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Case Report

Challenges with Takayasu arteritis in pregnancy: a case report

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ABSTRACT

Takayasu's arteritis (TA) is a rare, chronic, inflammatory, large vessel vasculitis (LVV) of unknown etiology characterized by narrowing, occlusion, and aneurysms of systemic and pulmonary arteries affecting especially the aorta and its branches. Pregnant patients are at increased risk of cardiovascular complications, including hypertension and congestive heart failure, which may jeopardize both maternal and fetal outcomes, so one should pay special attention to these patients. We present a case of 30-year-old female G5P3L2A1 with 9 months of amenorrhea with previous 2 LSCS with chronic hypertension and superimposed pre-eclampsia with oligohydraminos. She was a known case of Takayasu arteritis, diagnosed since she was 16 years old. Her lower limb pulses were palpable with absent upper limb pulses. On Echocardiography there was dilated aortic root, moderate MR and LVEF was 55%. She proceeded to have emergency LSCS under spinal anesthesia in view of severe pre-eclampsia with previous 2 LSCS at 35 week and delivered alive healthy female baby weight 2.1 kg. A life-threatening maternal cardiovascular complication is observed in more than 5% of pregnant women with TA. Hypertension is present in 90% cases of Takayasu arteritis. A good control of TA disease activity and arterial hypertension before conception and during pregnancy is important to improve feto-maternal outcomes. Pregnancies in the setting of TA should be considered high-risk, requiring a close interdisciplinary collaboration of rheumatologists, nephrologists and obstetrician's specialists involved in care of TA.

Keywords: Takayasu arteritis, Pregnancy, Pre-eclampsia, Pulseless disease

INTRODUCTION

Takayasu's arteritis (TA) is a rare, chronic, inflammatory, large vessel vasculitis (LVV) of unknown etiology characterized by narrowing, occlusion, and aneurysms of systemic and pulmonary arteries affecting especially the aorta and its branches in a progressive pattern, which can lead to secondary hypertension, retinopathy, cardiac pathology, stroke, and death at an early age.[1,2] It is also known as young female arteritis/pulseless disease/ aortoarteritis mainly affecting women of childbearing age.[3] The diagnosis is based on the combination of clinical history, physical examination, clinical suspicion, and vascular imaging techniques.[2]

Pregnant patients are at increased risk of cardiovascular complications, including hypertension and congestive heart failure, which may jeopardize both maternal and fetal outcomes, so one should pay special attention to these patients. Maternal and fetal prognosis can be improved by multidisciplinary collaboration of obstetricians, cardiologists, and neurologists.

CASE REPORT

We are reporting a case of 30-year-old female G5P3L2A1, a known case of Takayasu arteritis with 9 months of amenorrhea with previous 2 LSCS with chronic hypertension and superimposed pre-eclampsia

with oligohydraminos. The diagnosis of TA was established when patient developed sudden weakness of upper limbs at 16 year of age. On investigation, there was a right front parietal infarct on computed tomography (CT) and complete occlusion at origin of both subclavian and carotids with reformation of collaterals on digital subtraction angiography thus corroborating with diagnosis of TA. She was taking oral prednisolone and low dose aspirin, but prednisolone was stopped after conception, as she was in remission phase.

Her past obstetrical history revealed first pregnancy complicated by pre-eclampsia and intrauterine fetal death at 8 month of gestation, 12 years back. Eleven year back, second pregnancy was managed by lower segment cesarean section (LSCS) in view of intrauterine growth restriction of baby at 36 weeks of gestation. Third pregnancy was complicated by severe oligohydramnios and cesarean was done at 36 week of pregnancy, 9 year back. On examination, upper limb pulses were not palpable, bilateral carotids were weak and her lower limb pulses were normal. Blood pressure was monitored on thigh using popliteal pulsations. She was on tab labetalol 200 mg 12 hourly for chronic hypertension. She was followed up in antenatal OPD every fortnightly, along with cardiologist and rheumatologist. All the routine investigations were within normal limit. Electrocardiogram (ECG) showed mild left ventricular hypertrophy. Fundus examination was normal. Echocardiogram revealed dilated aortic root, moderate MR, moderate aortic regurgitation, and LVEF was 55%.

She was admitted at 32 weeks of pregnancy in view of superimposed preeclampsia on chronic hypertension. On admission her BP was 180/90mm Hg in lower limb. Her antihypertensives were increased progressively to tab labetalol 200 mg 6 hourly and tab nifedipine 10 mg 6 hourly. Her obstetric ultrasound showed a single live fetus with oligohydramnios (AFI-4 cm). Her Doppler velocimetry was normal. She proceeded to have emergency LSCS under spinal anesthesia in view of severe preeclampsia with prev 2 LSCS at 35 week and delivered a live healthy female baby with weight of 2.1 kg. The patient remained hemodynamically stable and was discharged uneventfully.

DISCUSSION

Takayasu's arteritis is a rare form of giant cell vasculitis involving inflammation in the walls of the large arteries: the aorta and its main branches, coronary and pulmonary arteries.[4] There is narrowing of the arteries due to inflammation, and this can reduce blood flow to many parts of the body. It was named after Mikito Takayasu, a Japanese ophthalmologist.[5] He first reported a case of 21 years old female, whose eye grounds exhibited coronary anastomosis, arteriovenous anastomosis around the papilla due to ischemia of cerebrovascular circulation.

