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A RARE CASE REPORT OF MOYAMOYA DISEASE WITH OCULAR ISCHAEMIC SYNDROME IN A 34 YEARS OLD INDIAN WOMAN.

Opthalmology

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ABSTRACT

A case of 34 years old Indian female with ocular ischaemic syndrome in both eyes with Moyamoya Disease reported as unusual presentation with its clinical diagnosis management and result. Came with a complain of Giddiness headache which were on and off for 4-6 months and did not relieve on routine medical therapy. The patient was investigated with CT Angiogram, diagnosed as Moyamoya disease and HRCT (High-resolution computed tomography) temporal bone and diagnosed as a chronic serous otitis media. Ocular ischemic syndrome is extremely rare in young patient with moyamoya disease may be particularly susceptible to the development of ocular ischemia due to the associated carotid occlusion. This rare instance of young female ocular ischemia in conjunction with moyamoya disease demonstrates the serious ocular and systemic sequelae of occlusive vascular disease.

KEYWORDS

moyamoya disease, ocular ischaemic syndrome.

CASE REPORT

In our tertiary care eye OPD patient presented with the complaint of diminution of vision since 1 year. On ophthalmic examination revealed best corrected visual acuity both eye 2/60 and both eye pupil 3mm regular in size and sluggish reactive to light. On fundoscopic examination both eyes vertically oval and slightly pale optic disc.

There was no history of strabismus, no history of trauma, any operation, No family history of such similar complaints, No history of weight loss or other symptoms suggestive of malignancy. Systemic examination within normal limits. No lymph node enlargement. Both eye IOP 17.3 mm hg & lacrimal passages patent both eyes. Rest ocular examination were within normal limits.

Complete work up revealed to rule out presence of any malignancy. All routine investigations were performed Hbsag negative, negative for HIV antibodies, all blood counts within normal limits and Ancillary lab results were unremarkable, patient showed a hypercoagulable state, and ANA, ASMA, Anti-DSDNA, and Anticardiolipin antibodies were negative patient had mildly elevated Serum Homocysteine levels 12.76.

Under conservative management patient received steroids at the discretion of the treating physician. Drug levels were monitored. IV Mannitol was reserved with deteriorating neurological status or increased intracranial pressure in the setting of haemorrhage. With vitreoretinal surgeon opinion Fluorescein angiogram has been done and diagnosis was confirmed as a right eye NVD and both eye peripheral nonperfusion ocular ischemic syndrome.

