Takayasu's arteritis in pregnancy: A case report

J A Locher,[1] MB ChB, FCOG (SA), MMed; **S Bhoora,**[1] MB ChB, FCOG (SA), Cert. Critical Care O&G (SA); **J Zamparini,**[2] MB ChB, FCP (SA), MMed, FRCP

[1] *Department of Obstetrics and Gynaecology, Charlotte Maxeke Johannesburg Academic Hospital and the University of the Witwatersrand, Johannesburg, South Africa*

[2] *Department of Medicine, Charlotte Maxeke Johannesburg Academic Hospital and the University of the Witwatersrand, Johannesburg, South Africa*

Corresponding author: *JA Locher (admin@drjlocher.co.za)*

Takayasu's arteritis (TA) is a large-vessel vasculitis predominantly affecting the aorta and its branches. It most commonly affects women in the second and third decades of life. Although analyses of case series and case studies have been done, no definitive consensus on optimal management has been reached. We describe the clinical course of an 18-year-old woman at 31weeks and 5 days' gestation with active TA Type III (with multiple areas of stenosis, including both renal arteries, and areas of aneurysmal dilatation), uncontrolled hypertension and uveitis, with a favourable maternal and fetal outcome through a multidisciplinary approach.

S Afr J Obstet Gynaecol 2026;32(1):e2228. https://doi.org/10.7196/SAJOG.2026.v32i1.2228

Takayasu's arteritis (TA) is a large-vessel vasculitis predominantly affecting the aorta and its branches. It most commonly affects women in the second and third decades of life.[1] Pregnancy itself does not appear to affect the overall course of the disease,[2] but disease activity appears to be independently associated with a poor pregnancy outcome; more than 5% of pregnant women with TA develop a life-threatening maternal cardiovascular event.[3] Cardiovascular changes in pregnancy, such as increased intravascular volume, may impair circulation and worsen or precipitate aortic regurgitation, severe hypertension and heart failure, with subsequent potential intrauterine growth restriction and abruptio placentae.[4] Although analyses of case series and case studies have been done,[2–5] no definitive consensus on optimal management has been reached. We describe the clinical course of an 18-year-old woman at 31 weeks and 5 days' gestation with active TA Type III (with multiple areas of stenosis, including both renal arteries, and areas of aneurysmal dilatation), uncontrolled hypertension and uveitis, with a favourable maternal and fetal outcome through a multidisciplinary approach.

Case report

An 18-year-old primigravida at 31weeks and 5 days' gestation with normal booking blood tests (HIV negative; haemoglobin = 13.0; rapid plasma reagin negative; Rh positive) was referred to our centre for management of hypertensive urgency in pregnancy. She had been diagnosed with Type III TA a year before after she presented to a secondary hospital with early-onset hypertension and uveitis. A computed tomography (CT) aortogram and peripheral arteriogram at diagnosis showed involvement of the distal thoracic and abdominal aorta, with irregular mural thickening and segmental luminal narrowing distal to the aortic arch and major branches, including the renal arteries, the origin of the coeliac artery (with stenosis and post-stenotic aneurysmal dilatation), the origin of the superior mesenteric artery, and the left common iliac artery. The patient had been initiated on azathioprine at diagnosis, but defaulted on follow-up for a year prior to representing with uncontrolled hypertension at 25 weeks and 5 days' gestation, at which time azathioprine was reinitiated, in addition to prednisone, methyldopa and amlodipine being administered. An echocardiogram at the time of re-presentation showed left ventricular hypertrophy (LVH), grade I diastolic dysfunction and a left ventricular ejection fraction (LVEF) of 51%.

