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Case Report

The silent signal of widened pulse pressure-aortic dissection in pregnancy

Kaithri Devi Subramaniam*, Shasheila M. Daud, Rafaie Amin

Department of Obstetrics & Gynaecology, Sarawak General Hospital, Kuching, Malaysia

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***Correspondence:**
Dr. Kaithri Devi Subramaniam,
E-mail: kaithrids@gmail.com

ABSTRACT

Aortic dissection in pregnancy is a rare but potentially life-threatening condition, often presenting with non-specific symptoms that can hinder timely diagnosis. Stanford type A dissections are especially perilous, with high risks of maternal and fetal mortality if not promptly treated with surgical intervention. A 35-year-old woman, G3P2 at 39 weeks' gestation, presented with sudden-onset epigastric pain. Her blood pressure was 103/43 mmHg, with a widened pulse pressure of 60 mmHg - a subtle yet significant indicator. The initial examination was unremarkable however an urgent echocardiography and CT aortography confirmed a Stanford type A aortic dissection, extending from the aortic root to the infrarenal abdominal aorta. An urgent interdisciplinary discussion (MDT) was convened. Given the complexity of the case, which required the expertise of a highly experienced cardiothoracic surgeon, a surgeon was flown in from Peninsular Malaysia. She was transferred to the Sarawak Heart Centre, where she underwent an emergency caesarean section, delivering a baby boy weighing 2.7 kg. This was followed by a 13-hour Bentall procedure and coronary artery bypass grafting. Although the surgery was technically successful, she sadly succumbed to postoperative heart failure seven hours later. This case highlights the importance of early recognition and timely intervention in aortic dissection during pregnancy. Although rare, pregnancy associated aortic dissection carries significant risks for both mother and fetus. Despite the tragic outcome, the swift and coordinated response by the multidisciplinary team is commendable and reflects their commitment in providing high-risk care in a challenging and resource-limited setting.

Keywords: Aortic dissection, Bentall procedure, Maternal mortality, Multidisciplinary management, Pregnancy, Widened pulse pressure

INTRODUCTION

Aortic dissection (AD) in pregnancy is an extremely rare but potentially life-threatening. The condition and has been reported in only 0.0004% of all pregnancies, accounting for 0.1-0.4% of all aortic dissections, but with a high mortality rate for mother and fetus.[1,2] Risk factors associated with aortic dissection include hypertension, collagen disorders such as Marfan syndrome, Ehlers-Danlos syndrome, bicuspid aortic valve, and Turner syndrome; inflammatory diseases leading to vasculitis such as giant cell arteritis, Takayasu arteritis, rheumatoid arthritis; and a family history of aortic dissection and preexisting aortic aneurysm.[3-6] Pregnancy per se is a risk factor of aortic dissection, likely secondary to the physiologic hemodynamic changes that lead to increased circulatory volume and elevated systemic blood pressure, and to the hormonal alterations that cause structural changes in the aorta.[7-9] Pregnant women with underlying acropathies such as Marfan syndrome are more susceptible to pregnancy-related aortic dissection.[7] Patients often present with non-specific symptoms that can hinder timely diagnosis. Pregnancy increases the risk of aortic dissection due to increased blood volume, cardiac output, and hormonal changes. In high-risk women, multidisciplinary preconception counselling is advisable however, many women at high risk of aortic dissection are not recognized early.[7,10] Stanford Type A dissection is especially perilous,

with a high risk of maternal and fetal mortality if not promptly treated with surgical interventions. Mortality is exceedingly high in acute Stanford Type A cases, rising by 1-2% per hour immediately after the onset of symptoms.[5] Given the high risk of mortality, immediate surgical intervention is crucial to ensure a better prognosis.

This case from Sarawak General Hospital, Malaysia, highlights the diagnostic difficulties and the logistic challenges of providing urgent cardiovascular care during pregnancy.

CASE REPORT

A 35-year-old woman Gravida 3 Para 2 at 39 weeks of gestation, and without apparent complications or remarkable family history, presented with sudden onset of epigastric pain radiating to her back. There was no history of exertional chest pain or shortness of breath. Following an uneventful development of pregnancy, this was her first visit to the hospital. Initial vital signs showed a blood pressure of 103/54 mmHg, pulse 90 beats per minute, oxygen saturation 97% on room air, and body temperature of 37 ºC. We noted that her pulse pressure was widened to 60 mmHg, which was a subtle but significant indicator. The physical examination showed a gravid woman in the left lateral knee chest position. No cardiac murmur or skeletal abnormalities were appreciated. As the symptoms were persistent and severe, with the subtle sign of widened pulse pressure, a decision was made for an urgent echocardiogram (ECHO).

