



Multimodal Imaging Characteristics and Functional Test Findings in a Case of Acute Macular Neuroretinopathy Accompanied by Behçet Disease

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ABSTRACT

Purpose: To report a case of acute macular neuroretinopathy (AMN) in Behçet Disease.

Case: A 23-year-old male presented with a complaint for central scotoma in his right eye. He had been diagnosed with Behçet Disease 3 years ago. Best-corrected visual acuity (BCVA) was 20/20. Anterior chamber and fundus examinations were unremarkable. Optical coherence tomography revealed a paracentral area of outer nuclear layer thinning. Infrared reflectance showed a well-defined, circular, hyporeflective area. Optical coherence tomography angiography revealed an area of capillary dropout in deep retinal capillary plexus corresponding to that hyporeflective lesion. Microperimetry test showed decreased macular sensitivity on the lesion area and the loss of the macular integrity. In multifocal electroretinogram, diminished amplitudes of the central cone responses were detected nasal to fixation.

Conclusion: Behçet disease is a cause of occlusive retinal vasculitis. Accompanied retinal microvascular disease may be a possible risk factor of AMN suggesting ischemic etiopathogenesis for AMN.

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Acute macular neuroretinopathy; Behçet disease; optical coherence tomography angiography; microperimetry; multifocal electroretinogram

Acute macular neuroretinopathy (AMN), firstly defined by Bos and Deutman¹, is a rare clinical entity characterized by reddish oval or wedge-shaped macular lesions causing paracentral scotomas. In 2013, Sarraf et al.² defined two variants of AMN according to the localization of the lesion, above and below the outer plexiform layer (OPL), depending on spectral domain optical coherence tomography. Recently, it was clarified that these two variants, paracentral acute middle maculopathy (PAMM) and typical acute macular neuroretinopathy, should be regarded as two distinct entities.^{3,4}

Acute macular neuroretinopathy is relatively rare and has a different demographic and risk profile from that of PAMM.³ Typical AMN lesion is characterized by a hyperreflective band appearance at the border of the outer plexiform layer and outer nuclear layer (ONL). This hyperreflective band typically progresses to thinning of the ONL over time.

We herein describe multimodal retinal imaging and functional test findings of a case with AMN secondary to accompanied Behçet Disease.

Case presentation

A 23-year-old male presented with a complaint for dark spot obscuring his central vision in his right eye for the last few months. He had been diagnosed with Behçet disease 3 years ago, but he did not describe any uveitis attacks. He was on azathioprine treatment (150 mg/day). However, he did not take his medication regularly. Best-corrected visual acuity (BCVA) was 20/20 bilaterally. Anterior chamber examination was unremarkable. There was neither anterior chamber activity nor vitreous haze. Fundus

examination of both eyes was normal. Spectral domain optical coherence tomography (Spectralis®, Heidelberg Engineering Inc., Heidelberg, Germany) revealed an area of outer nuclear layer thinning located nasal paracentrally (Figure 1). Infrared (IR) reflectance showed a well-defined, circular, hyporeflective area nasal to the fovea (Figure 2). Optical coherence tomography angiography (Avanti RTVue XR® with AngioVue® software; Optovue Inc., Fremont, USA) revealed an area of capillary dropout in deep retinal capillary plexus corresponding to the hyporeflective lesion on IR reflectance indicating an ischemic situation (Figure 3). On the deep retinal capillary slab, the vessel density in the parafoveal nasal area was 49.1% which was 61.4% at the unaffected eye. Macular integrity assessment (MAIA) microperimetry (Centervue, Padova, Italy) test showed decreased macular sensitivity on the lesion area and the loss of the macular integrity (Figure 4). Multifocal electroretinogram (ERG) (MonPackOne, Metrovision, Perenchies, France) showed diminished amplitudes of the central cone responses nasal to fixation (Figure 5). Fluorescein angiography (Heidelberg Retina Angiograph 2®; Heidelberg Engineering, Heidelberg, Germany) was unremarkable for the lesion but showed peripheral vascular leakage on both eyes (Figure 6). The patient did not describe any change in the character of the paracentral scotoma since its initial onset. At time of the first presentation, the lesion was in the chronic phase of the disease in which thinning of the outer nuclear layer had already occurred. Therefore, scotoma was thought to be permanent. The patient was advised to continue azathioprine therapy on a regular basis for Behçet disease.

