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Takayasu's Arteritis with Immature Bilateral Cataract

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Abstract

Takayasu arteritis is an inflammatory disease of large- and medium-sized arteries, with a predilection for the aorta and its branches. Advanced lesions demonstrate a panarteritis with intimal proliferation.Lesions produced by the inflammatory process can be stenotic, occlusive, or aneurysmal. All aneurysmal lesions may have areas of arterial narrowing. Patients with aortitis syndrome exhibit various ocular changes.Cataract has seldom described as the initial manifestation of Takayasu arteritis .Here we are presenting case of 14 year old female patient with arteritis who developed bilateral immature cataract before the initiation of steroid treatment.

Keywords: Takayasu Arteritis; Cataract; Prednisolone; Immunosuppresants; Angiography.

Introduction

Takayasu's arteritis or aortic arch syndrome or pulseless disease is a large vessel granulomatous vasculitis affecting often young or middle-aged women of Asian descent (1,2). It has a worldwide distribution with the greatest prevalence in Asians (3-5). The disease mainly affects the aorta and its main branches, as well as the pulmonary arteries. Females are affected about 8-9 times more than males. The symptoms and signs of the disease begin between 15 and 30 years of age. Due to obstruction of the main branches of the aorta, including the brachiocephalic artery, and the left subclavian artery, takayasu's arteritis can present a decrease or absence of upper extremities pulses and for the same reason it is also called pulseless disease (6, 7). Some patients develop an initial inflammatory phase characterized with malaise, fever, night sweats, weight loss, arthralgia, and fatigue. Initial inflammatory phase is often followed by the symptoms of vascular insufficiency manifesting arms or leg claudication, hypertension due to renal artery stenosis, and neurological manifestations like lightheadedness and syncope.

The neurological symptoms of the disease vary depending on the distribution and degree of the blood vessel involvement (8-10). The one rare but important feature of the takayasu's arteritis is ocular involvement in the form of visual field defects, visual loss, or retinal hemorrhage (11, 12). The most important laboratory findings are anemia and marked elevation of the ESR or C- reactive protein (1, 2). The gold standard of diagnosis is vascular study by arterial angiography (DSA), Magnetic resonance angiography (MRA), computed tomography angiography (CTA). Inflammation, granuloma, and fibrosis causes stenosis (93%), occlusion (57%), dilatation (16%), aneurysm (7%) in subclavian artery (93%), common carotid artery (58%), renal artery (38%), vertebral artery (35%) (13-18). Here we want to present a case of Takayasu's Arteritis with immature cataract which is a rare presentation.

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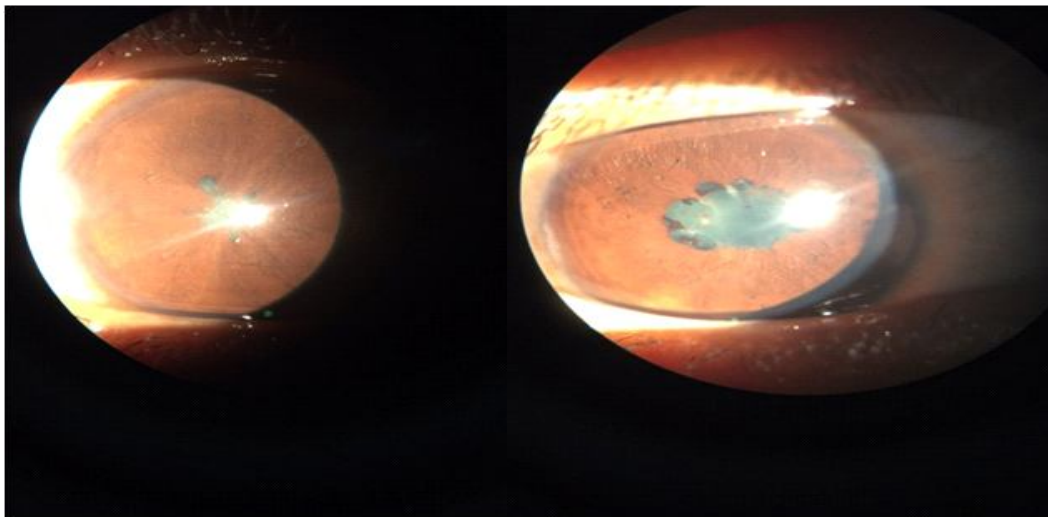
Case Presentation

