Bilateral ischemic optic atrophy as a presentation of Takayasu arteritis in a 14 year old boy

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ABSTRACT

Takayasu's arteritis (TA), also known as "pulseless disease", is a rare chronic large vessel vasculitis of unknown origin. Most commonly it occurs in females (M: F: 4:1). Although commonly thought of an adult disease, the initial manifestation frequently appears during adolescence. A case report of a 14 year old hypertensive boy having history of focal seizures, right sided hemiparesis and sudden painless loss of vision is presented herewith. His bilateral radial and brachial pulses were not palpable and bruit was auscultated over left carotid artery. He was. Fundoscopy revealed bilateral ischemic optic atrophy. Takayasu's arteritis presenting with bilateral ischemic optic atrophy in an adolescent male child has rarely been reported.

Key words: takayasu arteritis, optic atrophy

INTRODUCTION

Takayasu's arteritis (TA) is a chronic large vessel vasculitis of unknown aetiology that predominantly involves aorta and its major branches. It has predilection for females with a reported 4:1 ratio in children and adolescents.[1] Indian origin aortoarteritis is a chronic granulomatous, necrotizing vasculitis, predominantly affecting the aorta with its branches.[2]

Tuberculosis, streptococcal infections, rheumatoid arthritis and other collagen vascular diseases have been debated as its aetiology in the past. Recently more emphasis has been given on an immunopathological cause.[3]The disease is classified based on the site of involvement into five types; the clinical manifestations occurs in accordance with the site involved.

The disease is global but studies have shown its very high incidence in Japan. Due to lack of population based study in India its geographical distribution and incidence is not very much ascertained but possibilities are that it could be high. Most of the patients, even in India who are diagnosed with this condition are women in their second or third decade of life. The occurrence of the disease in young children and infants is extremely rare with only a few cases reported all over the world.

CASE REPORT

A 14 year old male child presented with complains of two episodes of focal seizures (tonic-clonic) six months back lasting for 5 – 10 minutes of post-ictal drowsiness. Weakness of the right upper and lower limbs followed which was recovered by physiotherapy. There was a sudden painless bilateral vision loss 3 months back. He also suffered easy fatigability, dizziness, malaise and syncopal attacks.

Clinically, he was afebrile. Bilateral radial and brachial pulsations were not palpable, however both popliteal and dorsalis pedis were palpable. Respiratory rate was 18/min, BP in the upper limbs could not be recorded, BP in right lower limb was 160/80 and left lower limb was 150/90. Systemic examination did not reveal any abnormality.

Lab reports show raised ESR and positive CRP. Ophthalmic examination revealed complete loss of vision in both eyes with absence of PR

and PL. Fundoscopy revealed both eye- disc vertically oval cup with 0.8 to 0.9 CDR(cup disc ratio) with pale NRR(neuro retinal rim), lamellar dot sign was present and macula was oedematous. Impression on ophthalmic evaluation was bilateral ischemic optic atrophy (figure 1, 2).

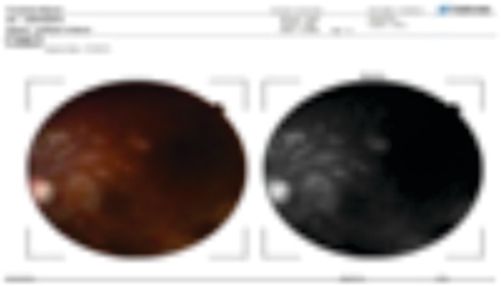

Fig.1. Showing disc pallor and venous tortuosity

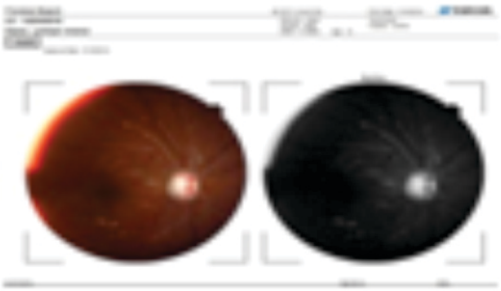

Fig.2. disc pallor and venous tortuosity suggestive of optic atrophy

TA presenting with bilateral ischemic optic atrophy in this age group is extremely rare. MR angiography was suggestive of left internal carotid artery and left common carotid artery narrowing with wall thickening and thickening of the wall of the arch of aorta. Right internal carotid artery and common carotid artery not visualised throughout the extent (figure 3,4,5) Impression of MR angiography was large vessel vasculitis which confirmed the diagnosis of takayasu's arteritis type 1.

