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CASE REPORT

Optical coherence tomography angiography findings in patients with systemic lupus erythematosus

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Abstract

Systemic lupus erythematosus (SLE) is a chronic autoimmune disorder that can affect eye, such as retina vascular occlusions are frequent with this disorder. We aimed to describe the optical coherence tomography angiography (OCTA) findings of SLE patients. We evaluated three SLE patients which one of them had retinal vein occlusion and active vasculitis in different eyes. Superficial capillary plexus, deep capillary plexus, and optic nerve head were evaluated using OCTA RTVue XR AVANTI. Two patients, with the lack of retinal pathologies, had no changes that were seen on OCT-A. Hypointense dark-grayish areas of retinal capillary non-perfusion/hypoperfusion, capillary telengiectasies, capillary rarefaction, and diffuse capillary network disorganization were seen on third patients' OCT-A images. OCT-A shows better visualization of perifoveal microvascular structures than fundus fluorescein angiography in eyes with active and chronic SLE.

Keywords: Capillary plexus; optical coherence tomography angiography; retinal vein occlusion; systemic lupus erithematosus; vasculitis.

Systemic lupus erythematosus (SLE) is a systemic, autoimmune, and connective tissue disorder that can harmfully affect multiple organ systems including the eye. Up to one-third of SLE patients experience some kind of ocular manifestation.[1] Different ocular structures can be affected by SLE; including the cornea, conjunctiva, episclera, sclera, retina, uveal tract, optic nerve, vasculature, and orbit.[2,3] The incidence of retinopathy in patients with SLE ranges from 3% to 29% and retinal vasooclusions which are sight threatening manifestation of SLE retinopathy, reported to cause severe visual loss in 55% of patients.[4,5]

Optical coherence tomography angiography (OCT-A) is a newly developed, dye-less, and imaging method that provides three-dimensional mapping of the retinal and choroidal microvasculature.[6] OCT-A employs amplitude or phase decorrelation technology with high-frequency and dense volumetric scanning to detect erythrocyte movement, allowing direct visualization of blood vessels in vivo.[7,8]

It was showed in a previous study that this novel method can be useful in vasculitic patients.[9] The aim of this study was to evaluate the SLE patients with having vasculitis or not with using OCT-A.

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Case Report

This study was conducted in Başkent University School of Medicine Zubeyde Hanım Research Hospital. SLE patients were evaluated using OCT-A, retrospectively. This study adhered to the tenets of the Declaration of Helsinki informed consent was obtained from all participants.

SLE patients were recruited from rheumatology clinic of Başkent University School of Medicine Zubeyde Hanım Research Hospital. All patients satisfied the updated revised criteria for the classification of SLE.[10]

We did not include the patients whose had significant media opacities, and trauma, other retinal diseases such as retinal vascular occlusive disease unrelated to BD, diabetic retinopathy, hypertensive retinopathy, central serous chorioretinopathy, and macular degeneration, and optic nerve diseases such as glaucoma or optic neuropathy.

Every patient underwent a complete ophthalmic examination that included the following: Best-corrected visual acuity measurement with the Early Treatment Diabetic Retinopathy Study charts, anterior segment examination, intraocular pressure (IOP) measurement by applanation tonometry, dilated-fundoscopy, FA (TRC-50IX, Retinal Camera, Topcon), OCT, and OCT-A.

OCT-A images were acquired with OCT-A (RTVue XR AVANTI; Optovue Inc, Fremont, CA). The AngioVue OCT-A system operated at 70,000 Ascans per second, using a light source centered on 840 nm and a bandwidth of 50 nm. The OCT-A volumes contained 304 304 A-scans with 2 consecutive Bscans that were captured at each fixed position before proceeding to the next sampling location. Split spectrum amplitude-decorrelation angiography was used to extract the OCT-A information.[11] Each OCT-A volume was acquired over 3 s, and 2 orthogonal OCT-A volumes were acquired to perform motion correction to minimize the motion artifacts arising from microsaccades and fixation changes.[12] Angiography information is displayed as the average of the decorrelation values when viewed perpendicularly through the thickness being evaluated. The modifications in reflectivity are directly related to blood flow. The horizontal and vertical scans were combined with a motion correction technology algorithm that compensates for the motion of the patient's eyes to create a 3D-volume of the retinal vascularization. A qualitative analysis and comparisons of the entire imaging data set were conducted in 3 min–5 min.