At the last visit, three months later than the first presentation, the patient's complain for paracentral scotoma persisted. His best-

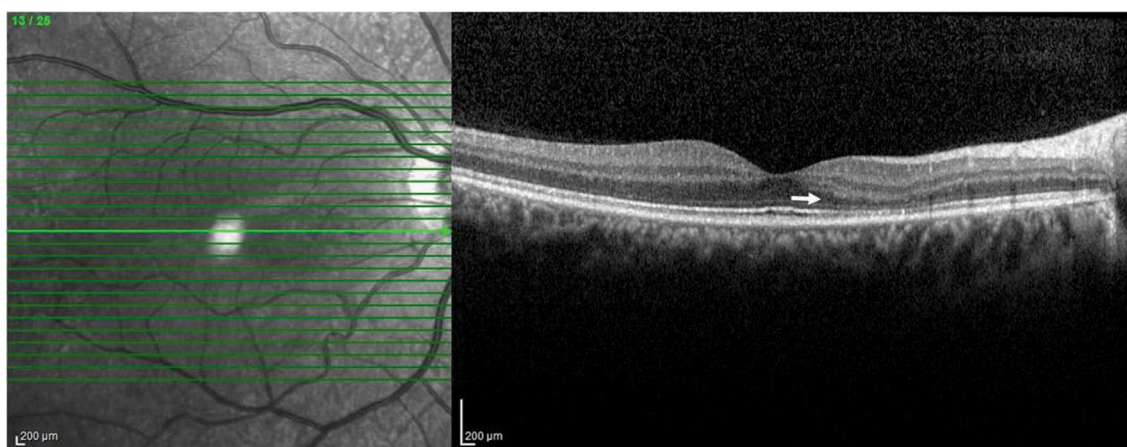


Figure 1. Spectral domain optical coherence tomography of the right eye revealed an area of outer nuclear layer thinning located nasal paracentrally (arrow).



Figure 2. Infrared reflectance imaging of the right eye showed a well-defined, circular, hyporeflective area nasal to the fovea (arrow).

corrected visual acuity (BCVA) was 20/20 bilaterally. Spectral domain optical coherence tomography revealed permanent thinning of outer nuclear layer and partial ellipsoid zone disruption, while optical coherence tomography angiography showed permanent capillary dropout in the nasal parafoveal area of deep retinal capillary plexus with a vessel density of 51.0% (Figure 7). Macular integrity assessment microperimetry test demonstrated permanent paracentral scotoma corresponding to hyporeflective lesion on IR reflectance with relatively improved scores in macular sensitivity and macular integrity (Figure 8).

Discussion

In this case report, the diagnosis of AMN was confirmed by multimodal imaging techniques including spectral domain optical coherence tomography, infrared reflectance, and optical coherence tomography angiography. Additionally, these multimodal retinal imaging methods were used in combination with different

functional tests including microperimetry and multifocal ERG. As a result of all these tests, we have observed how well the imaging methods and functional tests overlap with each other. To the best of our knowledge, there is only one case series reporting the association of Behçet disease and AMN in literature.⁵

Behçet disease, firstly described by Hulusi Behçet in 1937, is a chronic multisystemic disorder characterized by relapsing inflammation of unknown etiology. The underlying pathogenesis is an occlusive an occlusive vasculitis that affects both the arteries and the veins in multi-organ systems.⁶ Behçet disease is the leading diagnosis of noninfectious uveitis in Turkey.⁷ The most common ocular involvement of this disease is panuveitis and retinal vasculitis and vitritis are the most common findings and are eventually observed in every eye with panuveitis or posterior uveitis.⁸

