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Acute Posterior Multifocal Placoid Pigment Epitheliopathy

(A case report with Literature Review)

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SUMMARY

Acute posterior Multifocal placoid pigment Epitheliopathy is a rare idiopathic disease. A case with typical features of this disease is reported here, along with some review of the literature about this interesting condition.

CASE REPORT

A 20-year-old male presented in the Eye OPD of Shaikh Zayed Hospital with the complaints of sudden decrease in the vision of the right eye within the last 2-3 days. This patient was otherwise quite healthy, and there was no history of any significant relevant disorder in the past.

General Physical Examination did not reveal any abnormality.

Ocular examination revealed the unaided visual acuity to be C.F at 1 foot in the right eye, and 6/6 in the left eye. Refraction did not improve the visual acuity in the right eye. The left eye was found to be perfectly normal. The right eye had an intra-ocular pressure of 10 mmHg. The Anterior Chamber had 1+ flare and occasional cells. The pupil was normal and the lens was clear. The vitreous had a mild flare and a few cells but was clear enough to allow a good fundus examination which revealed multiple, small, 1/4 to 3/4 disc diameter, greyish-white lesions scattered throughout the posterior pole. The macula had a definite oedema clinically. A diagnosis of Acute posterior Multifocal placoid pigment Epitheliopathy (APMPPE) was made. Intra-venous fluorescein angiography confirmed the diagnosis. There was early hypofluorescence, with late hyperfluorescence of the lesions.

The blood counts and the chest X-Ray were normal. A complete battery of tests, usual for idiopathic uveitis patients, was not deemed necessary in view of the typical clinical and fluorescein angiographic picture.

A course of systemic steroids was given to this patient for a period of two weeks on empirical grounds. At the end of this period, the macular oedema had resolved and the visual acuity in the right eye improved to 6/9. This patient did not report for further follow-up.

DISCUSSION

Acute posterior multifocal placoid pigment epitheliopathy (APMPPE) was first described by Gass in 1968[1]. Many other reports followed[2-3]. Later reports include some studies of the long term visual out come, including the progressive fundus changes during the recovery phase[1]. A comparison is made here between our case and the more typical and recognized disease pattern in an effort to highlight the variability in the clinical presentation.

Mild episcleritis, anterior uveitis and vitritis has often been noticed in APMPPE cases but none of these is a constant feature of this condition. The fundus picture is however quite characteristic and often a single fundus examination is enough to suggest the diagnosis.

The clinical appearance of the fundus lesions is described as multiple, small, cream-coloured or yellow-white lesions scattered through-out the posterior fundus. The lesions in our case were quite typical, except for their color which was more on the grayish side, perhaps due to racial pigmentation differences. The disease remained unilateral in our case. Both eyes are usually involved although simultaneous bilateral involvement is not essential. Both sexes are equally affected. Adolescents or

young adults are more affected. The onset of symptoms often occurs about two weeks after a prodromal influenza-like illness.

The fundus lesions usually resolve in 2-6 weeks but leave permanent pigment alterations. If fovea is not directly involved, the visual prognosis is usually good[4].

There is no specific treatment for this condition

During the active stages, fluorescein angiography shows early hypofluorescence with late hyperfluorescence of the lesions. The early hypofluorescence is due to the blocking effect of the placoid lesions. The exciting light, as well as the emitted, fluorescent light is blocked by the lesions to produce the generalized effect of the hypofluorescence of the lesions. The late hyperfluorescence of the lesions is produced due to the staining of the lesions by the fluorescein. During the late phase, fluorescein is was had away from the rest of the choroid, which loses its fluorescence while the lesions, now stained with fluorescein, become hyperfluorescent.

Fluorescein angiography after resolution shows early hyperfluorescence of the lesion sites but no late staining. The effect it produced due to local pigmentary changes at the lesion sites. The retinal pigment epithelium is believed to be thinned out and more translucent at the lesion sites[5]. This appears as hyperfluorescent area due to the showing-through of the choroidal fluorescence during the early phase of the angiography. This hyperfluorescence fades out with the choroidal flow. There is no late staining or leakage of the eye. This late fluorescein angiography pattern is quite similar to the typical window-defect due to any other cause.

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