OCT-A software was used to automatically segment the retinal layers and generates en face projection images after adjusting the level of the segmented layer on the B-scans to best visualize the region of interest. OCT-A was also capable of visualizing PED through semi-automated segmentation of the outer retina and subretinal or sub-RPE space using a volumetric SD-OCT data set. The OCT-A software was able to remove retinal vessel shadowing by subtracting vessels seen above the outer plexiform layer from the outer retina OCT-A image. In addition, OCT-A images were manually segmented into four layers: Superficial and deep plexus (to show retinal vasculature), outer retina, and choriocapillaris. Specific desired locations of retinal segmentation were identified by meticulously scrolling through OCT-A images and their corresponding OCT B-scans. The outer border of each segment was then individually adjusted to align with Bruch membrane.

Three female patients were included in this study. The median age was 40 (range 34–42 years) First patient was SLE patients for 15 years, uses hydroxyquinolone and she had no ocular involvement of SLE. Her vision was 20/20 on both

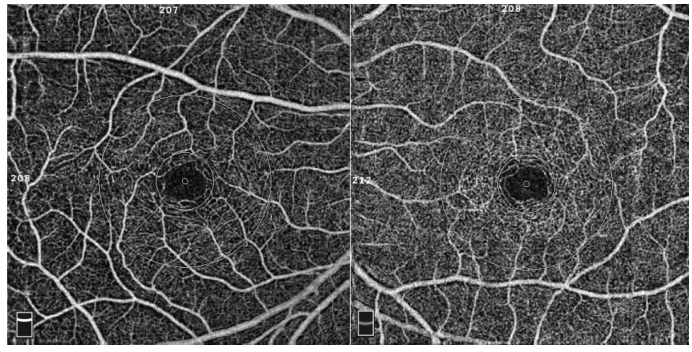

Fig. 1. An example of superficial capillary plexus (left image) and deep capillary plexus (right image) image within normal limits.

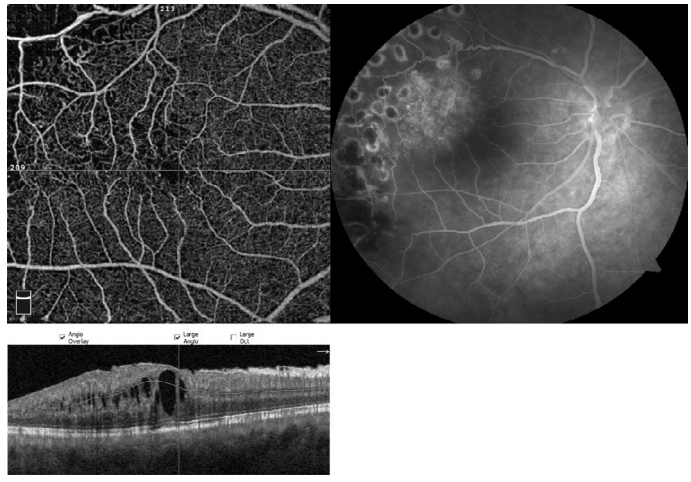

Fig. 2. Superficial plexus image (left image) and fundus fluorescein angiography image (right image) of the third patients right eye. Hypointense dark-grayish areas of retinal capillary nonperfusion/hypoperfusion, capillary telengiectasies, capillary rarefaction, and diffuse capillary network disorganization were seen.