The ECHO was reported to have moderate to severe aortic regurgitation with suspicion of dissecting aneurysm with an ejection fraction of 55%. She was then subjected to an urgent CT aortogram, which was reported to have extensive Stanford A aortic dissection from the root of the aorta extending to the infrarenal abdominal aorta, with the distal end of the intimal flap appearing at the L1 level (Figure 1 and 2). The aortic root measurement was 4.2 cm by 4.6 cm. There was no evidence of a leak or active bleeding.

An urgent multidisciplinary team (MDT) discussion, consisting of an obstetrician anaesthesiologist, cardiothoracic surgeon, transfusion medicine specialist, and neonatologist was convened. It was decided that she needs an urgent caesarean section followed by heart surgery following stabilisation. She was deemed not suitable to fly to Peninsular Malaysia for this complex surgery. In the absence of local cardiothoracic surgical expertise, a cardiothoracic surgeon was flown in from Peninsular Malaysia the very same day. She was transferred to Sarawak Heart Centre, where she underwent an emergency lower segment caesarean section with B-Lynch sutures, delivering a baby boy weighing 2700 g with an initial Apgar score of 1/3/4 intubated. A prophylactic B-Lynch suture was done in anticipation of post-partum haemorrhage with the ongoing bypass. This was followed by a 13-hour Bentall procedure and coronary artery bypass grafting. Following the surgery, she was transferred to the intensive care unit with for close monitoring and observation. Although surgery was technically successful, she sadly succumbed to postoperative heart failure seven hours later.

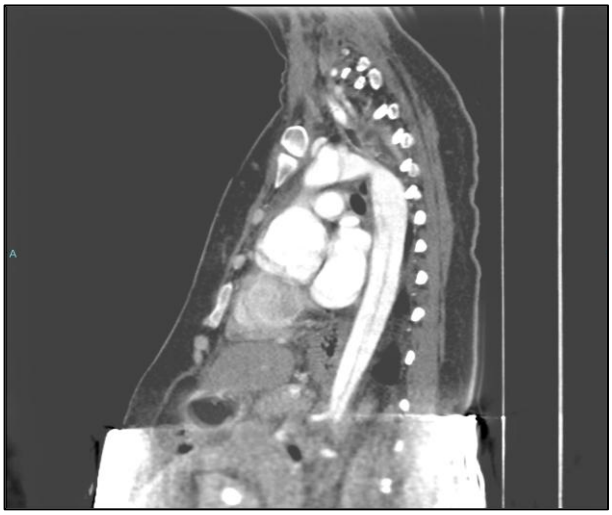

Figure 1: Aortic dissection from the root of the aorta.

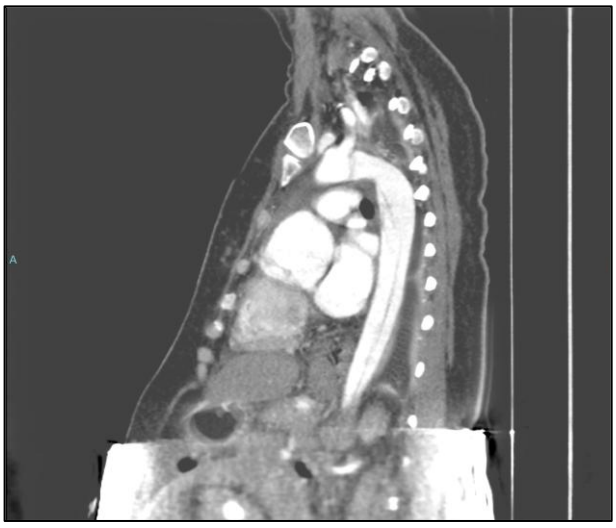

Figure 2: Aortic dissection extending to the infrarenal abdominal aorta.

DISCUSSION

Aortic dissection during pregnancy is a rare but potentially life-threatening event. Despite known predisposing factors, mainly a history of heritable thoracic aortic disease, it has been previously reported to occur even in patients without risk factors.[12] Pregnancy-related aortic dissection is rarely reported in the literature, and non-syndromic, sporadic cases like this are uncommon. Despite variations in timing and clinical presentation, all reported cases underscore the need for a high index of suspicion for aortic dissection and the importance of comprehensive physical and non-invasive maternal

assessments during pregnancy. Yuans et al study illustrated that hereditary fibrillinopathies represent the principal risk factors leading to AD in pregnancy, with Marfan syndrome being the most common.[13] Although connective tissue disorders, aortic root diameter of 40 mm or more, hypertension, bicuspid aortic valve, coarctation, and so on can predispose pregnant women to the occurrence of aortic dissection, some believe that pregnancy alone, with no underlying causes, is an independent risk factor for aortic dissection. Evidently, our patient's family history in the present case appeared unremarkable, and she had no prior history of hypertension.[14] The occurrence of an extensive Stanford type A dissection in this patient highlights that such catastrophic events can occur idiopathically, emphasizing the need for high clinical suspicion in similar presentations.

