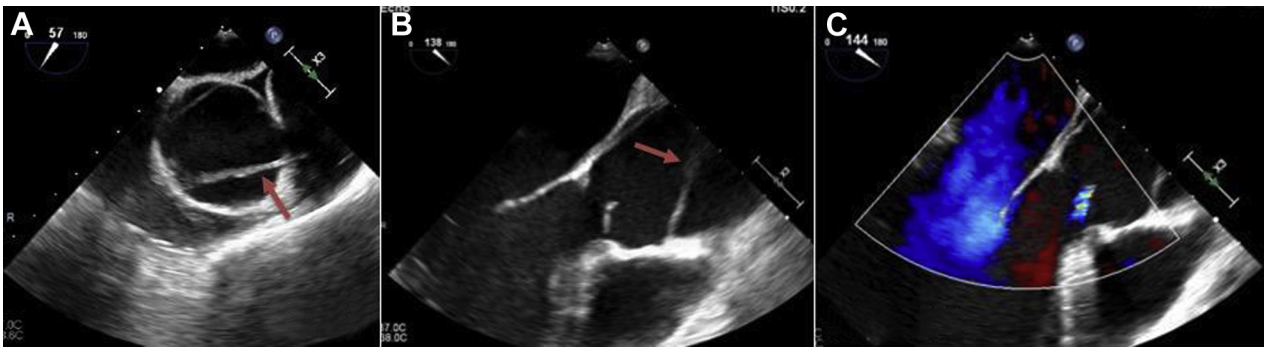
An acute aortic dissection in pregnancy is an uncommon but catastrophic event that can result in both maternal and fetal demise if not diagnosed and treated promptly. The risk of aortic dissection increases 5-fold in pregnancy compared with the nonpregnant state (3). This is because of a combination of hemodynamic and hormonal factors. An increase in heart rate, stroke volume, and cardiac output as seen in normal pregnancy can increase aortic wall stress. Meanwhile, estrogen suppresses the synthesis of both collagen and elastin, resulting in

FIGURE 1 Cardiac Computed Tomography



Type A dissection. **Arrow** points toward the dissection flap.

FIGURE 2 Intraoperative Transesophageal Echocardiogram



(A) Short-axis view of proximal ascending aorta demonstrating aortic dissection flap **(arrow)**. **(B)** Long-axis view of proximal ascending aorta demonstrating aortic dissection flap **(arrow)**. **(C)** Mild aortic regurgitation.

weakening of the aortic wall (4). More recently, oxytocin also has been implicated as a potential contributor to aortic events in pregnancy, as seen in mice models (5).

Both type A and type B aortic dissections have been described in pregnancy. Most aortic dissections are described during the last trimester of pregnancy and in the immediate postpartum period (6). To date, much of the literature around aortic dissection in pregnancy surrounds Marfan syndrome. The risk of aortic dissection during pregnancy with Marfan syndrome is increased when aortic dimensions are larger than 4 cm or when progressive enlargement is seen during pregnancy (7). Current guidelines therefore give a broad recommendation for a normal vaginal delivery for all pregnant women with any underlying aortopathy and an aortic dimension <4 cm, and a cesarean delivery for those with an aortic dimension >4.5 cm, implying that an aortic dimension <4 cm is of low risk, whereas that >4.5 cm is high risk (8). These guidelines oversimplify this rather complex disease process, failing to take into account the specific underlying aortic pathology and the family history. Aortic dissections have been described in pregnant patients with normal-sized aortas in Marfan and Loeys Dietz syndromes (9,10). Irrespective of pregnancy, one study demonstrated that patients with Marfan syndrome who did not have aortic complications were found to have a mean aortic dimension of 3.3 cm, which is well below what one would consider normal dimensions assuming an average height and body surface area in these patients (11). It is therefore clear that we as yet do not have all the sufficient tools to accurately predict the occurrence of aortic dissection in pregnancy in a woman with an underlying aortopathy. Aortic dimensions alone are evidently an inadequate guide for risk stratification for pregnancy, and additional factors such as underlying aortopathy and family history should be incorporated into preconception counseling.

FOLLOW-UP

The patient is currently 2 weeks following surgery and is awaiting follow-up in both the COB clinic and the heritable aortopathy clinic.

CONCLUSIONS

The risk of aortic dissection in pregnancy in a woman with underlying aortopathy can sometimes be difficult to predict, as aortic dissections can occur with normal aortic dimensions. It is therefore apparent that we need to incorporate other factors, including the underlying aortic pathology and family history, to appropriately risk stratify these women.

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