Peripartum acute aortic dissection: A case report & review of literature

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ABSTRACT Acute aortic dissection is a rare clinical entity that mainly affects patients older than 50 years. It is unusual in younger patients and its presence has been traditionally associated with trauma, Marfan syndrome, bicuspid aortic valve and pregnancy. We present here, a case of a 30 year old pregnant female with acute aortic dissection type A (De Bakey II), without family history of connective tissue diseases and signs of Marfan syndrome.

Key words: Acute aortic dissection; Peripartum.

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Introduction

Type A aortic dissection (AD) is the presence of dissection proximal to the left subclavian artery (Stanford classification). According to De Bakey classification, type II is the presence of the dissected area confined to the ascending aorta. Aortic dissection is considered surgical emergency and it has been well documented, if untreated the mortality rate is extremely high. It has been estimated that mortality approaches 1% per hour for the first 48 hours and exceeds 80% during the first month (1). This type of dissection must be operated emergently and the extent of the reconstruction is dictated by the presence of aortic valve insufficiency and the proximity of the dissection to the aortic arch.

Case report

A 30 year old Hispanic female with no significant past medical history and currently 28 weeks pregnant, presented to the emergency room for severe substernal chest pain radiating to her back. The pain started 12 hours prior to presentation and was continuous, non reproducible, not associated with dyspnea and had no aggravating or relieving factors. There was 18 mm difference in the systolic blood pressures of bilateral upper extremities. Due to this clinical presentation, the suspicion of aortic dissection was very high. So a CT aortogram was done to rule out aortic dissection, which revealed an ascending aortic dissection starting from level of sinuses of valsalva to the distal ascending aorta and a large pericardial effusion highly suggestive of hemopericardium Fig 1.

A bedside 2D echo was performed emergently, which confirmed a Type A-dissection involving ascending aorta with impending cardiac tamponade.

A decision was made to take the patient to operating room for emergent surgical intervention. Initially the baby was delivered by caesarian section. Later a sternotomy incision was made and repair of aortic dissection was carried out. The patient was found to have a bicuspid aortic valve and was replaced by a prosthetic valve. Postoperative course remained benign and patient recovered fully.

Discussion

Aortic dissection during pregnancy is potentially lethal to both mother and fetus. The management is complex and depends on the type of dissection and gestational age. The Stanford classification system divides aortic dissections into two types: type A, involving the ascending aorta regardless of the entry site location, and type B, involving the aorta distal to the origin of the left subclavian artery. Type B dissections occurring during pregnancy are very rare. The treatment is medical with strict control of blood pressure as previously reported (1). Type A dissections require emergency surgery (2). There are cases in the literature detailing surgical repair at all stages of pregnancy and the post-partum period (3-10). Following a 12-year review of acute aortic dissection complicating pregnancy

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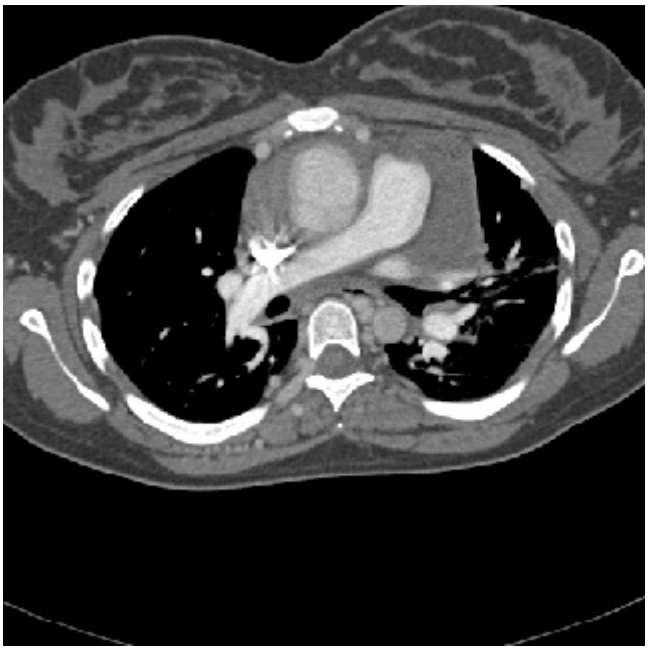