On arrival at our centre, the patient complained of exertional dyspnoea, fatigue and palpitations but no chest pain, headache, dizziness or syncope. She also reported dull pain in both eyes of about 1 month's duration, with intermittent blurred vision. Blood pressure was noted to be markedly elevated, with systolic and diastolic blood pressures of 170 - 190 mmHg and 80 - 90 mmHg, respectively, despite antihypertensive therapy as noted earlier. Oxygen saturation, respiratory rate and pulse rate were within normal ranges and general examination was normal. All pulses were present on cardiovascular examination; however carotid, brachial, radial, popliteal and dorsalis pedis pulses were diminished on the left. A pan-systolic murmur was noted at the mitral area, radiating to the axilla. Renal bruits were auscultated bilaterally, with sounds on the right louder than on the left. Carotid bruits were also auscultated bilaterally, with the left sounding louder than the right. The cardiac apex was displaced to the sixth intercostal space in the anterior axillary line. The symphysis–fundal height was 31 cm, the fetal lie was left longitudinal, with cephalic presentation. All other systems were normal.

Obstetric ultrasound showed an estimated fetal weight of 1 519 g (below the 10th percentile for gestational age) and an umbilical artery resistance index of 0.63 (normal for gestation). The patient was transferred to the obstetric high-care unit and initiated on a labetalol infusion as per local protocols, as well as a magnesium sulphate infusion for neuroprotection of the fetus and also on the suspicion of superimposed pre-eclampsia, given an elevated urine protein–creatinine ratio of 0.050. Laboratory tests for renal, hepatic and haematological function were normal. Repeat echocardiogram confirmed the previous normal LVEF as well as mild mitral and aortic regurgitation, LVH and grade II diastolic dysfunction. An electrocardiogram showed LVH changes of increased R-wave amplitude from the augmented voltage left arm. The patient was not placed on anticoagulation treatment owing to refractory hypertension, with an anticipated imminent delivery. Prednisone and azathioprine were continued under the advice of our rheumatology service.

Four days into admission, cardiotocography was non-reactive. In light of this, as well as the uncontrolled blood pressure, risk of aneurysmal rupture and risk of abruptio placentae, the decision to perform a caesarean section was made following discussion with the patient. Because prednisone had been administered for four days, and also one month prior, additional steroids for fetal lung maturation were not administered. Caesarean section was performed under general anaesthesia, which was decided on instead of spinal anaesthesia because of anticipated severe haemodynamic changes and possible resultant organ ischaemia. Invasive haemodynamic monitoring was performed. Surgery was uneventful, with a good neonatal outcome, with a paediatric team being present at delivery; an alive male was born weighing 1 620 g with Apgar scores of 9, 10 and 10. The neonate was transferred to the neonatal transition unit for monitoring because of low birth weight.

Post operatively the patient was transferred back to the obstetric high-care unit, where her blood pressure improved slightly, and the labetalol was decreased. Methyldopa was stopped post delivery. Blood pressure was labile after surgery, with frequent titration of labetalol infusion needed; however, by the third day post partum, the patient was weaned off intravenous antihypertensive agents and maintained on methyldopa and amlodipine.

A repeat CT aortogram showed multi-level dilatation and narrowing (string-of-pearls sign) of the thoraco-abdominal aorta (Figs 1 - 3); no aneurysmal dilatation was seen but long-segment circumferential wall thickening from the origin of the descending aorta to the infrarenal short-segment stenotic region was noted. Other findings included dilatation of the major branches of the abdominal aorta with proximal stenosis (Fig. 3), penetrating aortic ulcers involving the descending thoraco-abdominal aorta at T6 and T11, and large right renal midpole complex cortical cysts. The left kidney was smaller than the right (9.7 cm v. 11.8 cm). Multidisciplinary team management of the patient was essential in producing the favourable maternal and fetal outcome, with obstetrics, critical care, rheumatology, vascular surgery, nephrology, anaesthesiology and paediatric expertise involved. The patient was discharged from the hospital in a stable condition 10 days post operatively for continued outpatient follow-up with rheumatology and vascular surgery. The neonate was discharged to the mother in a stable condition once the discharge goal weight of 1 800 g was reached. The mother was counselled on contraception options and to book early at our high-risk antenatal clinic prior to conception.

Ethical considerations

Written informed consent was obtained from the patient prior to publication of this case report and all accompanying images. Ethical approval was not required for this single case report.

