Multimodal Imaging of A Patient With Bilateral Paracentral Acute Middle Maculopathy

Kubra OZDEMIR YALCINSOY[1], Sibel DOGUIZI[1], Merve INANC TEKIN[2], Mehmet Yasin TEKE[3]

ABSTRACT

A 45-year-old woman presented with a 10-day history of paracentral scotoma in the right eye and 30-day history of paracentral scotoma in the left eye. Her best-corrected visual acuity (BCVA) was 20/200 in the right eye and 20/25 in the left eye. Both anterior segments examination results and intraocular pressure levels were normal. Fundus examination revealed superior parafoveal whitening and a few intraretinal exudations in the right eye, and cotton wool spots, intraretinal hemorrhage and exudates on the nasal parafoveal area in the left eye. Spectral domain optical coherence tomography (SD OCT) findings revealed that there were parafoveal hyperreflective plaque at the middle retinal layers in the right eye, thinning of the nasal parafoveal inner retinal layers in the left eye. OCT angiography (OCTA) revealed bilateral mild attenuation of the superficial capillary plexus (SCP), patchy areas of attenuation and capillary dropout of the deep capillary plexus (DCP). After six weeks, SD OCT demonstrated subsequent thinning with attenuation of the inner and middle retinal layers in the right eye, similar to the left eye. In this report, we documented bilateral paracentral acute middle maculopathy at different stages in patient. Multimodal imaging can be used in the evaluation of ischemic changes in retinal vascular diseases and in the diagnosis of PAMM. This disease may occur in association with systemic disorders or related retinal vascular diseases, but it should be considered that it may also occur idiopathically.

Key Words: Optical coherence tomography, Optical coherence tomography angiography, Paracentral Acute Middle Maculopathy.

INTRODUCTION

Paracentral acute middle maculopathy (PAMM) is a recently defined manifestation presenting with hyperreflective band-like lesions within the inner and middle retinal layers on spectral domain optical coherence tomography (SD-OCT). It is thought that often limited ischemia or infarction of the parafoveal deep retinal capillary plexus.[1] At first time in 2013, it was described by Sarraf et al. as the variant of acute macular neuroretinopathy.[2] PAMM has been reported with various retinal vascular diseases and systemic disorders using multimodal imaging, may also arise as idiopathic.[3-5]

In this case report, we aimed to present bilateral paracentral acute middle maculopathy in a woman with typical multimodal imaging and clinical findings.

CASE

A 45-year-old healthy woman presented with a 10-day history of paracentral scotoma in the right eye and 30-day history of paracentral scotoma in the left eye. There was no systemic or ocular disease, drug use, eye trauma, recent invasive medical procedure, any recent flu-like illnesses in the medical history of the patient. She was also a non-smoker and was not a coffee drinker. Her blood pressure was 110/70 mm Hg at the time of examination. Her best-corrected visual acuity (BCVA) was 20/200 and 20/25, in the right and left eyes, respectively. Color vision, motility and pupillary responses were normal. Bilateral anterior segment examination results and intraocular pressure levels were also normal. Dilated fundus examination revealed superior parafoveal whitening and several intraretinal exudations in the right eye, consistent with branch retinal artery occlusion, and cotton wool spots, intraretinal hemorrhage and exudates on the nasal parafoveal area in the left eye (Figure 1A-1B). Fundus fluorescein angiography (FA) in early-phase revealed delayed perfusion of superior parafoveal branch retinal artery; FA in late-phase showed only mild late arteriolar

1- MD, University of Health Sciences Ulucanlar Eye Training and Research Hospital, Department of Ophthalmology, Ankara,Turkey

2- MD, Ercis State Hospital, Van, Turkey

3- MD, Associate Professor of Ophthalmology, University of Health Sciences Ulucanlar Eye Training and Research Hospital, Ankara, Turkey

Received: 31.05.2019
Accepted: 23.08.2019
Ret-Vit 2020; 29: 270-273
DOİ:10.37845/ret.vit.2020.29.49

Correspondence Adress:
Kubra OZDEMIR YALCINSOY
University of Health Sciences, Ulucanlar Eye Education and Research Hospital, Kale Mahallesi, Ulucanlar Caddesi, No:59, 06250, Altındağ /Ankara /Turkey
Phone: +90 312 312 6261
E-mail: kubraozdemir250@gmail.com

