Case Report

Takayasu's arteritis during pregnancy: A case report

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ABSTRACT

Takayasu's arteritis (TA) is also known as aorto arteritis having an incidence is 13/per million population. Here, we report the case of a 30-year-old woman gravida 2, para 1, who was admitted for safe delivery at 38 weeks of pregnancy. Clinical examination revealed an absence of radial pulse and blood pressure (BP) in both upper limbs and lower limbs BP recording showed 150/90 mm Hg with a pulse rate of 112/min. She was a known case of TA diagnosed during her first pregnancy. An elective cesarean section was done and a healthy male baby weighing 3.2 kg was delivered. The postnatal period was uneventful. She was discharged on the 8th day. A magnetic resonance imaging (MRI) angiogram was done before the second pregnancy. There was complete occlusion of the left common carotid artery from the origin with occlusion of the left internal carotid artery and there was collateral refilling of the left anterior cerebral artery and middle cerebral artery from the posterior communicating artery. The left vertebral artery showed circumferential wall thickening with hypointense flap within.

Key words: Arteritis, Caesarean section, Magnetic resonance imaging angiogram

akayasu's arteritis (TA), also known as "young female arteritis" or pulseless disease/Aorto arteritis, is a rare and chronic inflammatory disease of large vessels mainly affecting reproductive age women of Asian origin having an incidence of 13/million population. It was first described by Mikito Takayasu, a Japanese Ophthalmologist, and Onishi in 1908 [1,2]. The main complications observed in TA are occlusion and aneurysm formation in systemic and pulmonary arteries as there is an increased risk of cardiovascular complications such as hypertension and congestive cardiac failure during pregnancy [1,2]. Hypertension, multiple organ dysfunction, and stenosis are possible complications during pregnancy. Aortic stenosis with reduced blood flow causes restricted intrauterine growth restriction and low birth weight [2]. Women with TA need to be advised to plan pregnancy ideally during the remission period. There is a need for an interdisciplinary approach for the care of these patients [1]. Insidious onset and non-specific early symptoms are responsible for diagnostic and therapeutic delay [3].

The present case is being reported due to the rarity of this disease. The present case was managed successfully during the antenatal and postnatal periods.

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CASE HISTORY

A 30-year-old woman, a known case of TA, was admitted to our hospital for safe confinement on March 22, 2023. Her last menstrual period date was June 25, 2022, and the estimated date of delivery was April 02, 2023. She was gravida two, 1st living healthy baby delivered by lower segment cesarean section (LSCS) in 2015 in the same hospital.

While doing the antenatal checkup during first pregnancy in 2015, there was an absence of radial pulse on the right upper limb and she was referred to Bangalore for further investigation. TA was diagnosed and she was on treatment for the same. LSCS was done due to non-progress of labor. There were no postnatal complications and was discharged from the hospital. Three years after the first delivery, she had attacks of severe headaches off and on. She consulted a clinician and was advised to get a computed tomography angiogram (Fig. 1). After injecting contrast through bolus tracking, images were obtained from carotid bifurcation to Aortic bifurcation, 0.6 mm slice thickness followed by volume rendered maximum intensity projection and MRP images. The ascending aorta, arch of aorta, descending aorta, brachiocephalic trunk, left common carotid, left subclavian, and left vertebral artery were normal. Right and left pulmonary arteries were normal. Abdominal aorta, coeliac trunk, superior mesenteric, and inferior mesenteric arteries were normal. Both renal arteries were normal

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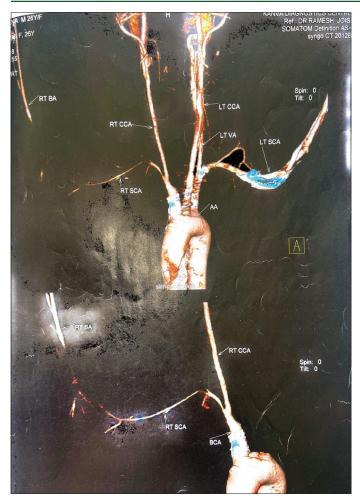


