Case Report

Takayasu arteritis in pregnancy

Priya Soma-Pillay, Adekunle Adeyemo, Farhana Ebrahim Suleman

Abstract

Takayasu arteritis is a chronic, granulomatous arteritis affecting large and medium-sized arteries. During pregnancy, maternal and foetal complications are largely as a consequence of maternal arterial hypertension. We present a case of a 35-year-old para one gravida two patient with Takayasu arteritis (group III disease) complicated by chronic hypertension and a severely dilated ascending aorta. Good blood pressure control during pregnancy is an important measure in reducing obstetric morbidity.

Keywords: Takayasu arteritis, pregnancy, hypertension, pre-eclampsia

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Takayasu arteritis is a chronic granulomatous arteritis affecting large and medium-sized arteries. The disease is characterised by inflammation of the blood vessels, resulting in destruction and distortion of the layered components of their walls. During the early stages of the disease, there are mononuclear cell infiltrations in the adventitia and granulomas with Langerhans cells in the media. This is followed by disruption of the elastin layer and subsequent massive medial and intimal fibrosis. These lesions result in segmental stenosis, occlusion, dilatation and aneurysmal formation in the affected vessels.[1]

Stenotic lesions predominate and have been reported in 90% of cases, while aneurysms are only reported in approximately 25%.[2] This is a disease of young adults with a peak onset in the second and third decades of life. A case series reported from South Africa of Takayasu arteritis in childhood demonstrated a 2:1 female-to-male ratio.[3] Patients with Takayasu arteritis may present with a variety of clinical manifestations, but arterial hypertension is the most common feature of the disease.[4]

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Case report

A 35-year-old para one gravida two patient, with a previous uncomplicated full-term delivery at the age of 16 years, was referred to the cardiac-obstetric unit at eight weeks' gestation. She had been diagnosed with Takayasu arteritis six years earlier, and tuberculosis two years previously, for which she was treated. She was hypertensive and her blood pressure was controlled with nifedipine, carvedilol and hydrochlorothiazide. The Takayasu disease was being treated with prednisone and azathioprine.

Further history revealed that her ascending aorta was dilated, and on evaluation by cardiothoracic surgeons, the lesion was considered to be inoperable. The patient was not using any contraception and this was a planned and wanted pregnancy. She had no other medical or surgical history of note.

On examination, the patient was apyrexial with a blood pressure of 120/70 mmHg in both arms and a pulse of 88 beats per minute. Cardiac examination revealed normal first and second heart sounds. No third or fourth heart sounds were heard and there was a one-quarter aortic regurgitation murmur. Respiratory and abdominal examinations were normal.

Ultrasound examination confirmed an intra-uterine pregnancy of eight weeks' gestation. The electrocardiogram was normal. On echocardiography, the patient had good left ventricular systolic function with no regional wall-motion abnormality. There was a tricuspid aortic valve with trivial aortic regurgitation. The ascending aorta was markedly dilated, measuring 5.7 cm. No dissection flap was seen.

The descending aorta and its branches had been evaluated by CT angiography two months prior to pregnancy. The ascending, arch and descending thoracic aorta were dilated (Fig. 1A, B) with marked mural thickening of the thoracic (2.3 cm) aorta (Fig. 2A, B). A laminated thrombus was found on the descending aorta. The renal arteries were patent. No abnormality was detected on fundoscopy.

Laboratory findings showed an elevated erythrocyte sedimentation rate of 56 mm/h and a C-reactive protein level of 9 mg/dl. The full blood count, renal function, electrolytes and urinalysis were normal.

The patient was managed by a multidisciplinary team of cardiologists, obstetricians and rheumatologists. After the initial investigations were performed, the patient was counselled about the aortic lesions. She was informed about the possibility of further dilatation or rupture of the aorta during pregnancy. The patient was offered a termination of pregnancy for medical reasons, which she declined.

