

MEWDS: ELECTROPHYSIOLOGICAL EVIDENCE OF RETINAL GANGLION CELL DYSFUNCTION

RENATO SANTOS SILVA^{1,2}, LUÍS FIGUEIRA^{1,2}, ELISETE BRANDÃO¹, MANUEL FALCÃO^{1,2},
AMÂNDIO ROCHA SOUSA^{1,2}, FERNANDO FALCÃO-REIS^{1,2}

¹Department of Ophthalmology, Hospital Sao Joao, Porto, Portugal
²Faculty of Medicine, University of Porto, Porto, Portugal

Introduction - Multiple Evanescent White Dot Syndrome (MEWDS), first described in 1984, is characterized by the appearance of altered visual fields, often with increased blind spot or paracentral scotoma, decreased visual acuity (VA), photopsias and myodesopsias, being more frequent in young female patients, from 15 to 50 years. It typically affects only one eye and has spontaneous resolution in 4 to 6 weeks.

In 2014, Hakayama, H and colleagues demonstrated, with OCT, the reduction of the retinal ganglion cell layer, which remained decreased after the complete resolution of the symptoms and total recovery of the ellipsoid. However, the functional evaluation of retinal ganglion cells has not been described so far in patients with MEWDS.

Material and Methods - Clinical and electrophysiological evaluation of a patient with MEWDS in the initial phase and 6 weeks after the beginning of the symptoms, with fluorescein angiography, indocyanine green angiography, OCT, autofluorescence, Goldman's kinetic perimetry and electrophysiological evaluation with flash electroretinogram (ERG), pattern ERG, multifocal ERG and pattern visual evoked potentials (VEP)

Results - A 17-year-old female patient with decrease VA and paracentral scotoma in the right eye, observed 4 days after the onset of symptoms, being evident the presence of multiple whitish spots in the peri-macular region and foveal granularity. In fluorescein angiography, there were multiple clusters of hyperfluorescence in early phase and, in the late phase, impregnation of the optic disc. Indocyanine green angiography showed multiple hypocyanescent points. In the OCT, ellipsoid disruption was observed. An increase in the blind spot was evident in Goldman's kinetic perimetry. Based on these findings the diagnosis of MEWDS was made (figure 1).

Electrophysiological evaluation revealed no changes in flash ERG or in the pattern PEV. In the multifocal ERG the decrease in the central response (R1) was evident, with the remaining rings within normal range. In the pattern ERG the N95 wave was decreased, with a normal P50 wave, compatible with retinal ganglion cell dysfunction (figure 2).

The clinical and electrophysiological evaluation was repeated 6 weeks after the onset of symptoms. At that time fluorescein angiography, indocyanine green angiography, OCT were normal, being only evident an increase of the blind spot although smaller than the initial one (figure 1). Electrophysiological assessment was normal, with reversal of pattern and multifocal ERG changes (figure 2).

