CHOROIDEREMIA: ONE-YEAR FOLLOW-UP WITH SCANNING LASER POLARIMETRY EXAMINATION

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ABSTRACT

Case report: We report the follow-up of a case of choroideremia who underwent three white-on-white automated visual field and three scanning laser polarimetry (SLP) examinations by means of a GDx VCC in the course of one year. A bilateral perimetric deterioration in indices and scotomas was found. As a result, retinal nerve fiber layer retardation parameters and maps changed on GDx VCC advanced serial analyses in both eyes.

Discussion: Serial analyses with GDx VCC may be used as objective and quantitative tests to assess the progression of chorioretinal dystrophies like choroideremia (Arch Soc Esp Oftalmol 2008; 83: 487-492).

Key words: Choroideremia, GDx, progression, chorioretinal atrophy, scanning laser polarimetry.
INTRODUCTION

Choroideremia (CRDM) is a chorioretinal dystrophy (CD) displaying X-linked recessive inheritance, characterized by reduced night vision, and a progressive and centripetal loss of the peripheral visual field.

Visual field scotomas are usually related to areas affected by clinically distinguishable chorioretinal atrophy (CA). These areas coalesce with time, and visual field constriction may be checked using perimetry.

Since CA increases delays in GDx (1) laser polarimetry (LP), the premise was adopted that time evolution for CRDM might be assessed with this technology.

CASE REPORT

Male, 53 year old patient, previously diagnosed of CRDM (nyctalopia, perimetric progressive constriction, well defined CA areas by fluorescein angiography (fig. 1), altered electroretinogram, normal plasmatic ornithine, and typical X-linked inheritance pattern) with no other medical background. Three comprehensive ophthalmological examinations were carried out by the same specialist in one

Fig. 1: Ocular fundus and fluorescein angiography for the right eye (top) and the left eye (bottom). The underlying choroidal vessels may be visualized through the areas patched with atrophy of the pigmentary epithelium.
year, including best corrected visual acuity (BCVA), automated white-white campimetry for the central 30º (Octopus 301, TOP strategy, G1 Haag-Streit, Koeniz, Switzerland), laser polarimetry (LP) (GDx with variable corneal compensation (VCC), software version 5.5.0; Carl Zeiss Meditec, Dublin, CA, USA), slit lamp examination, Goldmann aplanation tonometry, and retinal examination. Four visual fields had been performed for the patient before the follow-up period, so a possible learning effect was discarded.

AVMC stayed bilaterally in the unit during the whole of the follow-up period. Deterioration was noticed for the scotomas represented in the maps, and for Bebie’s curves. Besides, the mean defect (MD) got worse during the serial examinations for both eyes. Reliability indices were acceptable (Fig. 3). Serial analysis using GDx VCC showed the corresponding bilateral-shaped changes for the temporal-superior-nasal-inferior-temporal (TSNIT) maps, the retinal nerve fibre layer (RNFL) maps, and the difference-from-baseline maps). Besides, absolute parameters increased, and the nervous fibre indicator (NFI) decreased. All GDx examinations received an ‘acceptable quality’ score (Q score ≥ 8) (fig. 3).

On the other hand, intraocular pressure remained at normal levels, and no significant changes were detected in biomicroscopy or ocular fundus.

**DISCUSSION**

Measuring the changes in the state of polarization of light traversing the eye is a good technique for researching ocular media. Commercial devices developed so far have focused on the diagnosis and follow-up for glaucoma.

However, laser polarimetry (LP) has been shown to be useful for other clinical purposes, such as locating the leaking point and the fluid area for central serous chorioretinopathy (2), checking the integrity of Henle fibres in neovascularization secondary to age-related macular degeneration (3), or quantifying posterior capsular opacification in non-glaucomatous patients (4). To the best of our knowledge, the current communication seems to be the first study in the literature about assessing the progression of choriotinal dystrophy (CD) by means of LP.

Follow-up was carried out by automated perimetry and GDx VCC. Automated perimetry is one...
Fig. 3: Automatic perimetry and GDx and VCC examination for the right eye (top) and the left eye (bottom). Note how scotomas either extended and/or got deeper, whilst new scotomas were visible. The mean defect (MD) got worse for both eyes. Note the progression of higher delay signal areas for TSNIT maps, RNFL (retinal nerve fiber layer), and difference from baseline. GDx parameters show a trend consistent with maps during follow-up.
of the most common tests for assessing CD progression, but results are dependent upon patient’s willingness to collaborate, and may be affected by the learning effect. Besides, patients are required to pay attention for a few minutes at least, so children and patients affected by neuropsychiatric pathologies are not suitable for this kind of test.

On the other hand, GDx VCC is an objective test, with no dependence on patient’s collaboration. The test takes only 0.8 seconds for each eye, and it has been proven to be a reproducible technique (5). The only important requirement is for patients to keep fixation.

Another standard test for CD follow-up is ERG. This is however performed by neurophysiologists in many hospitals, so that it may not be available as often as required. Another drawback of this technique is the large variability of results between different visits.

Though new studies are necessary with large numbers of patients (perhaps multicentric studies, due to the small amount of cases and the slow evolution of atrophies), the current case study may suggest that LP through GDx VCC may be a suitable technique for assessing the evolution of progressive CD, such as CRDM.

**REFERENCES**