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CASE REPORT

Unilateral electronegative ERG in a presumed central retinal artery occlusion

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Luiz H Lima[1]
Wener Cella[2,4]
Claudia Brue[5]
Stephen H Tsang[2,3]

[1]Federal University of Sao Paulo (UNIFESP), Sao Paulo, Brazil; [2]Department of Ophthalmology, [3]Bernard and Shirlee Brown Glaucoma Laboratory, Edward S Harkness Eye Institute and Departments of Opthamology, Pathology and Cell Biology, Columbia University, New York, NY, USA; [4]Department of Ophthalmology, University of Brasilia, Brasilia, Brazil; [5]Department of Opthamology, University Politecnica delle Marche, Ancona, Italy

Abstract: A unilateral electronegative electroretinogram (ERG) was seen in a 94-year-old man with presumed central retinal artery occlusion. Goldmann perimetry revealed central scotoma in the right eye and no abnormalities in the left eye. Full-field ERG in the right eye described a reduction of the b-wave with a relative preservation of the a-wave which is characteristic of electronegative ERG. Hence, our case illustrates that ERG testing is essential for the work-up of individuals with suspected retinal vascular disorders.

Keywords: central retinal artery occlusion, electronegative ERG, inner retina, spectral domain optical coherence tomography

Introduction

Patients with acute central retinal artery occlusion (CRAO) typically relate a history of painless visual loss occurring over several seconds followed by classic signs of retinal infarction in the fundus. Spontaneous recanalization frequently occurs in many patients making CRAO a challenging diagnosis.[1] In such cases, electroretinogram (ERG) can be a helpful ancillary test for CRAO diagnosis. As the retinal artery supplies the middle and inner layers of the retina, the physiological consequence of CRAO is a predominant reduction of the scotopic maximal ERG b-wave with preservation of the a-wave.[2] A negative full-field ERG usually describes International Standard for Clinical Electrophysiology of Vision standard maximal response in which the amplitude of b-wave is smaller than the minimally attenuated a-wave.[3–5] This reduced b-wave is usually associated with a number of congenital and acquired conditions such as X-linked juvenile retinoschisis,[6,7] congenital stationary night blindness,[8,9] CRAO,[10,11] birdshot chorioretinopathy,[12–14] paraneoplastic and autoimmune retinopathies,[15–17] and retinal toxicity.[18,19]

We report an atypical presentation of CRAO in which a unilateral negative ERG was valuable in diagnosis and the initiation of systemic work-up.

Case report

A 94-year-old ophthalmologist noted central vision deterioration in the right eye over the past two years. Medical history was significant for an 18-year history of systemic hypertension, for which the patient was being treated with beta-blocker (atenolol) and diuretic (hydrochlorotiazide) drugs. Screening procedures for malignancy including a careful medical history, a chest X-ray, and liver enzymes were negative. There was neither history of retinotoxic medication use, nor family history of eye disease. Ultrasound examination of the carotid arteries was performed and showed tiny atherosclerotic plaque

Correspondence: Stephen H Tsang
Bernard and Shirlee Brown Glaucoma Laboratory, Edward S Harkness Eye Institute, Columbia University, 160 Fort Washington Avenue, Room 513, New York, NY 10032, USA
Tel +1 212 342 1186
Fax +1 212 342 7942
Email gene.targeting@gmail.com

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without hemodynamically compromised lesion. Two years before his presentation, his ophthalmologist recorded 20/60 vision in the right eye and 20/20 in the left eye.

