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Case Report

Takayasu arteritis in pregnancy

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of pregnancies in TA patients is a challenge for the obstetrician, the rheumatologist and the cardiologist.

Case report

A 22 yr old female patient, G2P1L1 reported in the ANC OPD at 14 weeks period of gestation (POG). 3 yrs back she underwent full term normal delivery and delivered a healthy male baby without any complication. She gives no history of high blood pressure during her last pregnancy. On examination, upper limb peripheral pulsations were not felt and blood pressure was not recordable in both upper limbs. Lower limb pulsation was present and blood pressure was 128/82 mm of Hg. Other systemic examination and biochemical and haematological parameters were within normal limits. Ultrasonography showed single live intra uterine fetus with corresponding POG.

Colour Doppler flow of bilateral upper limbs in early second trimester revealed left common carotid artery long segment stenosis with adequate distal flow. Diffuse wall thickening in proximal parts of the branches of great vessels and monophasic flow with reduced velocities bilateral forearm arteries. In view of pregnancy, no radiation based investigations were done. However patient was asymptomatic throughout her pregnancy. MRI facility was not available during this period of time.

She was followed up with weekly blood pressure measurement, ultrasonography fortnightly for fetal growth. Her antenatal period remained uneventful and at 38 weeks POG she reported with labour pains with cervical dilatation of 08 cm. She underwent full term vaginal delivery without any complications.

Introduction

In 1908, Mikito Takayasu, a Japanese ophthalmologist, reported the case of a 21-year-old woman with retinal arteriovenous anastomoses, syncope, and absent upper extremity pulses.¹ Takayasu arteritis (TA), also known as occlusive thromboangiopathy, is a primary systemic vasculitis of unknown etiology, predominantly affecting women² during their reproductive years (female to male ratio is 4:1). It has variable geographical distribution with the greatest prevalence in Asians, especially Japan, and the Orient. Optimal management for pregnant patients with this disease has not yet been established. Due to the manifold cardiovascular complications that can occur in the course of the disease, management

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Postnatally patient underwent CT angiography which revealed that there is concentric intimal thickening of the left common carotid in the proximal 2/3 with stenosis of its lumen (Fig. 1) and the right subclavian artery is occluded immediately distal to the Rt VA origin (Fig. 2). The left subclavian artery shows significant concentric wall thickening and diffuse luminal narrowing distal to origin of Lt VA. The distal 1/3 of the left subclavian shows complete occlusion of its lumen for a short segment. CT angiography findings were suggestive of Takayasu's Arteritis with bilateral Subclavian and Common Carotid Artery wall thickening.

Discussion

Takayasu's arteritis (TA) is a chronic inflammatory vascular disease of unknown origin. Although it can affect other segments of the aorta as well as pulmonary and renal arteries but it often affects the aortic arch and its main branches.³ TA1 is characterized by intimal leukocytary infiltration and myofibroblast proliferation almost invariably leading to vascular stenosis. Sometimes aneurysm formations occur due to the release of metalloproteases. Clinical features are related to the affected artery: subclavian and iliac arteries involvement present with limb claudication; carotid involvement may induce vertigo; renal arterial lesions are associated with arterial hypertension; while some patients can progress to aortic insufficiency and congestive heart failure.⁴ The inflammatory reaction in TA responds to glucocorticoids which are the drugs of first choice in pregnancy. If prednisone treatment fails, azathioprine should be considered. Clinical presentation may be insidious and diagnosis is often delayed.

Fig. 1 – MIP coronal image of CT angiography showing concentric wall thickening of the LCCA with stenosis of the lumen.

Fig. 2 – Occlusion of right subclavian artery.

In 1990, the American College of Rheumatology proposed⁵ six diagnostic criteria, at least three of them are required for classifying a patient as having TA (Table 1). The natural course of TA is chronic and progressive, with a reported survival range of 1–15 years. In our patient she did not have any symptoms and blood pressure was within normal range throughout the pregnancy.

Arteriography, either traditional or magnetic resonance angiography, is the gold standard for delineating abnormal vessels in TA. Typical findings include an irregular intimal surface, stenosis of the aorta or its branches, poststenotic dilations, saccular aneurysms, or the typical narrowed, “rat-tail” appearance. High-resolution duplex ultrasound technology may be used to evaluate and monitor disease in the common carotid and subclavian arteries; however, this imaging modality is not useful for evaluation of the aorta.⁶ Most cases of TA during pregnancy have been reported in patients with a known diagnosis prenatally. We consider our case to be unique and quite rare because the diagnosis was made during pregnancy with no presenting symptom. In our case, the diagnostic options were limited secondary to her pregnancy and post delivery CT angiography confirms our diagnosis.

Pregnancy does not significantly affect the inflammatory activity of the disease. Wong et al studied 19 pregnancies in 11

Table 1 – Clinical diagnosis of Takayasu arteritis criterion.^a

Age at disease onset ≤40 yr
Claudication of extremities
Decreased brachial artery pulse
Systolic blood pressure difference >10 mm Hg between 2 arms
Bruit over subclavian arteries or aorta
Arteriographic evidence of narrowing or occlusion of aorta, its primary branches or large arteries in the proximal upper or lower extremities

^a Diagnosis requires at least 3 of the 6 criteria.

patients and did not find any evidence of acute inflammatory exacerbation.⁷ Although the majority of women with TA do well in pregnancy and labor,⁴ those with secondary hypertension and cardiac involvement may be negatively impacted. The majority of the problems stem from exacerbation of the preexisting chronic hypertension and/or development of superimposed preeclampsia. Despite aggressive medical treatment, uncontrolled hypertension may require termination of pregnancy. High-risk patients may be affected by intrauterine growth restriction, which is principally determined by the severity of hypertension and the extent of arterial disease, especially abdominal aortic involvement.⁷

Management of TA in pregnancy may require multiple drug therapy including prolonged steroids, immunosuppressives like azathioprine, antihypertensive drugs, anticoagulants, and antibiotics. Prophylactic antibiotics should be used especially if aortic regurgitation is present to avoid infective endocarditis and puerperal sepsis.⁸

In conclusion, successful outcome is known in TA, but require close monitoring of mother and fetus. Hypertension, preeclampsia and IUGR are the most frequent problems in this group of patients and a comprehensive prepregnancy evaluation and a multidisciplinary approach to these patients in a tertiary care center can have a favorable outcome.

Conflicts of interest

All authors have none to declare.

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