CASE REPORT

Undiagnosed Takayasu Arteritis in Pregnancy

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ABSTRACT

Takayasu arteritis is a chronic inflammation involving large vessels and it often occurs in young women of childbearing age. We described a case of a 29- year- old lady with previous history of proliferative ischemic retinopathy was noted to have low upper limbs blood pressure and weak upper limb pulses postpartumly. An urgent CT angiogram of thorax revealed features suggestive of large vessel vasculitis with involvement of ascending arch, descending aorta and its main branches, corresponding to type II TA . She was diagnosed to have Takayasu arteritis post delivery, and she underwent a successful pregnancy without intrapartum and postpartum complications. High index of suspicion must be given for pregnant patient who have persistent low blood pressure and weak pulse for early detection to avoid severe complications.
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INTRODUCTION

Takayasu arteritis (TA) is a chronic, inflammatory, progressive, idiopathic disease that causes narrowing, occlusion, and aneurysms of systemic and pulmonary arteries affecting mainly the aorta and its branches (1). It occurs in the women of childbearing age, with the mean age of second and third decade of life with greater prevalence in Asian women. It is more common in woman than man (8:1), and the peak incidence is in the second and third decades (2). Although TA affects women of childbearing age, literature on pregnancy is lacking. We reported a case of an undiagnosed TA who survived pregnancy and delivered a healthy baby successfully.

CASE REPORT

A 29 –year- old primigravida lady at 39 weeks of gestation was electively admitted for induction of labor in view of intrauterine growth restriction with oligohydramios. She was diagnosed with proliferative ischemic retinopathy since 2014 under ophthalmology follow up and was started on tapering dose of prednisolone. Further questioning, she had history of hospital admission 3 years ago for frequent syncopal attack which lasted for 3 minutes and regained consciousness spontaneous. She denied any joint pain, mouth ulcers or any other connective tissue disease symptoms. She has no history of thrombosis in the past. Autoimmune worked out were all negatives. She was discharged with impression of proliferative ischemic retinopathy without systemic involvement. Her antenatal followed up revealed persistent low blood pressure. Her lowest documented blood pressure was 54/30mmHg. Despite this, she remained asymptomatic.

On arrival to the antenatal ward, she appeared well and not in respiratory distress. She was afebrile. Her blood pressure was 90/60 mmHg, tachycardic with pulse of 110 bpm. There was no skin rashes noted. Cardiorespiratory systems were unremarkable. Per-abdomen examination revealed a fundal height corresponding to 39 weeks with audible fetal heart sound.

Initial full blood counts, renal and liver function tests were all normal. Urinalysis revealed no proteinuria or RBC. ESR 8 mm/hr and CRP 0.7 mg/L. Repeated screening of antinuclear antibodies (ANA), double stranded DNA (dsDNA), c-ANCA and p-ANCA were negatives. Lupus anticoagulant, anticardiolipin IgM/IgG, anti beta2-glycoprotein and infective screening were also negatives. Her echocardiogram revealed normal ejection fraction, normal chamber size with normal valves and regional wall motion.

She was later induced with prostin. However, her fetus developed fetal distress by showing pathological type II deceleration in CTG. Emergency caesarean section was performed. Intraoperatively, she was noted to have low upper limb systolic blood pressure ranging 70-110mmHg, she was given fluid resuscitate and multiple boluses of IV Phenylephrine and ephedrine. There was no massive blood loss nor rapid drop in haemoglobin. Estimated blood loss was about 400mls. The case was referred to medical team for hypotension.

Upon medical review, there was a huge discrepancy in blood pressure between upper limb and lower limb associated with weak upper limb peripheral pulses. Left carotid bruit was present. An urgent CT angiogram of thorax revealed features suggestive of large vessel vasculitis with involvement of ascending arch, descending aorta and its main branches, corresponding to type II TA (Figure 1 and 2).

