Original Article

Takayasu Arteritis In Pregnancy

Dr. Avisha Malu[1], Dr. Dipak S Kolate[2], Dr. Meenal Patvekar[3], Dr. Kale Dhana Laxmi[4*]

[1]Junior Resident, Dept of Obstetrics and Gynaecology, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Pimpri, Pune, India,Email Id:maluavisha@gmail.com

[2]Associate Professor, Dept of Obstetrics and Gynaecology, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Pimpri, Pune, India, Email Id: drdkpune@gmail.com

[3]Professor, Dept of Obstetrics and Gynaecology, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Pimpri, Pune, India, Email Id:mpatvekar@gmail.com

[4*]Junior Resident, Dept of Obstetrics and Gynaecology, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Pimpri, Pune, India, Email Id:ldhana96@gmail.com

***Corresponding Author: -** Dr. Kale Dhana Laxmi

*Junior Resident, Dept of Obstetrics and Gynaecology, Dr. D.Y. Patil Medical College, Hospital and Research Centre, Pimpri, Pune, India. Email Id:ldhana96@gmail.com
DOI: 10.47750/pnr.2023.14.S01.88

Abstract

Takayasu's arteritis (TA) is a rare, systemic, chronic inflammatory arteriopathy of large and medium vessels. It affects women of reproductive age and causes narrowing, occlusion, and aneurysms of systemic and pulmonary arteries, especially the arch of the aorta and its branches. A can lead to an increased risk of cardiovascular complications and neurological manifestations especially during conception, further impacting both maternal and perinatal outcomes. Here, we are presenting a rare case of a TA in pregnancy in a 29-year-old primigravida with postpartum neurological sequelae, who was closely monitored and managed at our tertiary care centre and ultimately enjoyed a favourable outcome. We noted the importance of detailed history, pre-conceptional counselling, multi-disciplinary care, and prompt management at the tertiary care institute.

Keywords: Takayasu's arteritis (TA), Maternal and perinatal outcome, multi-disciplinary care

INTRODUCTION

TA is also known as "young female arteritis," and "pulseless disease" and is a rare and chronic inflammatory disease of the large vessels like the aorta and its branches, coronaries, and pulmonary artery causing progressive inflammation. [1] The disease mainly affects women of the reproductive age group. The incidence of TA is 2.3 per million persons per year, with male to female ratio being 1:9. [3] TA leads to several dreadful events including aneurysm formation and occlusion in systemic and pulmonary arteries. During pregnancy, there is an increased risk of cardiovascular complications such as hypertension, pre-eclampsia, congestive heart failure, aortic aneurysm and dissection, IUGR, and IUD. Pregnancy-associated physiological changes can further aggravate underlying active TA. Therefore, peri-conceptional counselling and early identification of the disease and its complications are the key to a successful gestational outcome. In addition, optimal outcomes during conception require a multidisciplinary approach with an obstetrician, radiologist, cardiologist, rheumatologist, and vascular surgeon.

CASE REPORT

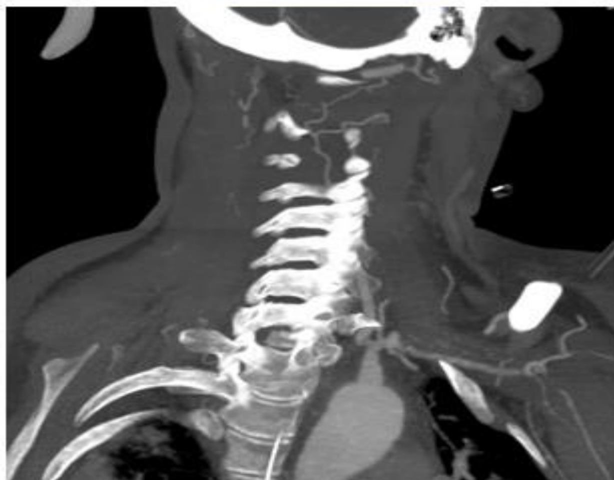
A 29- year-old primigravida, booked ANC, at 37 weeks of gestation presented at the labor room with a complaint of pre-labor rupture of membrane (PROM) with gestational diabetes mellitus (GDM) on insulin since 1 week. On examination her general condition was fair, afebrile, pulse-88 bpm, BP-120/80 mm of Hg. Cardiovascular and respiratory examinations were normal. She did not have any cardiovascular or neurological complaints on admission and did not give a history of any disease or treatment in the past.

