A Challenging Case of Takayasu Arteritis in Pregnancy with Successful Fetomaternal Outcome

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Introduction

Takayasu arteritis is a chronic inflammatory arteritis affecting large vessels. It occurs before age 40 with upper extremity vascular involvement. MR angiography is the most important investigation and done without gadolinium contrast in pregnancy. Treatment is with steroids which is safe in pregnancy. Here, we are presenting a case of Takayasu arteritis in pregnancy which was carried till term with a successful fetomaternal outcome.

Case Report

A 26 year old G2P0+1 presented to NRSMCH outdoor at 32 weeks gestation being referred from a private practitioner as a case of Takayasu with echo report showing left atrium enlarged and probable left coronary artery aneurysm.. She had no complaints at that time. She gave history of ovulation induction with letrozole. On examination, her vitals were a pulse rate of 70/min on right upper limb which was low volume and 90/ min on left side which was high volume. Pressure was 110/70 mmHg on right side and 220/80 mmHg on left side. She had carotid artery bruit and bilateral renal artery bruit. Obstetric examination showed uterus size corresponding to period of amenorrhea, 32 weeks with regular heart sound of 136 bpm. Patient was admitted and was dealt with a multidisciplinary approach with opinions from cardiology, rheumatology and anaesthesiology.

Her blood pressure in all 4 limbs was monitored strictly twice daily and was maintained at 208/54 mmHg in left upper limb and 116/70mmHg in right upper limb and antihypertensive drugs dosage was adjusted at regular intervals to suit her needs. She was started on labetalol 100 mg three times a day increased to 400 mg three times a day and subsequently nifedepine 30 mg twice a day was added as pressure was not under control. Patient had no complaints of any premonitory symptoms of pre-eclampsia throughout pregnancy. Alternate day urine dipstick was done to rule out any new onset proteinuria and it was negative throughout pregnancy. Complete blood count, liver and renal function tests were repeated weekly and were normal. Takayasu specific imaging investigations were done and results are given in Figure 1. All these showed her to have Stage V disease. Ophthalmology examination was done and it revealed few dot haemorrhages in both eyes. Foetal monitoring was done with ultrasonography and colour Doppler every 3 weeks and showed normal growth and liquor

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Figure 1: Imaging investigations in antenatal period

MR Neck Angiography	Thickening in arch, right brachio cephalic artery, bilaterial common carotid artery Occlusion of left subclavian artery 40-50% stenosis left proximal mid common carotid artery No signal in vertebral artery Possiblity of Aorto arteritis
MR Renal Angiography	Diffuse mild disease in abdominal aorta Bilateral renal atery stenosis Occluded SMA 90-99% stenosis at celiac ostium
Repeat Echo	Concentric LVH LVEF 65% Grade 2 Diastolic Dysfunction Normal values No RWMA at rest
Colour Doppler	Narrowing in the lumen of bilateral renal arteries Acceleration time increased in bilateral sites RAR – 0.82 Diffuse wall thickening in bilateral common carotid arteries Palvus tardus waveform in bilateral intersegmental arteries

volume. She was put on Tab. Prednisolone 7.5 mg once a day and Tab. Aspirin 75 mg once a day from 32 weeks as per rheumatology advice.

Pregnancy was carried till 37 weeks gestation and since she was Stage V disease termination was by elective Lower segment caesarean section which was under combined spinal epidural as per anaesthesiology advice. Her intra op period was uneventful and her pressure was maintained at 236/92 mmHg. She delivered a healthy 2.250 kg baby girl. From anaesthesiologist side, she received dexamethasone 8 mg intravenous three times a day starting from intra operative period and continued postoperatively for 2 weeks.

Post op, patient was followed up in High dependency unit and her vitals were similar to antenatal period. Epidural top up was continued with infusion ropivacaine 0.15 % diluted in 50 ml NS at 5ml/hour for upto 60 hours post operatively. She was advised for follow up at cardiology and rheumatology outdoor post discharge and mother and baby was discharged in stable condition. She was discharged with labetalol and nicardia and Inj. Dexamethasone 6 mg intravenous three times a day for 2 weeks followed by oral prednisolone as per anaesthesiology advice.

Through telephonic conversation, we came to know that patient did not take any medicines as advised and did not come for follow up. One year after her discharge she developed cerebrovascular accident with weakness of the left side and at present she is recovering from that.

Discussion

Takayasu arteritis is a rare, chronic, inflammatory, progressive, idiopathic arteriopathy, affecting young women of reproductive age group, causing narrowing, occlusion and aneurysm of systemic and pulmonary arteries, aorta and its branches. Its incidence is reported to be 13 cases per million population with Asian predominance.[1]

Prognosis of Takayasu arteritis in pregnancy is worsened by comorbid severe renovascular hypertension, cardiac involvement or pulmonary hypertension. Involvement of abdominal aorta portends worse perinatal outcome. It can complicate pregnancy by causing pregnancy related hypertension, preeclampsia, abruption, congestive cardiac failure, progressive renal involvement, foetal growth restriction, preterm birth and foetal death. In our patient severe systolic hypertension was seen which was managed with a holistic approach with opinions from cardiologist and rheumatologist.

Clinical diagnosis is by using American College of rheumatology 1990 criteria where if 3 out of 6 criteria is fulfilled, the diagnosis is made. It includes age under 40 at disease onset, claudication of extremities, decreased brachial artery pulses, systolic blood pressure difference > 10 mmHg between arms, bruit over subclavian arteries or aorta and angiogram abnormalities.[2] In our patient, all these criteria were fulfilled. Imaging options in pregnancy include angiogram, echo and Doppler studies all of which were done in our patient.

Management in pregnancy is by routine antenatal check up with serial monitoring of blood pressure, renal function, cardiac status and preeclamptic screening. In our patient, these were followed. Foetal surveillance is by daily foetal movement count, foetal biometry, biophysical score and foetal Doppler. Cardiologist and rheumatologist opinion was taken and she was put on Prednisone and aspirin.

Vaginal delivery is usually preferred in these patients.[1] Induction of labour is not indicated and spontaneous onset of labour is awaited. Second stage of labour is cut short by instrumental delivery. Lower segment caesarean section is preferred from stage IIb and onwards of Numano classification to prevent cardiac decompensation due to increased blood volume and blood pressure observed during uterine contractions and increased cardiac output observed in labour. As our case was Numano Stage V, elective Lower segment caesarean section was performed.

Epidural anaesthesia provides stable hemodynamic and pain relief which was used in our patient.[1] It is associated with gradual onset of sympathetic block and decrease in pressure. There is smooth control of pressure in intraoperative and postoperative period. Regional anaesthesia is associated with sympathetic blockade and subsequent drop in pressure and compromised regional circulation due to stenosed arteries. There is risk of aortic dissection on table. In such a case patient presents with a sudden ripping pain, breathlessness or loss of consciousness. It is treated with beta blockers, narcotics, opiates, pericardiocentesis to relieve the pressure and urgent surgery.[3]

Conclusion

The optimal management of pregnant patients with this disease has not yet been defined. The course of the disease seems to be neither affected nor worsened by pregnancy. Although unfavourable foetal and maternal outcomes has been reported, our case had a favourable fetomaternal outcome. With an interdisciplinary collaboration, the prognosis of both can be improved.

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