

Pregnancy with Takayasu's Arteritis

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Abstract

Takayasu's Arteritis is more common in women than men (8:1) with the peak incidence in the second and third decades, although a minority may present in their teens. The exact etiology remains unknown but it may have an autoimmune basis. It affects the aorta and its branches, leading to stenosis, thrombosis and the formation of aneurysms. During pregnancy it is associated with a number of fetal and maternal complications. The mode of delivery in patients with TA is determined by the haemodynamic status of the mother. A multidisciplinary collaboration of rheumatologists, cardiologist along with obstetricians is necessary to improve maternal and fetal outcome.

Keywords: Takayasu's Arteritis, Fetal maternal complications, Mode of delivery

Introduction

Takayasu's arteritis (TA) is an inflammatory condition that affects medium and large size arteries, with an annual incidence of 1.2–2.6 cases per million.⁽¹⁾ It is not uncommon to encounter this disease during pregnancy. Although the inflammatory activity of the disease is thought to be unaffected by pregnancy,⁽²⁾ vascular complications such as gestational hypertension, heart failure, cerebro-vascular accidents, intrauterine growth restriction (IUGR), intrauterine fetal demise,⁽³⁾ anesthetic complications like hypotension and rupture of aneurysm can occur. It can precipitate cerebral hemorrhage and cardiac dysfunction.

It has been reported that survival rates are 80% to 90%.⁽⁴⁾ Poor outcome depends chiefly on the early onset of the disease and presence of complications like hypertension, aortic regurgitation, and aneurysm, and a rapidly progressive course.

Here we are discussing few cases of TA with pregnancy reported and managed in a tertiary care center of South India.

Case 1

Mrs. N 37yr old lady presented with history of giddiness and left arm tingling, intermittent claudication and discrepancy of BPs since 2011. Her right upper limb Blood pressure (BP) was 156/80 mm Hg and Left Upper limb BP was 140/70mm Hg. Coronary and Peripheral angiogram showed left anterior descending artery had a distal cork screw appearance and was occluded further distally. Left subclavian artery was occluded at the origin; vessel beyond occlusion was seen filling by collaterals from vertebral artery. Peripheral angioplasty with recanalization with stenting of left subclavian artery and superior mesenteric artery was done on 2014. She was started on oral antihypertensives and steroids. She presented to antenatal clinic at 21 weeks of gestational age with chronic hypertension. BP was optimized with

dose adjustment of antihypertensive medication. It was decided that BP measurement and drug modulation would be done based on the systolic and diastolic pressure measurement in the right upper limb. She was monitored in the antenatal period by several departments including Obstetrics, Medicine, Rheumatology and Cardiology. Her antenatal routine investigations and hypertensive workup were normal. At 36 weeks 3 days gestation, she delivered a girl baby of 3 kgs. Intra operative period was uneventful. Post operatively she was stable and her BP was within normal limits. She was discharged well.

Case 2

Mrs. NJ a 30 year old lady, G2P1L1 at 34 weeks with chronic hypertension, IUGR and TA presented with premature rupture of membranes. Induction of Labour was done in view of raised C - reactive protein and risk of chorioamnionitis. Her right upper limb BP is 150/80 mm Hg and left upper limb BP is 100/80 mm Hg. She had a preterm vaginal delivery. She was on antihypertensives, steroids and immunomodulators. Her postnatal period was uneventful. During post natal period her BP was within normal limits with drugs.

Case 3

Mrs. V 26 year lady, primigravida at 37 weeks gestation with chronic hypertension on antihypertensive presented to the labour ward. Her BP in right upper limb was 136/100mm Hg and left upper limb was 170/110mm Hg. In view of discrepancy in BP she was evaluated for TA, Doppler studies showed moderate left hydronephrosis, abdominal aortic ectasia with irregular stenosis at diaphragmatic level with bilateral renal artery stenosis (right>>left). Echocardiogram was also done and showed Left ventricular hypertrophy. She had spontaneous onset of labor and delivered a healthy 2.58kg baby by caesarean section for non reassuring fetal status. Post-operative period was uneventful and

her BP was under control with drugs. Rheumatology and cardiology consult was sought and planned for stenting.