All routine investigations were under normal limits and the patient was taken for LSCS (lower segment cesarean section) in view of PROM and CPD (cephalo-pelvic disproportion). The intraoperative period was uneventful, she delivered a healthy male child of 2.8kg. Postoperatively the patient was kept under observation, vitals were stable and the patient was shifted to the ward, and the antibiotics were initiated as per protocol. Suddenly she started complaining of dizziness followed by up rolling of her eyes, tongue bite, and jerky movements of all limbs. She became unresponsive after the episode of seizure. Her Pulse-94 bpm in her right arm and 60 bpm in the left arm, BP-200/120 bpm in the right arm, spo2-70%, and vascular bruit present in the left carotid and femoral arteries. Code blue was activated, and the patient was intubated and shifted to SICU immediately.

All the above findings simulated eclampsia, but how could it be, some pieces of the story must be definitely missing. On further repeated inquiry, the husband revealed the history of TA since 2016.
Further investigations revealed: -
CRP-24.8mg/l, ESR-77mm/hr, ANCA- negative, ANA-Weak Positive,
Fundal examination – Normal
Chest X-RAY – Normal

2D ECHO- IVC dilated >16mm, dilated DTA, cardiac chamber size and wall thickness normal, and left ventricular systolic function normal

Figure 1



CT Angiography of Neck and Brain (Figure 1)**:** Circumferential wall thickening in aortic arch measuring 2.7 mm. Circumferential wall thickening of left subclavian artery extending from its origin proximally to visualized left axillary artery causing mild luminal narrowing (2mm).
Circumferential wall thickening (2mm) of left common carotid artery extending from its origin till its bifurcation causing mild luminal narrowing.

The brachiocephalic trunk and proximal right subclavian artery show mild dilatation. ?? pseudo aneurysm. The right common carotid artery shows marked circumferential thickening (4.9 mm) causing 50% stenosis in its entire extent up to carotid bifurcation. Right ECA AND ICA (cervical portion) appear smaller in caliber. The cavernous portion of the right ICA shows mild narrowing.
Findings were suggestive of TA likely type 2 TA.

MRI Brain and venogram –
Reversible cerebral vasoconstriction syndrome and the possibility of atypical posterior reversible encephalopathy syndrome.
The patient was evaluated by the cardiologist, neurologist, and rheumatologist started on Metoprolol and steroids extubated in 24 hours and stabilized.

DISCUSSION

Women with TA are known to have poor obstetric outcomes, as witnessed in our case. However, favourable pregnancy outcomes have also been reported in various studies, poignantly many reports from developing countries show a high prevalence of severe complications [1] as observed in our case. Type V with predominant abdominal aorta is most common in Indian patients. [4]

Patients with TA may be asymptomatic or may present with signs and symptoms of fatigue, myalgia, arthralgia, dizziness, low-grade fever, claudication, and visual defects. [3] On examination, there may be a feeble or absent peripheral pulse on one side of the body, blood pressure variation in both limbs, and bruit on the affected side. Clinical diagnosis is braced by high levels of acute-phase reactants (ESR & CRP), and radiological demonstration of vessel stenosis. The imaging modalities include color doppler, Computerized Tomography (CT), magnetic resonance (MR) angiography, and PET scan. CT scan is associated with radiation exposure and contrast use is not safe during pregnancy. The gold standard for diagnosis is the biopsy of the affected vessel. Diagnosis of TA relies on clinical presentation, characteristic structural arterial abnormalities, and evidence of inflammatory vasculopathy on imaging or histopathology.