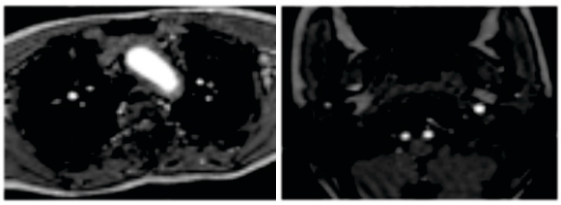

Fig.3,4. MR Angiography axial view of the thorax and neck showing thickening around the arch of aorta

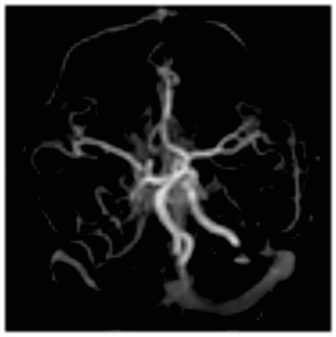

Fig.5. MR Angiography of the brain showing thickening of the left internal carotid and the absence of the right internal carotid artery throughout its course

DISCUSSION

TA, also known as "pulseless disease", "occlusive thromboaortopathy" and "Martorell syndrome", was first systematically described by a Japanese Ophthalmologist Mikito Takayasu. The disease remains an enigma as the exact cause of the disease is still not elucidated. It is a well known yet rare form of large vessel vasculitis.

In a study, the author has reported that the most frequent presentation in childhood is hypertension (82.6%), followed by headaches (31%), fever (29%), dyspnoea (23%), weight loss (22%) and vomiting (20.1%) [2, 16, 25–54]. Non-specific symptoms such as abdominal pain (16.6%) and vomiting can herald TA in children.[4]

In yet another study it was observed that arterial hypertension was the commonest mode of presentation followed by systemic

symptoms. Other related symptoms were due to ischemia and consisted of abdomen, chest, and limb pain. Inflammation markers were always abnormal. Angiography was performed in all cases; left subclavian artery and common carotid artery were more frequently involved. Renal artery stenosis was observed in two patients.[5]

Emely Z karam et al, described the unusual as well as typical ocular findings in patients with takayasu's arteritis. All patients had clinical and angiographic evidence of pulseless disease. Typical findings included retinal venous congestion, fluoroscein staining, capillary drop-out, microaneurysms and arteriovenous shunting. Unusual findings included cotton wool spots, anterior ischemic neuropathy and retinal emboli. The study concluded that Takayasu's disease can cause a variety of retinal manifestations. Less common findings being cotton wool spots, anterior ischemic optic neuropathy and retinal emboli.[6]

Rajashree Khot et al, reported a similar case of 22 years old patient with sudden loss of vision in both eyes 5 days prior to admission. This case was unique for it was a male patient who presented earlier i.e. 3rd decade with unusual presentation of bilateral anterior ischemic optic atrophy. In literature chronic retinal hypoxic changes and ischemic ocular inflammation resulting from cervical vascular occlusion have been extensively described. The eye signs originally described by Takayasu i.e. new vessel formation in the retina giving a wreath-like appearance are seen in long standing and chronic illness. Anterior ischemic optic neuropathy has rarely been reported in this condition, and only in conjunction with other ophthalmic findings.[7]

Laboratory investigations are usually non-specific such as raised erythrocyte sedimentation rate (ESR) in 50% cases, increased serum C-reactive protein (CRP) and normocytic normochromic anemia. The raised ESR, CRP and anemia reflect underlying inflammatory process.[8]

Eye disease in TA is an infrequent, late complication. Marked hypoperfusion of the posterior ciliary artery leads to ischemia of the retina, choroid, and anterior segment. This results in ischemic glaucomatous neuropathy, and later, optic atrophy without local arteritis.[9]

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