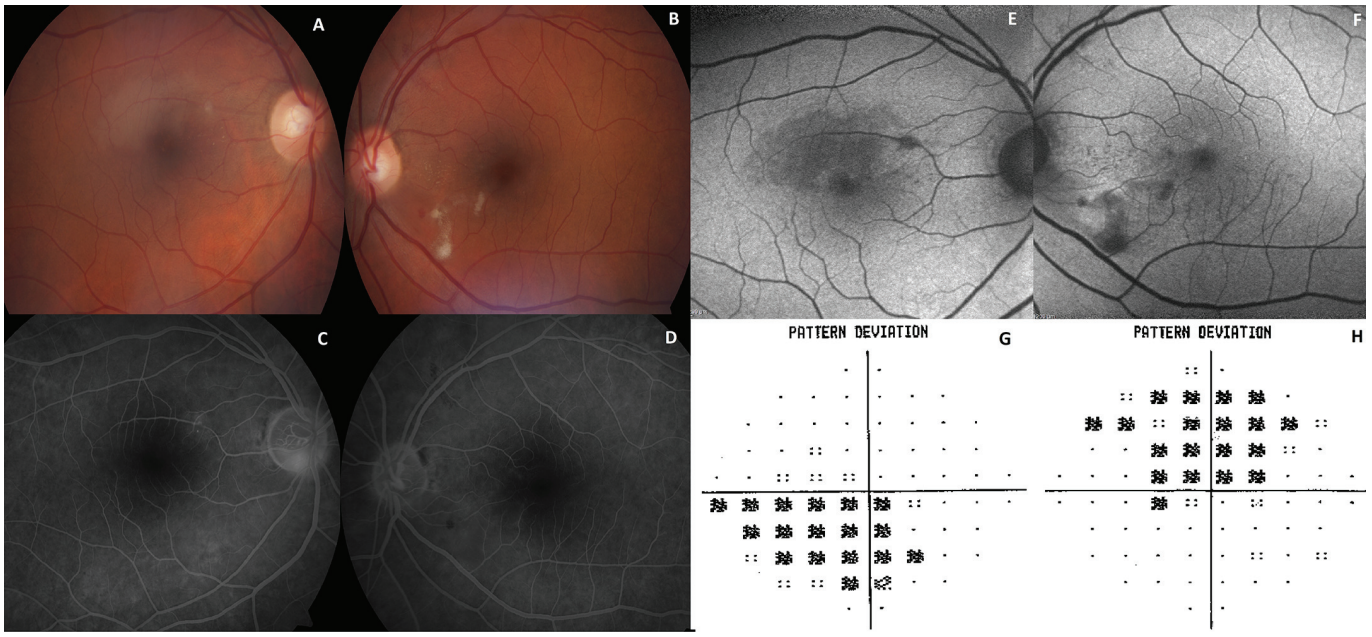

Figure 1. *(A-B) Color fundus photography showing superior parafoveal whitening and a few intraretinal exudations in the right eye, and cotton wool spots, intraretinal hemorrhage and exudates on the nasal parafoveal area in the left eye. (C-D) Fundus fluorescein angiography(FA) in late-phase showing only mild late arteriolar leakage in a small area corresponding to the retinal whitening observed clinically in the right eye. FA findings in the left eye was unremarkable. (E-F) Blue light fundus autofluorescence (BLFAF) demonstrated bilateral dark-gray lesions in the distribution of the retinal whitening observed clinically in both eyes. (G-H) Initial Humphrey visual field 10-2 pattern deviation for the right and left eyes, showing central/paracentral scotomas.*

leakage in a small area corresponding to the retinal whitening observed clinically in the right eye (Figure 1C). FA findings in the left eye was unremarkable (Figure 1D). Blue light fundus autofluorescence (BLFAF) demonstrated bilateral dark-gray lesions in the distribution of the retinal whitening observed clinically in both eyes (Figure 1E-1F). The Humphrey Field Analyzer 10-2 degree visual field examination showed bilateral paracentral scotoma (Fig 1G-1H). SD OCT findings revealed parafoveal hyperreflective plaque was present at the middle layers of the retina of the right eye at the level of the INL corresponding to the retinal whitening area observed clinically (Figure 2A-2B), thinning and mild distribution of inner retinal layers in the left eye (Fig 2D-2E). OCT angiography (OCTA) revealed mild attenuation of the superficial capillary plexus (SCP), patchy areas of attenuation and capillary dropout of the deep capillary plexus (DCP) (Fig 3A-D).

A complete investigations was started to investigate this bilateral macular ischemic vasculopathy. There were no systemic risk factors associated with PAMM such as hypertension, hypotension, migraine and pregnancy. An extensive laboratory work up was performed for autoimmune connective tissue diseases, noninfectious and infectious inflammatory diseases, hemoglobinopathies and thrombophilia screen. These laboratory tests were within normal limits. Serologic investigations for Treponema