Clinical assessment in Takayasu arteritis was measured by the 'historic' National Institute of Health (NIH) criteria or so-called Kerr criteria [5] which are now realized to be insufficient. Thus, The Indian Takayasu Clinical Activity Score

(ITAS2010), based originally on the Birmingham Vasculitis Activity Score (BVAS), was developed and validated as the current composite index to assess disease activity in Takayasu arteritis. [6] Indian Takayasu clinical activity scoring (ITAS): A clinical scoring system formulated by the Indian rheumatology association vasculitis group in March 2010, to study disease activity. Any new clinical manifestations of flare that have occurred over the previous three months are documented. A score ≥2 is considered an active disease ITAS [7]. A scoring, ITAS including acute phase reactants (ESR, CRP). Our patient had an unsupervised pregnancy presented directly during 3rd trimester in stage II TA and in active disease ITAS score being 10 points.

Pregnancy in women with Takayasu's arteritis is associated with feto-maternal morbidity. Maternal complications like hypertension, pre-eclampsia, eclampsia gestational Diabetes, Premature rupture of membranes, Prolonged labor, Jaundice, Polyhydramnios, Post-Partum Haemorrhage, and Ante-Partum Haemorrhage. Fetal complications like Low Birth, Preterm, Congenital malformation, Meconium-stained liquor, Neonatal sepsis, and Neonatal jaundice.

Various studies have asserted hypertension as the most common complication and the need for targeted treatment for high blood pressure, the same way in our case.
Similar studies by Garikapati et al. [8] observed hypertension in 90% of their patients and Singh et al. [9] found in 90% of patients renal involvement in addition to hypertension.

Controlling BP during pregnancy may be difficult due to the physiological changes in this period. Thus, any patient with TA should plan to conceive only when the BP and disease are stable or the disease is in the remission phase. It is also vital to adjust the antihypertensive medication and avoid angiotensin-converting enzyme inhibitors or angiotensin inhibitors.[1] On the other hand, uncontrolled hypertension during pregnancy has been associated with abortion, stillbirths, aortic dissection, cardiac and renal insufficiency, stroke, and maternal death.

Also noteworthy is that although pregnancy does not have much effect on the progression of TA, the disease usually becomes apparent in 2nd and 3rd trimesters. Several studies concluded that TA pregnancies have favourable outcomes, especially when in the remission phase. It should be remembered that relapse of the disease is not uncommon in pregnancies, even in puerperium as in our case. Pregnancy is also associated with alteration in hematologic and cardiac functions, therefore cardiovascular injury and thromboembolic events are feared complications of TA.[1]

The management of TA is aimed to control active phase inflammation, prevention, and treatment of complications, and revascularization of vessels surgically by the interdisciplinary team including a physician, obstetrician, vascular surgeon, and hematologist. Antenatal check-up includes serial BP monitoring, renal and cardiac functions along with routine antenatal care. Fetal surveillance is also imperative with biophysical profile, daily fetal movement count, biometry, and doppler as per need.