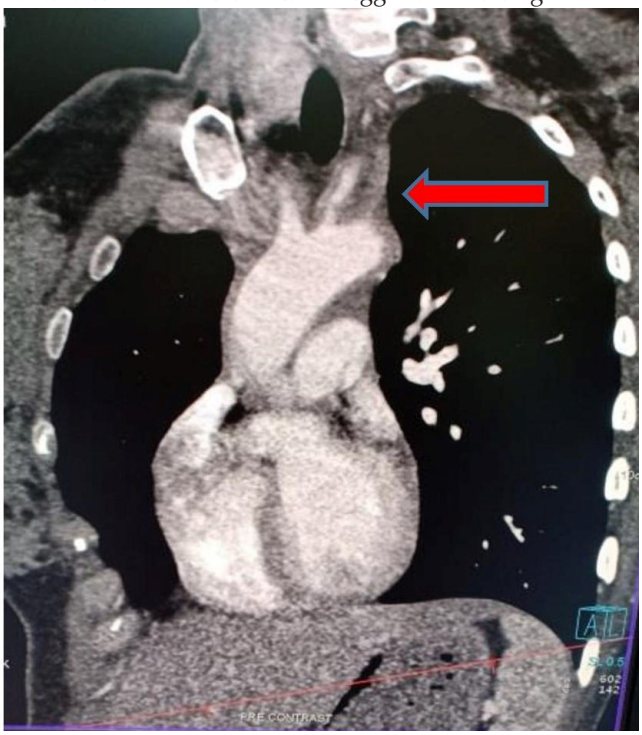

Figure 1: CT angiogram showed severe circumferential arterial wall thickening resultant in total occlusion of left subclavian artery (arrow)

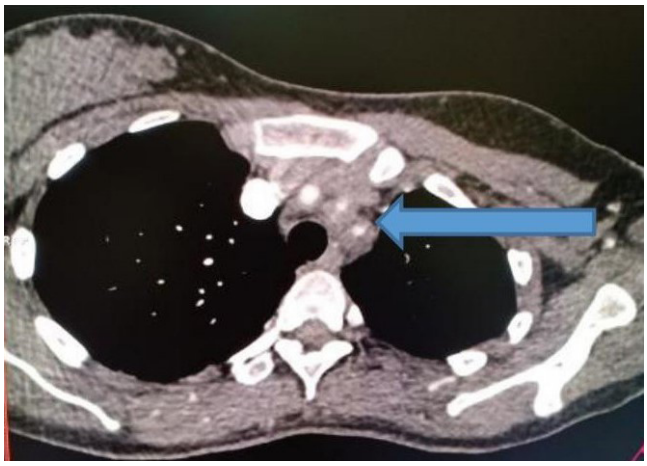

Figure 2: Transverse view of thoracic aorta at level of arch of aorta reviewed that there is filling defect mainly seen in the left subclavian artery (arrow)

Her labour progressed well and she delivered live boy girl weighing 2.02 kgs with Apgar score of 9/10. Her intrapartum and postpartum period were uneventful. There was no active intervention from cardiothoracic team. She was started on high dose steroid 1mg/kg, and discharged well. She was currently well on tapering dose of prednisolone and Azathioprine 150 mg daily. Azathioprine was chosen instead of Methotrexate / Mycophenolate Mofetil as steroid sparing in this case because the patient was on-going breastfeeding. She was planned for PET scan once she stopped breastfeeding.

DISCUSSION

Takayasu arteritis, is an uncommon, constantly recurring vasculitis that frequently acts on female of fertility age. This 'pulseless disease' is distinguished by inflammation of the abdominal and thoracic aorta namely renal, subclavian, vertebral, carotid and brachiocephalic arteries. It may also involves the pulmonary as well as coronary arteries (2). It is an autoimmune disease with unknown aetiology.