Many risk factors including preceding flulike illness, use of oral contraceptives, ocular trauma, caffeine consumption, epinephrine injection, pseudoephedrine, hypovolemia, and pregnancy-induced hypertension were defined for AMN.^{9,10} However, the exact pathogenesis of AMN remains unknown. It has been speculated that AMN may have resulted from a simultaneous hypoperfusion more proximally at the level of the ophthalmic artery that leads to reduced perfusion of both the deep capillary plexus and choriocapillaris.³ Therefore, the disease may be referred to as a “capillaropathy” which has not a specific treatment. Regarding this underlying mechanism, it is not surprising that the occurrence of AMN in a case with Behçet disease in which the most common retinal finding is occlusive retinal vasculitis. Presumed vascular pathogenesis in AMN may also be supported by the OCTA changes in the level of deep capillary plexus.

In conclusion, our case brings attention to accompanied retinal vasculitis as a possible risk factor of AMN suggesting ischemic etiopathogenesis for the development of AMN lesions. At the same time, it emphasizes the importance of using multimodal imaging methods and functional tests in combination for more accurate evaluation of the effect of the disease on visual function.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the article.

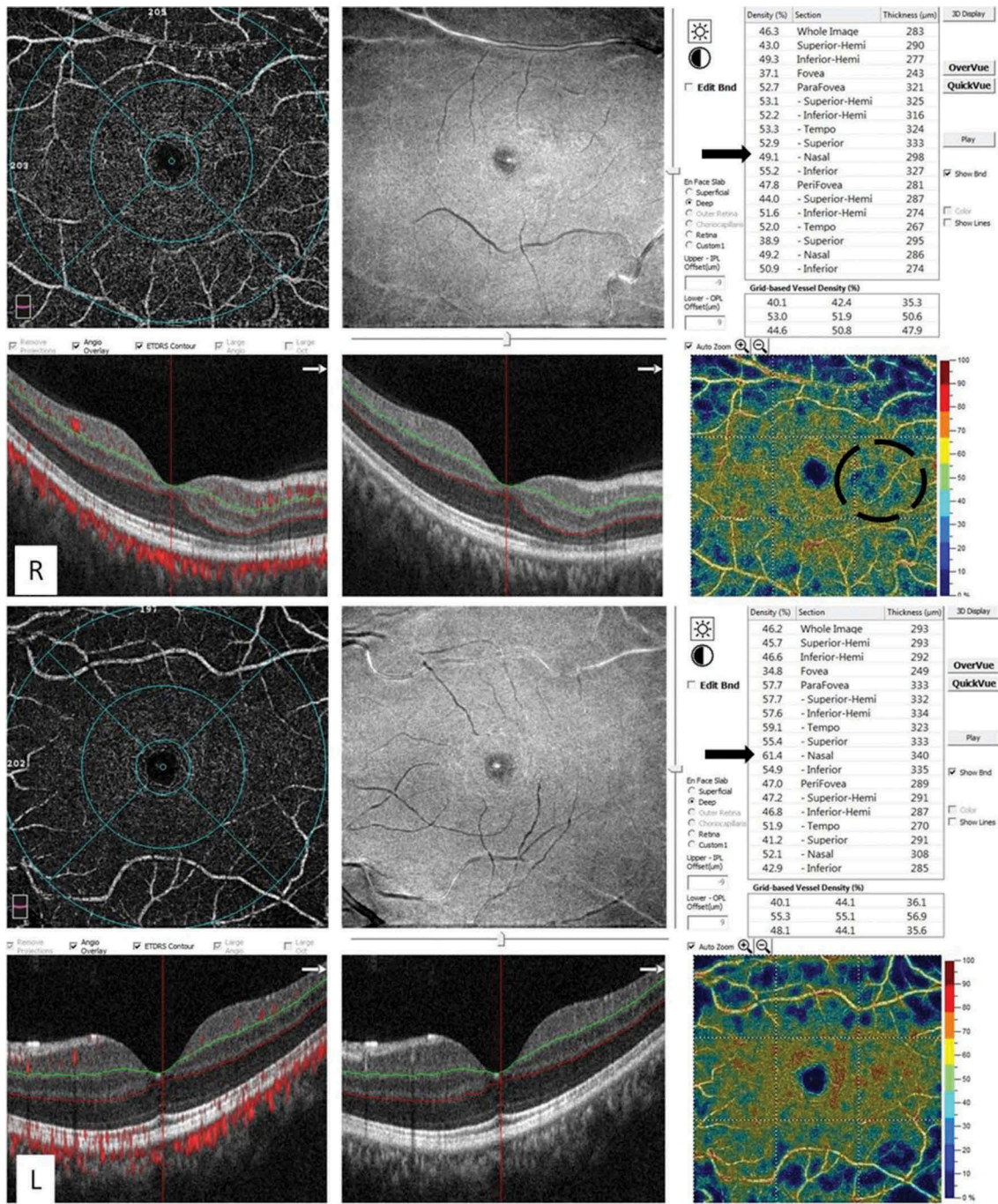


Figure 3. Optical coherence tomography angiography of the right eye revealed an area of capillary dropout in deep retinal capillary plexus (dashed circle) corresponding to the hyporeflective lesion on IR reflectance. On the deep retinal capillary slab, the vessel density in the parafoveal nasal area was 49.1% which was 61.4% at the unaffected eye (arrows). Right (R), left (L).