The pregnancy was managed further by the multidisciplinary team and the patient was treated with prednisone (10 mg daily) and azathioprine (150 mg alternating with 200 mg daily) for Takayasu disease, methyl-dopa for hypertension, aspirin

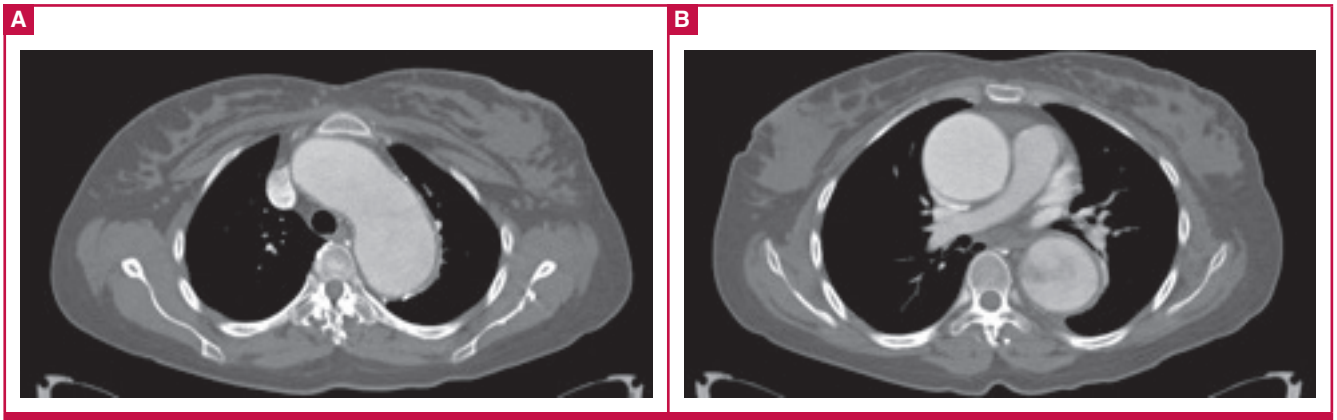

Fig. 1. Axial CT post-contrast images in the arterial phase demonstrating aneursymal dilatation of the arch of the aorta (A) and the ascending and descending aorta (B). Note the turbulent flow in the descending aorta.

and calcium gluconate for prevention of pre-eclampsia, and therapeutic enoxaparin for the thrombus in the descending aorta.

First-trimester foetal aneuploidy screening was negative and the foetus was also structurally normal. The patient was seen every two weeks at the cardiac-obstetric unit for evaluation of blood pressure, urinalysis and foetal growth. An echocardiogram was done monthly to evaluate the aorta for disease progression.

The antenatal course was uneventful. The blood pressure was well controlled (around 130/70 mmHg) with appropriate foetal growth. The patient was delivered by elective caesarean section with spinal anaesthesia at 34 weeks' gestation. A healthy baby weighing 2.3 kg with good Apgars was delivered. The mother was observed in a high-dependency unit for 24 hours after delivery where her blood pressure remained well controlled. After delivery, treatment with prednisone, azathioprine and enoxaparin was continued, methyl-dopa was stopped and nifedipine re-started and stool-softeners were also prescribed.

The patient was discharged five days after delivery. Repeat CT angiography and echocardiogram at the six-week postnatal visit was unchanged.

Discussion

Takayasu arteritis was first described in 1908 by the Japanese ophthalmologist who observed retinopathy in the absence of peripheral pulses. Autoimmunity, sex hormones (more common in females) and a genetic predisposition of the human leucocyte antigen have been proposed as possible causes.[5]

The disease is classified clinically into stages depending on the presence of complications such as hypertension, retinopathy, aneurysms and aortic insufficiency: group I, uncomplicated disease; group IIa, single complication with uncomplicated disease; group IIb, severe single complication with uncomplicated disease; group III, two or more complications with uncomplicated disease.[6] Our patient had group III disease.