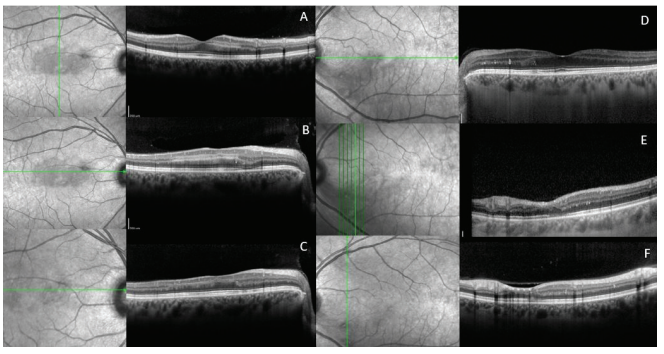

Figure 2. *(A-B) Spectral domain optical coherence tomography reveals a hyperreflective plaque-like band at the junction of the outer plexiform layer and inner nuclear layer (INL) in the right eye corresponding to the retinal whitening area observed clinically. (C) After six weeks, SD OCT findings showed subsequent thinning of the same area in the right eye. (D-E) SD OCT revealed thinning and mild distribution of inner and middle retinal layers on the nasal parafoveal area in the left eye. (F) After six weeks, SD OCT showing progressed atrophic findings on the nasal parafoveal area in the left eye.*

pallidum, Borrelia burgdorferi, Bartonella henselae, Rickettsia, Mycoplasma pneumoniae and Chlamydia trachomatis were also all negative. Brain magnetic resonance imaging (MRI) and MRI angiography were

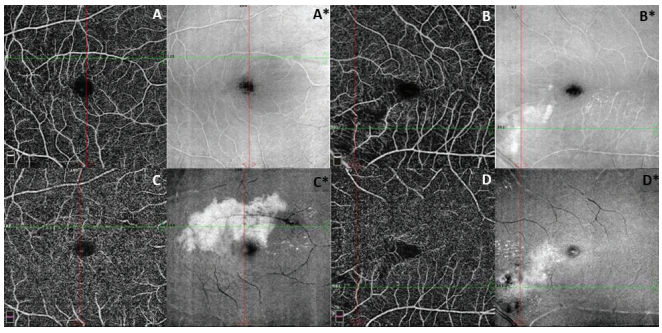

Figure 3. *OCT angiography (OCTA) revealed mild attenuation of the superficial capillary plexus (SCP) (A), patchy areas of attenuation and capillary dropout of the deep capillary plexus (DCP) (C) in the right eye. En face OCT at the level of the superficial retina was unremarkable (A*), at the level of middle retina demonstrated band-like hyperreflectivity (C*) in the right eye. OCTA revealed patchy areas of attenuation and capillary dropout of the SCP (B) and DCP (D) in the left eye. En face OCT demonstrated hyperreflectivite areas at the level of the superficial (B*) and middle retina (D*) in the left eye.*

performed and normal results were obtained. Additionally, the audiometric test of the patient was normal.

The patient was diagnosed with the different phases of PAMM in both eyes. After six weeks, the BCVA increased from 20/200 to 20/40 in the right eye and remained at 20/25 level in the left eye. SD OCT findings showed that the plaque on the right eye has become atrophic similar to the left eye (Figure 2C-2F).

DISCUSSION

In 2013, PAMM was described by Sarraf et al. as the new variant of acute macular neuroretinopathy (AMN). PAMM is diagnosed by detecting hyperreflective band-like lesions in middle layers of retina in SD OCT imaging. Two types of AMN lesions have been described with SD OCT. Type 1 refers to hyperreflective bands in the OPL/INL region with subsequent INL thinning. Type 2 is hyperreflective bands in the OPL/ONL region with subsequent ONL thinning.[2] We considered our case as type 1 disease. In type 1 disease, deep capillary ischemia can be diagnosed by a characteristic hyper-reflective lesion at INL in SD OCT imaging. Characteristic hyperreflective lesion of PAMM at INL level is seen in the acute phase and atrophied INL in chronic phase.[3] FA is currently the gold standard to image retinal ischemia. This imaging modality allows visualization of the SCP in excellent detail, however, the intermediate and DCP can not be evaluated. Both SCP and DCP can be evaluated with OCTA.[6,7]

Multimodal imaging; including color photographs, near-infrared reflectance, FA, SD-OCT, En face OCT and OCTA are used for PAMM diagnosis.[4,6,7] OCTA is latest developed imaging technique and is able to visualize and delineate the extent of ischemia in the DCP in patients with PAMM.[6,7,8] OCTA showed normal or mild attenuation of the SCP and diffuse patchy areas of attenuation and pruning capillary dropout of the DCP.[6,7] In our patient we found similar OCTA findings.

