APPLICATION OF A LASER FLARE-CELL METER TO EXAMINE BLOOD–AQUEOUS BARRIER FUNCTION IN A PATIENT WITH RETINITIS PUNCTATA ALBESCENS

Muh-Shy Chen, Ching-Chung Chang, I-More Fan, Man-Sim Cheng, and Ping-Kang Hou

Abstract: A laser flare-cell meter was used to examine blood–aqueous barrier function in a 21-year-old male patient who had typical retinitis punctata albescens with white dots scattered throughout large areas of the fundus in both eyes. Fluorescein angiography showed multiple areas of hyperfluorescence over dot lesions and window defects. Electroretinogram demonstrated subnormal amplitude and electrooculogram showed a reduced light-peak/dark-trough ratio. Goldmann perimetry revealed constricted peripheral fields. Laser photometry showed an increased aqueous flare intensity in both eyes compared to results in 10 normal subjects. Quantitative measurement of the aqueous flare intensity by a laser flare-cell meter might indicate abnormalities in the blood–aqueous barrier in patients with retinitis punctata albescens.

In 1882, Mooren first described retinitis punctata albescens as characterized by numerous discrete white dots throughout large areas of the fundus with maximum density in the equatorial region but sparing macular regions [1]. Lauber recognized that there were two forms of this disorder: progressive and stationary [2]. The progressive type has been called albipunctate dystrophy or progressive albipunctate dystrophy, while the stationary form has been called fundus albipunctatus or stationary albipunctate degeneration.

Retinitis pigmentosa has been shown to cause dysfunction of the blood–aqueous barrier [3]. We evaluated the function of the blood–aqueous barrier in a patient with retinitis punctata albescens to determine whether this disease has similar characteristics to retinitis pigmentosa.

Laser photometry is a non-invasive technique to assess blood–aqueous barrier function [4, 5]. We used a laser flare-cell meter to test for abnormalities in the blood–aqueous barrier. This is the first report to confirm the breakdown of the blood–aqueous barrier in patients with retinitis punctata albescens.

Case Report

The patient, who was a 21-year-old man with a history of night blindness since childhood, was examined in October 1999 due to progressively decreased vision. There was no family history of night blindness or consanguinity. His medical history was unremarkable.

Examination showed a visual acuity of 20/40 in both eyes with a correction of +1.0 D +2.5 D x 70° in the right eye and +1.5 D +1.25 D x 100° in the left eye. Biomicroscopy showed clear cornea and media in both eyes. Ophthalmoscopic examination showed numerous discrete, small, white dots throughout large areas of the fundus, especially in the midperipheral fundus (Fig. 1). Fluorescein angiography showed

![Fig. 1. Numerous discrete, small, white dots distributed throughout large areas of the fundus.](image-url)
multiple areas of hyperfluorescence over dot lesions and window defects, probably resulting from atrophy of the retinal pigment epithelium (Fig. 2).

Examination of color vision with the Farnsworth-Munsell 100-Hue test revealed a total error score of 288 in the right eye and 356 in the left eye, with no discernible axis for each eye. Dark adaptation studies with Goldmann-Weeker's adpatometer revealed that the final rod threshold was elevated 2.6 log units in the right eye and 2.8 log units in the left eye. Goldmann perimetry revealed constricted peripheral fields in both eyes. Electroretinography showed subnormal photopic and scotopic amplitudes in both eyes and electrooculography showed a light-peak/dark-trough ratio of 1.25 in the right eye and 1.23 in the left eye.

The aqueous flare intensity was measured using a laser flare-cell meter (FC 1000, Kowa, Tokyo, Japan) 30 minutes after pupillary dilation with 0.5% tropicamide and 5% phenylephrine hydrochloride. Five measurements in the mid-portion of the anterior chamber were taken to obtain the mean value. There was a marked increase in the aqueous flare values in both eyes: 12.3 ± 3.5 (mean ± standard deviation) photoncounts/ms in the right eye and 15.8 ± 3.8 photoncounts/ms in the left eye. In 10 age-matched control subjects, the aqueous flare intensity averaged 4.32 ± 0.86 photoncounts/ms.

Discussion

Retinitis punctata albescens is characterized by the presence of numerous discrete white dots scattered throughout the fundus without involving the foveal region. Fluorescein angiography can demonstrate hyperfluorescence over dot lesions and multiple areas of window defects, probably resulting from retinal pigment epithelium atrophy [6]. The ophthalmoscopic and fluorescein angiographic findings in our patient were identical to those in other reports [6–8]. Lauber divided the disease into two forms [2]. The first form is progressive albipunctate dystrophy, a disease with increasing constriction of visual fields, deterioration of central vision, anomalies of color vision, abnormal electrophysiologic function including both electroretinography and electrooculography, some optic atrophy, and occasionally some pigmentary changes. The second form is stationary albipunctate degeneration or fundus albipunctatus, a disease with a stationary or benign evolution, little or no constriction of visual fields, good central visual acuity, normal electroretinogram and mild abnormality gradually returning to normal levels after prolonged adaptation, normal electrooculogram, and the presence of white dots in the fundus without pigmentary changes [7]. The presence of night blindness, typical ophthamoscopic findings, and abnormalities in dark adaptation, visual field, electroretinogram, and electrooculogram, as seen in this patient, indicate the progressive form of retinitis punctata albescens.

The clinical features of retinitis punctata albescens must be differentiated from those of fundus flavimaculatus and dominantly inherited drusen. Krill and Klien have shown that dominantly inherited drusen, fundus flavimaculatus, and fundus albipunctatus have very similar retinal functional profiles and have categorized these three characteristics as floeked retina syndrome [8]. However, patients with the progressive form of retinitis punctata albescens, such as this patient, have abnormal retinal function and can be differentiated easily.

Patients with retinitis pigmentosa have been demonstrated to have dysfunction of the blood–aqueous barrier [3]. However, no studies of the function of the blood–aqueous barrier in retinitis punctata albescens have been reported. Usually, indirect measurements, inferes from fluorescein angiography, or anterior chamber coefficients of fluorescein leakage have been used as an index of blood–aqueous barrier permeability [9]. However, fluorophotometric analysis with systemic fluorescein administration is complicated by rapid metabolism of the dye and the adverse effects of fluorescein. Laser photometry is a feasible noninvasive method to assess the function of the blood–aqueous barrier [4, 5]. The laser flare-cell meter used in this study consists of a helium-neon laser slit lamp and binocular microscope equipped with a photomultiplier and a personal computer to analyze the data. The instrument can measure protein concentration in the aqueous humor in vivo by measuring light scattered from aqueous protein.

The aqueous flare intensity in this patient was elevated about 2.9 times in the right eye and about 3.7 times in the left eye compared to normal values obtained from age-matched control subjects. The increase in the aqueous flare intensity in this patient showed alteration of the blood–aqueous barrier associated with retinitis punctata albescens. The sites of the breakdown of the blood–aqueous barrier remain unknown, but are possibly at the stroma of the iris root and in the iris vessels [10]. The changes may be mediated by chemical mediators [11], and may also be influenced by neurogenic or humoral factors [12]. Meanwhile, the proteins released by the breakdown of the blood–retinal barrier may diffuse anteriorly into the anterior chamber directly, or these released factors may have a direct effect on the vascular permeability of the anterior segment. To the best of our knowledge, this study is the first report of the evaluation of blood–aqueous

Fig. 2. Fluorescein angiography showing hyperfluorescence over dot lesions and multiple window defects.
barrier function in retinitis punctata albescens with a laser flare-cell meter.

References

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