

Case Reports

Successful Outcome of Pregnancy in two Cases of Takayasu Arteritis

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Abstract:

Introduction: *Takayasu arteritis is a vasculitis of unknown cause. It is a disease of involvement of vessels of branches of the aortic arch. It can affect women of reproductive age and may present with pregnancy. Pregnancy may have a favorable outcome if diagnosed and appropriately managed from the onset of the disease and enter pregnancy in the remission phase and with medication. But, if it is diagnosed for the first time during pregnancy outcome might be worse.*

Case reports: *Here presented two case reports of diagnosed Takahashi Arteritis with term pregnancy; who entered pregnancy in remission phase and on medication. The prior preparation leads the pregnancy to a successful outcome without any residual effect on fetomaternal health.*

Conclusion: *Early diagnosis of the disease and proper management of the progress of the disease by intervention is essential for a good prognosis. A close follow-up and multidisciplinary approach during pregnancy make pregnancy a safer journey for a woman with Takahashi Arteritis.*

Keywords: *Takayasu Arteritis, Pregnancy*

Introduction:

Takayasu Arteritis (TA) is an inflammatory major artery vasculitis that primarily affects the aorta, branches, and pulmonary arteries.^{1,2} TA typically affects women of reproductive age⁵, but males have also been discovered to be affected, but disease symptoms vary from person to person. The second and third decades are the period of life when women suffer from TA.^{2,5} It has been reported worldwide, with Asian women having the highest occurrence.^{3,4}

Although the cause and pathology of TA remain unknown, much has been learned about the illness since M. Takayasu, a Japanese ophthalmologist, first documented it in 1908.^{6,7} Researchers are still looking into the disease's immunological and psychological elements despite this. Nonetheless, research into the disease's immunological and genetic elements is

ongoing.⁸ TA is a granulomatous vasculitis of large and medium-sized arteritis that progresses. In most cases, the aorta and its branch arteries show uneven thickening, narrowing aneurysms, intimal stenosis, aneurysms, and occlusion.⁹ The condition affects the coronary, pulmonary, and renal arteries. The clinical manifestations of TA vary depending on which blood arteries are damaged. Most patients have reduced or absent pulses in their upper extremities, which is why it is sometimes known as "pulseless sickness," and they also suffer numbness and coldness in their fingers.^{1,8} TA causes blockage and aneurysm formation in the systemic and pulmonary arteries, among other problems. It has two clinical phases: an active inflammatory phase and a chronic phase. Fever, lethargy, malaise, lack of appetite, weight loss, headaches, dizziness, arthralgia, myalgia,

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claudication, skin rashes, and life-threatening hemoptysis, heart failure characterize the early inflammatory stage of TA. Late symptoms are caused by fibrous thickening of artery walls, which leads to vascular obstruction and ischemic heart disease.^{10,11} The late chronic symptoms are determined by the location of the stenosis and the time needed to diagnose the disease and treat it.

According to the American College of Rheumatology, at least three of six criteria must be met for a confirmed TA diagnosis. Age 40 years or older, claudication of the limbs, decreased brachial artery pulse, BP differential of 10 mmHg between arms, bruit over the subclavian artery or aorta, and angiography abnormalities- aortic occlusion or constriction.

Classification of the disease is carried out depending on the involvement of blood vessels –

Table-I
New angiographic classification of Takayasu arteritis, Takayasu conference 1994

Type	Vessel Involvement
Type I	Branches from the aortic arch
Type II-a	Ascending the aorta, the aortic arch, and its branches
Type IIb	Ascending aorta, aortic arch and its branches, descending thoracic aorta
Type III	Thoracic descending aorta, abdominal aorta, and/or renal arteries
Type IV	Abdominal aorta and/or renal arteries
Type V	Combined features of types IIb and IV

Here, two cases of Takayasu Arteritis with Pregnancy with the successful fetomaternal outcome are narrated.

Case Report:

Case 1:

A-26 year-old 2nd gravida with 39+ weeks of pregnancy was admitted to the hospital with active labor pain. During the evaluation, her pulse was felt on her left hand and was feeble. We learned that she was a known case of Takayasu's arteritis (TA) from history. She had no history of other co-morbidities like Inflammatory bowel disease (IBD) and Sinusitis. Previous medical reports revealed that at 21 years of age, she had presented with fever, headache,

dizziness, loss of appetite, dyspepsia, chest pain, loss of weight, and polyarthralgia, myalgia in the initial phase.