This autoimmune disease typically presented in the young adult group, mainly before the age of 40. Recent ACR / EULAR 2022 Classification Criteria for Takayasu Arteritis (TAK) has increased the cut off age from < 40 to < 60 years old (3). In between symptoms of onset and diagnosis is often detained (2). Though TA typically affects young female in the fertility age group, the research review pertaining this disease is still limited. Disease remission is utmost important during pregnancy, in order to ensure good fetal and maternal outcome. Fertility or pregnancy itself are not affected by TA. In a retrospective study reported by Hidaka et al. 24 patients out of 26 pregnancies revealed newborns of normal growth and development (70%) (4). As T2 helper cell cytokine centralized at the fetomaternal surface during pregnancy, this will enhanced improvement in symptoms and disease activity in TA (2). Lower CRP and ESR also suggested reduced inflammatory activity and clinical stability. Such finding was similar to our patient. Despite improvement of symptoms during pregnancy, there are increased risks of cardiovascular complexity in gravid females namely high blood pressure, congestive heart failure, aortic regurgitation and myocardial infarction (2). It is vital to monitor blood pressure closely in pregnant and non-pregnant TA patients, to avoid episodes of hypo – hypertension. The most common complication in gravid females is hypertension, as this can lead to cerebral haemorrhage in both mother and fetus.

Although pregnancy does not interlobe with disease sequence, it has a few negative impacts such as

miscarriages, preeclampsia, intrauterine growth restriction (IUGR), intrauterine death and abruption (5). Our patient presented with IUGR and oligohydramnios, hence admitted for induction of labour. Aetiology of IUGR may be due to impaired blood flow to placenta. (4)Vascular damage on the maternal side restricting fetal nutrition but not manifesting itself in placental histology could also played a role. Pregnancy induced hypertension in this case is still a possible cause of IUGR. Blood pressure in TA patient must be measured at both upper and lower limbs to detect discrepancy in BP recordings. As in our case BP recordings is higher at lower limb than upper limb.

There was no established optimal management for pregnant patients with TA.Glucocorticoids is known to suppress or reduced systemic inflammation of the disease (5). However, half of the patients are irresponsive to steroidal treatment and need steroid sparing agents such as methotrexate or azathioprine (5). As reported by Hoffman et al, initial remission was demonstrated in 81% of patients treated with methotrexate and steroids, however relapsed was observed in more than half within 34 months (5). Methotrexate is contraindicated in pregnancy, thus posed a clinical challenge to treating physician.

Our patient had persistent hypotension during her antenatal follow up. Since she remained asymptomatic, there was no suspicion of TA at all. It is important to note that this might not be her actual 'hypertension' at all as there was no signs of chamber enlargement, which might suggested that her systemic blood pressure was not elevated.

The course of disease seems to be neither affected nor worsened by pregnancy. So far, there was no reported maternal deaths directly related to pregnancy. Planning for another pregnancy requires early interdisciplinary collaboration of rheumatologists, cardiologists and obstetricians as to ensure a good maternal and foetal prognosis. Controlling all risk factors especially BP should be aggressive from early pregnancy. Prophylaxis antenatal low dose aspirin as well as elective delivery of the baby might also be considered.

Lack of compassionate in the knowledge of TA can have destructive outcomes for both parties. Physician should be aware of the clinical signs and symptoms, diagnosis, and further care of TA patients. Multidisciplinary approach is essential to ensure good outcome in both baby and the mother. To our best of knowledge, this was the first case reported of undiagnosed TA in a pregnant lady presented with persistent hypotension who went through a successful delivery without any intrapartum complications.

CONCLUSION

Although Takayasu is uncommon disease, it commonly affects woman of childbearing age. Due to its rarity, the full impact on pregnancy is not well described. They can be presented with either hypotension or hypertension, however, it is important to note that peripheral blood pressures may not be accurate. A single brachial blood pressure measurement is inadequate because of frequent subclavian involvement, therefore additional ankle blood pressure measurement in Takayasu arteritis is recommended. The initial diagnosis of ocular ischemia in our patient should prompt the clinician for further physical examination of peripheral pulses, hence the diagnosis of TA. Further investigations for signs of uncontrolled systemic hypertension should be obtained including echocardiography and echocardiogram. Optimal direction of care in patients with Takayasu Arteritis is not always clear but should include managing inflammation and monitoring for the vascular complications of arterial narrowing and hypertension/ pre-eclampsia. A fraternization of all subspeciality namely neurologists, rheumatologists, cardiologist and obstetricians is important for a maximum benefit in the prognosis of fetal and maternal.

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