Discussion

TA is a systemic, granulomatous, inflammatory large-vessel vasculitis, mainly involving the aorta and its major branches, causing stenosis, occlusion and aneurysmal dilatation of large arteries.[6] A relatively rare disease, with a reported incidence in North America of 2.6 cases per million per year, it predominantly affects women of childbearing age,[1] making the interaction between pregnancy and TA important.

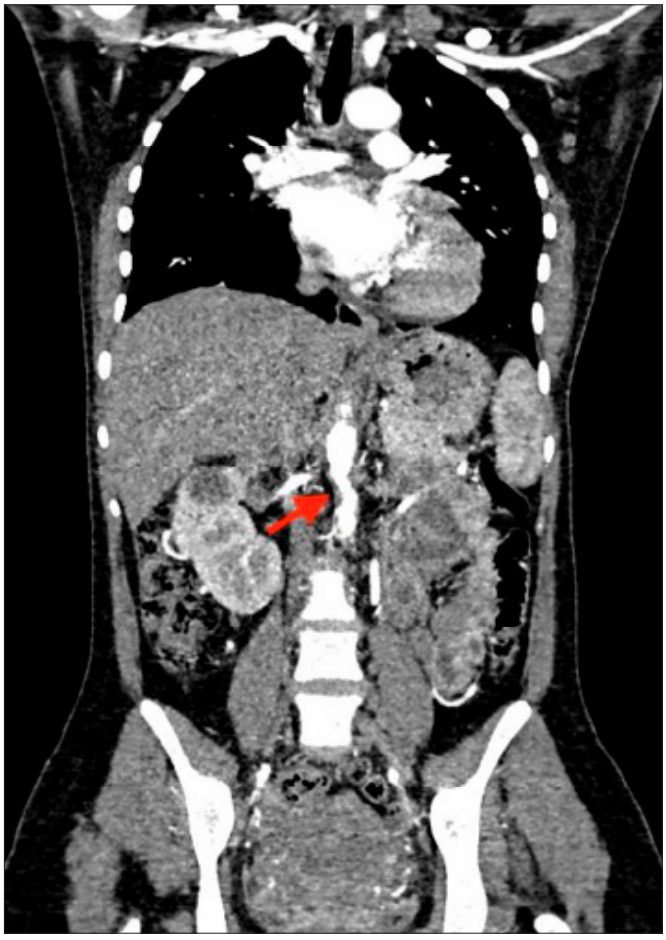

Fig. 1. CT angiogram showing narrowing of the abdominal aorta below the level of the renal arteries