Case 4

Mrs. K 25 year old lady, primigravida at 36 weeks gestation with TA with chronic hypertension presented with pre term labour and delivered a by Caesarean section which was done for breech presentation. Post natively her BP was normal and discharged well and planned to continue steroids and immunomodulators. Prior to pregnancy she had multiple episodes of hospital admission for both limb claudication, her right upper limb BP was 170/96 mm Hg and left upper limb BP was 90/60 mm Hg, she was investigated and TA was confirmed. Later she underwent stenting of left subclavian artery and aorta and balloon angioplasty for right external iliac artery and common femoral artery.

Case 5

Mrs. P 26 year old lady, G2A1 at 37 weeks of gestation with TA and chronic hypertension. On examination BP on right upper limb was 180/40 mm Hg and left upper limb BP was not recordable, both lower limbs BP was 190/70mm Hg. Prior to pregnancy she was admitted with syncopal attacks, giddiness and claudication of limbs for which she had angioplasty and stenting of left renal artery, recanalization of left subclavian artery and stenting, angioplasty of left common carotid artery and Plain old balloon angioplasty to instent restenosis of left renal artery was done. She was under regular follow up with rheumatologist and cardiologist for aortic regurgitation. She had regular antenatal checkup. At 37 weeks of gestation she had elective caesarean, both mother and baby were well after surgery. She was monitored in high dependency unit for post operative monitoring.

Case 6

Mrs. R 25 year old lady, G2A1 at 39 weeks and 2 days with TA with moderate to severe aortic regurgitation. Four years back she was evaluated for TA. Her right upper limb BP was 100/50 mm Hg and left upper limb BP was not recordable, her right lower limb BP was 140 systolic and left lower limb was 146mm Hg. Her angiogram showed severe aortic regurgitation, dilated ascending aorta and occlusion of the right common carotid artery, aneurismal disease, stenotic and aneurismal disease of left common carotid artery, distal left subclavian artery occlusion, distal occlusion of the right subclavian artery and left renal artery stenosis. Stenting of left common carotid artery aneurysm and angioplasty with stenting of the left renal artery was done. Her regular antenatal checkup was normal and had an elective caesarean section and delivered a healthy baby boy 2.6 kg. She was monitored in Intensive Care Unit postoperatively. She was

discharged on antihypertensives, steroids and immunomodulators.

Case 7

Mrs. S 26 year old lady, primigravida at 35 weeks 3 day gestation with IUGR and preterm labour with TA. Her right upper limb BP was 120/80 mm Hg and left upper limb BP was 130/86 mm Hg. She was on steroids and immunomodulators and had bilateral carotid stenting. She had a preterm vaginal delivery of a healthy baby boy of 2.2kg.

Discussion

TA was first described in 1908 by the Japanese ophthalmologist Mikito Takayasu.⁽⁵⁾ It is an arteritis with inflammatory and mononuclear cell infiltrates that invariably leads to fibrosis, scarring, and degeneration of the elastic lamina. It has variable geographical distribution with the greatest prevalence in Asians, especially Japan.⁽⁶⁾ Women are affected in 80–90% of cases with a mean age of presentation in the second and third decade of life, reflected in a synonym for TA as “young female arteritis”.⁽⁷⁾ TA has higher incidence during the childbearing years and is usually detected on examination by decreased or absent peripheral pulses, discrepancies of blood pressure between the upper and lower extremities, and by the presence of arterial bruits. The maternal risk of pregnancy in association with TA is mainly attributed to the arterial hypertension,⁽⁸⁾ heart failure and cerebral vascular accidents. The major risk for the fetus is intrauterine growth restriction and in utero fetal demise.⁽⁹⁾ The risk of fetal death is greatest in the third trimester, especially in those with uncontrolled hypertension.⁽⁹⁾ The mode of delivery in patients with TA is determined by the hemodynamic status of the mother and for obstetrical indications. Labour and vaginal delivery with or without epidural anaesthesia has been shown to be safe, as long as blood pressure is controlled.⁽¹⁰⁾ Elevated or uncontrolled blood pressure may be a risk factor for vaginal delivery because increased sympathetic tone from labor pain or straining during labor can exacerbate hypertension. It has been demonstrated that during intrapartum uterine contractions, patients with TA experience marked elevation of systolic blood pressure as compared to control patients without TA.⁽¹¹⁾ Evidence shows instrumental deliveries to shorten the second stage of labour may reduce the occurrence of cerebral haemorrhage.⁽¹²⁾ In cases of uncontrolled elevated systolic BP, caesarean section may be safer.⁽¹⁾ Combined spinal–epidural anaesthesia or epidural alone have been reported successfully caesarean delivery. However, anaesthetic management can be complicated and put the patient at greater risks. Hypotension resulting from anaesthesia can lead to cerebral ischemia. Therefore, caesarean section should not be carried out routinely.⁽¹⁾