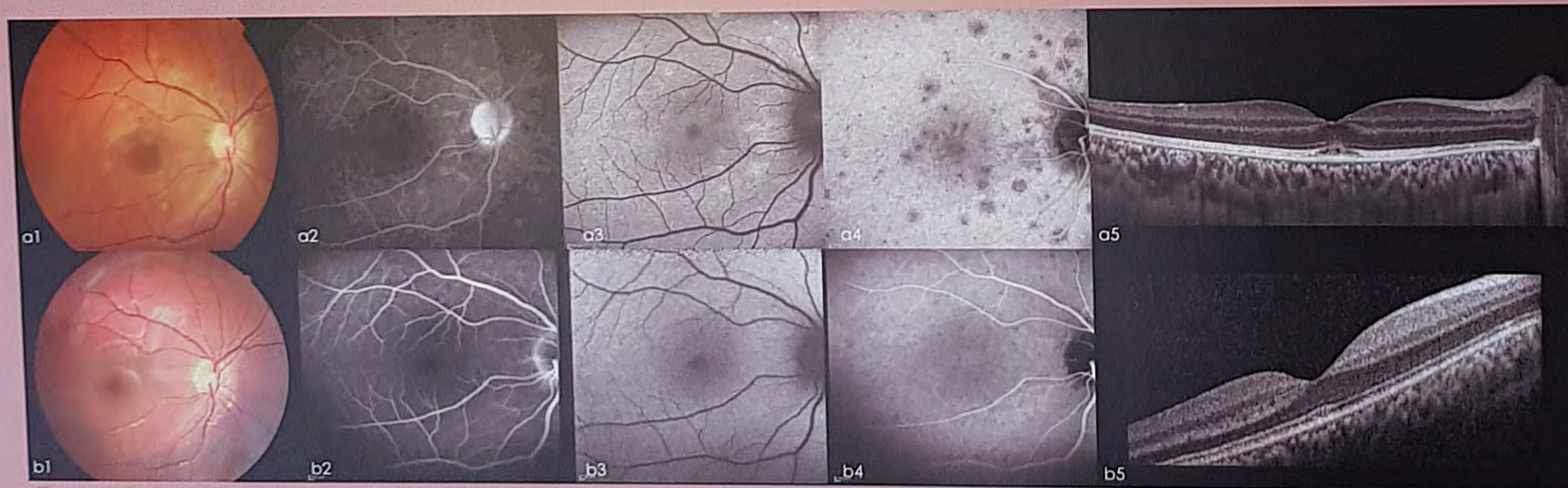


Figure 1 - Images from the top row [a] show examinations performed on the day of the first observation, 4 days after the onset of symptoms. A number of whitish spots and foveal granularity are observed in retinography (a1), optical disc impregnation in fluorescein angiography (a2), areas of hyperautofluorescence with hypoautofluorescent point (a3), multiple hypocyanescent points in indocyanine green angiography (a4), and disruption of ellipsoid in the OCT (a5). The images from the lower row [b] corresponded to the observations made 6 weeks after the onset of symptoms, with no changes in retinography (b1), fluorescein angiography (b2), autofluorescence (b3), indocyanine green angiography or in the OCT (b5).

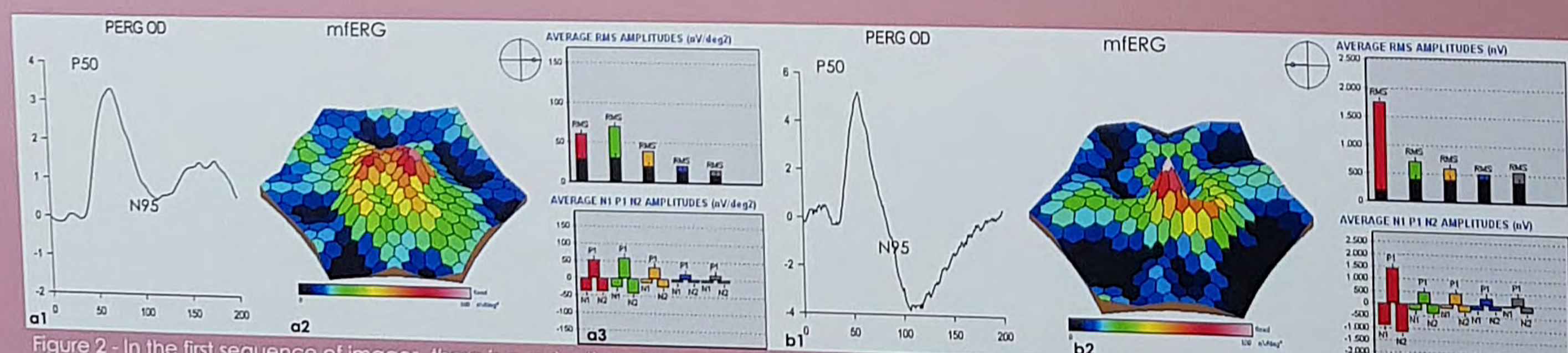


Figure 2 - In the first sequence of images, there is a reduction of the N95 wave in the pattern ERG (a1), which indicates dysfunction of the retinal ganglion cells, and decrease of the central response in the multifocal ERG (a2 and a3), both present 5 days after the onset symptoms. The second sequence shows the return to normal of ERG pattern (b1) and multifocal ERG (b2 and b3)

Conclusion - To date there is no publication regarding the pattern ERG and evaluation of retinal ganglion cell function in patients with MEWDS, this being the first case described in the literature.

The reduction of the N95 wave observed in the pattern ERG performed 5 days after the onset of symptoms suggests retinal ganglion cell dysfunction. The decrease in the retinal ganglion cell layer described in 2014 by Hakayama appears to have a functional repercussion. However, unlike the structural change that is maintained over time, the function of retinal ganglion cells seems to recover after 6 weeks.

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