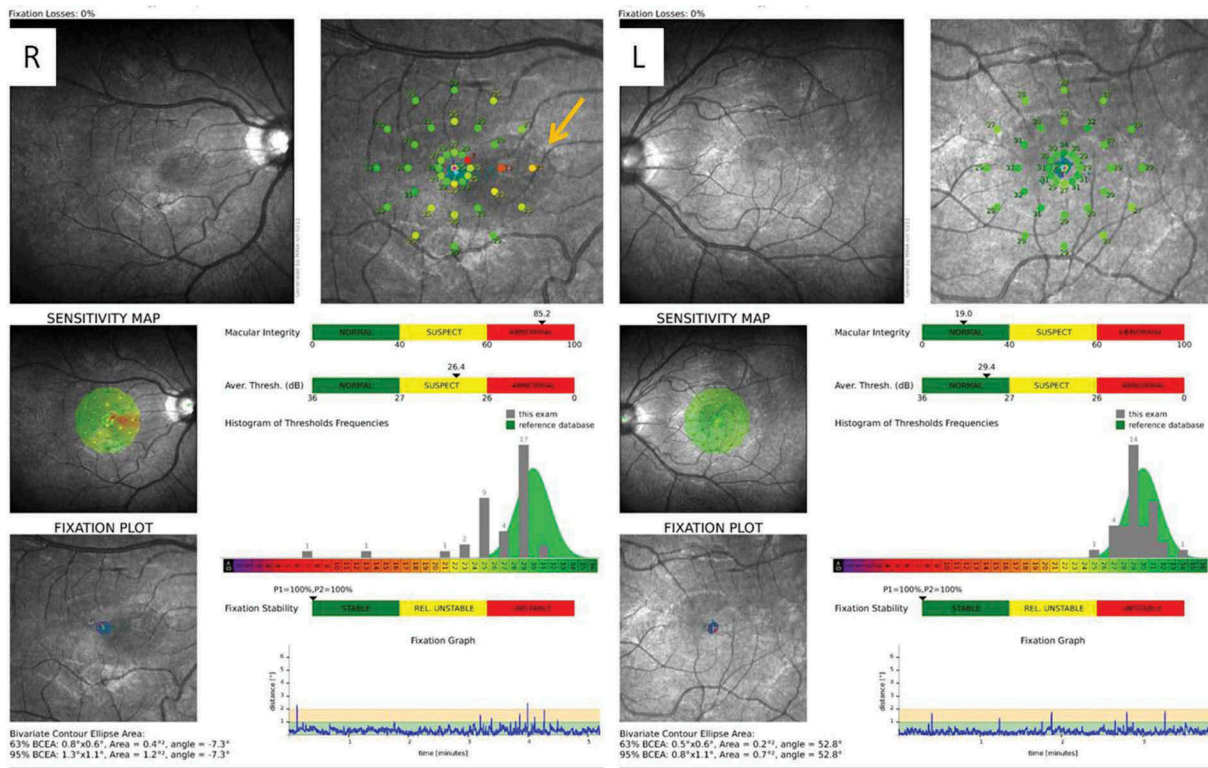


Figure 4. Macular integrity assessment microperimetry test of the right eye showed decreased macular sensitivity on the lesion area and the loss of the macular integrity. Right (R), left (L).

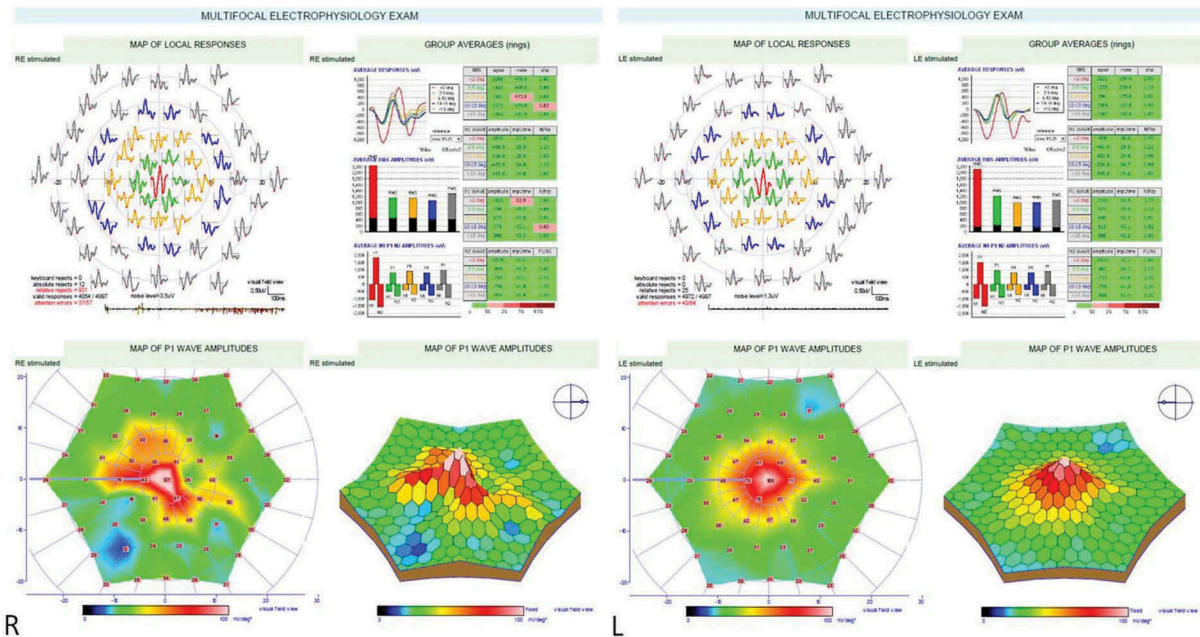


Figure 5. Multifocal electroretinogram of the right eye showed diminished amplitudes of the central cone responses nasal to the fixation. Right (R), left (L).



Figure 6. Fluorescein angiography showed peripheral vascular leakage and limited ischemic areas on both eyes. Right (R), left (L).

