

Successful Fetomaternal Outcome in a Case of Takayasu Arteritis

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Abstract: *Takayasu arteritis is a rare type of vasculitis, a group of disorders that causes large vessel inflammation.¹ The disease can lead to narrowed or blocked arteries, or to aneurysm and tear. It may lead to complications like high blood pressure, heart failure, stroke, transient ischaemic attack, and aneurysm in the aorta¹. A healthy pregnancy is possible with the disease but it is a medical challenge. It usually complicates the latter half of the pregnancy and causes hypertension, organ dysfunction, and fetal growth restriction.² Pregnancy with TA is a medical challenge as, the results by the American College of Rheumatology highlight the serious concerns ie the miscarriage rate of 11% and intrauterine death of 1%, preterm delivery rate was 15%, 16% of pregnancies had IUGR and 28% of patients required delivery by LSCS. Here presenting a case of G4P3IUFD3 with previous 1 LSCS with Takayasu arteritis with chronic hypertension with late - onset severe IUGR. Following a multidisciplinary approach, she delivered an alive live born low - birth - weight baby. Her postpartum course remained uneventful. Despite advancements in cardiovascular management and new drugs, the optimal management for pregnant patients with this disease remains elusive.*

Keywords: Lower segment caesarean section (LSCS), Intrauterine growth restriction (IUGR), Takayasu Arteritis (TA), Gravida (G), Para (P), Intrauterine death (IUD)

1. Introduction

Takayasu's arteritis, also known as pulseless disease/ aortoarteritis/ "young female arteritis", is a rare chronic inflammatory progressive large vessel vasculitis of unknown etiology affecting young women of the childbearing age group.¹⁻⁴ It is predominantly seen in women of Southeast Asia, Areas of Africa, and Southern America.⁵ Its incidence is reported to be 13 cases per million population.^{6,7} It leads to narrowing, occlusion, and aneurysm of systemic and pulmonary arteries in the body, primarily affecting the aorta and its branches^{3,7}.

Pregnancy as such has no effect on the evolution of the disease, however, its peak incidence is in the second and third trimesters. However, during pregnancy, there is an increased risk of cardiovascular complications such as hypertension and congestive heart failure. Thus, such patients need special attention during the peripartum period as there is an increased risk of development of complications such as hypertension, multiple organ dysfunction, and stenosis obstructing regional flow leading to restricted intrauterine fetal growth and low birth weight in babies¹¹⁻¹³. Delay in diagnosis is quite common, so patients often conceive without prior knowledge of having TA or having initiated specific treatment against it.²

Ideal management for pregnant patients with this disease still poses a stringent challenge, an interdisciplinary collaboration of obstetricians, cardiologists, rheumatologists and neurologists is often necessitated for an optimal maternal and fetal prognosis. Here is the case discussed to enlighten the obstetricians on the fetomaternal outcome and

management of this rare and uncommon clinical entity, a pregnant female with active TA.

2. Case history

A 28years old female married for 7 years, spontaneous conception with 3 months of gestational age with a known case of chronic hypertension, currently not on medication (as was non - compliant to medications) was referred in view of Renal artery color doppler dated 16/11/21 suggestive of bilateral renal artery stenosis at the origin. Atherosclerotic changes in the form of intimal thickening and calcification in the intima were noted in the abdominal aorta and proximal superior mesenteric artery with surrounding soft tissue edema. Aortoarteritis

On examination, the General condition of the patient was fair with a pulse rate of 74/min, BP being 160/100mmhg in the left arm and 140/90 in the right arm with negative urine albumin and premonitory signs and symptoms with normal DTR, SPO2 99% on RA, Absent pallor/ icterus/ edema

The findings of CVS and RS were within normal limits
On P/A examination: soft, no gtr with the scar of previous lscs +
P/S: cervix, and vagina healthy
P/V: uterus 12weeks
b/lfornices free, non - tender

She was admitted for further evaluation and management on admission, all routine investigations were within normal limits after which we proceeded with a specific examinations after discussion with medicine faculty. USG

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obs + A + P was done S/O SLIUG of 12 weeks 2 days with hemodynamically significant Bilateral Renal Artery Stenosis. CRP was negative, urine albumin negative, ESR 32

