

Multifocal electroretinography in type 2 idiopathic macular telangiectasia

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Abstract

Background To characterize the electroretinographic response of the macula by multifocal electroretinography (mfERG) in patients with type 2 idiopathic macular telangiectasia (MacTel).

Methods A prospective study of mfERG in patients with type 2 MacTel was conducted from April 2009 to November 2009. mfERGs were recorded using a visual evoked response imaging system (MonElec2, Metrovision, Perenchies, France). The International Society for Clinical Electrophysiology of Vision (ISCEV) guidelines were followed. Patients with type 2 MacTel confirmed by fundus fluorescein angiography without subretinal neovascularisation were included. For recording purposes, 61 stimulus hexagonal elements were used. The first-order kernel

mfERG responses were analyzed. Individual mfERG responses for the hexagons were grouped into concentric rings centered on the fovea for analysis ($< 2^\circ$, $5\text{--}10^\circ$, $10\text{--}15^\circ$ and $> 15^\circ$). Student's *t*-test and Mann-Whitney U test and linear regression analysis was performed with STATA ver 11.1 (StataCorp, College Station, TX, USA).

Results Twenty eight eyes of 14 patients and 20 eyes of ten normal controls were included in the study. The mean log-MAR visual acuity of the patients was 0.51 (Snellen equivalent 20/63). The mean N1 amplitude (nv/deg²) of patients were significantly reduced compared to controls and were as follows: 8.91 ± 14.00 vs 43.44 ± 9.55 ($p < 0.0001$) in less than 2° , 9.24 ± 10.47 vs 22.00 ± 3.87 ($p < 0.0001$) in $5\text{--}10^\circ$, 8.57 ± 10.02 vs 15.24 ± 1.89 ($p < 0.0001$) in $10\text{--}15^\circ$, and 7.03 ± 6.52 vs 12.47 ± 2.62 in $> 15^\circ$ ($p < 0.001$). The mean P1 amplitude (nv/deg²) was also significantly reduced in patients compared to controls and was as follows: 27.66 ± 37.44 vs 96.20 ± 12.41 ($p < 0.0001$) in less than 2° , 22.61 ± 19.38 vs 53.78 ± 9.79 ($p < 0.0001$) in $5\text{--}10^\circ$, 18.75 ± 20.21 vs 35.22 ± 4.16 ($p < 0.001$) in $10\text{--}15^\circ$, and 17.10 ± 12.54 vs 25.71 ± 3.93 ($p < 0.001$). The implicit time of N1 and P1 were also delayed significantly in all the rings. The mean central foveal thickness assessed by optical coherence tomography (OCT) scan was $84.78 \pm 45.12 \mu\text{m}$. There was poor correlation between mfERG amplitudes or implicit times with either the visual acuity or OCT central thickness.

Conclusion mfERG showed significant reduction in amplitudes and implicit times of the waveforms in patients with type 2 MacTel in all the rings, suggesting a more generalized affection of the macula. The maximum reductions were seen in the $< 2^\circ$ rings. Although there was poor correlation between the visual acuity and the amplitudes of the waveforms, mfERG is a useful investigative modality for functional assessment of macula in type 2 MacTel patients.

Keywords mfERG · PFT · IMT · IJRT

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Authors have full control of all primary data, and we agree to allow Graefes' Archive for Clinical and Experimental Ophthalmology to review their data upon request.

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Introduction

Idiopathic juxtafoveal retinal telangiectasis (IJRT) was first defined in 1982 by Gass and Oyakawa as a unilateral or bilateral disease associated with incompetent retinal capillaries only in the perifoveal or juxtafoveal area [1]. In 1993, Gass and Blodi presented a revised classification, staging, and hypothesis on the pathogenesis of IJRT which was based largely on clinical examination and fluorescein angiography [2].

