Journal of Surgery and Surgical Specialities
Volume 1, Issue 1, March 2025; pp: 31-35
eISSN: 2455-2852 DOI: 10.65129/surgical.v1i1.21

CASE SERIES

The Arterial Odyssey: Navigating Pregnancy in Takayasu Arteritis - A Case Series

Kakulavaram Sandhya and S. Valarmathi*

Department of Obstetrics and Gynaecology, Madurai Medical College, Madurai – 625020, Tamil Nadu, India; valar1206@gmail.com

Abstract

Background: Takayasu Arteritis (TA) is a chronic, idiopathic, granulomatous vasculitis primarily affecting the aorta and its branches. The disease predominantly affects young women of reproductive age, creating unique challenges during pregnancy. **Objective:** To describe the clinical profile, disease characteristics, and pregnancy outcomes in women with Takayasu arteritis, and to evaluate factors associated with adverse maternal and fetal outcomes. **Methods:** This retrospective case series was conducted at Government Rajaji Hospital, Madurai, over one year. Six pregnant women diagnosed with Takayasu arteritis based on American College of Rheumatology (ACR) criteria were included. Data was collected from the case sheets and high-risk registers. **Results:** The mean age was 24.1 years (range 21–28). Three women (50%) had active disease during pregnancy. Hypertension was the most common maternal complication (87%), with severe preeclampsia observed in two cases. Fetal complications included fetal growth restriction, stillbirth, and miscarriage. One patient developed azathioprine-induced pancytopenia postpartum. Cesarean section was performed in three cases for obstetric and maternal indications, while one patient had an instrumental vaginal delivery. **Conclusion:** Pregnancy in Takayasu arteritis is a high risk and requires multidisciplinary management. Preconceptional optimisation of disease activity, strict blood pressure control, and the use of safe immunosuppressive therapy are critical for improving maternal and foetal outcomes.

Keywords: Hypertension, Pregnancy Outcomes, Takayasu Arteritis, Vasculitis

1. Introduction

TA is a rare, chronic granulomatous vasculitis of the aorta and its major branches, predominantly affecting young women of reproductive age. Owing to absent or diminished pulses, it is also termed "pulseless disease"[1]. The disease typically presents in two phases:

Inflammatory phase – with nonspecific symptoms such as fever, malaise, and weight loss.

Occlusive phase – with limb claudication, unequal arm blood pressure, vascular bruits, and hypertension.

Diagnosis is based on the ACR criteria, and angiographic classification defines the arterial segments involved. Abdominal aorta and renal artery disease are strongly associated with adverse pregnancy outcomes. Pregnancy in TA is high risk, complicated mainly by hypertension, preeclampsia, Intrauterine Growth Restriction (IUGR), and stillbirth. Maternal risks include severe hypertension, aortic aneurysm, dissection, and stroke. Management requires multidisciplinary care, preconception counselling, optimisation of blood pressure, and use of safe drugs such as corticosteroids and azathioprine. Early diagnosis and careful monitoring are essential for favourable maternal and fetal outcomes[2].

2. Aim and Objectives

To describe the clinical profile and disease characteristics of Takayasu Arteritis in pregnancy.
To assess the impact of disease activity and vascular lesions on pregnancy outcomes.
To evaluate maternal and fetal outcomes.

*Author for correspondence

Article Received on: 05 Jan 2025 Revised on: 25 Jan 2025 Accepted on: 03 March 2025

To identify factors associated with adverse pregnancy outcomes.

3. Review of Literature

TA is a rare, chronic, large-vessel vasculitis of the aorta and its major branches. It predominantly affects women under 40 years of age, thereby posing unique challenges in reproductive health. The disease typically progresses from an early systemic inflammatory phase to a later occlusive phase, characterised by claudication, absent or diminished pulses, vascular bruits, and hypertension[1].

The American College of Rheumatology (1990) criteria remain the cornerstone for diagnosis, requiring at least three of six features, while the angiographic classification (Types I–V) defines the extent of vascular involvement. Renal and abdominal aortic lesions have consistently been associated with poorer pregnancy outcomes.

Pregnancy in TA is considered high risk. Published series report hypertension as the most frequent maternal complication, occurring in 30–80 % of cases, followed by preeclampsia in 10–15 %. Maternal risks such as stroke, aortic dissection, and heart failure are less common but potentially life-threatening. On the fetal side, studies have demonstrated increased rates of miscarriage (15–20 %), intrauterine growth restriction (20–30 %), preterm birth, and stillbirth compared to the general obstetric population. Importantly, active disease at conception and renovascular involvement are recognised predictors of adverse outcomes[3].

Medical management during pregnancy emphasises the use of corticosteroids and azathioprine, which are considered safe, while teratogenic agents such as methotrexate and mycophenolate mofetil are strictly contraindicated. In refractory disease, calcineurin inhibitors like tacrolimus or cyclosporine have been employed. Antihypertensive therapy with labetalol, nifedipine, or methyldopa forms the backbone of management, alongside low-dose aspirin for preeclampsia prevention.