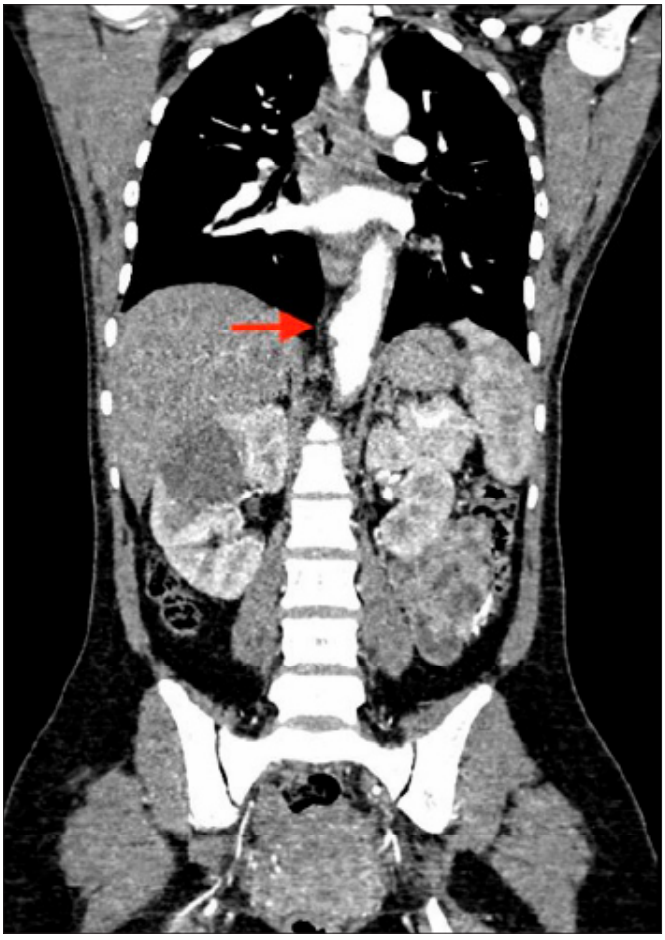

Fig. 2. CT Angiogram showing aneurysmal dilatation of the thoracic aorta

Its aetiology is unknown and its pathogenesis seems mainly due to abnormal cell-mediated immunity, but other molecular and genetic abnormalities may contribute.[6,7] Efforts to identify the antigen(s) that trigger the autoimmune response have been unsuccessful, but it is likely that viruses or bacteria, by a molecular mimicry mechanism, initiate or propagate the response.[7] It is divided into five types depending on vessel involvement.

The majority of patients present with hypertension, resulting from renal artery stenosis, and the differential of TA must be considered in pregnant women with hypertension, especially if decreased or absent pulses are present. The presence of bruits, particularly affecting the carotid, subclavian and abdominal vessels, as well as limb claudication, blood pressure discrepancies and end-organ or limb ischaemia, should also prompt a search for TA. Heart failure associated with hypertension, aortic regurgitation and dilated cardiomyopathy is also seen. Neurological features secondary to hypertension or ischaemia, such as dizziness, focal neurological signs and seizures, may occur.[1]

Fever and elevated inflammatory markers, including C-reactive protein and erythrocyte sedimentation rate, may be seen on presentation, but given the nonspecific clinical and laboratory findings, the gold standard for diagnosis is CT or magnetic resonance (MR) angiography, which identifies vascular wall thickening and enhancement early on, and arterial stenosis, occlusions and aneurysms as the disease progresses.[1,8] Doppler ultrasound is useful for the assessment of vessel wall inflammation.[1]

Assessment of TA disease activity is difficult as there is no effective outcome measure reflecting significant ongoing arterial wall inflammation, with a need for biomarkers.[9] Currently only CT and MR angiography can be used as an accurate follow-up tool;[1] however, recent trials have shown benefit in the use of positron emission tomography-CT to assess disease activity.[10]