Recently, Yannuzzi proposed a new term for this disease, referring to it as idiopathic macular telangiectasia (MacTel) [3]. He described a new classification of MacTel comprising of two types. Type 1 or aneurysmal telangiectasia consists of multiple capillary, venular, and arteriolar aneurysms in both superficial and deep retinal circulations. It is associated with minimal ischemia, and does not have secondary neovascularization. Type 2 MacTel manifests in the middle-aged to elderly, and the visual acuity is usually good [4]. The macula shows a perifoveal gray halo due to loss of local retinal transparency, with cystic appearance of the fovea. Type 2 is the more common form of MacTel characterized by bilateral involvement and later onset than group 1, grouped into 2A, acquired, and 2B, congenital. Unlike in type 1 patients, type 2 patients show no hemorrhages, aneurysms, or lipid accumulation. We showed in our previous study that the temporal macula was most commonly involved [5]. We also showed that the distance of the parafoveal telangiectasia from the center of the foveal avascular zone (FAZ) could be up to 2,530 μm . The sight-threatening complications of type 2 MacTel can be a result of either nonproliferative (exudation and foveal atrophy) or proliferative disease [subretinal neovascularization (SRN) or fibrosis].

The pathogenesis of this disease is still not known, though there is speculation that impaired transport and/or storage of lutein and zeaxanthin may play a role. A central depletion of macular pigment in patients with type 2 MacTel has recently been established. This depletion of macular pigments have been studied non-invasively by confocal blue-reflectance, autofluorescence and macular pigment reflectometer [6–9]. Anatomic alterations on optical coherence tomography (OCT) have been also reported [10, 11]. Bottoni et al. described blue-reflectance and autofluorescence changes in correlation to the OCT findings in type 2 MacTel [7]. Functional deficit due to damage have been revealed by microperimetry and fine matrix mapping [12–16].

Currently, the gold standard for diagnosis of this disease is fluorescein angiography [17]. Multifocal ERG (mfERG) is an emerging modality for assessing retinal function in various retinal disorders. However, there is no information regarding macular function in type 2 MacTel patients using noninvasive objective investigative modality like multifocal

response can be derived from the multifocal electroretinogram, which concurrently stimulates a large number of retinal locations [18]. Numerous studies have reported the effects of various retinal diseases on the local responsiveness of the retina, confirming the ability of mfERG to detect and map small dysfunctional regions [9, 19–21]. To the best of our knowledge, there is no literature available on the mfERG responses in MacTel. Analyzing mfERG responses in MacTel could help in understanding the origin and pathophysiology of the disease, which could guide possible management. We hypothesized that since MacTel is a disease localized to the macula, the local electroretinographic response may be reduced, and hence we performed mfERG in these patients to prove our hypothesis.

Methods

A prospective study of mfERG in patients with type 2 MacTel was conducted from April 2009 to November 2009 at LV Prasad Eye Institute, Hyderabad, India. Twenty-eight eyes of 14 patients (study group) and 20 eyes of ten normal controls were included in the study. Patients with a clinical diagnosis of type 2 MacTel and confirmed by fundus fluorescein angiography without subretinal neovascularisation were included. All patients underwent a comprehensive eye examination, which included recording the best-corrected visual acuity (BCVA), intraocular pressure by applanation tonometry (IOP), slit-lamp biomicroscopy and a detailed fundus examination. mfERGs were recorded at each visit using a visual evoked response imaging system (MonElec2, Metrovision, Perenchies, France). The International Society for Clinical Electrophysiology of Vision (ISCEV) guidelines were followed [22]. All patients had clear media before the recordings. Data collection was performed with approval from the Institutional Review Board.

For recording the mfERG, a central red square was used as a fixation target, and good fixation was ensured throughout. Pupils were maximally dilated using 1 % tropicamide eye drops. The stimulus matrix consisted of 61 scaled hexagonal elements displayed on a monochrome monitor driven at a 75 Hz frame rate. The radius of stimulus array subtended was $20^\circ \times 20^\circ$ at a viewing distance of about 27 cm, and each element was independently alternated between black ($<5 \text{ cd/m}^2$) and white (200 cd/m^2) using a binary m-sequence. The cornea was anesthetized with proparacaine hydrochloride eye drops and the mfERG was recorded using the L.V. Prasad Eye Institute (LVP) electrode [23]. A ground electrode was attached to the ear lobe. Mean stimulus luminance was adjusted to 110 cd/m^2 . An individual recording was divided into short segments of 30 s each. The signals were fed into an amplifier with a bandwidth of 0.5–100 Hz.

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