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A Rare Case of Large Vessel Vasculitis Leading to Vision Loss

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Abstract: ***A rare case of large vessel vasculitis coexistent with middle cerebral artery infarct is described with brief discussion in a fourteen year old young female. Patient under follow up and is maintaining sound health.***

Keywords: Takayasu arteritis, middle cerebral artery infarct, vasculitis

1. Introduction

The disease is also called 'pulseless disease', since peripheral pulses are often absent due to vascular obstruction. Takayasu's arteritis (TA) is a granulomatous large vessel vasculitis of unknown etiology.The term TAKAYASU ARTERITIS was named in the honour of Japanese ophthalmologist – Mikito Takayasu (1905).Takayasu arteritis has been rarely reported in childhood[1]. Male to female ratio was 1:4.4.Takayasu arteritis (TA) is a large vessel vasculitis affecting mainly the aorta and its major branches near origin.Chronic inflammation leads to stenosis and occasionally aneurysm formation. We are presenting a case which is having a atypical presentation in the same individual. The clinical manifestations are variable with some patients presenting with systemic features such as fever of unknown origin, arthralgia and myalgias without any clinical evidence of vascular involvement. The management profile and the follow up is discussed.

2. Case Report

A fourteen year old female patient presented to the ophthalmic wing of Shantiram Medical College and General Hospital, Nandyala, with complaints of diminution of vision since one year. The patient complained of diminution of vision in BE since one year which was insidious in onset, gradually progressive in nature (RE > LE).

She had a history of fatigue and malaise since two years. History of fever and night sweats three years back. Symptomatic treatment was taken. History of fatigue since three years. History of syncopal attacks on fully extended neck of about two to three attacks per day two years back. The frequency of these episodes has now decreased. History of lumbar puncture done two years back. History of CVA (right MCA infarct) two years back. The patient took treatment (tab. Phenytoin 100mg + tab. Levipil 250mg) for two years and had now stopped treatment since eight days. The patient is on cap. B-complex OD + tab. Iron OD since two years. Birth history – Antenatal, natal and postnatal history was uneventful. Immunization history – Immunized till date. Developmental history – There was no delay in the achievement of milestones. All the milestones were attained. Hers was a third degree consanguineous marriage. There was no history of similar complaints in her siblings and family members. Clinical examination revealed patient is conscious, cooperative, coherent, well oriented to place, time and person, afebrile. Pulse rate was 70 beats per minute, regular rhythm, reduced volume. Pulse rate was felt in lower limb (Femoral, Popliteal and Dorsalis Pedis) on both sides. Pulse rate was not felt in upper limb (Carotid, Brachial and Radial arteries). BP was 90/70 mm Hg was felt in lower limbs on both sides, rather than upper limbs. There is 10mm Hg difference between Right LL and Left LL. BP was recorded in the supine position with no bruit or added sounds heard. RR was 22 breaths per minute.

On local examination, head posture and facial symmetry were normal. Extraocular movements were full in all directions. Distant visual acuity was noted in RE – perception of light and projection of rays in all quadrants, while LE was 6/60. No improvement was seen with pinhole and with refraction. Patient did not appreciate any of the colours and contrast. On ocular examination, anterior segment was normal in both eyes, pupils both were mid-dilated, fixed, sluggish reacting to light, lens status was total cataract in RE and clear in LE. Intraocular pressure was measured to be 10 mm Hg and 12 mm Hg in the RE and LE respectively. Due to the presence of dense media opacity (mature cataract) in RE, fundus examination was not possible. In the LE, the following findings were noted. Media was clear, disc showing pallor and neo vascularization, vessels were dilated tortuous arterioles & venules with multiple talengectasias, AV anastomosis and 360 degree sclerosed vessels in the periphery with normal foveal reflex seen. B-scan showing no evidence of retinal detachment with anechoic echoes.After correlation of clinical features and ocular examination, the patient was clinically diagnosed to have ocular ischemia in the LE and is subjected to detailed systemic evaluation. Patient was referred to department of cardiology for further evaluation.After complete clinical examination, patient was advised for following investigations. 2D echo imaging report showed aortic gradient 36 cubmm. ? Coarctation of aorta or large vessel vasculitis. CT aortography revealed concentric intimo-medial thickening with near total occlusion is noted in brachio cephalic trunk, left common carotid and left subclavian arteries. Multiple collaterals are noted in neck, chest wall and upper abdominal wall. Delayed phase shows mild enhancement of above said occluded vessels, secondary to reformation to collaterals. Ascending aorta, arch of aorta and descending aorta are normal in course, calibre and shows good contrast filling. She was diagnosed to have Type I Takayasu's Arteritis (angiographic classification) with multiple collaterals in neck, upper abdominal wall and chest walls with right middle cerebral artery infarct with RE mature cataract. Patient was further advised for cataract

surgery in RE under guarded visual prognosis and with moderate cardiac risk.

