Takayasu Arteritis in Pregnancy: A Rare Case Report

Iata Assudani

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Takayasu arteritis is a rare type of vasculitis, a group of disorder that causes large vessel inflammation. 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 ischeamic attack and aneurysm in aorta. 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 feotal growth restriction. Pregnancy with TA is a medical challenge as the results of American college of rheumatology highlights the serious concerns ie 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 (ACR). 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 a live born female child with low birth weight. Her postpartum course remained uneventful. Despite advancements in cardiovascular management and new drugs, the optimal management for pregnant patients with this disease remains elusive.

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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 a live born female child with low birth weight. Her postpartum course remained uneventful. Despite advancements in cardiovascular management and new drugs, the optimal management for pregnant patients with this disease remains elusive.

I. INTRODUCTION

Takayasu arteritis (TA), also known as "young female arteritis", is a rare and chronic inflammatory disease of large vessels. The disease mainly affects women of reproductive age and Asian origin[1]. Moreover, TA leads to several complications including occlusion as well as aneurysm formation in systemic and pulmonary arteries. Its incidence is reported to be 13 cases per million population[2]. Pregnancy as such has no effect on the evolution of the disease, however, its peak incidence is in second and third trimesters.

Thus, such patients warrant special attention during peripartum period owing to likelihood of development of complications such as hypertension, multiple organ dysfunction and stenosis hindering regional flow leading to restricted intrauterine fetal growth and low birth weight in babies [3-5]. Delay in diagnosis is quite common, so patients often conceive without prior knowledge of having TA or having initiated specific treatment against it [6] Ideal management for pregnant patients with this disease still poses a stringent challenge, an interdisciplinary collaboration of obstretricians, cardiologists, rheumatologists and neurologists is often necessitated for an optimal maternal and fetal prognosis. Here is the case described to enlighten the obstretricians on fetomaternal outcome and management of this infrequent but not uncommon clinical entity encountered nowdays.

II. CASE HISTORY

A 28 years old female married since 7 years, spontaneous conception with 3 months of gestational age with known case of chronic hypertension non-compliant to medications was referred in view of renal artery color doppler suggestive of bilateral renal artery stenosis at origin. Atherosclerotic changes in the form of intimal thickening and calcification in the intima noted in the abdominal aorta and proximal superior mesenteric artery with surrounding tissue edema ? Aortoarteritis.

On examination, general condition of the patient was fair with pulse rate of 74/minute, BP being 160/100mmof hg in the left arm and 140/90mm of hg in right arm with negative urine albumin and pre-monitory signs and symptoms with

normal DTR, Spo2 99% on RA, absent pallor/ icterus/ edema.

Findings of CVS and RS were within normal limits

On P/A examination: soft, No GTR scar of previous LSCS+

P/S: cervix vagina healthy

P/V: uterus 12 weeks

B/L fornix free, non tender

She was admitted for further evaluation and management.

On admission, all routine investigations were within normal limits.

USG 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 with contrast.

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

USG KUB with renal doppler S/O hemodynamically significant B/L renal artery stenosis (right>left)

Carotid artery doppler unremarkable

MR Angiography 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.

2D echo S/O normal study with left ventricular ejection fraction 60%

Fundoscopy was done: Within Normal limits

USG obs NT scan done was S/O SLIUG 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.

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

Repeat rheumatology refernce 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 for emergency LSCS at 31 weeks of gestation in view of USG S/O uteroplacental and fetoplacental insufficiency with prev LSCS 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 post-operative monitoring which was uneventful. Interventional radiologist reference done in view of definitive management of Takayasu arteritis and was advised PET CT and stenting after funds are available.

Patient was discharged with baby on day 36 PNC.

III. DISCUSSION

Takayasu arteritis was first described in 1908 by 2 Japanese opthalmologists, 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 hormone (more common in young females) and genetics (demonstrated by the predisposition of the human leukocyte antigen-HLABW52).

Disease progression typically occurs in various stages from acute inflammatory arteritis to lymphocytic infiltration, intimal thickening, elastic tissue destruction, fibrosis and patchy minimal narrowing of arteries.

Depends on angiographic classification there are five types based on the involvement arteries [1,7] .

Type I involves branches of aorta

Type IIa involves ascending aortoaortic arch and its branches.

Type IIb involves type IIa and thoracic descending aorta.

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

Type IV involves abdominal aorta, renal arteries or both.

Type V involves entire aorta and its branches. 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 [8] . The increased intravascular volume seen during pregnancy may impair circulation and exacerbate maternal hypertension, aortic regurgitation, and congestive heart failure [9].

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 Takaysu 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% [10,11,12].

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 [13]

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[14]. But this is currently not available in our hospital.

However, the gold standard for diagnosis still remains as vessel biopsy [10] which could not be performed in our case.

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 [15]. 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 Doppler [16]

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 [17-19].

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 [20]. 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 previous lscs 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 revasvularisation surgery.