Takayasu's arteritis can lead to weak pulse or loss of pulse in arms, legs and organs. For this reason, it is also known as "pulseless disease." The disease progression occurs in a triphasic pattern.[6] Initial phase is the systemic or pre-pulseless period, characterized by constitutional symptoms such as low-grade fever, night sweats, malaise, arthralgia, anorexia and weight loss. This progresses to second phase which is vasculitis stage where constitutional symptoms are associated with features of vascular involvement like tenderness or pain over vessels (angiodynia). The final phase results into late, fibrotic, occlusive, quiescent, or "burnt-out" phase and characteristic features of TA related to arterial occlusion or stenosis appears. Only a few patients have such a temporal progression. The acute phases of the disease remit spontaneously in about 3 months or may progress insidiously for months to years into the chronic phase.

Takayasu arteritis causes different clinical conditions depending on the sites of constriction such as arm claudication, decreased arterial pulses, visual loss, stroke.[7] Hypertension is present in 90% cases of Takayasu arteritis.[8]

According to the American college of rheumatology 1990 criteria, three out of 6 criteria must be fulfilled for the diagnosis of Takayasu arteritis.[9] 1. Age under 40 at disease onset, 2. Claudication of extremities, 3. Decreased brachial arterial pulses, 4. Systolic blood pressure difference of more than 10 mmHg, between arms, 5. Bruit over subclavian arteries or the aorta, 6. Angiogram abnormalities: occlusion or narrowing of entire aorta, its primary branches or large arteries in the proximal upper or lower extremities.

If these criteria are satisfied, there is 90.5% sensitivity and 97.8% specificity.

Sometimes patients with Takayasu arteritis may have no symptoms and detected for the first time in pregnancy. Like other rheumatologic diseases it may remain asymptomatic until pregnancy, when it may become more aggressive, putting both the mother and baby at risk. TA should be kept as an important differential diagnosis for severe hypertension in pregnancy. Physical examination is usually sufficient to alert to the presence of this disease. Headaches, a pulselessness of unilateral or both radial arteries, a blood pressure discrepancy, vascular bruit or limb weakness should be looked at in all cases of severe hypertension.[10,11]

In a diagnosed patient of Takayasu arteritis, a good control of disease activity and arterial hypertension before conception and during pregnancy is helpful to improve the outcome of both mother and foetus. Pregnancy does not exacerbate the disease; but TA has several adverse implications on pregnancy like abortions, sustained refractory hypertension, superimposed PE, IUGR, oligohydraminos, abruption, IUD, congestive heart failure, and progression of renal involvement. The

four most important complications during pregnancy are Takayasu's retinopathy, secondary hypertension, aortic regurgitation, and aneurysm formation.[3] Acute inflammation destroys the arterial media and lead to aneurysm formation.[12]

Baseline investigations should include full blood count, renal function tests and C-reactive protein. Doppler ultrasound and magnetic resonance imaging (MRI) of carotids and other major vessels involved should be performed to assess the severity of disease and echocardiogram to assess the ejection outflow. Uterine artery scans at 24 weeks of gestation can provide a prediction for uteroplacental insufficient. Pregnancy can be continued till term and induction of labor can be considered in the presence of superimposed pre-eclampsia or hypertension. Elective LSCS is indicated for obstetric indication and severe disease (retinopathy, arterial aneurysm and aortic regurgitation).[13] Second stage of labor can be shortened by low forceps delivery or vacuum extraction to reduce the risk for cerebral hemorrhage.[14]

In our case, the main problem was severe refractory hypertension. Management was a real challenge, which makes a multidisciplinary approach necessary. Increases in blood pressure can cause rupture of the aneurysms, aortic dissection. However, the control of blood pressure is not always achieved easily because of difficult brachial BP measurements with pulseless upper extremities.[11] In case, despite aggressive medical treatment, patient's systolic BP levels reached 200 mmHg and decided to terminate the pregnancy by cesarean section to avoid further feto-maternal morbidity and mortality.

Careful anesthetic surgical planning is fundamental. Perfusion maintenance is the main concern for these patients. In patients with compensated TA complications, monitoring remains same as of routine cesarean sections.[15] Patients should be monitored in an intensive or semi-intensive care unit for 24 hours to avoid postoperative hypoperfusion or hypertensive complications.

Drug therapy needs a careful assessment of the risk/benefit ratio for the patient. Low dose steroid (usually prednisolone) is the mainstay of treatment during pregnancy and if she is in remission, cytotoxic drugs can be withdrawn as in our case. Careful BP monitoring, medication and regular obstetric USG with fetal Doppler improves feto-maternal outcome.[4]

CONCLUSION

A life-threatening maternal cardiovascular complication is observed in more than 5% of pregnant women with TA. A good control of TA disease activity and arterial hypertension before conception and during pregnancy is important to improve feto-maternal outcomes. Our experience indicates that BP levels in TA and pregnancy play an important role in feto-maternal outcomes. Pregnancies in the setting of TA should be considered high-risk, requiring a close interdisciplinary collaboration of rheumatologists, nephrologists and obstetrician's specialists involved in the care of TA.

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