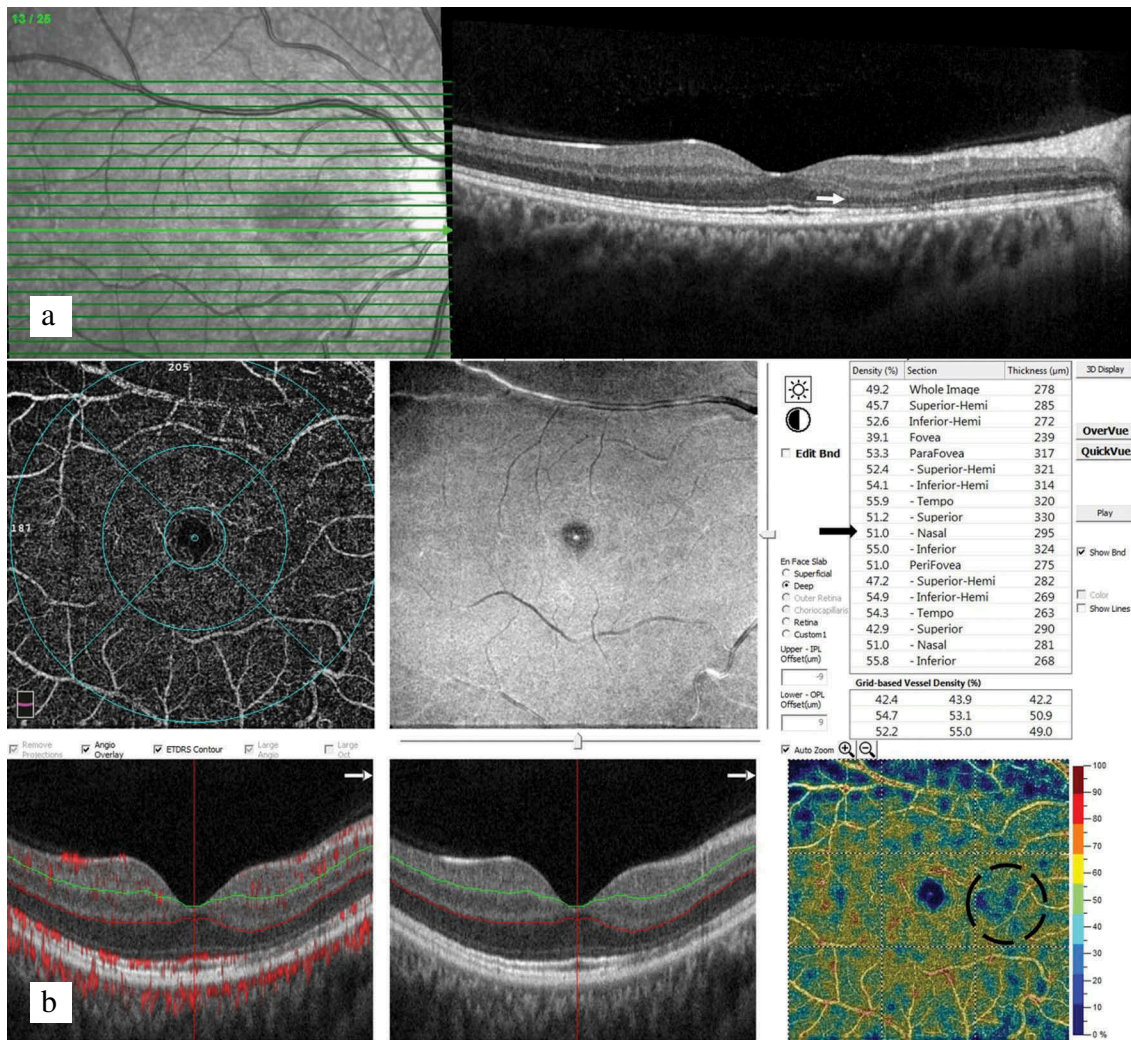


Figure 7. Multimodal imaging findings of the right eye at the last visit: (a) Spectral domain optical coherence tomography demonstrated permanent thinning of outer nuclear layer (arrow) and partial ellipsoid zone disruption. (b) Optical coherence tomography angiography showed permanent capillary dropout (dashed circle) in the nasal parafoveal area of deep retinal capillary plexus with a vessel density of 51.0% (arrow).

