Takayasu’s Arteritis and Pregnancy

ABSTRACT

Introduction: Takayasu’s arteritis (TA) is a rare clinical entity characterized by the progressive obliteration of the aortic arch and the main vessels arising from it and, in some cases of the thoracic and abdominal aorta and its main branches.

Case: We report a case in which diagnosis of TA was made 6 years ago and present pregnancy was successfully managed.

Conclusion: Multidisciplinary management is essential for satisfactory clinical outcome during pregnancy and their blood pressure should be strictly controlled for a favorable maternal and fetal outcome and mode of delivery should be planned.

Keywords: Arteritis, Intrauterine growth retardation, Oligohydramnios.


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INTRODUCTION

Takayasu’s arteritis (TA) is a rare clinical entity characterized by the progressive obliteration of the aortic arch and the main vessels arising from it and, in some cases of the thoracic and abdominal aorta and its main branches. The basic disease appears to be unaffected by pregnancy, inflammatory activity and hemodynamic status improves with pregnancy. We report a case of Takayasu’s arteritis with pregnancy which was successfully managed.

CASE REPORT

A 25 years old female known case of TA presented to obstetrics and gynecology outpatient department (OPD) at 26 weeks gestation. The diagnosis of TA was established 6 years back when patient developed sudden hemiparesis. Computed tomography (CT) head revealed right front parietal infarct. Digital subtraction angiography revealed complete occlusion at origin of both subclavian and carotids with reformation of collaterals thus corroborating with diagnosis of TA. She was put on oral steroids and low dose aspirin. Her past obstetrical history revealed first pregnancy complicated by pre-eclampsia and intrauterine fetal death at term 5 years back. Second pregnancy was complicated by intrauterine growth restriction of baby so, it was managed by lower segment cesarean section (LSCS) 3 years back at 36 weeks of gestation. In present pregnancy, patient had presented at 26 weeks of gestation. At her first antenatal visit, patient was well aware of her medical status, counseling regarding the complications and prognosis was done. Patient was totally asymptomatic. On examination, upper limb pulses were not palpable, bilateral carotids were weak and her lower limb pulses were normal so blood pressure was monitored on thigh using popliteal pulsations. Her lower limbs blood pressure was 118/78 mm Hg. All the routine investigations were within normal limit. Electrocardiogram (ECG) showed mild left ventricular hypertrophy. Fundus examination was normal. Echocardiogram revealed moderate aortic regurgitation with normal left ventricular dysfunction. She was followed up in antenatal OPD every fortnightly, along with cardiologist and rheumatologist. Her blood pressure was normal throughout the pregnancy. Pregnancy progressed well under supervision until 36 weeks of gestation when severe oligohydramnios was diagnosed on sonography. Lower segment cesarean section was planned at 36 weeks period of gestation under epidural anesthesia. She delivered a healthy female baby weighing 2.6 kg. There were no intra and postoperative complications. Patient was discharged on 7th postoperative day.

DISCUSSION

Takayasu’s arteritis (TA) is a primary arteritis of unknown cause, which occurs in the women of childbearing age group. The mean age of presentation of TA is usually the second and third decade of life with a greater prevalence in Asian women, it occurs sporadically throughout the world. It is more common in women than in men (8:1).
Takayasu’s disease was first described in 1908 by two Japanese ophthalmologists Takayasu and Onishi, who observed retinopathy occurring with absent limb pulses. Nowadays, it is recognized as a rare (2–3 per million) inflammatory disease of the vascular tree, principally affecting major vessels, such as the aorta, its main branches and the pulmonary arteries. The disease preferentially involves the elastic arteries including the aorta and its major branches. Typically histopathological changes consist of disruption of the elastic fibers of the vessel wall, inflammatory cell infiltration with occasional granulomatous reaction, fibrotic thickening of the adventitia and intimal thickening.

The disease is characterized classically by a triphasic course, commencing with an initial active phase of constitutional symptoms, such as malaise, weight loss, fever, myalgias and arthralgias, and is associated with an increase in acute phase reactants. This progresses to a second stage with symptoms of cerebral, visceral or extremity ischemia before resulting in a final burnt-out quiescent phase of fibrosis. Unfortunately, these typical symptoms are seen only in 33% of patients and severity and progression of vessel involvement are extremely variable.

Fertility is not adversely affected and pregnancy does not appear to exacerbate the disease, inflammatory activity and the hemodynamic status may improve with pregnancy although development of blood pressure is not uncommon in pregnancy. However, pregnancy should be considered in remission period. Baseline investigations should be taken as part of the evaluation and should include full blood count, C-reactive protein and renal function tests. Apart from baseline investigations 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 tract around 28 weeks gestation.

A multidisciplinary team that includes an obstetrician, midwife, radiologist, hematologist, anesthetist, cardiologist and rheumatologist should provide antenatal care. All the women should be booked as early as possible. Maternal complications are superimposed preeclampsia, congestive cardiac failure; cerebral hemorrhage, progressive renal impairment and fetal complications include growth restriction.

All the women should receive regular growth scans. Uterine artery scans performed at 20 and 24 weeks of gestation provide a predictor for uteroplacental function and help to identify women with high risk pregnancy.

Pregnancy can continue till term and induction of labor is considered in the presence of superimposed pre-eclampsia or hypertension. Elective LSCS is indicated for severe disease (retinopathy, arterial aneurysm and aortic regurgitation). Hypertension in the second stage of labor is a risk factor for cerebral hemorrhage; shortening this stage by low forces delivery or vacuum extraction appears to be a reasonable solution.

The anesthetic management of a parturient suffering from this arteritis is important involvement of anesthesiologist at an early gestation to make plan depending upon needs and risks involved regarding the regional anesthesia.

Drug therapy requires a careful assessment of the risk/benefit ratio for the patient. Low dose steroid (usually prednisolone) is often the mainstay of treatment during pregnancy and if she is in remission, cytotoxic drugs are withdrawn. In event of blood pressure, antihypertensive should be started at an early gestation for better maternal and fetal outcome.

CONCLUSION

Fertility is not adversely affected and pregnancy does not appear to exacerbate the disease, TA should have a medical screening prior to conception. Medical management of a pregnant patient with TA does not differ significantly from nonpregnant patient. Multidisciplinary management is essential for satisfactory clinical outcome during pregnancy and their blood pressure should be strictly controlled for a favorable maternal and fetal outcome and mode of delivery should be planned.

REFERENCES