Table 1. Demographic data and ophthalmologic examinations of the patients

	Patient 1	Patient 2	Patient 3
Age (years)	34	40	42
Sex (F/M)	F	F	F
Systemic lupus erythematosus duration	15	New diagnosed	New diagnosed
Vision (R/L)	20/20–20/20	20/20–20/20	20/200–16/20
Intraocular pressure (R/L) (mmHg)	12/13	14/13	12/12
Anterior segment (R/L)	N/N	Dry eye	N/+1 cell
Fundus	N/N	N/N	BRVO/Vasculitis

F: Female; M: Male; R: Right; L: Left; N: Normal; BRVO: Branch Retinal Vein Occlusion.

eyes, her IOP was 12 mmHg at the right eye and 13 mmHg at left, she had no pathological situation either at anterior segment or posterior segment. Second patient was newly diagnosed, she does not use any systemic medication for disease; however, she had dry eye syndrome and uses lubricant eye drops. Her vision was also 20/20 on both eyes. Her right IOP was 14 mmHg and left was 13 mmHg. Her anterior and posterior segment was also totally normal. These two patients' spectral domain-OCT (SD-OCT), fundus fluoresein angiography (FFA), and OCT-A (Fig. 1) evaluations were normal. Third patient was also newly diagnosed, she had a history of branch retinal vein occlusion at right eye 1 year ago from diagnose and she had an active vasculitis at the left eye during the study. This patient has 20/200 vision at the right eye and 16/20 at the left. Her IOPs were 12 mmHg in both eyes. Anterior segment of right eye was normal, on the left eye 1+ cell was detected. She had cystoid macular edema at the right eye on SD-OCT image and laser spots were seen at superior hemisphere on fundus photography. On FFA, there was leakage to cystoid spaces as smokestack pattern on the right eye (Fig. 2). On the left eye, some retinal hemorrhagies were seen on fundus examination and dye leakages were seen around the vessels on FFA (Fig. 3, Table 1). On OCT-A image of the superficial capillary plexus (SCP) of the right eye, hypointense dark-grayish areas of retinal capillary non-perfusion/hypoperfusion and capillary telengiectasies were seen (Fig. 2). In the same eye, OCT-A shows in the deep capillary plexus (DCP), larger grayish areas of capillary nonperfusion/hypoperfusion, capillary rarefaction, and diffuse capillary network disorganization. In the left eye, there were perivascular grayish-dark areas seen around the vessels in both superficial and DCP and also perifoveolar capillary network disruption was seen (Fig. 3).

Discussion

Retinal vascular changes are frequent in patients with SLE. [13] Quite severe vision losses were reported in the previ-