A 14 year old female child was admitted to the paediatric ward with complaints of loss of vision, history of Lightheadedness and syncope since 5 years and also history of easy fatigability present. There was a gradual loss of vision with 2-3 syncopial episodes per month.There is also history of claudication pain on both upper limbs.On Examination she was conscious and oriented, radial and brachial pulses were not palpable on both sides and BP was not recordable, Carotid pulses were feeble and femoral pulses were normal, with bilateral femoral bruits. The pedal pulses were equal and palpable bilaterally, BP was 110/80 in both lower limbs. On eye examination showed abilateral immature cataract with searching nystagmus is seen and visual acuity is restricted to finger counting. Slit lamp examination of Right eye showed seclusio papillae,irregular pupils, complicated cataract, fundal glow was poor details could not be seen due to cataract , On left eye slit lamp examination showed occlusion papillae, irregular pupils, complicated cataract, fundal glow was poor and details could not be seen due to cataract.B-scan of both eyes failed to show Retinal detachment. Cardiac examination is normal and Other system examination is also normal.CBC was done showing Hb 10.9, TLC 8100 , Platelets 2.84lacks, LFT and KFT were normal, PT APTT INR are normal with ESR is raised at 28. MR Aortogram showed stenosis of lesions.

DSA Aortogram with Carotid Angiography done through trans-femoral artery approach showed

a. More than 90% stenosis in right common carotid artery, right vertebral and right internal carotid artery in proximal part from its origin.
b. More than 80% stenosis in left common carotid artery just after its origin.
c. More than 70% stenosis at left subclavian artery origin.
d. Left vertebral artery is feeding most of intracranial circulation
e. Abdominal artery, Bilateral renal arteries and coronary arteries are normal.

X ray Chest and ECG is showing features of left ventricular hypertrophy , 2D Echo and USG abdomen were normal



Slit Lamp pictures of cataract

Right eye cataract extraction with posterior chamber intraocular lens implantation under nil visual prognosis was done.

Discussion

A total of 5,000 patients have been registered to date. Worldwide incidence is estimated at 2.6 cases/million/year(13) with the majority of cases being reported from south-east Asia and the Indian sub-continent. It is primarily a disease of females (96%), as illustrated by Morimaki et al(14) . In India, mean age of onset of symptoms was 24.0 (± 8.8) years with a female preponderance (63%)(15).The age of our patient was 14 years. This case is also a female. The disease presents with non-specific features including fever, weight loss, arthralgia, myalgia, malaise, and anaemia. As inflammation progresses, stenotic lesions develop and patients present with

diminished or absent pulse (84.96%), vascular bruits (80 - 94%), hypertension (33 - 83%), retinopathy (37%), aortic regurgitation (20.24%) and less commonly as neurological manifestations and pulmonary artery involvement. There is paucity of pediatric literature on TA from our country.Our child also presented with absent pulse with retinopathy.

The commonest clinical feature at the time of presentation is hypertension. However, it should be noted that a pre-pulseless stage precedes the onset of hyper-tension in this condition. The clinical features during the prepulseless stage are non specific and include fever; night sweats, malaise, arthralgia, myalgia and skin rash., Second stage is of vascular insufficiency(12). Development of hypertension in these patients is multifactorial in origin. Retinopathy in TA can be hypertensive or hypotensive, but classical Takayasu's retinopathy is hypotensive which results from neovascularisation secondary to arterial hypotension(13).