The patient has undergone cataract surgery under local anaesthesia with high risk consent and moderate cardiac risk was explained to the patient. Phacoemulsification with posterior capsule intraocular lens implantation was done. After the cataract surgery, fundus examination of RE observedas media was clear, disc was pallor, neovascularization at disc, sclerosed vessels, foveal reflex dull, ischemic retina from mid periphery to periphery.

After cataract surgery in RE, staging of the disease was done: RE – Type I – Takayasu's Retinopathy Stage IV and LE – Type I – Takayasu's Retinopathy Stage III. Patient was advised and treated for panretinal photocoagulation in BE, first better viewing eye (LE) followed by RE, Tab. Ecospirin AV 75/10 OD, Tab. Wysolone 50mg BD in divided doses, Tab. Methothrexate 15mg once a week, Tab. Folic acid 5mg alternate day. After six months in ophthalmological examination, there was improvement in vision in RE (6/60) and in LE also. Anterior segment examination was normal bilaterally. Intraocular pressure was measured to be 16 mm Hg and 12 mm Hg in both eyes respectively. Fundus examination showed both eyes pale disk with neovascularization with 360 degrees sclerosed vessels and s/p PRP spots seen in mid periphery to periphery indicating stable fundus. Patient now continued with Tab. Wysolone 5mg OD.

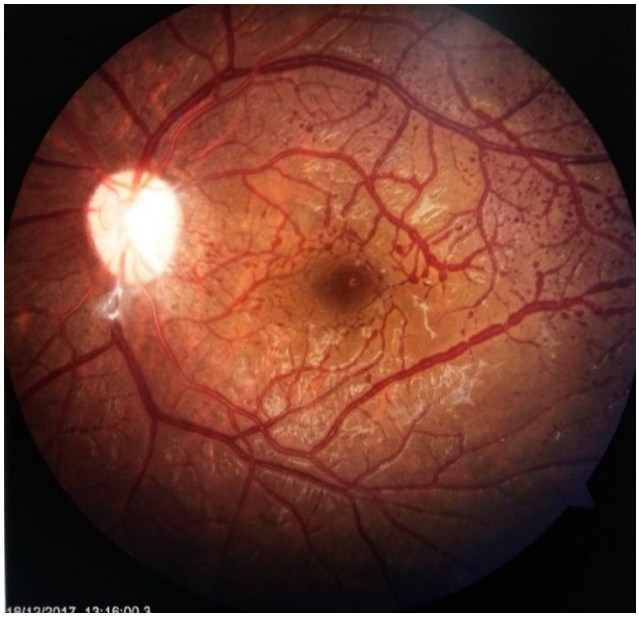

Figure 1: Fundus photograph of LE at presentation showing disc pallor and neo vascularization, vessels were dilated tortuous arterioles & venules with multiple talengectasias, AV anastomosis and 360 degree sclerosed vessels in the periphery with normal foveal reflex indicating Type I – Takayasu's Retinopathy Stage III.

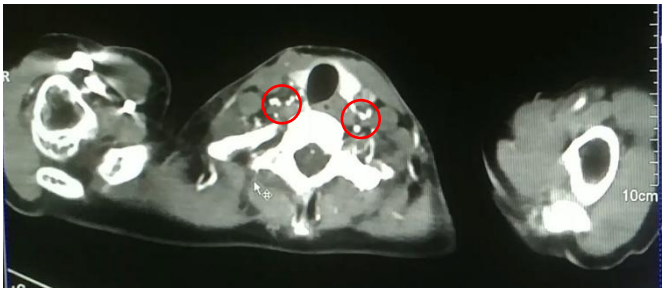

Figure 2: CT aortography showing multiple collaterals in the neck, chest wall and upper abdominal wall.

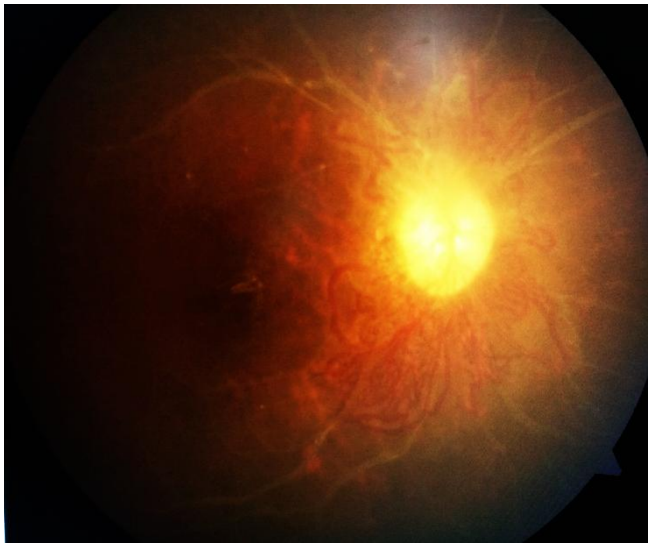

Figure 3: Fundus photograph of RE after cataract surgery showing disc pallor, neovascularization at disc, sclerosed vessels, foveal reflex dull, ischemic retina from mid periphery to periphery indicating Type I – Takayasu's Retinopathy Stage IV.

