

Acute type B aortic dissection in a pregnant woman with undiagnosed Marfan syndrome: A case report and review of the literature

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ABSTRACT

Aortic dissection during pregnancy is rare but can be life-threatening to both the mother and the foetus. Marfan syndrome is a major risk factor for acute aortic dissection during pregnancy. Here, we present the case of a woman who had not been diagnosed with Marfan syndrome prior to pregnancy and who developed acute type B dissection at 32 weeks of gestation. The maternal hemodynamic status was stable, and foetal well-being was ensured. However, under conservative treatment, the dissection extended to the descending aorta, reaching the bilateral iliac artery 2 days later. Due to foetal distress, preterm delivery was performed via caesarean section. The primary treatment of type B aortic dissection is conservative medical treatment, with the goals of hemodynamic stabilisation, minimising the extent of the dissection and decreasing the risk of rupture. However, type B aortic dissection, even the uncomplicated type, in pregnant women may require early and aggressive obstetric interventions to improve maternal and foetal prognoses.

1. Introduction

Marfan syndrome is a systemic connective tissue disorder that involves the eyes, bones, and cardiovascular system. Cardiovascular complications, predominantly aortic dissection (AoD), are associated with Marfan syndrome and increase the mortality rate. Acute aortic dissections are classified into those that involve the ascending aorta (Stanford type A) and those that do not (Stanford type B). Stanford type B AoD does not typically require urgent surgery, unless the patient develops complications (e.g. end-organ malperfusion, refractory pain, rapidly expanding false lumen, and impending or frank rupture). The primary treatment for uncomplicated type B AoD is a conservative medical management that focuses on the strict control of heart rate and blood pressure. Beta-blockers reduce contractility and HR, which effectively decreases blood pressure and aortic wall stress [1]. The pooled 30-day mortality rate is 2.4% [2]. Type A AoD is managed surgically in both non-pregnant and pregnant women. In contrast, type B AoD has no established management during pregnancy. Here, we report a case of a pregnant woman who had not been diagnosed with Marfan syndrome but presented with acute type B dissection at 32 weeks of

gestation.

2. Case Presentation

A 38-year-old woman conceived her first baby via in vitro fertilisation and embryo transfer. She was 170 cm tall and weighed 42 kg prior to pregnancy. She had never been diagnosed with heart disease and had no family history of AoD or sudden death. She had smoked 15 cigarettes a day for 23 years. At 32 weeks and 3 days of gestation, she experienced sudden and severe pain in her chest and back, for which she visited the emergency room. Her blood pressure was 117/66 mmHg, and heart rate (HR) was 69 beats per minute (bpm). Contrast computed tomography (CT) revealed a type B AoD, starting at the aortic arch (Fig. 1). Transthoracic echocardiography (TTE) revealed a Valsalva sinus dilation of 38 mm (Fig. 2), without mitral valve prolapse or aortic valve regurgitation. With the addition of systemic findings (positive wrist signs, hindfoot deformity, dorsal ectasia, protrusio acetabuli, and some facial features), the patient was diagnosed with Marfan syndrome [3].

A multidisciplinary team evaluated her clinical course constantly. The treatment aimed to reach less than 120 mmHg systolic blood

Abbreviations: AoD, Acute aortic dissection; bpm, beats per minute; CT, computed tomography; HR, heart rate; TTE, Transthoracic echocardiography.

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Fig. 1. Computed tomography scan with 3D reconstruction showing aortic dissection starting at the arch (arrow)