Figure 1: Computed tomography angiogram

with normal enhancement of both kidneys. Aortic bifurcation and common iliac arteries were normal. Circumferential wall thickening was seen involving the right subclavian artery causing significant luminal narrowing of about 70–80%. Right axillary and proximal brachial arteries were occluded. The brachial artery was reformed in the upper 1/3 of the arm. Numerous chest collaterals were noted on the right side. Circumferential wall thickening was seen involving the right common carotid artery causing about 30% narrowing. The right vertebral artery was diffusely narrow in caliber and showed faint contrast opacification.

She was advised to follow treatment inj Actemra 400 mg 6 injections. During 2019, she was advised the same treatment 4 injections. In January 2020, she had attacks of severe headache, suggesting cerebrovascular lesions. She was advised inj Actembra again and inj Clexene for 3 days and was advised magnetic resonance imaging (MRI) brain which showed acute infarcts in the left frontal lobe, predominantly involving the cortical and subcortical region. Lacunar fluid intensity lesions were seen in the genu of the corpus callosum as well as in the right caudate nucleus (squeal to old infarcts). No hemorrhages were noted. The rest of the neuroparenchyma was normal. Bilateral caudate nucleus, putamen, globus pallidus, and internal capsule were normal. Both lateral and third ventricles were normal. MRI angiogram showed an occluded left internal carotid artery. Collateral filling through the left middle cerebral and anterior cerebral from the posterior

communicating artery shows mild narrowing near the PCOM origin. Hypoplastic A1 and A2 segment of the right anterior cerebral artery were seen. The distal anterior cerebral artery was not visualized, right vertebral artery hypoplastic. Rest all vessels were normal.

There was a circumferential wall thickening with a thin flap within the lumen of the left proximal vertebral artery. The right vertebral artery was hypoplastic, left common carotid artery was occluded from the origin with occlusion of the cervical part of the internal carotid artery. The left external carotid artery showed collateral filling. The right common carotid artery showed circumferential narrowing. Carotid bulb was normal. The right internal carotid artery showed a thin linear flap within the lumen in the petrous segment. The rest of the internal carotid artery was normal.

After undergoing all the investigations, acute infarcts in the left frontal lobe predominantly involving cortical and subcortical regions were noted. Complete occlusion of the left common carotid artery from origin with occlusion of the left internal carotid artery. There was collateral refilling of the left anterior cerebral artery and middle cerebral artery from the posterior communicating artery. Lacunar infarcts were shown in the genu of corpus callosum as well as right caudate nucleus. The left vertebral artery showed circumferential wall thickening with hypointense flap within. She discontinued all treatment due to financial reasons and was on Ayurvedic medication for 2 years. Later, she discontinued all treatment.

On general physical examination, radial pulse and blood pressure (BP) were not felt on both upper limbs. The pulse rate was 112/min and BP was 150/100 mm of Hg in the lower limb. Cardiovascular and respiratory system examinations were normal. Obstetric examination findings were uterus 38 weeks and relaxed with cephalic presentation, fetal heart sound was 130/min. She was on the following treatment—Tab Prolomet 25 mg daily, Tab Ecosprin 150 mg daily, Tab Azothioprim 50 mg daily, and Tab Metpure 25 mg daily. A bilateral upper limb arterial Doppler study was done on September 12, 2022. There was diffuse wall thickening with severe diffuse narrowing of the second and third parts of the right subclavian artery, causing monophasic flow in arteries distal to it.

Elective LSCS was done on March 23, 23 and a healthy male baby weighing 3.2 kg was delivered. During LSCS, liquor was clear there was a true knot of umbilical cord seen. There were no other specific findings. The postnatal period was uneventful. She was discharged on the 8th day.