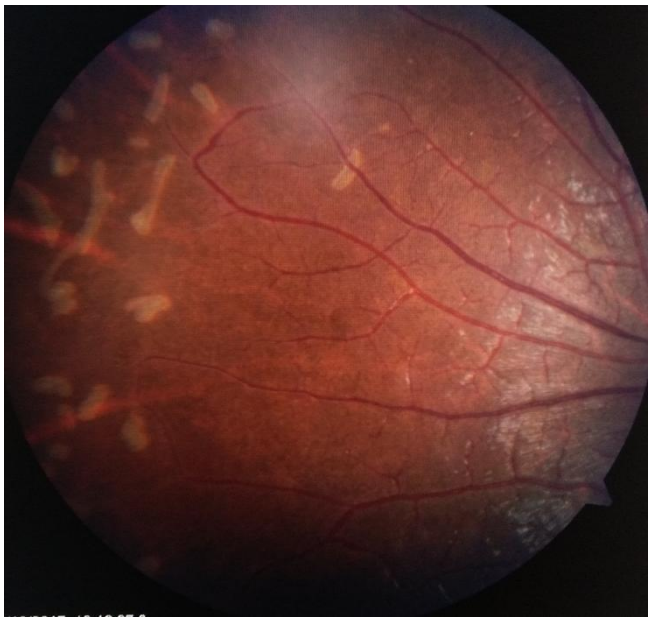

Figure 4: Fundus photograph of LE after three sittings of PRP showing PRP spots at mid periphery.

3. Discussion

Takayasu's arteritis is an uncommon inflammatory disease of the large and medium sized arteries [2]. Ocular

presentation of TA is reported to vary between 8.1%and 68% of the patients[3]. The ocular findings of the disease are related with carotidartery obliteration which leads to hypoperfusion of all theeye structures. Reduced blood flow in the carotid arteriescauses Takayasu's retinopathy which is related to the chronicischemia and occurs in the late phase of the disease [4]. Takayasu's arteritis is a disease that affects large vessels; however, previous reports presented the involvement of small retinal vessels.

Takayasu arteritis has been rarely reported in childhood. A common clinical mode of disease presentation in our patients was arterial hypertension, together with nonspecific symptoms (headache, fatigue, myalgia, weight loss). Symptoms due to ischemia, which are frequent in adults, have been seldom reported in children. However, although true claudication (upon effort) was present only in one patient, we have observed ischemic findings (chest, limb, and abdominal pain) in three of our four patients. The disease is also called 'pulseless disease', since peripheral pulses are often absent due to vascular obstruction;

Genetic susceptibility to Takayasu's arteritis has been extensively studied. There are heterogeneous population data regarding HLA associations in TA. HLA B-52 and DR-2 are associated with Takayasu's arteritis in Japan, HLAB-52 and B-5 association is also reported from Korea and India [5], whereas HLA B-39 is frequently found in Mexican Takayasu's arteritis patients. Further characterisation of HLA association in Takayasu's arteritis is being studied in order to identify alleles or epitopes responsible for the susceptibility to this disease.

The criteria for the diagnosis of Takayasu's arteritis as suggested by Ishikawa[6] are shown in Table 1.

Conventional or digital subtraction angiography has been considered the gold standard for the diagnosis of Takayasu's arteritis. Angiography shows luminal irregularity, vessel stenosis, occlusion, dilatation or aneurysms in the aorta or its primary branches.

Neurofibromatosis of the abdominal aorta and some other causes of mid-aortic syndrome may produce an identical angiographic picture in children. Based on angiographic morphology, Takayasu's arteritis is divided into type I (involving aortic arch and its branches), type II (thoracoabdominal aorta and its branches) and type III (involving lesions of both type I & II). Involvement of pulmonary arteries in addition to any of the above types is grouped as type IV [7].