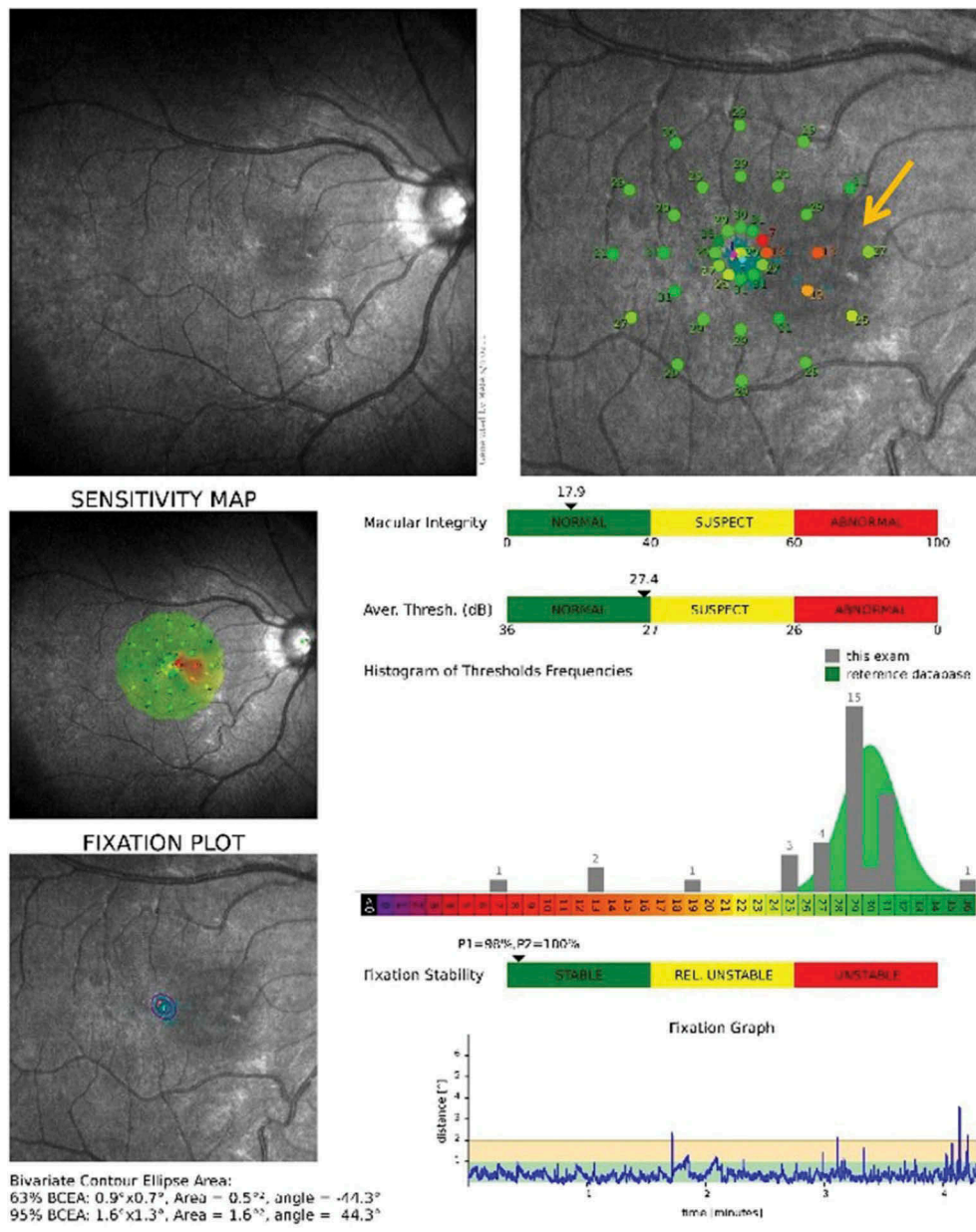


Figure 8. Macular integrity assessment microperimetry test of the right eye at the last visit. It showed permanent paracentral scotoma corresponding to the hyporeflective lesion on IR reflectance with relatively improved scores in macular sensitivity and macular integrity.

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