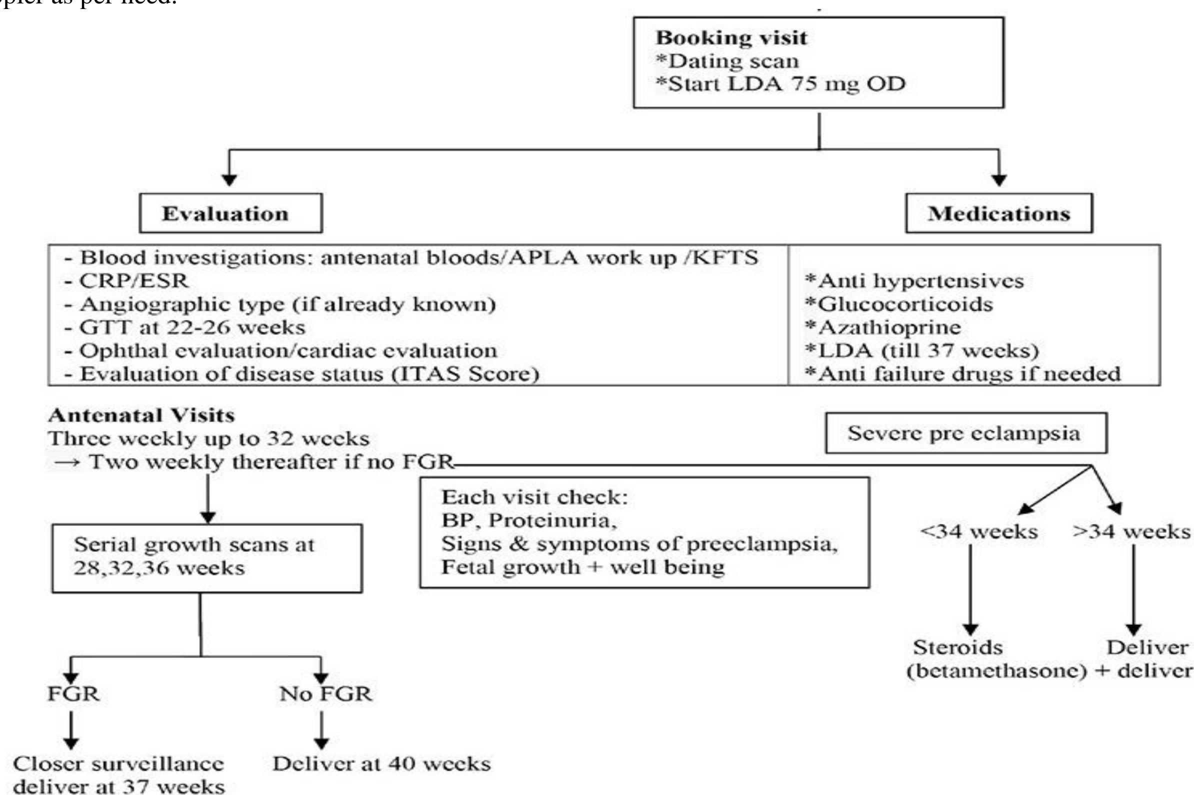


Figure 2: - Algorithm for obstetric management in women with TA **[13]**

Vaginal delivery with epidural analgesia is the preferred mode of delivery in stages I, and II. In patients with higher stages, cesarean section is preferred to prevent cardiac decompensation during labor.

Incorporation of the Wong's prognostic scoring system can be useful to predict outcomes and the need for high-priority care. The study from eastern India also found a similar significance of the Wong score. [10] The other study from Chandigarh, India also found that abdominal aorta involvement (one of the components of Wong's score) portends poorer obstetric outcomes [11] also noteworthy that the Wong score >4 predicts low birth weight and the use of this simple tool can improve perinatal outcomes. In our case, wong's score was 5.

Table 1 Wong's prognostic scoring for neonate born to mother with Takayasu's arteritis

Score	Involvement of abdominal aorta	Trimester when treatment started	Highest Mean arterial pressure in 3rd Trimester	Super-imposed pre-eclampsia
0	No	1st	< 100	None
1	Yes	2nd	101–130	3rd Trimester
2	Yes + Renal involvement	3rd	> 130	1st-2nd Trimester

Table 1: - Wong's prognostic scoring for neonates born to mothers with TA [**14**]

Our case was an eye opener towards the lack of pre-conceptional counselling, misleading history, irregular ANCs, education, and awareness regarding TA and its effect on pregnancy and its effect on maternal and fetal morbidity as also concluded in a study

The main objective of periconceptional counselling is to assess disease activity; optimal control of blood pressure and change over to safer drugs. [20] On the contrary in our case, poor outcomes could be partially attributed to a lack of patient awareness about the need for periconceptional counselling, planned pregnancy as well as the prolonged diagnosis to pregnancy interval. Unfortunately, this is not an uncommon scenario in the developing world where women do not have easy access to specialized health care.

Close antenatal, intrapartum, and postpartum monitoring, with strict control of hypertension, and immediate surgical intervention whenever required, could prevent maternal deaths, as was observed by Lakhi and Jones [6] and Shafi et al. [8] in their case reports.