Our Case alsohad ,typicalTakayasu's retinopathy with cataract.Cataracts have been described in association with TA, mostly after many years of evolution, rarely antedating other manifestations. They have been attributed to the patients' age and chronic treatment with steroids . In one series in which cataracts were described in 15 eyes from 65 patients, the mean age of the patients was 50.2 years, with a mean time of disease evolution of 16.8 years [19].But our child has cataract at a very young age without any start of steroid therapy which is atypical. In unpublished observations, we have seen cataracts in seven out of 83 patients (8.4%), mostly related to the above stated factors.

Angiography is still the gold standard technique for the diagnosis of vascular luminal abnormality and is now being replaced by digital subtraction angiography (DSA). A classification based on angiographic findings has been proposed (Table I). Type III is the most common type found in south-east Asia and Africa and is called as 'middle aortic syndrome' (53 - 76%).In abdominal aorta, the renal artery is the most frequently involved branch, often bilaterally8 . Renal artery stenosis is seen in 20 - 75% of all patients presenting with hypertension1 .Infrarenal aorta or iliac vessels are rarely involved. Under newer modalities, colour coded dopplersonography can facilitate an accurate diagnosis of the disease by the characteristic appearance9 ; computerised tomography and magnetic resonance angiography are a newer non-invasive approach to vascular disease including Takayasu's arteritis. The most important aspect of treatment is control of the inflammatory process, hypertension, and treatment of the complication if any.

Table I: Angiographic classification of Takayasu's arteritis (Takayasu conference, 1994)6

Type	Vessel involvement
Type I	Branches from the aortic arch
Type IIa	Ascending aorta, aortic arch, and its branches
Type IIb	Ascending aorta, aortic arch and its branches, thoracic descending aorta.
Type III	Thoracic descending aorta, abdominal aorta, and / or renal arteries.
Type IV	Abdominal aorta and / or renal arteries.
Type V	Combined features of Type IIb and IV.

N.B.: Involvement of the coronary or pulmonary arteries should be designated as C (+) or P (+) respectively.

Surgery seems to increase the long-term survival of the patient with stage III disease11 .

The mortality and morbidity data are limited due to rarity of disease, but a study from India showed a cumulative survival rate at 5 years after onset to be 91%(16). The mortality is mainly due to complications, mostly cerebrovascular disease, and cardiac failure. The incidence of rupture and bleeding complication of aneurysmal Takayasu's arteritis is low.

Medical treatment of Takayasu's arteritis includes use of immunosuppresants. The preferred drug is prednisolone which is given in high doses initially *(i.e.,* 1-2 mg / kg / day) and then tapered off after the activity parameters (especially the ESR) have come down. Long term low dose prednisolone is usually continued indefinitely(17). Use of additional immunosuppresants (azathioprine / methotrexate) has been tried by some workers, but is largely empirical(18). Our child was started on oral prednisolone for 1month and slowly tapered over 1 month. Since Takayasu's arteritis is a rare disease, exact guidelines for use of these drugs are not available in litera-ture. Role of antitubercular therapy is in those patients who either have coexisting tuberculosis or when there is possibility that steroid therapy can result in exacerbation of old tuberculosis. Platelet inhibitors can be used in patients having risk of cerebral infarction or ischemic changes of other organs due to occlussive arterial lesions. In patients with evidence of increased blood coagulability, anticoagulants are to be used.

Conclusion

Although cataract is rarely reported in TA, its appearance can be abrupt and proceed to visual loss.

It seems that in cases that are not related to age or treatment, the presence of cataracts confers an adverse visual prognosis, as severe TR seems to be invariably present . Therefore, we emphasize the importance of considering TA in the differential diagnosis of young patients presenting with severe cataracts, especially when bilateral. Subsequent adequate diagnostic workup can establish TA as the cause of cataracts and evaluate other sites of involvement amenable to proper treatment and follow-up.

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