Visual acuity at presentation was 20/80 in the right eye and 20/20 in the left eye. The patient's central vision in the right eye progressively deteriorated but stabilized over six months, resulting in acuity of 20/100. Anterior segment examination was unremarkable, except for mild superficial punctate keratitis, a few guttata, more prominently on the left cornea, and pseudophakia. There was no iris neovascularization in both eyes. The funduscopic examination revealed hard drusen on the macular area in both eyes, and atrophic retinal pigment epithelium (RPE) changes with pigmentary clumps on the macular area of left eye. Minor retinal artery narrowing was observed in both eyes without optic atrophy (Figure 1A and 1B). Fundus fluorescein angiography (FA) was unremarkable in the right eye and revealed transmission defect in the posterior pole compatible with RPE abnormalities in the left eye (Figure 1C and 1D). FA choroidal phase was 6 seconds in the right eye and 5 seconds in the left eye; FA arteriovenous phase was 14 seconds in the right eye and 11 seconds in the left eye. Spectral domain optical coherence tomography (SD-OCT) demonstrated reduced thickness of the inner retina in both eyes but more prominently in the right eye (Figure 2). The thickness of the inner retina (from retinal nerve fiber layer to inner nuclear layer) was measured 600 μm, 1,000 μm, and 2,000 μm nasally and temporally from the fovea. We also measured the inner retina

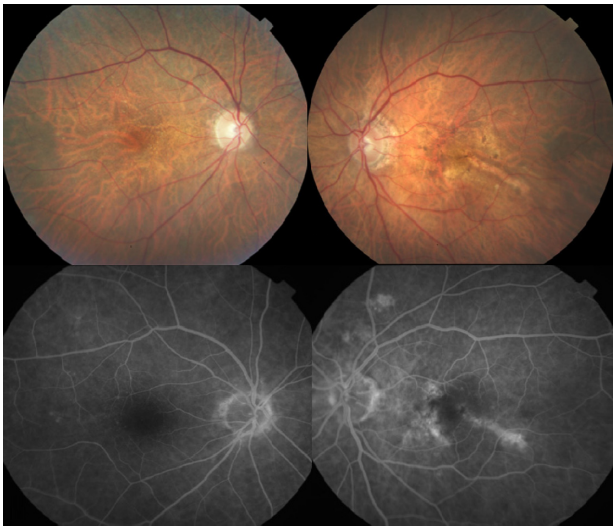

Figure 1 Color fundus photographs shows few drusen on the posterior pole in both eyes, and retinal pigment epithelium (RPE) atrophy with pigmentary clumps on the macular area of left eye. Minimal retinal artery narrowing can be observed in both eyes. Fundus fluorescein angiography depicts window defect in the posterior pole compatible with RPE atrophy in the left eye in the venous phase. There was no apparent delayed fluorescein filling in both eyes.

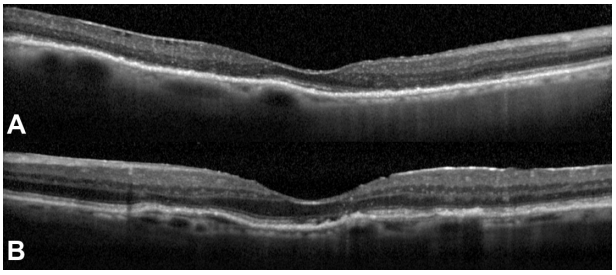

Figure 2 Spectral domain optical coherence tomography (SD-OCT) revealed reduced retinal thickness in the right eye (**A**) and left eye (**B**). Note the decreased thickness of the inner retina in right eye is more advanced than the left eye.

thickness at the same locations described previously in 10 normal age-matched control eyes. The measurements were performed using the 1,000 μm caliper available in the OCT-SLO Spectralis, Heidelberg Retina Angiograph 2 (Heidelberg Engineering, Dossenheim, Germany). Our measurements showed that the inner retina thicknesses were decreased at 600 μm, 1,000 μm, and 2,000 μm nasally and temporally from the fovea in the patient's right eye in comparison to the left eye and normal control eyes (Figure 3). Goldmann perimetry demonstrated central scotoma in the right eye and no abnormalities in the left eye. Full-field ERG in the right eye was attenuated in both scotopic and photopic responses and revealed a b-wave smaller than the a-wave amplitude, ie, electronegative maximal response. In the left eye, minimal scotopic and photopic attenuation was observed (Figure 4). Western blot analysis of 1:500 dilutions of the patient's serum did not show any reactivity against retinal protein extract.