CONCLUSION

The current case scenario adds further evidence to the importance of detailed history and past health records as well as the medical management of TA during pregnancy, the peripartum period. It also denotes the necessity of pre-conceptional counselling.

Emphasizing early diagnosis, pre-conceptional counselling, conceptions during the remission phase of TA and a multidisciplinary approach for TA patients has proven pivotal role to reach optimal and favourable maternal and fetal outcomes.

Keeping in account the variability of individual response to the disease, including individual disease extension, and disease activity the risk of pregnancy and its effect on the mother and the fetus should be individually assessed prior to the planning of pregnancy.

It is the duty of the medical professional to educate the patient about the expected pregnancy outcomes and associated risks, Improve awareness, and the need for pre-conceptional counselling.

REFERENCES

1. Pregnancy with Takayasu Arteritis and Heart Disease, Venkateswaran S, Journal of postgraduate Gynecology and Obstetrics.
2. Johnston SL, Lock RJ, Gompels MM. Takayasu arteritis: a review. J Clin Pathol. 2002; 55(7): 4816.
3. Takayasu Arteritis, Trinidad B, Surmachevska N, Lala V. Takayasu Arteritis. 2022 Aug 8. In: StatPearls [Internet]. Treasure Island (FL): StatPearls Publishing; 2022 Jan–. PMID: 29083666.
4. Goel R, Danda D, Joseph G, Ravindran R, Kumar S, Jayaseelan V, et al. Longterm outcome of 251 patients with Takayasu arteritis on combination immunosuppressant therapy: single Centre experience from a large tertiary care teaching hospital in southern India. Semin Arthritis Rheum. 2018;47(5): 718–26.
5. Kerr GS, Hallahan CW, Giordano J, et al. Takayasu arteritis. Ann Intern Med 1994; 120:919–929.
6. Misra R, Danda D, Rajappa SM, et al. Development and initial validation of the Indian Takayasu Clinical Activity Score (ITAS2010). Rheumatology (Oxford) 2013; 52:1795–1801.
7. Mekinian A, Resche-Rigon M, Comarmond C, Soriano A, Constans J, Alric L, et al. Efficacy of tocilizumab in Takayasu arteritis: a multicenter retrospective study of 46 patients. J Autoimmun 2018; 91:55–60
8. Garikapati K, Kota LN, Kodey PD. Pregnancy in Takayasu arteritis maternal and fetal outcome. Int J Reprod Contracept Obstet Gynecol 2017; 5: 2596-600.
9. Singh N, Tyagi S, Tripathi R, Mala YM. Maternal and fetal outcomes in pregnant women with Takayasu aortoarteritis: does optimally timed intervention in women with renal artery involvement improve pregnancy outcome? Taiwan J Obstet Gynecol 2015; 54:597–602

10. Mandal D, Mandal S, Dattaray C, Banerjee D, Ghosh P, Ghosh A, et al. Takayasu arteritis in pregnancy: an analysis from eastern India. Arch Gynecol Obstet. 2012;285(3):567–71.
11. Suri V, Aggarwal N, Keepanasseril A, Chopra S, Vijayvergiya R, Jain S. Pregnancy and Takayasu arteritis: a single Centre experience from North India. J Obstet Gynaecol Res. 2010;36(3):519–24
12. Doria A, Bajocchi G, Tonon M, Salvarani C, Pre-pregnancy counselling of patients with vasculitis, Rheumatology, Volume 47, Issue suppl_3, June 2008, Pages iii13 -iii15, https://doi.org/10.1093/rheumatology/ken152
13. Liji Sarah David et al Obstetric and perinatal outcomes in pregnant women with Takayasu's arteritis: single centre experience over five years, J Turk Ger Gynecol Assoc 2020; 21: 15-23.
14. Latika Gupta et al Poor obstetric outcomes in Indian women with Takayasu arteritis, Advances in Rheumatology, (2020) 60:17.