PAMM has been reported to be associated with a wide range of ischemic retinal vascular disorders such as nonproliferative diabetic retinopathy, central retinal vein occlusion, retinal arter occlusion, systemic hypercoagulate status.[2] Ischemic retinal vasculopathies can also be seen in cases of non-infectious inflammatory disorders such as autoimmune connective tissue diseases, systemic lupus erythematosus, Wegener's granulomatosis, polyarteritis nodosa and antiphospholipid syndrome, Susac Syndrome. Extensive laboratory studies for infectious and noninfectious inflammatory diseases in our patient were all negative. Haider et al[10] reported a case of PAMM with susac syndrome, occur with clinical symptoms in the form of encephelopathy, visual and hearing disorders triad, usually seen in young women. Susac Syndrome is a rare vasculopathy and it is diagnosed by the detection of characteristic MRI findings, the audiometric test neurosensorial hearing loss and retinal branch artery occlusion.[11,12] In our patient due to occasional headache complaints, MRI, MR angiography, the audiometric test and neurology consultation were performed and all results were within normal limits. In addition,a case of bilateral PAMM in a pregnant female patient with systemic Behçet's disease (BD) has been reported by Papadaki at al.[13] The ocular and systemic signs of our patient were not typical of BD.[14]

Chen at al[5] examined nine patients with PAMM diagnosis associated with sickle cell retinopathy, carotid artery stenosis, occlusive retinitis, post traumatic Purshcher retinopathy. Also Paula et al[9] reported PAMM in a hypotensive young pregnant woman. Hypotension and at the same time hypercoagulability in the pregnancy contributed to caused to genesis of ischemia. In our patient all of these associated conditions did not detected.

Yanyan et al[3] reported idiopathic PAMM patient in a Chinese young man whom all systemic risk factors were investigated but all were negative. Yanyan et al. suggested that possible causes of idiopathic PAMM could be caused by small blood vessel vasoconstriction, vasoconstrictor substance and sympathetic effects.[3,7]

We documented bilateral paracentral acute middle maculopathy in a woman with typical optical imaging

and clinical findings. Despite extensive laboratory studies, radiological imaging and tests, we have not detected any specific systemic disease or associations. In conclusion, multimodal imaging can be used in the evaluation of ischemic changes in retinal vascular diseases and in the diagnosis of PAMM.

REFERENCES

1. Tsui I, Sarraf D. Paracentral acute middle maculopathy and acute macular neuroretinopathy. Ophthalmic Surg. Lasers Imaging Retina 2013;44:33-5.

2. Sarraf D, Rahimy E, Fawzi AA et al. Paracentral acute middle maculopathy:a new variant of acute macular neuroretinopathy associated with retinal capillary ischemia. JAMA Ophthalmol 2013;131:1275–1287.

3. Chen Y, Hu Y. The optical imaging of idiopathic paracentral acute middle maculopathy in a Chinese young man and review of the literature. Photodiagnosis Photodyn Ther 2017;19:383-387.

4. Ghasemi Falavarjani K, Phasukkijwatana N, Freund KB et al. En Face Optical Coherence Tomography Analysis to Assess the Spectrum of Perivenular Ischemia and Paracentral Acute Middle Maculopathy in Retinal Vein Occlusion. Am J Ophthalmol 2017;177:131-138.

5. Chen X, Rahimy E, Sergott RC et al. Spectrum of Retinal Vascular Diseases Associated With Paracentral Acute Middle Maculopathy. Am J Ophthalmol 2015;160(1):26-34.

6. Yu S, Pang CE, Gong Y et al. The spectrum of superficial and deep capillary ischemia in retinal artery occlusion. Am J Ophthalmol 2015;159:53–63.

7. Casalino G, Williams M, McAvoy C et al. Optical coherence tomography angiography in paracentral acute middle maculopathy secondary to central retinal vein occlusion. Eye (Lond) 2016;30(6):888-93.

8. Turgut B. Retinal Hastalıklarda Optik Koherens Tomografi Anjiografi. Güncel Retina 2017;1(1):69-74.

9. Paula EP, Adiel GS, Justis PE, Optical Coherence Tomography Angiography of Acute Macular Neuroretinopathy/ Paracentral Acute Middle Maculopathy. JAMA Ophthalmol 2015;133(12):1478–1480.

10. Haider AS, Viswanathan D, Williams D et al. Paracentral Acute Middle Maculopathy In Susac Syndrome. Retin Cases Brief Rep 2017 [Epub ahead of print]

11. Olszanecka-Glinianowicz M, Hrycek A. Susac syndrome as an example of a rare vasculopathy. Pol Arch Med Wewn 2009;119(1-2):80-2.

12. Azevedo AGB, Lima LH, Müller L et al. Anatomical and functional correlation in Susac syndrome:multimodal imaging assessment. Int J Retina Vitreous 2017;Oct 16;3:39.

13. Papadaki M, Lefebvre P, Janssens S et al. Bilateral retinal ischemic vasculopathy in a pregnant patient. Retin Cases Brief Rep 2015 Spring;9(2):185-9.

14. Tugal-Tutkun I, Onal S, Altan-Yaycioglu R et al. Uveitis in Behçet disease: an analysis of 880 patients. Am J Ophthalmol 2004;138:373–380.