Discussion

Our patient presented with a history of systemic hypertension treated with antihypertensive medications and vision loss of

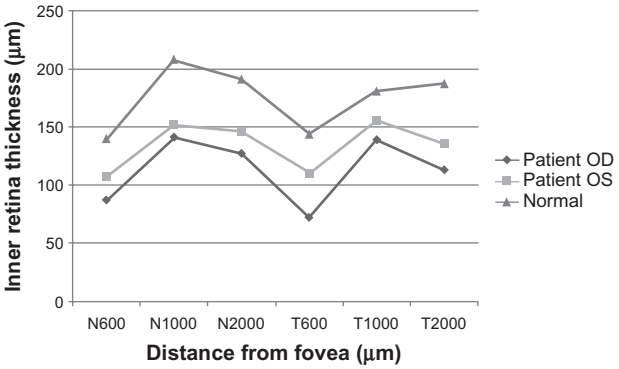

Figure 3 The thickness of the inner retina was established as the distance between retinal nerve fiber layer and inner nuclear layer and was measured using the 1,000 μm caliper available in the OCT-SLO Spectralis. Ten age-matched normal eyes were used as controls. Observe the decrease in the nasal and temporal inner retinal thickness in the right eye in comparison to the left and normal control eyes.

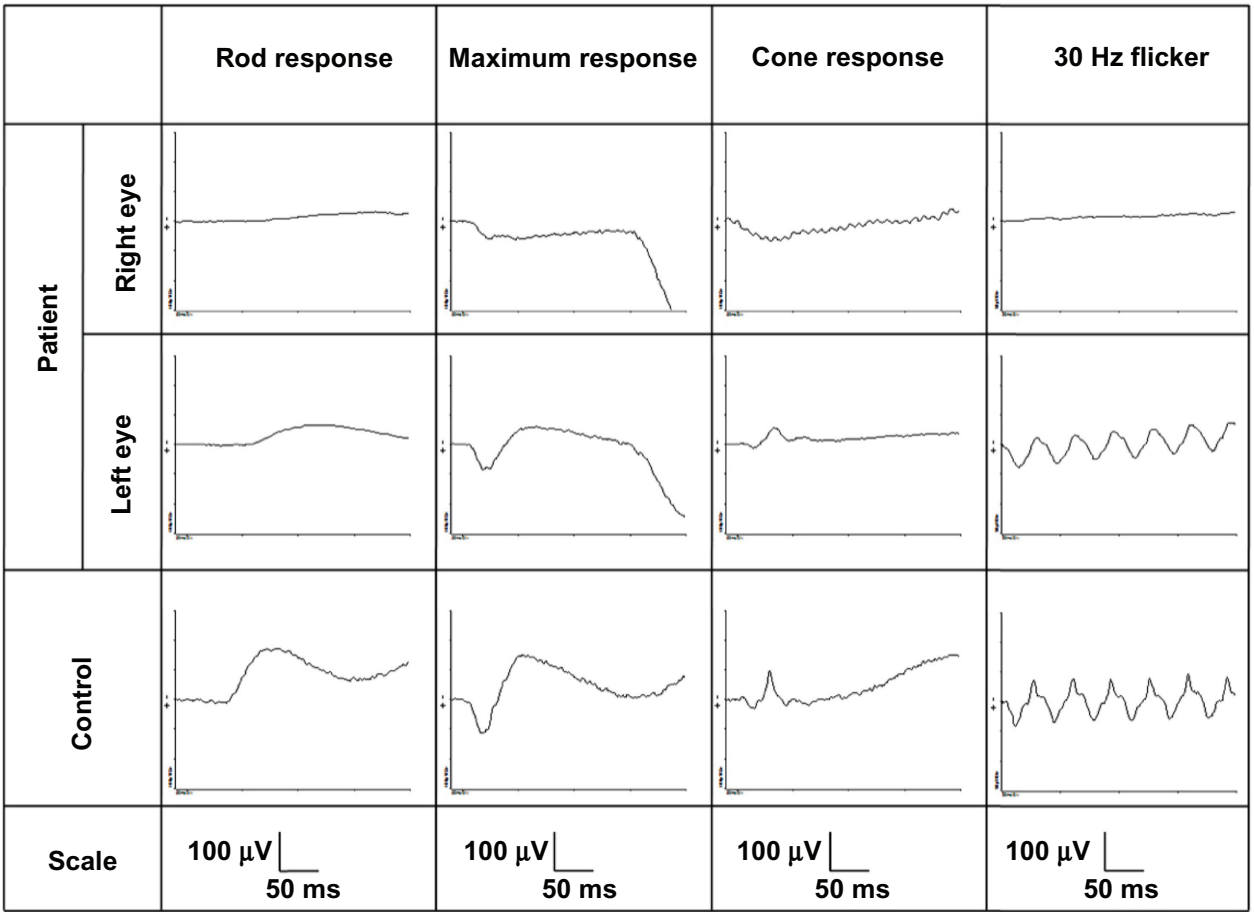

Figure 4 Full-field electroretinogram (ERG) showing electronegative ERG in the right eye and minimal generalized dysfunction in the left eye. Scotopic rod specific ERG has b-wave amplitude of 27 µV and in the right and 71 µV in the left. Maximal ERG a- and b-wave amplitudes were 59 and 9 µV in the right eye and 83 and 130 µV in the left eye. Maximal ERG a-wave implicit times were 22 milliseconds in the right and 19 milliseconds in the left. Transient photopic ERG b-wave amplitudes were extinguished in the right and 68 µV in the left. Photopic 30 Hz flicker ERG were extinguished in the right but had implicit times and amplitudes of 50 µV and 30 milliseconds in the left.

unknown origin with minimal abnormalities in the clinical and imaging studies. The ERG revealed a unilateral negative waveform. Based on his history, imaging examinations, and on the ERG result, CRAO and autoimmune retinopathies were considered as differential diagnosis. Although autoimmune retinopathies, including cancer-associated retinopathy and melanoma-associated retinopathy, can cause electronegative ERG, they are bilateral. Indeed, autoimmune retinopathy patients tend to have antiretinal antibody activity often with different antibodies found on immunoblots.