This case highlights the importance of early recognition and timely intervention in aortic dissection during pregnancy. It is important to have high clinical vigilance in patients presenting with atypical symptoms. Recognizing atypical signs, such as widened pulse pressure, was key in shifting focus to cardiovascular pathology. Early and accurate diagnosis is essential to prevent maternal mortality in such high-risk but often overlooked conditions.

Symptoms of AD can be nonspecific (e.g., chest pain, back pain, dyspnea, syncope, and weakness on one side of the body), and they mimic common problems in pregnancy. With the rarity of AD in pregnancy, this diagnosis may not be on the differential for providers in the community.[15] The patient's presentation of sudden epigastric pain radiating to her back, although common in various obstetric or gastrointestinal conditions, should prompt consideration of cardiovascular aetiologies, especially when accompanied by subtle clinical signs like widened pulse pressure. It has been reported that pain was the common symptom or complaint at the onset of AD, with back pain accounting for 55% of all presentations and chest pain accounting for 12%.[13]

In pregnancy, classical signs of aortic dissection can be masked or attributed to benign causes; hence, maintaining a high index of suspicion is vital. The physical examination, revealing a widened pulse pressure of 60 mmHg, although subtle, was a key finding prompting further advanced investigations. Thoraco-abdominal artery angiography is the "gold standard" of invasive aortography; however, exposure of the embryo to contrast agent is a concern. Iodinated contrast agents can suppress foetal thyroid function. Transthoracic echocardiography is a practical, non-invasive, bedside, and timely recommended diagnostic tool for unstable patients for whom there is a high degree of suspicion for aortic dissection.[16] Aortic dissection in pregnancy is a complex clinical scenario, difficult to diagnose and difficult to treat, considering the survival of both mother and fetus.[13] The treatment for AD in pregnancy is dependent on the type of AD and the gestational age of the fetus. In patients with acute type A dissection, surgery with aortic repair is necessary.[17] The reason for urgent caesarean section for the pregnant woman with AD was to avoid hemodynamic stress, progressive aortic expansion, and aortic rupture.[13] The decision to perform surgery, leaving the fetus in utero versus caesarean delivery prior to aortic surgery, depends on the gestational age of the fetus. There is no defined gestational age where delivery is recommended prior to surgery, but the recent ACC/AHA guidelines set this threshold at 26 weeks.[17] As our patient is advanced in her gestation, there is no dilemma to leave the baby in utero prior to repair. As suggested by the review by Wei Guo Ma et al in the third trimester, primary caesarean section followed by aortic repair performed in a single operative session is recommended.[18]

In her case, after an MDT discussion between an obstetrician anaesthesiologist, cardiothoracic surgeon, transfusion medicine specialist, and neonatologist the operation planned was caesarean section followed by heart surgery. Due to the lack of local expertise in Sarawak, a cardiothoracic surgeon was urgently flown from Peninsular Malaysia. The logistical challenges inherent in this case specifically, the absence of a local cardiothoracic surgeon and the geographical barrier of the South China Sea dividing Sarawak from the nearest cardiac center stressed the need for a contingency plan.

This scenario highlights the potential value of a rapid-response mobile surgical team, an approach successfully modelled by Zhu et al and colleagues for acute aortic emergencies in China.[14] While the establishment of such a specialized team must weigh the cost-benefit ratio against the infrequency of these critical cases, its utility in delivering timely emergency care to unstable patients in geographically challenging regions warrants serious consideration. She underwent a caesarean section followed by 13-hour Bentall procedure and coronary artery bypass grafting. Although technically successful, the unfortunate maternal mortality due to postoperative heart failure stresses the severity and high mortality associated with such cases.

CONCLUSION

Aortic dissection during pregnancy is a rare but potentially life-threatening event. Clinicians must maintain a high level of suspicion in pregnant patients with chest and back pain and continue to pursue appropriate imaging for potentially life-threatening conditions, even when initial testing is unrevealing. This case highlights the vital importance of early recognition and timely intervention in aortic dissection during pregnancy.

Although rare, pregnancy-associated aortic dissection carries a significant risk for both mother and fetus. Despite the tragic outcome, the swift and coordinated response by the multidisciplinary team is commendable and reflects

their commitment to providing critical, high-risk care in a challenging, resource-limited setting.

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