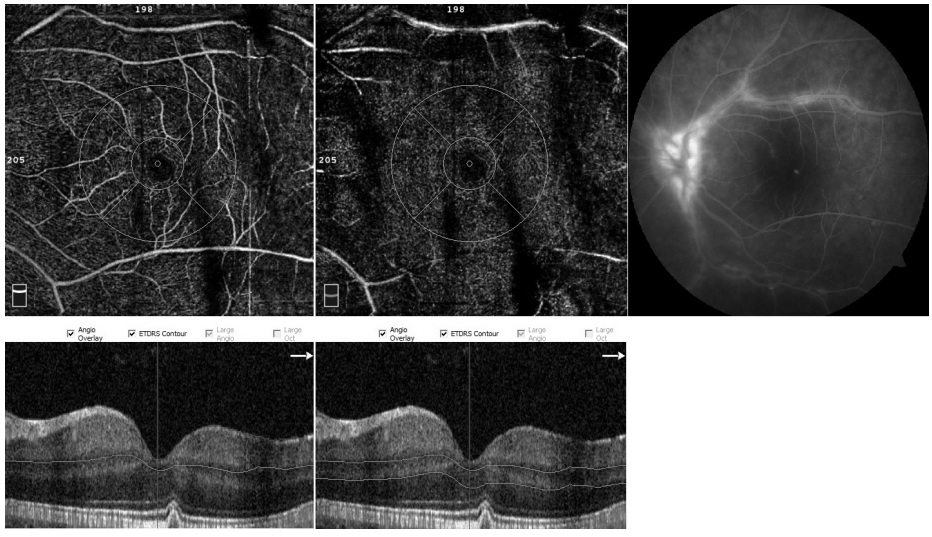

Fig. 3. Superficial (left image) and deep capillary plexus (middle image) and fundus fluorescein angiography image (right image) of the third patients left eye. Perivascular grayish-dark areas were seen especially in superficial capillary network image, consistent with fundus floresein image of inferior arcuate.

ous studies, especially according to retinal artery and vein occlusions.[14,15] The most common findings described are cotton wool spots, retinal hemorrhages, vascular abnormalities such as dilatation, tortuosity, narrowing, and vasculitis and optic disc edema.[4] FFA may reveal focal or diffuse areas of dye leakage even before vascular damage is clinically demonstrated.[16] However, it is difficult to obtain improved imaging of these perifoveal microvascular alterations, mainly due to the limited duration of the early frames, problem in focusing, media opacities, light scattering, and early dye leakage from the capillaries.[17] In addition, there were some reports about SLE retinopathy follow with OCT.[18,19]

Although effectiveness of the OCT on visualizing macular pathologies as edema was known, it is very restricted on retinal microvascular imaging. OCT-A is a noninvasive technique for rapidly acquiring depth-resolved images of the retinal and choroidal vasculature without the need for contrast agent injection.[20] It was reported in the previous studies that it is useful technique for imaging retinal microvasculature in several diseases such as Behçet disease, retinal vein occlusion, diabetic retinopathy, and sickle cell retinopathy.[9,21–24]

In our study, the patient had branch retinal vein occlusion on the right eye and active vasculitis on the left eye. Superficial capillary findings on OCT-A of the right eye were capillary arcade disruption, dark-greyish areas with reduced capillary density with capillary network disruption corresponding to low perfusion areas and capillary telengiectasies. Deep capillary findings on OCT-A of the right eye were capillary network disruption, dilation of deep superior macular capillaries with hyper signal, larger grayish areas of capillary non-perfusion/hypoperfusion, and cysts. Similar findings of retinal vein occlusions on OCT-A were reported in several previous studies.[17,23,25] In these studies, foveal avascular zone enlargement, capillary non-perfusion occurrence, greyish non-hypo perfusion areas, microvascular abnormalities appearance, vascular congestion signs, disruption in capillary network, and central cysts findings have been reported about branch or central retinal vein occlusion on OCT-A. The authors reported that cystoid spaces and dark grayish areas appear more common in DCP than in superficial and these spaces were the distinct from the dark grayish areas because they have well demarcated borders on the en face image. Therefore, this can be an indicator of the larger involvement of the DCP compared to the SCP in RVO due to anatomic feature.[17] The authors suggested that these findings about DCP is the superiority of the OCT-A to FFA. Because, it is difficult to focus on the different retinal layers, including the DCP, in the available short period of time with FFA imaging and hyperfluorescence from leaking vessels masks the perifoveal non-perfused areas.

On the left eye fundus examination of the patient, there were a vasculitic appear with retinal hemorrhages and perivascular sheeting. On the FFA, perivascular staining, capillary leakage and optic disk hyperfluorescence were seen. SCP findings on OCT-A of the left eye were perivascular grayish-dark areas seen around the vessels which have perivascular staining on the FFA. In addition, probable disruption in capillary network on the perifoveal area was seen. DCP findings were same as the superficial findings; however, perivascular areas were more darker. Khairallah et al.[9] have been evaluated the Behçet uveitis with using OCT-A and they reported that presence of grayish non-perfused/hypoperfused areas was the most common OCT-A finding in their study.