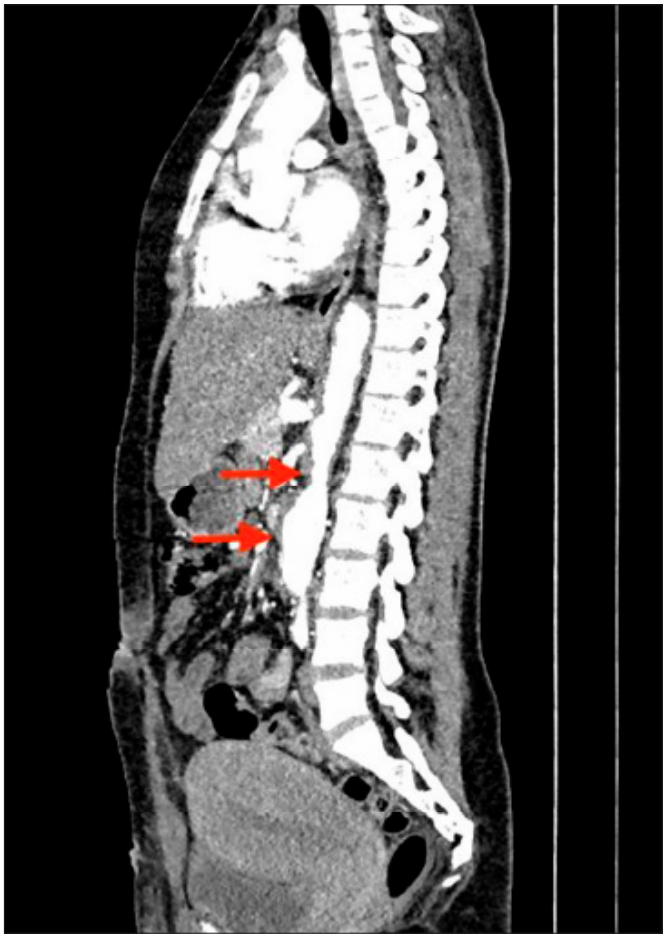

Fig. 3. CT angiogram showing narrowing (top arrow) and dilatation (lower arrow) of the abdominal aorta

Disease activity appears to be independently associated with poor pregnancy outcome;[3] however, pregnancy does not appear to affect the overall course of the disease.[2] More than 5% of pregnant women develop a life-threatening maternal cardiovascular complication.[2] Cardiovascular changes in pregnancy such as increased intravascular volume may impair circulation and worsen or precipitate aortic regurgitation, severe hypertension and heart failure, with subsequent potential intrauterine growth restriction and abruptio placentae.[4] Other maternal complications in one case series included superimposed pre-eclampsia, progressive renal impairment and heart failure.[1] The severity of maternal vessel involvement and hypertension could approximately predict fetal outcome, with poor perinatal outcome specifically associated with abdominal aortic involvement.[1]

Preconception counselling is essential in women with TA in order to time conception to periods of disease regression, stop cytotoxic drugs and ensure optimal disease control.[11] In addition, women should present to a high-risk antenatal unit or clinic as early as possible following conception.

Steroids are widely accepted as the first-line treatment option in patients with TA, and these drugs should be continued in pregnancy. Case series and uncontrolled studies have shown the addition of other 'steroid-sparing' immunosuppressive drugs to be beneficial.[12] Methotrexate, mycophenolate mofetil and cyclophosphamide have recognised teratogenic effects and should be stopped before conception as per published guidelines; however, cyclophosphamide can be used for life- or organ-threatening complications during the second and third trimester.[13] Leflunomide also has detrimental fetal effects and should be stopped 24 months before conception; else cholestyramine washout should be done.[13] Azathioprine, intravenous immunoglobulin, hydroxychloroquine, tacrolimus and cyclosporine appear to be safe in pregnancy.[13,14] Low-dose aspirin is recommended in women with TA and should be started before 16 weeks of gestation for the prevention of new-onset or superimposed pre-eclampsia and hypertension should be managed as per local and international guidelines.[15]

Little information is available regarding the best mode of delivery, but vaginal delivery with or without epidural anaesthetic has been recommended as safe if blood pressure is controlled.[2,15] Patients with TA often have severe increases in systolic blood pressure during uterine contractions and therefore regular blood pressure monitoring should be performed[15] and adequate analgesia given. Uncontrolled hypertension in the second stage of labour is a risk factor for cerebral haemorrhage and aortic dissection, which may be prevented by shortening this stage by forceps or vacuum delivery.[1,3] Caesarean section has been suggested in the case of more severe forms of TA as increased blood pressure and blood volume from uterine contractions could lead to cardiac decompensation.[15]

Conclusion

This case report emphasises the complexity of managing TA in pregnancy, and the importance of early recognition, blood pressure control, and coordinated multidisciplinary care. Despite the challenges of active disease, severe hypertension and extensive vascular involvement, favourable maternal and fetal outcomes were achieved through a multidisciplinary approach, with input from obstetricians, maternal–

fetal subspecialists, obstetric physicians, rheumatologists, cardiologists, vascular surgeons, critical-care specialists and specialist anaesthetists. Optimal outcomes in such high-risk cases depend on preconception counselling, continuity of immunosuppressive therapy and delivery planning in a tertiary setting with access to specialised care. Continued documentation of such cases contributes to refining evidence-based management strategies for this rare but high-risk condition in pregnancy.

Declaration. None.