With regard to delivery, vaginal birth is preferred in stable cases, whereas cesarean section is reserved for obstetric indications or severe vascular/cardiac complications. Epidural anaesthesia is usually recommended due to better hemodynamic stability[3].

4. Materials and Methods

Study Design: Retrospective case series.
Setting: Government Rajaji Hospital, Madurai.
Duration: 1 year.
Sample Size: 6 pregnant women.
Inclusion Criteria: Pregnant women with Takayasu arteritis diagnosed using the ACR criteria and managed during pregnancy.
Exclusion Criteria: Non-pregnant TA cases and patients with other autoimmune/vasculitic disorders.
Data Source: Case sheets and high-risk registers.

5. Results (Including Observations)

A total of six pregnant women with Takayasu arteritis were included in this retrospective case series. The clinical profile, angiographic classification, management, and outcomes were analysed.

Demographic and Clinical Characteristics: Table 1
Age range: 21–28 years.
Parity: Five patients were primigravidae; one was a second gravida with a previous abortion.
Time of diagnosis: Two patients were diagnosed before conception, one at 8 weeks, two during early pregnancy (12–20 weeks), and one at 32 weeks of gestation.
Symptoms at presentation: Hypertension was the most common symptom (4/6). Claudication was noted in three cases. Two patients were asymptomatic and diagnosed on routine evaluation.

Pulse Abnormalities and Blood Pressure:
Absent or feeble pulses were observed in all patients: absent femoral in one, absent radial in four, feeble radial in one, and carotid bruits in two.
Blood pressure recordings showed severe hypertension in five patients (systolic ≥160mmHg in at least one limb). One patient had a significant inter-limb Blood Pressure (BP) discrepancy.

Comorbidities:
Four patients had chronic hypertension.
Three patients had hypothyroidism, and one additionally had moderate anaemia.

Angiographic Classification:

Table 1. Demographic and clinical characteristics

Parameters	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Age	27	22	21	28	25	22
OBS Code	G2a1	Primi	Primi	Primi	Primi	Primi
Time of Diagnosis	8 Weeks	32 Weeks	20 weeks	Prepregnancy	12 Weeks	Prepregnany
Present in Symptoms	Hypertension Claudication	Hypertension	Asymptomatic	Claudication	Asymptomatic	Claudication
Pulse Abnormalities	Absent Femoral	Absent Radial, Carotid bruit	Absent Radial	Absent Radial, Carotid Bruit	Feeble Radial	Absent Radial
BP at Our Institution (1st Visit)	rt ul-160/100, rt ll nr, lt ul 150/80, lt ll 150/90	rt ul-160/90, rt ll 150/90, lt ul nr, lt ll 150/90	rt ul 130/80, lt ul 150/90	rt ul 160/100, lt ul nr	rt ul 130/80, lt ul 150/70	rt ul 140/80, lt ul 170/100
Comorbidities	Hypothyroid Chronic Hypertension	Hypothyroid Chronic Hypertension	Moderate anemia hypothyroid chronic hypertension	Hhronic hypertension	None	Chronic Hypertension

Table 2. Disease classification and treatment

	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Angiographic Classification	Type 2 a	Type 1	Type 2 a	Type 4	Type 1	Type 2 b
Disease Activity	Stable	Active	Active	Active	Stable	Active
Immuno Suppressive Therapy	Steroids + Aza	Steroids + Aza	Steroids + Aza	Steroids + Aza	Steroids	Steroids + Aza
Anti Hypertensives	Labetalol + Nifidipine	Labetalol	Labetalol	Labetalol + Nifidipine	---	Labetalol + Nifidipine

Type 1: Two cases.
Type 2a: Two cases.
Type 2b: One case.
Type 4: One case.
Renal/abdominal involvement (Type 4 and 2b) was associated with adverse outcomes.

Disease Activity: Table 2
Stable disease: 2/6 (33%). Active disease: 4/6 (67%).
Active disease at conception correlated with poor maternal and fetal outcomes.

Medical Management: Table 2
Immunosuppressive therapy: Five patients (83%) received steroids plus azathioprine; one patient (17%) was managed on steroids alone.
Antihypertensives: Labetalol was used in all hypertensive cases, with nifedipine added in three patients.
Drug toxicity: One patient developed azathioprine-induced pancytopenia, managed successfully with Drug withdrawal, transfusions, and Granulocyte Colony Stimulating Factor (G-CSF) support.
Maternal Complications: Table 3.
Hypertension was observed in five patients (87%). Severe preeclampsia occurred in two cases.
Uncontrolled hypertension persisted in two cases despite therapy. No maternal deaths were reported.

Fetal and Neonatal Outcomes: Table 3.

