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**Case Report**

# Takayasu's arteritis in pregnancy

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## ABSTRACT

We had the opportunity to manage a pregnant lady who presented for the first time at 34 weeks of amenorrhea with absent pulses in upper limbs on examination. She underwent Magnetic Resonance Angiography to confirm the diagnosis of Takayasu's arteritis. Luckily she did not have complications of Takayasu's such as hypertension, cardiac failure, neurological symptoms, visual disturbances. She proceeded to have a normal vaginal delivery after aggressive management which included a multidisciplinary team involving obstetrician, cardiologist, physician and rheumatologist.

**Keywords:** Takayasu's arteritis, Pregnancy, Pulseless disease

## INTRODUCTION

Takayasu's arteritis is a chronic idiopathic vasculitis affecting aorta and its primary branches, commonly seen in young women of Asian or oriental descent during child bearing age. Incidence is 2.6 cases/million/year.[1] The basic disease appears to be unaffected by pregnancy, but high arterial pressure and pre-eclampsia constitute main maternal complications, resulting in intrauterine growth restriction.

## CASE REPORT

A 22 year old primigravida with 8 months of amenorrhea came for first antenatal check up. She had no medical complaints till date. On physical examination only her lower limb pulses were palpable with absent carotid and upper limb pulses. Her BP could be recorded only in lower limb which was 90/70 mmHg. There was no jugular venous distension. On auscultation first and second heart sounds were normal with no murmur. No bruit heard on auscultation of carotid, subclavian artery and aorta. Suspecting peripheral arteritis we enquired about history suggestive of intermittent claudication, fever, arthralgia when she gave history of fever 3 yrs back with joint pain which subsided with treatment. She also gave history of on and off giddiness and severe pain in the upper limbs on exertion. Clinically she corresponded with period of amenorrhea of 34 weeks.

Suspecting Takayasu's arteritis (pulseless disease) we proceeded to investigate her further. Initial laboratory values revealed markedly elevated C-reactive protein (2.4mg/dL) and ESR (120mm/hr) with normal Complete blood count, liver and renal function tests, urine analysis. Electrocadiography showed normal sinus rhythm. Chest X-ray and fundoscopy were normal. Foetal USG with Doppler velocimetry revealed normal growth and no uteroplacental insufficiency.

Considering the vascular and cardiac implications, a cardiologist's opinion was taken. Echocardiography showed normal left and right systolic function. To search for aetiology of peripheral arteritis, specific investigations were carried out such as dsDNA, APLA, ANA, cANCA, pANCA , which were negative. FNAC of palpable axillary lymph nodes revealed granulomatous lymphadenitis. Upper extremity Doppler showed monophasic forward flow in arteries and carotids were not visible bilaterally with formation of multiple collaterals. Renal artery Doppler was normal.

To know the extent of occlusion of great vessels we performed Magnetic Resonance Angiography of neck and

brain without contrast which revealed complete occlusion of brachiocephalic, distal left common carotid, left subclavian and vertebral arteries with formation of collaterals.

**Figure 1: Magnetic resonance angiography showing occlusion of branches of aortic arch.**

We decided to do angiography of thoracic and abdominal aorta after delivery. Based on American society of Rheumatology classification criteria, the patient was diagnosed with Takayasu's arteritis and started on steroids (Tab prednisolone) after consulting a rheumatologist. Patient was also started on empirical antitubercular treatment as majority of cases of Takayasu's in India are associated with Tuberculosis. The patient underwent a preanaesthetic check up to prepare the line of management in the event of a caesarean delivery.

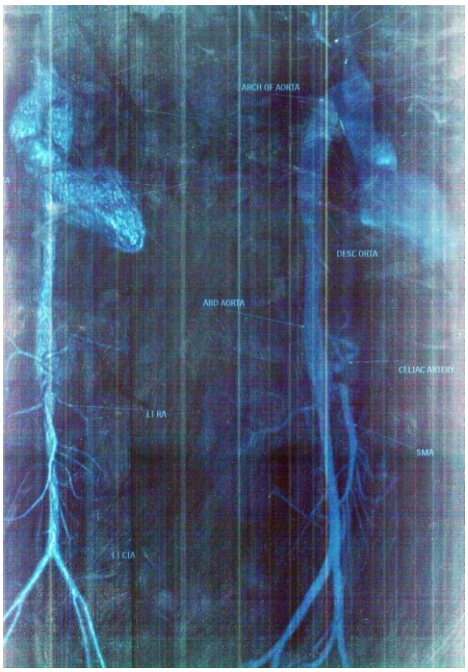


**Figure 2: CT angiography of thorax and abdomen showing narrowing of distal descending thoracic aorta and infrarenal abdominal aorta.**

The patient continued to be under close surveillance till she went in spontaneous labour at 39 weeks. Her vitals were monitored using non-invasive monitors. Fluid intake was monitored to avoid excess fluids. She progressed normally till full dilation when vacuum was applied to cut short the second stage of labour. A healthy male baby weighing 3.2 kg was delivered. In order to prevent congestive cardiac failure 10 mg inj Lasix was given IV after checking systolic BP which was 100 mm Hg in right thigh. In spite of adopting active management of third stage of labour patient had atonic PPH which was managed by giving inj prostodin (250 microgram) and tab misoprost (1000 microgram PV). Inj methergin was not given because of its vasoconstrictor effect.