On examination, blood pressure was measurable only in the left hand, and pulse was feeble in the left hand. On investigation, ESR was (117 mm/hour), and CRP was elevated with mild anemia. Her antinuclear antibody (ANA) and Anti-cyclic citrullinated peptide (Anti-CCP) were negative.

On evaluation, F-18FDG-PET CT (Computed Tomography) chest showed increased FDG (Fluorodeoxyglucose) uptake and the diffuse circumferential wall thickening in ascending aorta, descending thoracic aorta extending to the D7 level. Similar FDG acid wall thickening was observed in the brachiocephalic trunk, common bilateral carotid, and left subclavian arteries. Luminal narrowing of the common bilateral carotid and the proximal left subclavian artery was observed. The patient also had cervical spondylosis 5 years back. We found that cervical vertebrae (3-5) show anterior bony beaking on X-ray film. She also gave a history of asthma. Her IgE level was found to increase. An Ultrasonography of the whole abdomen was done at that time, where we got to know that she has fatty liver (Grade I). She was non-hypertensive, and no neurological deficit was present on presentation. She was discharged on Steroids, Methotrexate, and Folinic acid. She was on regular follow-up with a rheumatologist.

Her past obstetrical history revealed first pregnancy was uneventful. Eventually, Echo with Colour Doppler was carried out 3 years back, showing trivial mitral regurgitation and velocity across the pulmonic valve at 1.0 m/Sec. Trivial Tricuspid Regurgitation was also found in the Doppler study. Velocity across the AV was 1.0 m/Sec.

Her blood pressure was standard throughout the pregnancy. Lower segment cesarean was done under spinal anaesthesia due to prolonged 1st stage of labor. She delivered a healthy male baby. But, during cesarean section after spinal anaesthesia, she developed hypotension, but the patient was alert, cooperative and her Glasgow Coma Score (GCS) was 15. Her uterus was well contracted. An Electrocardiogram was performed at the bedside, revealing regular rate, rhythm, and character. Her fluid management was done. After resuscitation, a feeble pulse was felt on her left hand, and her blood pressure was normal.

On 3rd POD, her check dressing was done, which was healthy. There were no postpartum complications. On the 6th day of puerperium, the patient was discharged after evaluation.

Case-2:

A 21-yr-old housewife from the local vicinity was admitted as a diagnosed case of Primi Gravida with 36+ weeks of Pregnancy with Less Fetal Movement for one day with Takayasu Arteritis. She Was a regularly menstruating woman, married for 2.5 years, and conceived spontaneously. It was her planned pregnancy, and she was on a regular check-up. She did all routine investigations which were within the standard parameter. She was duly immunized against tetanus.

During antenatal care, the patient was under the supervision of an obstetrician and cardiologist. Her pregnancy was uneventful. Two days before admission, she developed sudden palpitation and was admitted to the cardiology department of Chittagong Medical College hospital. She has a history of Takayasu arteritis, diagnosed at age ten. At age ten, she developed chest pain, and her blood pressure was high. So, she was admitted to a superspecialized tertiary care hospital for evaluation. And after the angiogram, interpreted as Takayasu arteritis with renal artery stenosis and stenosis of the abdominal aorta. The percutaneous coronary intervention was done at the interventional cardiology department of another hospital, and her stenosis was corrected by stenting. After that, she was on regular follow-up, and from that time onwards, and on Nifedipine and Aspirin throughout her pregnancy. After admission, the patient was examined thoroughly. The patient was anxious, had decubitus on choice, mildly anemic. Pulse was 98 beats per minute on the right hand and 82 beats per minute on the left hand. Blood pressure was 170/70 on the right hand and 120/80 on the left hand.

On abdominal examination, all fetal parameter was found normal. The cardiac evaluation by ECG and echocardiography was performed, and the findings were normal (ejection fraction 67%). Consultation from an in-house senior anaesthetist was sought for possible surgery for termination of pregnancy. The decision was to continue pregnancy up to 37 completed weeks. At 37 weeks, LSCS was done under spinal anaesthesia for obstetric reasons. She delivered a healthy male baby. Apgar's score was 8/10, weighing 2.6 kg. Her postpartum period was uneventful. Again,

a cardiology consultation was done and advised her to replace antihypertensive with Labetalol thrice daily. As she was well, she was discharged on the 8th day of puerperium.