REFERENCES

1. Matsumura A, Moriwaki R, Numano F – Pregnancy in takayasu arteritis from the view of internal medicine. Heart Vessels. 1992;7 (suppl):120124.
2. M.J.koster, E.L. Matteson, and K.J. Warrington, “recent advances in the clinical management of giant cell arteritis and Takayasu arteritis” Current opinion in rheumatology, vol 28 no3, pp 211-217, 2016 [Google Scholar]
3. Y. Beilin and H. Bernstein, “successful epidural anaesthesia for a patient with Takayasu’s arteritis presenting for caesarean section” Canadian journal of anaesthesia, vol 40, no.1, pg 64-66, 1993 Publisher Site | Google Scholar.
4. S. Kathirvel, S. Chavan, V.K. Arya et al, “anesthetic management of patients with Takayasu arteritis : a case series and review” anaesthesia and analgesia, vol 93, no. 1 pp 60-65, 2001.
5. V. suri, N. Agrawal, A. Keepanasseril, S. Chopra, R. Vijayvergiya, and S. jain “Pregnancy and takayasu arteritis: a single centre experience fron north india” Journal of obstretrics and gynecology research, vol 36, no 3, pp519-524, 2010.
6. A.P.L. Assad, T.F.Da silvia, E. Bonfa, and R.M.R. perreria, “maternal and neonatl outcomes in 89 patients with Takayasu arteritis: comparision before and after the TA diagnosis”, the journal of rheumatology, vol 42, no 10, pp 1861-1864, 2015.
7. Wilke Ws – large vessel vasculitis (giant c ell arteritis, takayasu arteritis). Baillieres Cl in Rheumatol. 1997;11:285-313 Kraemer B, Abele H, Hahn M, Rajab T, Kraemer E, Wallweiner D et al.
8. Kraemer B, Abele H, Hahn M, et al. A successful pregnancy in a patient with Takayasu’s arteritis. Hypertens Pregnancy. 2008;27:247–252.[PubMed] [Google Scholar]
9. Matsumara A, Moriwaki R, Numano F. Pregnancy in Takayasu arteritis from the

view of internal medicine. Heart Vessels Suppl. 1992;7:120–124. [PubMed] [Google Scholar]
10. A successful pregnancy in a patient with Takayasu's arteritis. Hypertens Pregnancy 2008; 27: 247-52. Sato EL. Hatta FS, Levy-Neto M. Demographic, clinical and angiographic data of patients with Takayasuarteritis in Brazil. Int J Cardiol. 1998;66:67-70.
11. Mohamed AP, Michael LC, Mikhail E. A case of Takayasuarteritis causing sub clavian steal and presenting as syncope. J Emer Med. 2011;40:158-61.
12. Langford AC, Fauci AS. The Vasculitis Syndrome S. In; Lango LD, Kasper DL, Jameson JL, Fauci AS, Hauser SL, LoscalZo J. Eds Harrison‟s principles of Internal Medicine 18th edition. Mc Graw Hill; 2015.
13. B. K. Sharma, S. Sagar, A. P. Singh, and S. Suri, “Takayasu arteritis in India,” Heart and Vessels, vol. 7, supplement 1, no. 1, pp. 37–43, 1992. View at: Publisher Site | Google Scholar
14. F. A. Oner, S. Z. Aydin, and H. Direskeneli, “Recent advances in Takayasu's arterit is,” European Journal of Rheumatology, vol. 1, pp. 24–30, 2015. View at: Google Scholar.
15. Takayasu’s arteritis in pregnancy: a rare case report from a tertiary care infirmary in India. Marwah S, Rajput M, Mohindra R, Gaikwad H, Sharma M, Topden S. Case Rep Obstet Gynecol. 2017:1–6. [PMC free article] [Pub Med] [Google Scholar]
16. Vasculitis or fibromus culardysplasia? Papandony MC, Brady SR, Aw TJ. Med J Aust. 2015;202:100–101. [PubMed] [Google Scholar]
17. Takayasu’s arteritis in pregnancy complicated by peripartum aortic dissection. Lakhi NA, Jones J. Arch Gynecol Obstet.2010;282:103-106. [PubMed] [Google Scholar]
18. F. A. Oner, S. Z. Aydin, and H. Direskeneli, “Recent advances in Takayasu's arteritis,” European Journal of Rheumatology, vol. 1, pp. 24–30, 2015. View at: Google Scholar.
19. Pregnancy and Takayasu arteritis: a single centre experience from North India. Suri V, Aggarwal N, Keepanasseril A, Chopra S, Vijayvergiya R, Jain S. J Obstet Gynaecol Res. 2010;36:519–524. [PubMed] [Google Scholar].
20. A. Ioscovich, R. Gislason, A. Fadeev, S. Grisaru-Granovsky, and S. Halpern, “Peripartum anesthetic management of patients with Takayasu's arteritis: case series and review,” International Journal of Obstetric Anesthesia, vol. 17, no. 4, pp. 358–364, 2008. View at: Publisher Site | Google Scholar.