We did CT angiogram thorax and abdomen after delivery which showed diffuse narrowing of distal descending thoracic aorta D7-D10 levels and mild diffuse narrowing of infrarenal abdominal aorta.

She had uneventful postpartum period and was discharged on day 8 on steroids and AKT. We plan to consult a cardiologist to decide whether the stenotic lesions would benefit by angioplasty 6 weeks after delivery.

## DISCUSSION

Aetiology of Takayasu's arteritis is unknown but autoimmune process and infection has been considered to play a role in pathogenesis. Tuberculosis has been particularly implicated in view of high prevalence of infection, past or present, in affected patients.[1]

Patient may be asymptomatic with impalpable pulse or may present with symptoms ranging from fever, fatigue, weight loss to life threatening haemoptysis, heart failure from aortic regurgitation, or catastrophic neurological impairment.[2] On physical examination the patient may have diminished or absent pulse, difference between BP in arms, presence of bruits over neck, supraclavicular area, abdomen and hypertension.

Diagnosis is confirmed by CT or MR angiography revealing the characteristic tapered luminal narrowing or occlusion that is accompanied by thickening of the vessel wall. To avoid the large amount of radiation needed for CT angiography, magnetic resonance imaging is preferred during pregnancy.[3]

The American society of Rheumatology has devised criteria for diagnosis of Takayasu's arteritis.[4]

- Age at disease onset ≤ 40 yrs
- Claudication of extremities.
- ↓ Brachial Artery pulse
- Systolic BP difference of > 10 mm Hg between arms
- Bruit over Subclavian Artery or Aorta.
- Aortogram abnormality.

Presence of three or more criteria confirms the diagnosis of disease with sensitivity of 90.5% and specificity of 97.8%.

Steroids have formed the mainstay of treatment for Takayasu's arteritis as they have a role in suppressing systemic symptoms and are known to reverse arterial stenosis. Approximately 50% of the patients treated with steroids respond to the treatment. Cytotoxic drugs like methotrexate, azathioprine are also used to treat Takayasu's arteritis but avoided during pregnancy. Subsequent treatment is limited to angioplastic or surgical correction of the stenosis or aneurysm in severe disease.

Fertility is not adversely affected and pregnancy does not appear to exacerbate the disease or the inflammatory activity and the hemodynamic status may improve with pregnancy. Whenever possible, women should be assessed preconceptually so that appropriate adjustments or stoppage of cytotoxic drugs to be made prior to conception.[5]

Antenatal care should be provided by a multidisciplinary team including obstetrician, radiologist, anaesthetist, cardiologist and rheumatologist. Maternal complications include superimposed pre-eclampsia, congestive cardiac failure, cerebral haemorrhage, progressive renal impairement.[5] Fetal complications include intrauterine growth restriction. Baseline investigations include ESR and CRP, Doppler ultrasound, MR imaging to assess severity of disease and echocardiogram to assess ejection fraction. Fetal growth should be monitored serially with ultrasound and colour Doppler.

Steroids can be used safely during pregnancy in the low doses.[6] Prednisolone is metabolised by the placenta and thus not transmitted to the fetus except at high doses. Pregnancy is allowed to continue till term and induction of labour is considered only in presence of superimposed pre-eclampsia or hypertension. Elective caesarean section is indicated for severe disease. The anaesthetic management is important and it is advisable to involve anaesthetist at an early gestation. Hypertension in the second stage of labour is a risk factor for cerebral haemorrhage, shortening this stage by use of forceps or vacuum extraction appears to be a reasonable solution.[7]

Contraception in the form of progesterone only pill, depot injections or intra-uterine devices can be recommended, but combined contraceptive pill is avoided as there is increased risk of thromboembolism.[5]

## CONCLUSION

Medical management of a pregnant patient with Takayasu's arteritis does not differ significantly from a non-pregnant patient. Multidisciplinary management is essential for satisfactory clinical outcome during pregnancy and their blood pressure should be strictly controlled for favourable maternal and fetal outcome and mode of delivery should be planned.

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