

Case Report

Pregnancy with active Takayasu's arteritis: A medical challenge

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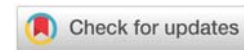
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Abstract

Takayasu arteritis is a large vessel vasculitis, and very rare in occurrence with pregnancy. It usually complicates the latter half of pregnancy and causes hypertension, organ dysfunction, fetal growth restriction. Its management during pregnancy is a medical challenge.

Here we present a case of 25-year old primigravida presented to us at 30 weeks with Takayasu arteritis and its successful management with good fetomaternal outcome.

Introduction

Takayasu arteritis (TA) is a large vessel vasculitis which was first described by the Japanese ophthalmologist Mikito Takayasu and Onishi [1]. This rare chronic inflammatory progressive vasculitis is also known as pulseless disease [2]. It mainly affects young women of childbearing age usually distributed in Southeast Asia, areas of Africa, and Southern America [3]. The average reported incidence is 13 cases per million populations [2,4].

The effect of TA on pregnancy and effect of pregnancy on TA is under study due to its rare presentation. Its peak incidence during pregnancy is seen in the second, third trimester, and during labor. Many a time the female is diagnosed to have TA during pregnancy only due to its complications. Common complications associated with TA in pregnancy are

hypertension, organ dysfunction, restricted intrauterine fetal growth, and low birth weight [2-5]. Here we discuss the management of a pregnant female with active TA, with limited published literature on same.

Case history

A 25-year-old 30 weeks primigravida was referred to obstetrics department for uncontrolled hypertension (BP 190/60mmHg) and left-sided chest pain, for which she was on labetalol (since 10 days). She had diagnosed case of TA 3 years back during the evaluation of intermittent pain in the left hands and neck. No history of hypertension or any neurological symptoms at that time. She was started on oral steroids, but she stopped taking medicine on her own after 1 year of diagnosis.

Examination

The detailed examination revealed wide pulse pressure

and differential blood pressure in both upper limbs with lower pressure in left arm. She also had bruits in the left subclavian and left common carotid artery region.

Investigation

On colour doppler examination there was approximately 80%–90% luminal narrowing of proximal left subclavian and 70% narrowing of the proximal left common carotid artery with reduction of blood flow velocity in distal part. The right sided subclavian and common carotid artery showed normal flow. At the same time reduced flow velocity and increased acceleration time noted in left side renal artery at hilum and polar branches suggestive of left renal artery stenosis. The right renal artery flow is normal. Her recent ESR was 52mm and CRP was 18.02 mg/Lt. She was diagnosed as a case of active TA. Disease activity of TA is quantified by an assessment tool called Indian Takayasu's Arteritis Activity Score (ITAS) 2010 scoring. She had 9 scores on Indian Takayasu's Arteritis Activity Score (ITAS) 2010 scoring on the basis of her renal involvement, hypertension, presence of bruits, pulse inequality.

Management

She was started on labetalol 200 mg and oral corticosteroids (tablet prednisolone 50 mg) twice daily in view of active disease. The low dose steroid therapy was continued as she responded well to this dose also her pre pregnancy weight was very low so calculated per day dose was less. After workup and stabilization of blood pressure, the patient was discharged and was supervised thereafter. At 38+1 weeks she was admitted again with labor pain. She was in stage II b (of complication to interpret management in labor) at the time of delivery. Injectable hydrocortisone was given in labor and trial of labor was given. The patient was taken for the cesarean section due to the arrest of cervical dilatation and she delivered a healthy baby of 3kg with a good Apgar score (9/9).

Follow up

She had raised BP records in the postpartum period also and was switched on oral steroids and antihypertensive.

Discussion

Takayasu arteritis is a disease of unknown pathology which is more common in females with a ratio of 1:4 [3]. There are hormonal, genetic, infective, immune-mediated, and other mechanisms to explain the pathophysiology of this disease [5,6]. One of the theories suggests association and causation with hormones like estrogen and progesterone which explains its incidence in the young female population [6].

The presenting complaints of the patients are usually fatigue, myalgia, arthralgia, low-grade fever, claudication, and visual defects. On examination one may find feeble or absent pulse on one side of the body, blood pressure variation in both the limbs, bruit on the affected side. Its diagnosis is made by clinical manifestations, high levels of acute-phase reactants (ESR & CRP), and radiological demonstration of vessel stenosis. The ITAS 2010 score is based on these above-mentioned criteria simplifies its severity assessment. The imaging modalities

include color doppler, Computerized Tomography (CT), magnetic resonance (MR) angiography, and PET scan.⁷ The main role of imaging is to look for severity of arterial narrowing and activity of disease. During pregnancy CRP and doppler are the helpful investigations as ESR normally increases during pregnancy. The CT scan is associated with radiation exposure and contrast use is not safe during pregnancy. The gold standard for diagnosis is biopsy [7].

Our patient was diagnosed as having an active TA based on ITAS 2010 score system and biopsy was planned for a later stage due to pregnancy. The disease stages as per the presence of complications are [8] stage I (no complications), stage IIa (patients having only one of the complications like hypertension, retinopathy, aneurysms, and aortic insufficiency, stage IIb (patients with only one of the complications, but the severe form), and stage III (more than one complication is present). Most of the cases described in the literature were well-supervised throughout pregnancy and were stable. This patient with unsupervised pregnancy presented directly during 3rd trimester in stage II.

The management of TA is aimed to control active phase inflammation, prevention, and treatment of complications and revascularization of vessels surgically by the interdisciplinary team. Antenatal check-up includes serial BP monitoring, renal and cardiac functions along with routine antenatal care. Fetal surveillance is also imperative with daily fetal movement count, gravidogram, biometry, and doppler as per need [4,9].

Corticosteroids are the drug of choice in the dose of 0.75–1mg/kg (pre-pregnancy weight)/day in active phase for 4 weeks followed by tapering down by 5–10% every 2 weeks to 5–10mg/day for long term. Other drugs like antihypertensives, antiplatelets, immune suppressors, and immune modulators are added as per need after risk/benefit assessment [4,9].

Vaginal delivery with epidural analgesia is the preferred mode of delivery in stage I, II. In the patients with higher stage, cesarean section is preferred to prevent cardiac decompensation during labor. During labor oral steroids are replaced by injectable hydrocortisone 200mg/day for 3 days. Our patient belonged to group IIa and was hemodynamically stable so a trial of labor was given to her. An emergency cesarean section was taken for obstetric indication. Infection and fluid overload should be avoided in the postpartum period. As the postpartum period is a hypercoagulable state thromboprophylaxis with low molecular weight heparin can be considered in patients judiciously.

In conclusion, even in the modern era management of active disease in pregnancy is still challenging. Medication for the treatment of TA should be given judiciously with risk-benefit analysis. Close monitoring of disease progression by the multidisciplinary team and feto-maternal surveillance along with termination of pregnancy at appropriate time helps to give good maternal and fetal outcomes.

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