Nephrology reference was done, advised Quantitative ESR and CRP, APWA, VDRL, MR Angiography of renal vessels and aorta without contrast

Doppler study of B/L upper limb S/O no significant abnormality

Finally, USG KUB was done by a senior radiologist with Renal Doppler S/O hemodynamically significant B/L Renal Artery Stenosis (Right>Left) Carotid Artery Doppler was unremarkable

MR Angiography was done as advised by nephrologist S/O Bilateral Renal Artery Stenosis at the Ostia, wall thickening and luminal irregularity involving the abdominal aorta, Pseudoaneurysm formation in the abdominal aorta in the left side involving the suprarenal portion, Nil flow detectable Signal involving the superior mesenteric artery, the image S/O large vessel vasculitis (Involving abdominal aorta, it's major branches and bilateral renal ostia) likely suggesting Takayasu Arteritis

To rule out any secondary cardiac changes 2d echo S/O normal study with left ventricular ejection fraction of 60%

Rheumatology reference done for definitive management advised, strict BP monitoring, continue antihypertensive for BP control, fundoscopy. Plan for detailed evaluation post delivery.

Fundoscopy to rule out any ophthalmic changes was done within normal limit

USG obs NT scan was done suggestive of single live intrauterine gestation of 13 weeks and 4 days

Patient started on tab lobet 200mg TDS and tab aspirin 75mg OD with twice daily BP charting at home

Patient was following up regularly for ANC visits and was admitted again in view of raised BP not controlling on antihypertensives at 26 weeks of gestation in March 2022

BP monitoring done and antihypertensives adjusted accordingly, started on tab nicardia 30XL QID and tab lobet 400 - 200 - 400 and tab prednisolone 20mg OD to reduce disease progress

Repeat rheumatology reference done, advised to control BP aggressively. She should not go into preeclampsia and eclampsia, further treatment after delivery as patient will require stenting.

Patient was taken up for emergency lscs at 31 weeks of gestation in view of USG S/O uteroplacental and fetoplacental insufficiency with prevlscs with short ICP after confirmation of NICU bed and ventilator

Baby cried immediately after birth and was admitted in NICU in view of Extremely preterm and very low birth weight of 1.1kg

Patient was shifted to CCU for postoperative monitoring which was uneventful Interventional radiologist reference done in view of definitive management of takayasu arteritis ans was advised PET CT ans stenting after funds are available

Patient was discharged with baby on day 36 of PNC

3. Discussion

Takayasu Arteritis was first described in 1908 by two Japanese ophthalmologists, Mikito Takayasu and Onishi, who observed retinopathy in the absence of peripheral pulses. The cause is unknown, but it seems to be related to autoimmunity, sex hormones (more common in young females), and genetics (demonstrated by the predisposition of the human leukocyte antigen – HLA BW52) Disease progression typically occurs in various stages from acute inflammatory arteritis to lymphocytic infiltration, intimal thickening, elastic tissue destruction, fibrosis and patchy luminal narrowing of arteries.

Depends on Angiographic classification there are five types based on the involvement arteries.^{14, 15}

Type I involves branches of aortic arch

Type IIa ascending aortoarch, and its branches

Type IIb involves Type IIa and thoracic descending aorta

Type III thoracic descending aorta, abdominal aorta, renal arteries, or combination

Type IV abdominal aorta, renal arteries, or both

Type V involves entire aorta and its branches

Takayasu Arteritis progresses through three stages.

Stage 1 (prevasculitic systemic stage) - constitutional symptoms like fatigue, malaise, giddiness, fever

Stage 2 (vascular inflammatory stage) - stenosis, aneurysms and vascular pain (carotidynia)

Stage 3 (burned - out stage) - fibrosis and generally associated with remission

The incidence of Takayasu arteritis during childbearing years is relatively high, the management of pregnancies with this disease is of great importance in clinical obstetrics. Pregnancy with Takayasu arteritis can be complicated by hypertension, as seen in our case, and worsening of cardiovascular hemodynamic status. Hypertension is a serious complication that can lead to intrauterine growth retardation, fetal hemorrhage, and maternal heart failure.¹⁶ The increased intravascular volume seen during pregnancy may impair circulation and exacerbate maternal hypertension, aortic regurgitation, and congestive heart failure.¹⁷