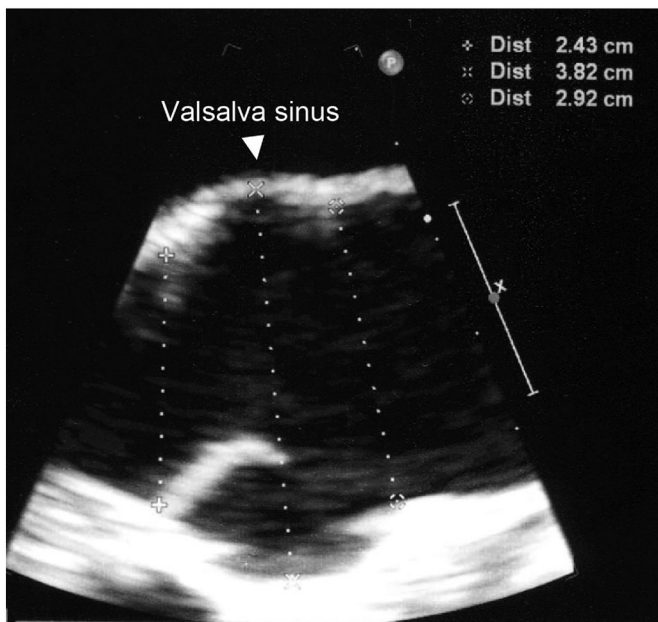


Fig. 2. Transthoracic echocardiography shows dilatation of the Valsalva sinus to 38 mm (arrowheads)

pressure and 60 bpm maternal HR [1]. She was not administered any medications, including opioids or cardiovascular agonists. Her systolic blood pressure remained at 100–110 mmHg, and HR was 60–70 bpm. Conservative management was administered for AoD because the dissection did not involve the ascending aorta, and there were no complications (including malperfusion). Cardiocography was performed frequently to ensure foetal well-being. Since both the patient and the foetal conditions were stable, it was decided to continue the pregnancy, considering foetal prematurity. Antenatal steroids were withheld because the delivery timing for the caesarean section had not been determined. However, on the third day of hospitalisation, the patient's sudden chest pain recurred. Her blood pressure could not be measured. She was transferred to the intensive care unit (ICU) and treated medically with opioid infusion. In the ICU, her blood pressure was 134/78 mmHg, and her HR was 54 bpm. TTE revealed no signs of type A dissection. However, transabdominal ultrasonography repeatedly detected foetal HR with prolonged deceleration down to 90 bpm, indicating foetal distress. Urgent caesarean section was performed under general anaesthesia, and a live, premature, male baby weighing 2070 g was delivered, with an Apgar score of 1/5.

Contrast CT imaging performed after the caesarean section demonstrated progression of the type B AoD, which had reached the right external iliac and left common iliac arteries (Fig. 3A, 3B). The three arterial branches in the left lower limb were poorly visualised compared with those on the right side (Fig. 3C). Additionally, contrast CT showed dissection of the origin of the left renal artery, resulting in poor perfusion of the left kidney (Fig. 3D, 3E). The patient did not undergo surgical repair after delivery. She developed renal dysfunction but did not require dialysis. She did not develop any motor or sensory dysfunction in her left lower extremity and increased her activities incrementally. The patient was discharged 31 days after surgery. She did not want genetic testing to be performed to confirm the diagnosis of Marfan syndrome. She was advised not to get pregnant nor did she intend to have another pregnancy. The baby was successfully discharged 46 days after birth.

3. Discussion

The most serious complication of Marfan syndrome in pregnancy is AoD, the incidence of which is reported to be approximately 4.0% [4]. The risk of AoD is substantially increased during pregnancy due to maternal hemodynamic changes and pathological changes to the aortic wall. Cardiac output is increased by 30%–50% between 28 and 32 weeks of gestation [5]. The levels of oestradiol, progesterone, and relaxin increase during pregnancy, all of which contribute to the loss of aortic wall integrity [6]. Foetal mortality in type A and B dissections has been reported to be 10.3% and 35%, respectively [7]. The higher incidence of foetal mortality in type B dissections may be due to the extension of dissections involving the internal iliac arteries, leading to uterine and placental hypoperfusion [7].