Patients with Takayasu disease should be managed in a high-risk obstetric unit. Pregnancy is not associated with disease progression, however there is a 60% risk of complications developing during pregnancy.[7] Maternal risks are attributed mainly to arterial hypertension, and the most important risks include development of pre-eclampsia, exacerbation of chronic hypertension, heart failure, and cerebral vascular accidents.[8]

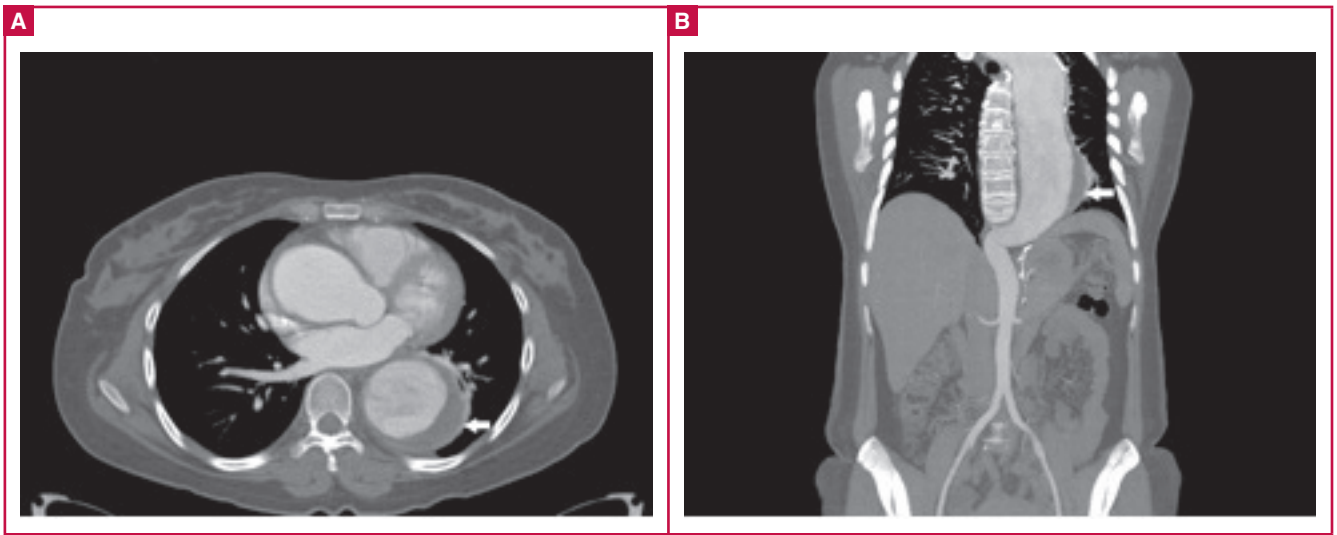

Fig. 2. Post-contrast CT images (A) in the axial post-contrast venous phase and (B) coronal curved reconstruction in the arterial phase demonstrating marked thickening of the wall of the thoracic aorta (arrows). Note the involvement of the thoracic aorta only, with sparing of the abdominal aorta.

Low-dose aspirin for pre-eclampsia prevention should be started before 16 weeks.

Lakhi and Jones reported a case of Takayasu arteritis complicated by aortic dissection in the peripartum period.[8] In this report, the patient's blood pressure remained elevated (160/91 mmHg) when she became symptomatic on the third postpartum day. In the 2003–2005 Confidential Enquiries into Maternal Deaths in the United Kingdom, aortic dissection was one of the leading causes of maternal death.[9] The deaths occurred mostly from failure to treat systolic hypertension.

Foetal complications such as growth restriction, miscarriage and foetal death have been reported in 60–90% of cases.[10] Foetal growth restriction is most likely the result of impaired placental blood flow caused by uncontrolled blood pressure and the involvement of the abdominal aorta and renal arteries. Another mechanism could be occlusion of the renal arteries, leading to an increase in renin production, with consequent increase in blood pressure.[11]

The mode of delivery is determined by the maternal haemodynamic status and by obstetric indications. Unfortunately there are very little data to guide clinicians as to the optimal mode of delivery. Labour and vaginal delivery with or without epidural anaesthesia is safe provided blood pressure is controlled.[8] Patients with Takayasu disease experience a severe elevation of systolic blood pressure during uterine contractions, compared to control patients, so regular monitoring of blood pressure is important during labour.[12] The second stage of labour should be shortened by the use of low forceps or vacuum delivery.

