Anaesthesia management of acute aortic dissection type B in Marfan syndrome complicating end-stage pregnancy

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ABSTRACT

Pregnancy in women with Marfan syndrome (MFS) is linked to approximately a 4.4% risk of acute aortic dissection (AAD). The natural history of pregnancy and the ability to deliver a viable fetus depends on the interaction between the pace of changes in the cardiovascular system and the advancement of pregnancy. We report the management of a type B acute aortic dissection in a woman in her 36th week of pregnancy. The anaesthesiologist has the unique task of managing complicated and mutually exclusive physiologic goals and acts as a consultant bridging the mother's well-being and fetal management.

Introduction

Marfan syndrome (MFS) is an autosomal dominant inherited defect of fibrillin production.[1] The most frequently cited anaesthesia-related considerations in MFS include: mitral valve dysfunction, aortic dissection, impaired systolic and diastolic function, abnormalities in chest anatomy, atlanto-axial subluxation and higher prevalence of dura ectasia.[1,2] Thus, each case of MFS has to be treated as unique by a multidisciplinary team. From an anesthesiology point of view, a pregnant MFS patient with much less prevalent acute aortic dissection (AAD) type B presents unique challenges.[2,4]

Case Report

A 27-year-old woman (para 0-1-1-1) in the 36th week of pregnancy checked into a regional emergency room because of sudden onset of mild chest pain. An interview demonstrated pre-existing MFS, gastrointestinal reflux, intermittent migraine headaches, and on-going tobacco abuse. At that time metoprolol had been prescribed for a slightly elevated blood pressure. At that time the patient's symptoms were attributed to heartburn considering negative finding on CXR and physical examination.

The patient returned three days later with pronounced complaints of a severe, constant abdominal and chest pain radiating to the back. She also complained of intermittent contractions. She was hypertensive (144/83 mmHg), tachycardic (100 min^{-1}) with palpable pulses on all extremities. Computer tomography showed type B AAD. Consequently, the patient was transferred to a tertiary medical centre.

Upon arrival at our centre, the patient's vital sign were stable. Her cervix was dilated to 4 cm with no contractions or vaginal bleeding. This was no progress in labour according to the documents from the referring centre. Amniotic membranes were intact. Fetal heart tracing showed a decreased beat-to-beat variability but it was reactive. Biophysical profile was assessed at six points. Review of the imaging showed AAD with origin just distal from the left subclavian artery and extending into the iliac vessels. Some extension of the aneurysm was seen in coeliac, superior mesenteric and renal arteries but blood flow to these organs was not compromised. Cardiothoracic consultant recommended pharmacologic management of the dissection. Subsequently, a right radial a-line was placed and an infusion of esmolol (~100 μg/kg/min) was started with target for systolic blood pressure of ~100 mmHg.[2] After these manoeuvres were achieved, fetal heart tracing slightly deteriorated. Upon consultation between the cardiothoracic, obstetric and anaesthesia team a decision was made that the patient should have a Caesarean section. Standard monitoring was accompanied by uninterrupted fetal heart monitoring. Continuous spinal anaesthesia was induced by using an 18G Touhy needle that was introduced into the spinal face at the level of L3-L4. After the clear outflow of spinal fluid was present an 18G epidural catheter was threaded to the length exceeding by 4 cm the depth at which spinal fluid was encountered. The epidural needle was gently withdrawn whereas the spinal catheter was secured in an aseptic way to the skin. After placing the patient in position, 0.4 ml of 0.75% spinal bupivacaine was introduced. This had to be repeated twice until sufficient height of loss of sensation to a pinprick (Th4 at nipple line) was obtained. No additional bolus of bupivacaine was given through the remainder of the procedure. Esmolol infusion was adjusted to maintain systolic blood pressure at 90 mmHg. Intermittently, phenylpherine (total dose during the 200 μg) was used to treat hypotension if accompanied by the patient's nausea and fetal bradycardia. Maternal haemodynamic remained stable (HR = 70–110 min^{-1}, SB = 90–120 mmHg). A viable fetus was delivered (APGAR score 9 and 10) and the spinal catheter was removed shortly afterward. The patient tolerated the procedure and was discharged from the hospital five days later.

Discussion

The majority of AAD in pregnant MFS patients are type A.[1,2] In

these cases surgical intervention is critical.[1,2] Type B of AAD is much less frequent.[1-2,4-6] It should be medically managed until relative maturity of the fetus is reached assuming there is no excessive risk to the mother.[1] In case of complications an immediate surgical repair is recommended since the false lumen can result in multiple ischaemic episodes involving the fetus, spinal cord, gastrointestinal system, kidneys and extremities.[2,3]

Some authors suggest that excessive mortality in MFS results from little or no prenatal counselling and lack of β-blockade.[2,6] If preventive medical treatment is implemented, MFS women have a maternal and fetal mortality similar to women without MFS.[2,6] Interestingly, Lind et al reported no increased mortality in pregnant MFS patients who were not treated with β-blockade, a result that questions a typical recommendation.[5] Additionally, non-selective drugs include fetal side-effects and decreased epinephrine-induced myometrial activity.[7] Hence, β1-selective blockers are preferable. There is no study examining the desired haemodynamic values after implementing β-blockade in pregnant patients but a heart rate below 80 beats-per-minute is suggested.[2,7] This is an important aspect of cardiovascular management since lowering the blood pressure to an arbitrary goal might result in impaired delivery of oxygen to the fetus while an aortic pressure that is too high, and heart rate to fast, increases the chance of a catastrophic haemorrhage.[2]

From the cardiovascular perspective the anaesthesiologist has to balance three goals: provide sufficient perfusion pressure to the mother's central organs, minimise risk of the dissection progression and provide optimal uteroplacental circulation. Meeting these targets is difficult since different optimal perfusion pressures and regulation mechanisms exist in a three circulation system.[7,8] Invasive measurement of blood pressure was used to assess the dissection force acting on the aortic wall.[2] Chest pain or change in the mother's mental status signified inadequacy of perfusion. Finally, continuous fetal heart rate monitoring provided with assessment of the fetal well-being.[7-9]

Epidural technique has been frequently used and recommended for C-section in Marfan patients.[9] However, a relatively high incidence of dura ectasia might result in a greater incidence of failure to achieve optimal pain control.[1,8] General anaesthesia abrogates the higher mental function required to monitor cerebral and coronary perfusion pressures. Continuous spinal anaesthesia provides superior pain control, acceptable haemodynamic stability and good fetal outcome if carefully titrated. The risk of spinal anaesthesia is hypotension, infection, and bleeding.[8] The latter complication is important if cardiopulmonary bypass will have to be used. In that case the intrathecal catheter should be retained until coagulopathy has been reversed with the risk of infection proportional to the length of time the catheter remains in place.[2] The management of the frequently observed spinal block-induced hypotension can be done with crystalloids or vasoactive pressor agents. In the case of our patient, we used phenylpherine since tachycardia was present. The current data are suggesting no clinically important benefit of using phenylpherine vs ephedrine.[10] Some authors also suggest that this technique has an unacceptably high risk of headache but we found this risk acceptable considering the overall clinical situation.[11]

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