rapidly resolved, and the patient’s visual symptoms improved, although the visual acuity remained the same. During a follow-up examination 6 years after the treatment, no recurrence of the CNV was observed, and the number and the size of drusen had not increased significantly in either eye (Fig. 2C, D).

Comments

The appearance of the fundus in our patient and her family members was consistent with those of dominantly inherited drusen, although an autosomal dominant inheritance was not completely established. The phenotype of dominant drusen is considered to overlap that of age-related maculopathy to some extent, and mutations in the genes causing dominant drusen are possibly involved in some cases of age-related maculopathy.

It has been reported that CNV occurs infrequently in younger patients with hereditary retinal dystrophies, such as Stargardt’s disease, Best’s disease, and retinitis pigmentosa, and CNVs have been occasionally observed in patients with dominant drusen. Because of the infrequent association of CNV with retinal dystrophies, the natural history and management of these lesions has not been established. By examining the published reports of cases of CNVs associated with retinal dystrophies, Marano et al. found a tendency for cases of retinal dystrophy to develop a circumscribed, low-active CNV that frequently leads to a focal subretinal scar without any treatment. However, we believe that if the lesion is extraretinal, or large and active, CNVs can be treated successfully by photocoagulation to improve subjective symptoms, as was found in this case and in some cases of age-related maculopathy.

Key Words: choroidal neovascularization, dominant drusen, photocoagulation

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References


A Case of Unilateral Occult Maculopathy with Normal-Tension Glaucoma

Recently, a case of bilateral occult macular dystrophy (OMD) accompanying normal-tension glaucoma (NTG) was reported. Here, we report a case of unilateral occult maculopathy with an otherwise normal fundus except for a glaucomatous optic disc with NTG.

Case Report

A 75-year-old Japanese man was referred to our clinic on 11 January 2003 with a 1-year history of a progressive decrease in vision in the left eye but otherwise in healthy general condition. His visual acuity was 20/20 in the right eye and 20/50 in the left eye. Intraocular pressure (IOP) was 18mmHg in both eyes. Ophthalmoscopic and fluorescein angiographic examination revealed a few small spots of pigmentary degeneration outside the fovea in both eyes and enlargement of the optic disc cupping with disc hemorrhage in the left eye; otherwise, no visible vision-threatening abnormalities were present, especially at the fovea (Figs. 1A, B). Goldmann perimetry also showed a mild peripheral nasal step in the right eye, and a relative paracentral scotoma in the left eye (Fig. 1C).

We diagnosed NTG in both eyes and controlled the patient’s IOP to below 14mmHg with latanoprost eye drops. During a 1-year treatment follow-up period, visual acuity decreased gradually to 20/200 in the left eye, whereas it did not change in the right eye. The relative paracentral scotoma had enlarged in the left eye at 1 year. There were no apparent changes or new findings in either fundus. The International Society for Clinical Electrophysiology of Vision (ISEV) standard of full-field electroretinogram (ERG) results showed normal scotopic (isolated rod), photopic (cone), 30-Hz flicker, and bright-flash responses (combined rod and cone) in both eyes (Fig. 2A). Multifocal ERG responses from 103 cortically scaled areas within the central 30° were recorded in both eyes. Severely attenuated responses and delayed implicit times were recorded from only the central areas in the left eye, mimicking occult maculopathy (Fig. 2B). The mean deviation of the central 30-2...
program of the Humphrey perimeter (Model 745, Zeiss-Humphrey, Dublin, CA, USA) was −1.07 dB in the right eye and −5.82 dB in the left eye ($P < 1\%$).

**Comments**

A diagnosis of acute zonal ocular outer retinopathy (AZOOR), a syndrome characterized by acute loss of vision and progressive scotoma with minimal funduscopic changes, was excluded in the present case because the patient showed normal full-field ERGs in both eyes and did not show pigment epithelial atrophy or intraretinal pigment migration on follow-up examination. The absence of any antecedent viral-like illness and the patient’s healthy general condition without any medical disorders, especially autoimmune diseases, also did not support a diagnosis of AZOOR in this case.

On the other hand, OMD is a rare form of macular degeneration characterized by an essentially normal fundus and fluorescein angiography findings, but progressive decline of visual acuity in both eyes. OMD patients have normal full-

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**Figure 1.** A Ophthalmoscopic and B fluorescein angiographic findings showing enlarged optic disc cupping with disc hemorrhage in the left eye (shown on the right), with an otherwise mostly healthy fundus, especially at the fovea. C Goldmann perimetry showed a mild peripheral nasal step in the right eye, and a relative paracentral scotoma in the left eye.