[20–22] The findings of generalized vascular narrowing, even though minimal, lack any sign of acute ischemia and the unilateral electronegative maximal ERG in the right eye were consistent with inner retinal dysfunction due to an old central arteriolar occlusion in the right eye. A reduction of the b-wave with a relative preservation of the a-wave of the scotopic maximal ERG is a characteristic of eyes with CRAO.[2,5] In addition, the reduced inner retina thickness more prominent in the right eye than in the left eye demonstrable by SD-OCT measurements strengthens the diagnosis of CRAO since CRAO is classically associated with a reduced thickness in the inner retinal layers.[23,24]

Although the classic retinal findings of CRAO have been described, a high degree of suspicion is needed to diagnose CRAO in some cases because of transitory abnormalities in the fundus appearance, FA findings for CRAO, the insensitivity of carotid Doppler imaging and echocardiography for associated lesions, and the subtlety of residual findings after CRAO. In such cases, mainly represented by the transient type, the diagnosis of CRAO is intricate and an electronegative ERG can support the diagnosis of artery occlusion.[1,2] CRAO is reported to be the most common cause of a unilateral negative ERG.[25]

We report an atypical case of CRAO without its characteristic angiographic findings, except mild retinal artery narrowing detected on his fundus and imaging exams, in which ERG was essential for diagnosis. Our case, therefore, illustrates that ERG testing is helpful for the work-up of individuals with suspected retinal vascular disorders.

Disclosure

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