Acknowledgements. We would like to acknowledge the staff of Charlotte Maxeke Johannesburg Academic Hospital for their dedication to patient care.

Author contributions. All authors contributed to conceptualising the case report and writing the manuscript. SB and JZ supervised the study.

Funding. None.

Conflicts of interest. None.

1. Johnston SL, Lock RJ, Gompels MM. Takayasu arteritis: A review. J Clin Pathol 2002;55(7):481-486. https://doi.org/10.1136/jcp.55.7.481
2. Hauenstein E, Frank H, Bauer JS, Schneider KTM, Fischer T. Takayasu's arteritis in pregnancy: Review of literature and discussion. J Perinat Med 2010;38(1):55-62. https://doi.org/10.1515/jpm.2009.120
3. Comarmond C, Saadoun D, Nizard J, Cacoub P. Pregnancy issues in Takayasu arteritis. Semin Arthritis Rheum 2020;50(5):911-914. https://doi.org/10.1016/j.semarthrit.2020.08.001
4. Kraemer B, Abele H, Hahn M, et al. A successful pregnancy in a patient with Takayasu's arteritis. Hypertens Pregnancy 2008;27(3):247-252. https://doi.org/10.1080/10641950801955741
5. Assad APL, Da Silva TF, Bonfa E, Pereira RMR. Maternal and neonatal outcomes in 89 patients with Takayasu arteritis (TA): Comparison before and after the TA diagnosis. J Rheumatol 2015;42(10):1861-1864. https://doi.org/10.3899/jrheum.150030
6. Serra R, Butrico L, Fugetto F, et al. Updates in pathophysiology, diagnosis and management of Takayasu arteritis. Ann Vasc Surg 2016;35:210-225. https://doi.org/10.1016/j.avsg.2016.02.011
7. Espinoza JL, Ai S, Matsumura I. New insights on the pathogenesis of Takayasu arteritis: Revisiting the microbial theory. Pathogens 2018;7(3):73. https://doi.org/10.3390/pathogens7030073
8. Gotway MB, Araoz PA, Macedo TA, et al. Imaging findings in Takayasu's arteritis. Am J Roentgenol 2005;184(6):1945-1950. https://doi.org/10.2214/ajr.184.6.01841945
9. Li Cavoli G, Mulè G, Vallone MG, Caputo F. Takayasu's disease effects on the kidneys: Current perspectives. Int J Nephrol Renov Dis 2018;11:225-233. https://doi.org/10.2147/IJNRD.S146355
10. Grayson PC, Alehashemi S, Bagheri AA, et al. 18 F-fluorodeoxyglucose-positron emission tomography as an imaging biomarker in a prospective, longitudinal cohort of patients with large vessel vasculitis. Arthritis Rheumatol 2018;70(3):439-449. https://doi.org/10.1002/art.40379
11. Itani R, Elmallahi N, Ramadan MAA, Al Ibrahim A. Pregnancy with Takayasu's arteritis: A case report and literature review. Cureus 2018;10(9):e3370. https://doi.org/10.7759/cureus.3370
12. Águeda AF, Monti S, Luqmani RA, et al. Management of Takayasu arteritis: A systematic literature review informing the 2018 update of the EULAR recommendation for the management of large vessel vasculitis. RMD Open 2019;5(2):e001020. https://doi.org/10.1136/rmdopen-2019-001020
13. Ross C, D'Souza R, Pagnoux C. Pregnancy outcomes in systemic vasculitides. Curr Rheumatol Rep 2020;22(10):63. https://doi.org/10.1007/s11926-020-00940-5
14. Natekar A, Pupco A, Bozzo P, Koren G. Safety of azathioprine use during pregnancy. Can Fam Physician 2011;57(12):1401-1402.
15. Soma-Pillay P, Adeyemo A, Suleman FE. Takayasu arteritis in pregnancy: Case report. Cardiovasc J Afr 2015;26(1):e14-e16. https://doi.org/10.5830/CVJA-2015-003

Received 15 May 2024. Accepted 14 April 2025.