One case of Fetal Growth Restriction (FGR). One stillbirth at 36 weeks.
One miscarriage at 8 weeks.
One Medical Termination of Pregnancy (MTP) at 8 weeks due to active disease. Two-term live births with normal birth weight (2.56kg and 3.1kg).
One preterm live birth at 36 weeks with low birth weight (1.7kg).

Mode of Delivery: Table 3.

Table 3. Fetal maternal outcome

Parameters	Case 1	Case 2	Case 3	Case 4	Case 5	Case 6
Maternal Complications	Hypertension	Severe Preeclampsia	Severe Preeclampsia	Uncontrolled Hypertension	None	Uncontrolled Hypertension
Fetal Complications	None	Fgr	Still Birth	None	None	Miscarriage
Mode of Delivery	Emergency Lscs	Emergency Lscs	Emergency Lscs	Mtp	Instrumental Delivery	---
Ga at Delivery	37 weeks	36 weeks	36 weeks	---	38 weeks	---
Birth Weight	2.56 kg	2.3 kg	1.7 kg	--	3.1 kg	--
Post Partum Complications	None	Drug-Induced Pancytopenia	None	None	None	None

Cesarean section (3/6): Indications were cephalopelvic disproportion, severe preeclampsia, and abruption placentae.
Vaginal delivery (1/6): Instrumental delivery under epidural anaesthesia. MTP (1/6): At 8 weeks of gestation. Miscarriage (1/6): Early pregnancy loss.

Postpartum Complications: Table 3.

One patient developed drug-induced pancytopenia postoperatively. No other major postpartum complications were noted.

6. Discussion

Pregnancy in women with TA is rare but poses significant challenges for both the obstetrician and rheumatologist. The disease is most prevalent in women of reproductive age, and the associated vascular lesions, hypertension, and immunosuppressive therapy can adversely affect maternal and fetal outcomes[4].

In the present series of six cases, hypertension emerged as the most frequent maternal complication, affecting 87% of patients, which is consistent with previous studies where hypertension was observed in up to 90% of pregnancies with TA. Severe preeclampsia was seen in two women, while uncontrolled hypertension persisted in two others despite therapy, underscoring the importance of early detection and strict blood pressure control.

Disease activity at conception was an important determinant of outcome. Half of our patients had active disease, and these cases were associated with adverse events such as fetal growth restriction, stillbirth, miscarriage, and need for medical termination of pregnancy. This observation is in line with published literature, which emphasises that conception during disease remission is associated with more favourable maternal and perinatal outcomes (Hidaka *et al.*)[5].

With regard to angiographic classification, abdominal and renal artery involvement (Types 4 and 2b) correlated with poorer outcomes, supporting the evidence that vascular distribution significantly impacts prognosis. Preconception surgical repair of affected arteries has been recommended in some reports to improve outcomes, although this was not performed in our series.

Pharmacological management was largely based on corticosteroids and azathioprine, both considered relatively safe in pregnancy. Five out of six women (83%) required combined therapy, while one was maintained on steroids alone. However, one patient developed azathioprine-induced Pancytopenia which was managed successfully with drug withdrawal, transfusion support, and granulocyte colony-stimulating factor. This highlights the importance of vigilant monitoring for drug toxicity. Antihypertensives used included labetalol and nifedipine, consistent with standard recommendations.

Fetal outcomes were variable. Two patients delivered healthy term neonates, but complications included one case each of intrauterine growth restriction, stillbirth, miscarriage, and early termination. This variability reflects the burden of active disease and renovascular involvement, which compromise uteroplacental perfusion and increase the risk of fetal compromise[6].

Mode of delivery was individualised: three women underwent cesarean section for obstetric or medical indications, while one achieved instrumental vaginal delivery under epidural anaesthesia. Vaginal delivery is feasible in stable disease without major cardiovascular

involvement, while cesarean is reserved for obstetric or high-risk vascular indications.

Overall, the findings reinforce the consensus that pregnancy outcomes in TA are largely determined by disease activity, blood pressure control, and vascular distribution of lesions. Preconception counselling with optimisation of disease status, timely switching to pregnancy-safe medications, and close multidisciplinary monitoring throughout gestation remain the cornerstones of management.

7. Summary and Conclusion

7.1 Summary

This case series analysed six pregnancies complicated by TA. Hypertension was the predominant maternal issue (87%), with severe preeclampsia and uncontrolled hypertension contributing to morbidity. Half the women had active disease at conception, correlating with adverse outcomes including fetal growth restriction, stillbirth, miscarriage, and medical termination. Most required steroids with azathioprine; one developed drug-induced pancytopenia. Deliveries included three cesarean sections and one vaginal birth.

7.2 Conclusion

Pregnancy in TA is high risk, largely influenced by disease activity, vascular involvement, and blood pressure control. Preconception counselling, safe immunosuppression, strict monitoring, and multidisciplinary care are vital for achieving favourable maternal and fetal outcomes[7].

8. References

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