Diagnosis of TA by American College of Rheumatology 1990

- Age at disease onset 40 years or younger
- Claudication of the extremities
- Decreased pulsation of one or both brachial arteries
- Difference of at least 10 mm Hg in systolic blood pressure between the arms
- Bruit over one or both subclavian arteries or the abdominal aorta
- Arteriographic narrowing or occlusion of the entire aorta, its primary branches, or large arteries in the proximal upper or lower extremities, not due to arteriosclerosis, fibromuscular dysplasia, or other causes.

For purposes of classification, a patient shall be said to have TA if at least 3 of these 6 criteria are present.

TA affects the aorta and its branches, leading to stenosis, thrombosis and the formation of aneurysms. Hence it is also called as aortic arch syndrome, pulseless disease or occlusive thromboaropathy.

TA can be classified according to presenting symptoms and also by angiographic findings.

Angiographic classification of Takayasu's arteritis

Classification is based on the vessels involved in the inflammatory process as seen on angiography.

- Type I includes Branches of the aortic arch.
- Type IIa includes Ascending aorta, aortic arch, and branches of the aortic arch.
- Type IIb includes Ascending aorta, aortic arch, and its branches and thoracic descending aorta.
- Type III includes Thoracic descending aorta, abdominal aorta, and/or renal arteries.
- Type IV includes abdominal aorta and/or renal arteries.
- Type V includes Features of types IIb and IV

BP control is of importance as increase can cause rupture of aneurysms and falls in BP can lead to cerebral ischemia in the mother. Most patients benefit from invasive BP monitoring or by monitoring peripheral BPs in multiple extremities as upper extremity pulses may be absent.⁽¹³⁾ Peripheral BP monitoring may not be accurate, complicating the treatment of hypertension in these patients. Antihypertensive agents that are safely used with proven success in pregnancy are labetalol, hydralazine, and alpha-methyldopa. Routine tests in pregnant patients with TA include serial ultrasound growth assessment, umbilical artery Doppler studies, and biophysical profile assessment from 24 weeks of gestation.⁽¹⁴⁾ Most cases of TA during pregnancy have been reported in patients with a known diagnosis prenatally.

Our cases were monitored with a multidisciplinary approach where along with the Obstetrician, the Obstetric Medicine unit; Rheumatology and Cardiology were also involved. During antenatal period they were

followed up periodically with a very strict BP monitoring and fetal growth monitoring. Fetal and maternal outcome in all cases were good.

Conclusion

TA should be regarded as a risk factor for aortic dissection, especially during peripartum period, as pregnancy itself predisposes to dissection. As the incidence of TA during childbearing years is relatively high, the management of pregnancies with this disease is of great importance in clinical obstetrics. Pregnancy with TA can be complicated with hypertension and hence BP checkup in both limbs and controlling the BP is the mainstay of the antenatal management. An interdisciplinary collaboration of rheumatologists, cardiologist along with obstetricians is necessary to improve maternal and fetal outcome.

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