Leal *et al*. recommend vaginal delivery for patients in groups I and IIa, as long as epidural analgesia is used for pain relief and the second stage of labour shortened by vacuum or forceps.[5] Caesarean section is recommended for patients in group IIb and III because the increased blood volume and blood pressure observed during uterine contractions may lead to cardiac decompensation.[5] Regional anaesthesia has been reported successfully for caesarean delivery.[13] This method also allows monitoring of brain perfusion through the patient's level of consciousness. Our patient had an elective caesarean section because, although the blood pressure was controlled, her aorta was severely dilated. This put her at a significant risk of dissection or rupture of the aorta.

Patients should be nursed in a high-care unit postoperatively to allow for early detection of hypoperfusion of organs and hypertensive complications. After delivery, maternal peripheral resistance and left ventricular workload increases. This physiological change may lead to the development of pulmonary oedema, heart failure, renal dysfunction or cerebral haemorrhage.[14] Use of immunosuppressive treatment may also increase the risk of puerperal infection.

Conclusion

Patients with Takayasu disease in pregnancy are at risk of several obstetric complications. These patients should be jointly managed during pregnancy by obstetricians, rheumatologists and cardiologists. Systemic hypertension must be aggressively treated to reduce the risk of complications.

References

1. Sadura E, Jawniak R, Majewski M, Czekajska-Chehab E. Takayasu arteritis as a cause of arterial hypertension. Case report and literature review. *Eur J Pediatr* 2012; **171**: 863–869.
2. Mason JC. Takayasu arteritis-advances in diagnosis and management. *Nature Rev Rheumatol* 2010; **6**: 406–415.
3. Hahn D, Thomson PD, Kala U, Beale PG, Levin SE. A review of Takayasu's arteritis in children in Gauteng, South Africa. *Pediatr nephrol* 1998; **12**: 668–678.
4. Dabague AL, Reyes PA. Takayasu's arteritis in Mexico: a 38 year clinical perpective through literature review. *Int J Cardiol* 1996; **54**: 103–109.
5. Leal PdC, Silveira FFM, Sadatsune EJ, Clivatti J, Yamashita AM. Takayasus's arteritis in pregnancy. Case report and literature review. *Rev Bras Anesttesiol* 2011; **61**: 479–485.
6. Ishikawa K. Natural history and classification of occlusive thromboaortopathy. *Circulation* 1978; **57**: 25–35.
7. Mandal D, Mandal S, Dattaray C, *et al*. Takayasu arteritis in pregnancy: an analysis from eastern India. *Arch Gynecol Obstet* 2011; **285**: 567–571.
8. Lakhi NA, Jones J. Takayasu's arteritis in pregnancy complicated by peripartum aortic dissection. *Arch Gynecol Obstet* 2010; **282**: 103–106.
9. Nelson-Piercy C. Saving mother's lives. The seventh report of the confidential enquiries into maternal deaths in the United Kingdom 2003–2005. London: RCOG Press, 2007.
10. Matsumura A, Moriwaki R, Numano F. Pregnancy in Takayasu arteritis from the view of internal medicine. *Heart Vessels* 1992; **7**: 120–124.
11. Suri V, Aggarwal N, Keepanasseril A, Chopra S, *et al*. Pregnancy and Takayasu arteritis: a single centre experience from North India. *J Obstet Gynaecol Res* 2010; **36**: 519–524.
12. Ishikawa K, Matsuura S. Occlusive thromboaortopathy (Takayasu's disease) and pregnancy. Clinical course and management of 33 pregnancies and deliveries. *Am J Cardiol* 1982; **50**: 1293–1300.
13. Williams GM, Gott VL, Brawley RK, Schauble JF, Labs JD. Aortic disease associated with pregnancy. *J Vasc Surg* 1988; **8**: 470–475.
14. Cunningham FG, Pritchard JA, Hankins GD, *et al*. Peripartum heart failure: idiopathic cardiomyopathy or compounding cardiovascular events? *Obstet Gynecol* 1986; **67**: 157–168.