The general management of complicated type B aortic dissection is surgical intervention, whereas that of uncomplicated type B AoD is conservative medical treatment, with strict blood pressure and HR control to limit the extent of the dissection and the risk of aortic rupture [8,9]. Type B AoD in pregnancy is rare, accounting for only 11%–21% of all cases [8]. The management strategies for AoD in pregnancy depend on factors such as complications, gestational age, and foetal conditions [10]. Medical treatment with aggressive maternal and foetal monitoring is recommended for pregnant women who develop uncomplicated type B AoD [11,12]. Our case was uncomplicated type B AoD conservatively treated with strictly controlled heart rate and blood pressure, and the management strategy adhered to certain guidelines [1,13]. Although blood pressure and HR was under control without any medication, the aortic dissection extended to the bilateral common iliac arteries approximately 2 days after the first dissection. Similar cases of type B AoD that progressed under medical treatment in the third trimester of pregnancy have been reported [14,15]. Since hemodynamic status and

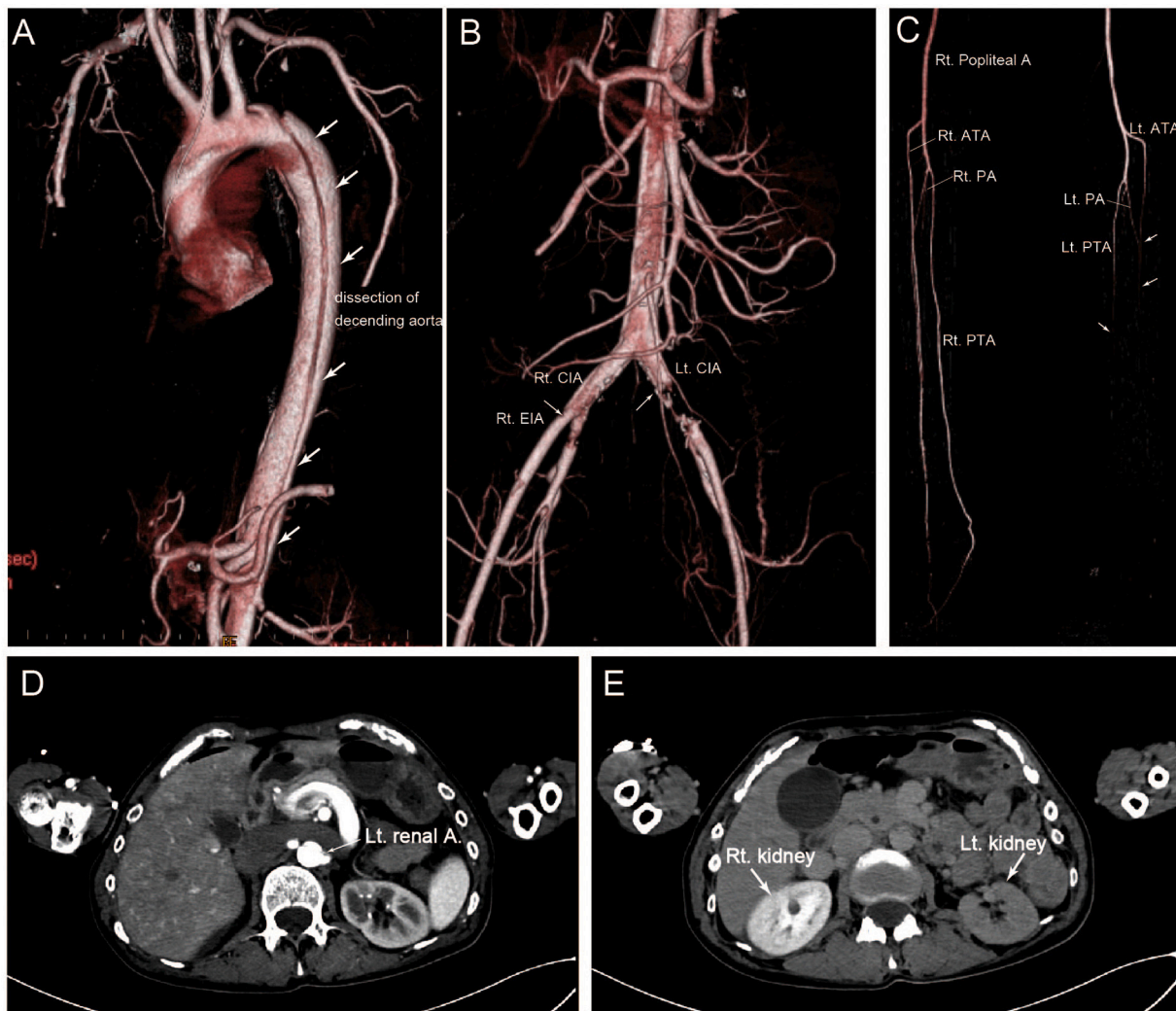


Fig. 3. Contrast computed tomography after caesarean section

(A) The aortic dissection begins just distal to the left subclavian artery and extends along the descending aorta (arrows).

(B) The dissection extends along the abdominal aorta to the aortic bifurcation, reaching the right external iliac artery and the left common iliac artery (arrows). CIA, common iliac artery; EIA, external iliac artery.

(C) The three arterial branches of the left lower limb (ATA, PA, and PTA) are poorly visualised (arrows) compared with those of the right limb. ATA, anterior tibial artery; PA, peroneal artery; PTA, posterior tibial artery.

(D) The aortic dissection extends to the origin of the left renal artery.

(E) Contrast perfusion in the left kidney was poorer than that in the right kidney.

the level of cardiovascular stress change frequently in pregnant women, aortic dissection may extend more easily than in non-pregnant women. Therefore, expanding the indications for interventions for type B dissection in pregnant women may reduce both maternal and neonatal mortality [8].

The optimal timing of delivery should be staged according to the clinical urgency of both maternal and foetal priorities. The survival rates for mothers and infants are based on the gestational age when type B AoD occurs [16]. If dissection occurs when foetal viability is expected, pregnancy termination may improve both the maternal and the foetal prognosis [17]. Antenatal steroids for foetal lung maturity could be considered if delivery is expected at less than 34 weeks