DISCUSSION

TA is a granulomatous vasculitis of large vessels that mainly affects the aorta and its proximal branches. In 1990, the American College and Rheumatology published the classification criteria and the disease took the name of TA. Its etiology is unknown but it seems to be related to infectious triggers that amplify the inflammatory response through cellular mechanisms and cause damage to the vascular wall with subsequent thickening, fibrosis, stenosis, thrombus formation, and aneurysms. The lesions are

Table 1: Comparison of cases reported by many authors

S No.	Author, year	Age, parity	Clinical presentation	Maternal complications	Mode of delivery	Fetal outcome
1	Itani et al. [1] 2018	31 year G2 P1	X-ray chest wide superior mediastinum CT dissecting aortic aneurysm	Known case of TA booked at 11 weeks of pregnancy	LSCS done	Live baby
2	Marwah <i>et al</i> . [2] 2017	25 years G2 P1 L1	Pain in the right lower limb	Controlled HTN, neurological squeal, IUGR	Vaginal delivery	Live baby
3	Arezzo et al. [3] 2021	32 years G2 P1	Known case of TA diagnosed in 2015. Aneurysm of aortic arch and ascending aorta	Persistent fever, paresthesia, HTN.	LSCS done	Live baby
4	Malhothra et al. [6] 2015	25 years G3P2.	Woman visited at 26 weeks and was asymptomatic. On examination upper limb pulses were not felt, bilateral carotids were weak.	Severe oligohydramnios at 36 weeks	LSCS done	Live baby
5	Goyal et al. [9] 2020	25 years G 1	Known case of TA. Now admitted with c/o Left sided chest pain HTN	Renal involvement ITAAS score 9 with	LSCS done	Live baby
6	Raka and Singh [10] 2022	G1 21 years	Cold, sneezing -15 days Briut heard over Right and left carotid Rt radial art.	Feeble radial pulse, BP readings variable in both arms.	LSCS done	Live baby
7	Present case 2023	30 years G2 P1	c/o headache O/E HTN	Known case of TA diagnosed during first pregnancy	LSCS done	Live baby

IUGR: Intrauterine growth restriction, LSCS: Lower segment cesarean section, BP: Blood pressure, TA: Takayasu's arteritis, CT: Computed tomography, HTN: Hypertension

usually asymptomatic and are discovered in the late stages of the disease when ischemic symptoms occur [4].

Systemic vasculides are rare autoimmune disorders characterized by necrotizing inflammatory infiltration of blood vessels and multisystem involvement. It is widely recommended that the woman suffering from autoimmune systemic disease need to plan their pregnancy during periods of remission or low disease activity to avoid unfavorable outcomes for both mother and fetus, as well as to prevent the severe response of the disease itself during this period [5-7].

The Hata Numeno classification is based on the extent of the disease and the vascular territory affected classified the disease as: Branches of aortic arch (I), ascending aorta, aortic arch and its branches (IIa), ascending aorta, arch of aorta, and its branches, descending thoracic aorta (IIb), abdominal aorta, renal arteries or both (IV), ascending aorta, aortic arch and its branches, descending thoracic aorta, renal artery, or both (V) [4,8]. Another classification, based on complications such as hypertension, retinopathy, aneurysm, and aortic insufficiency, categorized the disease as having no complications (Stage I), one of the complications (Stage II a), one of these complications with severe form (Stage II b), more than one complication (Stage III) [2]. The present case belongs to Type I as per the Hata Numeno classification because there is the involvement of aortic arch branches only. Based on complications, the case is Stage I as there were no complications. Table 1 shows the literature review of the previous cases [9,10].

CONCLUSION

TA affects women of childbearing age but there is no exacerbating effect on pregnancy. However, women with active disease during pregnancy with extensive aortic involvement present with unfavorable obstetric outcomes. Inadequate control of vasculitis leads to an increased risk of maternal and fetal complications. Close monitoring of the disease progression by the multidisciplinary team gives good maternal and fetal outcome.

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