The disease causes various clinical conditions depending on the sites of constriction such as arm claudication, decreased arterial pulses, visual loss, stroke, aortic regurgitation, Hypertension, congestive cardiac failure. Hypertension is seen in 90% cases Takayasu arteritis. The clinical patterns of TA differ at the acute and chronic periods. In the acute

period, systemic symptoms prevail, while in the chronic period, insidious ischemic - destructive signs are more prevalent. These signs appear together with stenosis at a rate of 85%, dilatation at a rate of 2%, and stenosis and dilatation at a rate of 13%.^{18, 19, 20} The symptoms range from fever, fatigue, and weight loss to life - threatening hemoptysis and heart failure.

Diagnosis is usually based on clinical manifestations, inflammatory markers (acute phase reactants), and arteriography demonstrating aortic stenosis and of its branches. Common features of active TA are fatigue, myalgia, arthralgia, and low - grade fever in initial stages and intermittent claudication, visual defects, and fainting attacks in later stages. Many may be diagnosed after clinical examination, when one or more peripheral pulses are not palpable or blood pressures vary in two limbs. However, computed tomography or magnetic resonance angiography can detect TA even before the development of severe vascular compromise as in our case.²¹

B Recently, 18 FDG - PET scan has been added as an adjunct imaging modality in the armamentarium of rheumatologists and cardiologists to diagnose LVV, with a pooled sensitivity and specificity of 70.1% and 77.2%, respectively.²²

However, the gold standard for diagnosis still remains as vessel biopsy which could not be performed in our case.²³

The management of TA is a multidisciplinary approach with the involvement of obstetricians, anesthesiologists, cardiologists, rheumatologists, and neonatologists. Ultimately, the aims encompass the control of inflammation, prevention, and treatment of complications like hypertension and occlusive or stenotic lesions.²⁴ The aims are control of inflammation, prevention and treatment of complications like hypertension and revascularization by percutaneous angioplasty, use of endoprosthesis, or surgical correction for occlusive and stenotic lesions.

When managing women of reproductive age with TA, preconception counseling is essential. In addition, such counseling will focus mainly on dosage adjustment, cessation of cytotoxic drugs, folic acid supplementation in the periconception period, and the optimal timing of pregnancy. Similarly, the pregnancy should be ideally planned in remission phase and patients are encouraged to pursue an early booking for regular antenatal supervision. In addition to routine antenatal visits, serial monitoring of BP, renal function, cardiac status, and pre - eclamptic screening is vital in such patients. Furthermore, fetal surveillance is also necessary and will include daily fetal kick count, gravidogram, serial fetal biometry, biophysical profile, and fetal Doppler²⁵ controlling BP during pregnancy may be difficult due to the physiological changes in this period. Thus, any patient with TA should plan to conceive when the BP and disease are stable. It is also vital to adjust the antihypertensive medication and avoid angiotensin - converting enzyme inhibitors or angiotensin inhibitors. On the other hand, uncontrolled hypertension during pregnancy has been associated with abortion, stillbirths, aortic

dissection, cardiac and renal insufficiency, stroke, and maternal death.^{26 - 28}

Antihypertensive drugs and antiplatelets can be started as per need, as was in the present case. TA may respond symptomatically to corticosteroid therapy (first line drugs) at a dose of 1 - 2 mg/kg/bodyweight for 4 weeks followed by slow tapering. However, chronic use of corticosteroids could lead to suppression of adrenal gland activity with inadequate release of endogenous corticosteroids in moments of stress, such as surgeries.²⁵ Also immune - suppressors including methotrexate and azathioprine are used.

Finally, vaginal delivery has proven to be the preferred mode of labor management for patients with TA. Additionally, epidural analgesia has been advocated for labor and delivery as well and delivery abbreviated by use of forceps. In our case, decision for emergency LSCS taken in view of USG S/O uteroplacental and fetoplacental insufficiency with prevlscs with short ICP and was uneventful.

Patient was monitored postoperatively in CCU and was transferred to medicine for further management, where patient continued on steroids as she was breast feeding and methotrexate was contraindicated and discharged with advise to monitor BP and follow up with BP charting after 3 months for revascularisation surgery

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