PHOTOGRAPHS

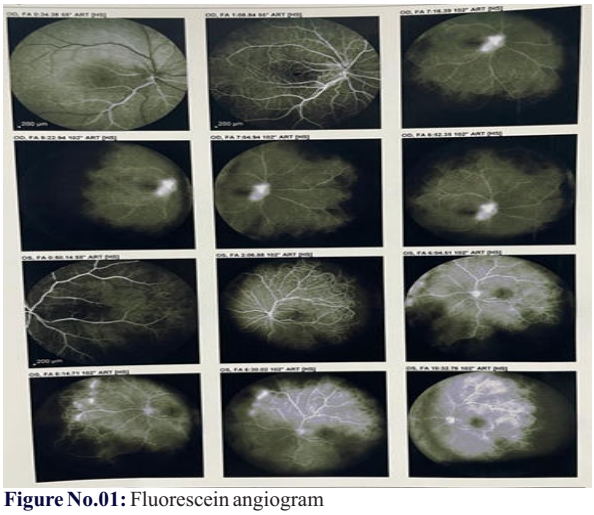

Figure No.01: Fluorescein angiogram

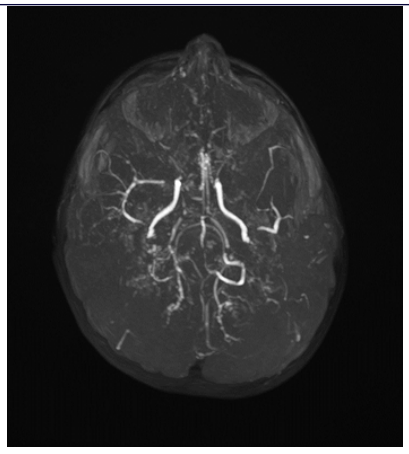

Figure No.02 Axial MRA

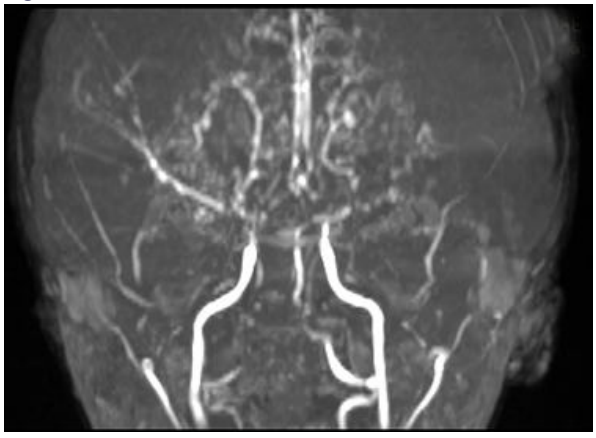

Figure No. 03 Coronal MR Angiogram

The retinal periphery was incompletely vascularized and new vessels extended into the vitreous. A small amount of vitreous hemorrhage was present inferiorly. The etiology of the retinal changes was unknown. Computed tomography (CT) and magnetic resonance imaging (MRI) were notable for non-visualization of the right internal carotid artery. The right carotid artery was presumed to be absent. The optic nerves were also slightly enlarged and have remained unchanged without treatment.

DISCUSSION

In 1963 Kearns and Hollenhorst reported a series of patients with "venous stasis retinopathy" associated with carotid stenosis.[1] In the

same year, Hedges also described retinal venous changes associated with carotid occlusion. [2] Knox reported cases of "ischemic ocular inflammation" in conjunction with carotid disease; many of these patients also exhibited the retinal features described by Kearns and Hollenhorst. Smith had already described the occurrence of neovascular glaucoma in the setting of carotid occlusive disease. [3] Today, the term "ocular ischemic syndrome" is used to encompass these many manifestations that result from chronic insufficiency of carotid or ophthalmic artery flow.

Panocular ischemia can cause pain and decreased vision. Anterior segment signs include episcleral venous congestion, corneal edema, uveitis, and cataract. Pupils may be dilated, sluggish, or nonreactive. Posterior segment signs include narrowed arterioles, dilated and irregular veins, neovascularization of the disk and/or retina, arteriovenous communications (anastomoses), spontaneous retinal arterial pulsations, and vitreous hemorrhage, presumably from neovascularization.[4-6]

Fluorescein angiography shows delayed and patchy choroidal filling, and an increase in arteriovenous transit time.[7] Retinal blood vessels may stain, and the optic disk may also show late staining. Areas of retinal capillary nonperfusion can frequently be identified.

OIS typically occurs in older patients.[7] Associated systemic diseases, in decreasing order of frequency, included systemic arterial hypertension, diabetes mellitus, ischemic heart disease, cerebrovascular accident, and peripheral vascular disease, all of which are diseases associated with atherosclerosis. [8] The ocular ischemic syndrome is thought to be a rare ocular manifestation of carotid atherosclerosis that usually occurs in elderly individuals who also have generalized atherosclerosis affecting other major organs.

Carotid occlusion can be associated with moyamoya disease, a central nervous system vasculopathy that occurs in all ethnic groups, but has been most frequently reported in the Japanese population. Moyamoya disease can occur at any age, but is most common in children.[9] Females may be more susceptible. Either unilateral or bilateral carotid stenosis with development of collateral vessels ("moyamoya vessels") is characteristic. Patients may be asymptomatic, or there may be severe neurological deficits. While adults more commonly have hemorrhagic events, children tend to present with ischemic events resulting in stroke. Moyamoya disease, a frequent cause of stroke in children, may be present for some time before symptoms occur.

Papavasileiou E et al., stated that although Moyamoya disease is a rare condition, it is important for the ophthalmologist to be cognizant of the ocular manifestations, as it can lead to irreversible vision loss and may be a harbinger of life-threatening CNS events. Considering that signs of the disease may be first observed in the eye before they are manifested in the cerebrovascular system, the ophthalmologist has an important role in the proper diagnosis and referral for further investigations. Collaboration between the ophthalmologist, neurologist, vascular surgeon, and primary physician is essential for appropriate management of the disease.[10]

CONCLUSION

Moyamoya disease is a rare condition, ophthalmic investigation related to Moyamoya disease include retinal vascular occlusion, visual field defects caused by ischemia or hemorrhage, optic disc abnormalities, and other fundoscopic findings. Retinal examinations would be beneficial for Moyamoya disease patients to reduce severe loss of vision.

ACKNOWLEDGEMENT

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