of gestation [18]. In contrast, prophylactic preterm caesarean section for Marfan syndrome may not always prevent type B aortic dissection [9]. In addition, caesarean section itself may cause hemodynamic changes and lead to an extension of the unrepaired aortic dissection [6]. Therefore, the maternal normal HR and intravascular volume status must be maintained both during and after caesarean section to avoid an increase in systemic vascular resistance. Sympathetic stimulation should be

minimised to prevent the exacerbation of unrepaired aortic dissection. Special consideration should be given to drugs that may have adverse hemodynamic effects, such as methylergometrine.

Marfan syndrome often remains undiagnosed prior to pregnancy and is recognised only after the onset of life-threatening complications during pregnancy or after delivery [19], as in our case. Approximately 75% of persons with Marfan syndrome have a positive family history [20]. Consequently, all patients should be checked for their family history so as not to miss the diagnosis of Marfan syndrome. Physical examination, including eye examination and echocardiography, is also important for diagnosing Marfan syndrome [21].

In summary, we report a case of a pregnant woman who had not been diagnosed with Marfan syndrome but who presented with acute type B dissection at 32 weeks of gestation. Under conservative treatment, the aortic dissection was observed to extend. For women with Marfan syndrome, management should be started prior to conception and pregnant patients should be treated by a multidisciplinary team. If type B AoD occurs when foetal extrauterine survival is expected, delivery by caesarean section may contribute to a better prognosis for both the

mother and foetus.

Contributors

Yumiko Miyazaki was involved in drafting the manuscript.

Makoto Orisaka supervised the preparation of the case report and revised the manuscript.

Masataka Kato contributed to patient management.

Hiroshi Kawamura collected the clinical data and helped with editing the manuscript.

Tetsuji Kurokawa conceived and generally supervised of this study and gave final approval of the version to be published.

Yoshio Yoshida conceived and generally supervised of this study and gave final approval of the version to be published.

All authors read and approved the final manuscript.

Conflict of interest

The authors declare that they have no conflict of interest regarding the publication of this case report.

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Patient consent

Written informed consent was obtained from the patient for publication of this case report.

Provenance and peer review

This case report was peer reviewed.

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