Case Report

Acute Idiopathic Maculopathy

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BACKGROUND: Acute idiopathic maculopathy is a relatively new clinical entity and an uncommon cause of sudden, severe, unilateral or bilateral, painless visual loss in young adults. The purpose of this article is to recognize the existence of this entity in South-East Asia and to highlight the condition which can be misdiagnosed because of relative non-familiarity of the general ophthalmologist with this condition.

CASE REPORT: We report a case of acute idiopathic maculopathy presented to the eye department, Baqai Medical University Hospital (Fatima Hospital) with sudden painless loss of vision in left eye of 50 years old house wife for the last 2-days. Examination revealed vision reduced to finger counting one-foot in her left eye. Both eyes were otherwise unremarkable except a serous elevation of the retina at the posterior pole of the left eye. B-scan revealed no sub-retinal mass while FFA led to the final diagnosis. Patient was reassured of transient nature of visual loss. Vision started improving after 2-weeks from the onset and recovered to 6/9 by the end of six-weeks.

CONCLUSION: Acute idiopathic maculopathy is also prevalent in our part of the world and it is prudent to include this condition in the differential diagnosis of all healthy, young adults presenting with sudden, painless visual loss and neural retinal detachment of the macula. Accurate diagnosis can be quite reassuring for the patient and save her from extensive work-up, unnecessary and sometimes harmful treatments.
history was also unremarkable except difficulty in near work.

On examination, best corrected visual acuity (BCVA) was 6/6 in right eye and FC 1/2-meter in left eye. Extra-ocular movements were full, free and painless. Pupils were briskly reacting to light; there was no RAPD.

On Slit-lamp examination, anterior segments of both eyes were unremarkable; there were no cells in the anterior chamber or anterior vitreous and no other sign of anterior uveitis.

Biomicroscopic posterior segment examination of the right eye was unremarkable (Fig. 1). A serous elevation of the retina at the posterior pole and few cells in the posterior vitreous was the only obvious finding in the left fundus (Fig. 2); there was no sign of retinochoroiditis, papillitis or vasculitis. Intraocular pressure by applanation tonometry was 12-mmHg in both eyes.

A normal B-scan US excluded choroidal thickening or a mass lesion behind the detached retina. A fundus fluorescein angiogram of the left eye was requested to determine the source of the sub-retinal fluid. There were multiple alternating dark (hypofluorescent) and white (hyperfluorescent) areas in the early phases of the angiogram (Fig. 3, 4). Late angiograms revealed hyperfluorescence due to both staining and pooling of the dye in the sub-retinal space (Fig. 5, 6). This characteristic angiographic pattern along with presentation with serous macular detachment leads to the diagnosis and helps differentiate from other conditions such as idiopathic choroidal neovascularization, posterior scleritis, central serous choroidoretinopathy (CSCR), retinal pigment epithelial detachment, amelanotic choroidal melanoma, choroidal metastasis and Vogt-Kayanagi-Harada syndrome.

Patient was reassured about the benign and self-limited nature of the disease and the possibility of complete visual recovery in few weeks time.

Visual recovery was noticed on second follow-up visit in two weeks time when vision in the left eye improved to 6/60. There was almost complete visual recovery at six-weeks from the onset of visual loss with visual acuity improved to 6/9 unaided in the affected eye. Fundus examination revealed a flat retina at the posterior pole. The other eye remains unaffected.

**DISCUSSION**

Acute idiopathic maculopathy is a rare and relatively new clinical entity first reported by Yannuzzi et al in 1991. They described a series of 9 cases from USA presenting with sudden, painless unioocular visual loss due to serous elevation of the macula associated with thickening of the underlying retinal pigment epithelium and coined the term “unilateral acute idiopathic maculopathy”. The visual symptoms typically occur following a prodrome of flu-like illness and resolved rapidly and almost completely with the resolution of exudative changes.

To date only 31 cases of this new entity have been reported in Ophthalmic literature. Fish et al reported the 10th case from USA in 1993 in which a shifting of the subretinal fluid represents a variation in the clinical presentation of this disease. Freund et al reported additional 17 cases from USA in 1996. They described new clinical findings such as eccentric macular lesions in four cases, sub-retinal exudation in two cases, bilateral involvement in few cases and associated mild papillitis in three cases. Qiu et al described the clinical and fluorescein angiographic features of 3 cases from China in 1998. Akduman et al described the progressive fluorescein and indocyanine green angiographic findings in a patient with acute idiopathic maculopathy from USA in 1999. Nakzawa et al described the clinical course and analyzed pathophysiology of a case of bilateral acute idiopathic maculopathy in a 33-year-old Japanese women in 2003. The last two cases of acute idiopathic maculopathy were reported by Adam et al from USA in 2004.

To the best of our knowledge no case of acute idiopathic maculopathy has been previously reported from the South-East Asia. Our case would be the 32nd case of acute idiopathic maculopathy and first reported case from this region.

With present knowledge and understanding acute idiopathic maculopathy is thought to be inflammatory process involving the outer neuroretina and retinal pigment epithelium. The condition belongs to a group of posterior uveitides of unknown aetiology and negative serological evaluation that affect otherwise healthy individuals of both sexes in second to sixth decade of life. They constitute an important cause of unilateral or bilateral visual disturbance in young individuals. These conditions produce characteristic ophthalmoscopic and fluorescein angiographic patterns, recognition of which is the mainstay in their diagnosis and management. They usually have a self-
limited course but exhibit variable prognosis depending on the individual disorder.

To date, no recurrence of the acute episodes have been reported. Development of choroidal neovascularization and disciform scarring has occurred in two cases and adversely affected the otherwise excellent long-term visual outcome.

Fig. 1: Right fundus: Normal disc, vessels and macula

Fig. 2: Left fundus: Serous elevation at the macula

Fig. 3: Right fundus: Normal fluorescein angiogram

Fig. 4: Left fundus: Early phase of fluorescein angiogram: Alternating dark (hypofluorescent) and light (hyperfluorescent) areas

Fig. 5: Left fundus: Late venous phase of fluorescein angiogram: Alternating dark (hypofluorescent) and light (hyperfluorescent) areas
Because of the self-limited nature of the condition and excellent visual recovery treatment neither seems prudent nor recommended. One of the patients in the original report who was treated with systemic corticosteroids developed milder pigmentary changes in the fovea in healed state; this by no means justify the use of systemic steroids.

CONCLUSION
Acute idiopathic maculopathy is also prevalent in South-East Asian countries and it is prudent to include this condition in the differential diagnosis of all healthy, young adults presenting with sudden, painless visual loss and neural retinal detachment of the macula in one or both eyes. Accurate diagnosis on the basis of typical clinical presentation and characteristic angiographic features can be quite reassuring for the patient and save him from extensive work-up and unnecessary, and sometimes harmful, treatments.

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