The diagnosis of TA is based on characteristic findings of diseased aorta and its major branches seen on angiography. This is demonstrated by luminal abnormalities such as stenosis or aneurysmal dilatation of the aorta, its major branches, and the pulmonary arteries. With regard to imaging studies, traditionally the angiographic patterns have been divided in: type I, affecting the aortic arch; type II, the thoracic and abdominal aorta; type III, the aorta both above and below the diaphragm; and type IV, the aorta and the pulmonary arteries. In our cases type I was the predominant pattern, whereas in two series type II was the predominant one [8,9].

Table 1: Criteria for diagnosing Takayasu's Arteritis

Criteria*	Definition
Obligatory criteria	
Age <40 year	Age < 40 year at diagnosis or at onset of "characteristic signs and symptoms" of one month duration in patient history.
Two major criteria	
Left mid subclavian artery	The most severe stenosis or occlusion present in the mid portion from the point one cm proximal to the left vertebral artery orifice to that three cm distal to the orifice determined by angiography.
Right mid subclavian artery lesion	The most severe stenosis or occlusion present in the mid portion from the right verterbral artery orifice to the point 3 cm distal to the orifice determined by angiography.
Nine minor criteria	
High ESR	Unexplained persistent high ESR>20 mm/h (Westergren) at diagnosis or presence of evidence in patient history.
Carotid artery tenderness	Unilateral or bilateral tenderness of common carotid arteries by physician palpation; neck muscle tenderness is unacceptable.
Hypertension	Persistent blood pressure 140/90mmHg brachial or > 160/90mmHg popliteal at age <40 year. Or presence of the history at age <40 year.
Aortic regurgitation or annuloaortic ectasia	By auscultation or Doppler echocardiography or angiography
Pulmonary artery lesions	By angiography or two dimensional echocardiography. Lobar or segmental arterial occlusion or equivalent determined by angiography or perfusion scintigraphy; or presence of stenosis, aneurysm, luminal irregularity or any combination in pulmonary trunk or in unilateral or bilateral pulmonary arteries determined by angiography.
Left mid common carotid lesion	Presence of the most severe stenosis or occlusion in the mid portion of 5cm in length from the point 2 cm distal to its orifice determined by angiography.
Distal brachiocephalic trunk lesion	Presence of the most severe stenosis or occlusion in the distal third lesion determined by angiography.
Descending thoracic aorta lesion	Narrowing, dilatation or aneurysm, luminal irregularity or any lesion combination determined by angiography; tortuosity alone is unacceptable.
Abdominal aorta lesion	Narrowing, dilation or aneurysm, luminal irregularity or any combination and absence of lesion in aorto-iliac region consisting of 2cm of terminal aorta and bilateral common iliac arteries determined by angiography; tortuosity alone is unacceptable.

* The proposed criteria consist of one obligatory criterion, two major criteria and nine minor criteria. In addition to the obligatory criterion, the presence of two major criteria, or one major and two or more minor criteria or four more minor criteria suggests a high probability of the presence of Takayasu's disease.

Table 2: Criteria for Active Disease in Patients with Takayasu Arteritis [7]

Constitutional features, such as fever, musculoskeletal pain (no other cause identified)
Elevated erythrocyte sedimentation rate (>20 mm / hr)
Features of vascular ischemia or inflammation, such as claudication, diminished or absent pulse, bruit, vascular pain (carotodynia), asymmetric blood pressure in either upper or lower limbs (or both)
Typical angiographic features.

New onset or worsening of two or more features indicates "active disease".

TA is a disease with severe prognosis, mortality rate being reported in children from 35 to 40% by five years [10]. It is therefore important to have a high index of suspicion and in doubtful cases a low threshold for diagnostic evaluation. We underline the possibility of TA in any young patient with unexplained arterial hypertension.

Uyama and Asyama [11] broadly classified TA into 3 types:
Type 1 – Ischemic ocular manifestation (Takayasu Retinopathy)
Stage 1 – Distension of veins
Stage 2 – Microaneurysm formation
Stage 3 – Arteriovenous anastomosis
Stage 4 – Complications - Retinal ischemia, NVI, NVE, VH
Type 2 – Features of mixed retinopathy
Type 3 – Retinal manifestation due to HTN which occurs due to involvement of renal and abdominal aorta.

Moreover, treatment options have been limited so far, with few reports focusing on immunosuppressive treatment (methotrexate, cyclophosphamide) in paediatric TA.

We conclude that TA is not such a rare disease in a paediatric rheumatology setting, and also that it has to be considered in cases of unexplained arterial hypertension or unexplained inflammatory syndromes without signs of localization. A thorough physical examination can lead to the correct diagnosis if pulses cannot be felt or if an abdominal bruit is heard, even if these are not constant findings. Since the disease can be progressive and life-threatening, an early recognition is vital in order to start immunosuppression, which proved to be very successful in our patients.

4. Competing interests

The authors declare that they have no competing interests.

5. Acknowledgements

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