Discussion:

80%-90% of TA cases are seen in women, and the age of onset is usually between 10 and 40 years.^{12,13} The sickness has international distribution, with the most significant incidence being in Asia.¹⁴ Furthermore, Japan stated an estimated one hundred fifty new instances of TA each year.¹⁵ In the USA and Europe, one to three cases new per year per million populace seem to be reported.¹³ Pregnancy doesn't impact the course of the disease itself, but incidence peaks in the second and third trimesters. Pregnancy is not usually associated with increased inflammation in the vascular system.^{8,15} pregnancy causes an increase in blood volume, which makes the management of pregnancy with Takayasu Arteritis quite challenging due to cardiovascular complications like hypertension, organ dysfunction, and perinatal mortality. During pregnancy, there's a raised chance of cardiovascular issues like hypertension and congestive heart failure.¹⁶ Stenosis decreases regional blood flow, which causes neonatal complications like prematurity, low birth weight, and increased incidence of caesarian section.^{9,17} On the other hand, uncontrolled hypertension at some point in the gestational period has been associated with aortic analysis, cardiac and renal insufficiency, stroke, maternal death, miscarriage, and stillbirth.¹⁸⁻²⁰ Also, raised blood pressure might cause IUGR and IUD.²¹ because the increased volume may impair circulation and exacerbate high blood pressure aortic regurgitation and congestive cardiac failure. BP management is of paramount importance because any amplification may rupture an aneurysm, resulting in hypotension and cerebral ischemia in the mother. Moreover, peripheral BP monitoring may additionally not be accurate and could complicate the remedy of hypertension in these patients. Most patients gain from invasive BP monitoring or the measurement of peripheral BP in more than one extremity because the upper extremity pulses might also be absent.²²

As pregnancy is risky, preconception counseling is a must for better pregnancy outcomes which provides scope for dosage adjustment and cessation of cytotoxic medication and folic acid supplementation in the periconceptional period. Also, BP control during

pregnancy may be difficult due to physiological changes. The optimal timing of pregnancy is the remission period when the BP and disease are stable. Therefore, it is also essential to adjust antihypertensive drugs and avoid the use of angiotensin-converting enzyme inhibitors or angiotensin inhibitors. Pregnant women with TA are encouraged for an early booking for regular antenatal supervision.²³

Case-1 presented with the symptom of acute inflammation with raised ESR, CRP, and anemia, reflecting the underlying inflammatory process. There is no specific serological test for the diagnosis of TA. But, these laboratory investigations are usually nonspecific for the diagnosis of TA. 18FDG-PET CT chest showed the typical feature of TA. According to classification, case-1 could be categorized as TA type IIb as there was diffuse wall thickening of ascending aorta, left subclavian artery, and descending thoracic aorta, and case-2 could be categorized as type IV as there was the involvement of renal artery and abdominal aorta before intervention.

A multidisciplinary approach to optimize the care of such patients in a tertiary care hospital is best for women, which was supported by a different study.⁸ The aim of management should be control of blood pressure, control of inflammation, prevention and treatment of any complications like hypertension and revascularization of the affected occlusive or stenotic vessel.²

During pregnancy, routine antenatal visits, serial monitoring of blood pressure, renal function, cardiac function, and preeclampsia screening are essential. Monitoring fetal status is also necessary and will include daily fetal kick count, serial fetal biometry, biophysical profile, and fetal Doppler.²³

Management of systemic and vascular inflammation is executed using corticosteroids such as prednisolone. Methotrexate, folinic acid, Cyclophosphamide azathioprine are used for immunosuppressant. In chronic stages of TA, endovascular revascularization procedures should be done. Procedures should be performed only after suppressing inflammation in the affected arteries. Surgical procedures carry risk, and the success rate depends upon the location and stage of stenosis of the blood vessel.¹ Despite the disease status, vaginal delivery is the preferred mode of delivery, and close labour management is crucial for patients with TA.

Additionally, epidural analgesia has been advocated for labour and delivery as well.

Although Takayasu Arteritis is associated with a pregnancy complication, tight preconception disease control, strict follow-up, and targeted high blood pressure treatment can result in positive pregnancy outcomes.¹⁰ Early diagnosis of the disease, closed antenatal monitoring and multidisciplinary care are necessary for a better outcome. Delayed diagnosis may lead to severe consequences.^{5,17}

Conclusion:

Pregnancy with Takayasu Arteritis does not exacerbate the disease if the patient enters pregnancy in the remission phase and with medication. For that, TA should have a preconception evaluation. Management of TA with Pregnancy does not differ much from with nonpregnant states. Multidisciplinary management is essential for good pregnancy outcomes.

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