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Case Report

Takayasu arteritis in pregnancy: case report of two cases and review of literature

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ABSTRACT

Takayasu arteritis is a rare medical disorder of unknown etiology. The primary vasculitis mainly affects women of reproductive age group. Even though the usual course is slow, sometimes it may be unpredictable. It usually involves the branches of the aortic arch. With advancement of medicine most of the patients enter pregnancy with the disease already being diagnosed and on medications. Even though pregnancy is a favourable state complications are bound to happen especially when the disease is diagnosed for the first time during pregnancy. We are presenting two cases of Takayasu arteritis during pregnancy with different clinical presentations.

Keywords: Takayasu arteritis, Pregnancy, Preeclampsia

INTRODUCTION

Takayasu arteritis is a chronic inflammatory vascular disease of idiopathic etiology.[1] The disease is predominant in women of reproductive age group from south East Asian countries.[2] The world-wide incidence is 6/1000. It is characterized by focal stenotic process involving large vessels such as aorta and its main branches. It has got a pre occlusive phase with non-specific symptoms and an occlusive phase with symptoms. Tuberculosis has been implicated in the pathogenesis of this disease. The state of the disease at the beginning of the pregnancy is an important prognostic factor. Multidisciplinary approach and close monitoring of the disease is important in the management during pregnancy.

CASE REPORT

Case 1

Mrs. G, 27 years short statured primi gravida was referred at 38 weeks with PROM in December 2103. She was booked at primary health centre. There was no significant medical or surgical illness in the past. There was no history of hospitalization any time during pregnancy. Immediately after admission she had two episodes of Generalized Tonic Clonic Seizures (GTCS) over 20 minutes interval. After the second episode she became unconscious and had abnormal breathing. Pulses were undetectable in the upper limb and BP was not recordable. Her heart rate was 140 per minute and she was not pale. She was intubated with the help of Critical Care Unit (CCU) residents. But her BP was 180/110 mmHg in the lower limbs. Uterus was term, acting mildly and it was not tense and tender. Fetal heart sound was 170 per minute. On P/V, cervix was 50% effaced and 2cm dilated, membrane was absent, vertex was at -2 station and pelvis was adequate for the baby. After the loading dose of $MgSO_4$ she developed hypotension to the extent of ionotropic requirement. As her blood pressure was very labile she was shifted to CCU for intensive monitoring. As her general condition was very poor to withstand the stress of anaesthesia and the fetal heart rate was reassuring immediate termination was deferred. She progressed satisfactorily after oxytocin. But her second

stage was lasted for 4 hours and delivered by low forceps. It was an alive female baby of 2.4 kg with APGAR of 7 at 1 minute and 9 at 5 minute, After 6 hours of postpartum she developed pulmonary edema due to malignant hypertension. After extubation on post natal day 3, she became drowsy and her CT brain showed Left intraventicular hemorrhage and features of hypertensive encephalopathy. It was managed conservatively. She was shifted to general ward after 5 days of stay in CCU where she was investigated and found to have Takayasu arteritis and was discharged from the hospital after 20 days. Her CT Angio showed narrow ostia of lumen of B/L subclavian arteries with evidence of wall thickening and anamolous origin of Right subclavian artery.

Case 2

Mrs X, 22 years G2A1 found to have Takayasu arteritis on evaluation when she presented with hypertension at 16 weeks. MRA showed occlusion of Left CCA & Left subclavian artery. As the disease was not in active phase and other work up was normal she was on methyl dopa for hypertension. Had spontaneous labour & delivered an alive female baby of 2.08kg with APGAR of 8&9. She was discharged on PND2 with T. atenolol. She had an uneventful course during pregnancy.

DISCUSSION

Takayasu arteritis is a chronic idiopathic inflammatory disease of the arteries which primarily involves aorta and its main branches as well as coronary and pulmonary arteries. Though 5 year survival rate exceeds 90% it has a high incidence of residual morbidity. Despite the term pulseless disease is a synonym for Takayasu arteritis common finding in these patients is asymmetric pulse.

It is a systemic disease with generalized and local symptoms. Death in these patients is mainly due to ischemia, congestive cardiac failure and stroke.[3] It should always be suspected in a young woman who has asymmetric or absent pulses or discrepancy in blood pressure between limbs. Diagnosis is usually confirmed by angiography either invasive or non-invasive which usually shows irregular vessel wall, stenosis, post stenotic dilatation, aneurysm formation, occlusion and evidence of collateral formation.

Even though pregnancy is not usually associated with an exacerbation of this condition it should only be considered during a phase of remission. Pre pregnancy counseling and hypertensive management are important before conception. Continuation of prednisolone or other immune suppressants are also important. There is increased risk of abortion, early onset severe preeclampsia and Fetal Growth Restriction (FGR). Hence low dose aspirin is a good prophylactic measure.[4] A close observation during pregnancy along with other specialists is mandatory since beginning. During the course of pregnancy maternal complications should be anticipated and include severe hypertension, early onset super imposed severe preeclampsia, congestive cardiac failure and progression of renal disease. Thus serial careful monitoring of blood pressure is essential and when conditions supervene especially with asymmetrical or absent pulses Takayasu arteritis should always be kept in mind. Fetal complications include abortions, FGR, preterm delivery and still birth can occur.

Apart from the state of the disease during early pregnancy, the magnitude of blood pressure elevation during the late gestational period is another determinant factor in the management of these pregnancies.

Along with routine antenatal visits serial monitoring of blood pressure, renal function, cardiac status and preeclamptic screening are also important. Fetal surveillance includes daily fetal kick count, gravidogram, serial fetal biometry, biophysical profile and fetal Doppler.

The desired mode of delivery is vaginal and caesarean section is only reserved for obstetric indication. Careful intra partum fetal monitoring is mandatory as there is increased risk for fetal distress and intra partum birth. The genetic predisposition of Takayasu arteritis should be kept in mind since a case of SIDS has been reported in babies of these patients. Close monitoring of these patients is also important in post-partum period due to circulatory redistribution.

CONCLUSION

Although the disease occurs worldwide most cases are reported from Asia. Patients who are having uncomplicated Takayasu arteritis and who are well monitored can have successful outcome. However complications are bound to occur in undiagnosed patients. Hence Takayasu arteritis should always be included in the differential diagnosis especially in patients with uncontrolled hypertension, early onset severe preeclampsia, renal insufficiency with asymmetrical pulses and blood pressure to avoid maternal and neonatal morbidity and mortality.

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