Fig 1. Ascending aortic dissection

Zeebregts and colleague's (11) have suggested the following management guidelines. In a pregnant woman with an acute type A dissection, treatment should be aimed at saving two lives. Before 28 weeks gestation, aortic repair with the fetus kept in utero is recommended. If the fetus is truly viable (ie after 32 weeks gestation), primary Caesarean section followed by aortic repair during the same operation is the treatment of choice. Between 28 and 32 weeks gestation there is a dilemma, with the delivery strategy determined by the fetal condition. The fetal and maternal mortalities for cardiovascular surgery during pregnancy are 20–30% and 2–6%, respectively (12).

Our patient's symptoms and examination findings were suggestive of aortic dissection and diagnosis was made in the least possible time. Due to the early diagnosis, our patient survived and made a full recovery. A number of studies have shown this to be unlikely. Me´sza´ros and colleagues showed a pre-hospital mortality of 21% (13). The mortality rate for untreated proximal aortic dissections increases by 1 to 3% per hour after presentation and is approximately 25% during the first 24 h, 70% at 1 week and 80% at 2 weeks. In our patient, the dissection progressed rapidly starting from level of sinuses of valsalva to the distal ascending aorta leading to huge hemopericoardium.

Although the clinical manifestations of acute aortic dissection are well described, the diagnosis is often overlooked in the pregnancy. A study over a 27-year period showed that misdiagnosis occurred in 85% of patients presenting with acute dissection (14). This is supported by a number of case reports in which the diagnosis was initially missed in the peripartum period. The most common presenting feature is sudden onset of severe back or chest pain (up to 96% of cases) that is characteristically stabbing, tearing or ripping. The pain is frequently migratory, generally following the path of propagation of the dissection. Signs on physical examination may reflect the location of the dissection and its extent.

Importantly, our patient was also noted to have a diastolic murmur and aortic regurgitation. Acute aortic valve incompetence accompanies 18–50% of proximal aortic dissections and is the second most common cause of death (after aortic rupture) in dissections. A bedside 2 D echo was performed which confirmed the Type A dissection involving the ascending aorta with impeding cardiac tamponade.

Although suggested by the clinical findings, the diagnosis of aortic dissection must be confirmed by investigation. Diagnostic modalities include chest radiography, echocardiography, contrast-enhanced computed tomography, aortography and magnetic resonance imaging. Although chest radiography lacks specificity it can be useful for the initial prediction of dissection when used in combination with the history and examination findings. The classic radiographic feature of mediastinal widening occurs in up to 50% of cases.

The main predisposing factor for aortic dissection is degeneration of the collagen and elastin in the intima media. Systemic hypertension is the main risk factor. Other well-established risk factors include hereditary connective tissue disease (eg Marfan's syndrome and Ehlers–Danlos syndrome), coarctation of the aorta, bicuspid aortic valve, aortitis and arch hypoplasia. On the basis that 50% of aortic dissections in women under 40 years of age occur in pregnancy or the puerperium, it is frequently stated that pregnancy is an independent risk factor for aortic dissection. The most common site of pregnancy-associated dissection is the proximal aorta, and aortic rupture usually occurs during the third trimester or first stage of labor.

In our patient neither clinical nor histological examination showed any evidence of hereditary connective tissue disease. Syphilis serology was normal and there was no documented evidence of hypertension. The only abnormal finding was a bicuspid aortic valve with idiopathic myxoid degeneration. This degeneration is defined as significant disruption of the valve fibrosa and its replacement by acid mucopolysaccharides together with cystic change. The underlying cause can be rheumatic, endocarditic, idiopathic, or connective tissue disorders. Idiopathic myxomatous degeneration is rare, with patients mainly in their fifties and sixties, although younger cases have been reported. There is a male preponderance. Most patients present with severe aortic regurgitation, with the diagnosis made after valve replacement.

To the best of our knowledge, this is the second reported case of pregnancy and acute aortic dissection occurring in association

with this valvular pathology in a non-Marfinoid patient.

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