In that study, OCT-A findings were more prominent in DCP, the authors suggested that this result is associated with vulnerability of DCP to ischemia due to not to be connected directly arterioles.

Other two patients', OCT-A images were not reveal significant findings compared to this patient. However, if we consider that the first patient have had SLE for 15 years, we may suggest that, in the lack of the vasculitic situation, long-term SLE disease does not make significant changes in retinal microvasculature.

Conclusion

We suggested that OCT-A can visualize retinal microvasculature better than in FFA, especially in deep retinal capillary plexus. This superiority seems to be valid both in acute and chronic cases.

Informed Consent: Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review: Externally peer-reviewed.

Authorship Contributions: Concept: S.E.; Design: .M.O.U.; Supervision: S.E.; Resource: S.E.; Materials: S.E.; Data Collection and/or Processing: M.O.U.; Analysis and/or Interpretation: M.O.U.; Literature Search: M.O.U.; Writing: M.O.U.; Critical Reviews: S.E.

Conflict of Interest: None declared.

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References

1. Preble JM, Silpa-Archa S, Foster CS. Ocular involvement in systemic lupus erythematosus. Curr Opin Ophthalmol 2015;26:540–5. [CrossRef]
2. Palejwala NV, Walia HS, Yeh S. Ocular manifestations of systemic lupus erythematosus: A review of the literature. Auto-

immune Dis 2012;2012:290898. [CrossRef]
3. Read RW. Clinical mini-review: Systemic lupus erythematosus and the eye. Ocul Immunol Inflamm 2004;12:87–99. [CrossRef]
4. Au A, O'Day J. Review of severe vaso-occlusive retinopathy in systemic lupus erythematosus and the antiphospholipid syndrome: Associations, visual outcomes, complications and treatment. Clin Exp Ophthalmol 2004;32:87–100. [CrossRef]
5. Jabs DA, Fine SL, Hochberg MC, Newman SA, Heiner GG, Stevens MB. Severe retinal vaso-occlusive disease in systemic lupus erythematous. Arch Ophthalmol 1986;104:558–63. [CrossRef]
6. Matsunaga D, Yi J, Puliafito CA, Kashani AH. OCT angiography in healthy human subjects. Ophthalmic Surg Lasers Imaging Retina 2014;45:510–5. [CrossRef]
7. Spaide RF, Klancnik JM Jr., Cooney MJ. Retinal vascular layers imaged by fluorescein angiography and optical coherence tomography angiography. JAMA Ophthalmol 2015;133:45–50.
8. Moult E, Choi W, Waheed NK, et al. Ultrahigh-speed swept-source OCT angiography in exudative AMD. Ophthalmic Surg Lasers Imaging Retina 2014;45:496–505. [CrossRef]
9. Khairallah M, Abroug N, Khochtali S, et al. Optical Coherence tomography angiography in patients with behçet uveitis. Retina 2017;37:1678–91. [CrossRef]
10. Hochberg MC. Updating the American college of rheumatology revised criteria for the classification of systemic lupus erythematosus. Arthritis Rheum 1997;40:1725. [CrossRef]
11. Jia Y, Tan O, Tokayer J, et al. Split-spectrum amplitude-decorrelation angiography with optical coherence tomography. Opt Express 2012;20:4710–25. [CrossRef]
12. Kraus MF, Potsaid B, Mayer MA, et al. Motion correction in optical coherence tomography volumes on a per A-scan basis using orthogonal scan patterns. Biomed Opt Express 2012;3:1182–99. [CrossRef]
13. Arevalo JF, Lowder CY, Muci-Mendoza R. Ocular manifestations of systemic lupus erythematosus. Curr Opin Ophthalmol 2002;13:404–10. [CrossRef]
14. Noma H, Shimizu H, Mimura T. Unilateral macular edema with central retinal vein occlusion ın systemic lupus erythematosus: A case report. Clin Ophthalmol 2013;7:865–7. [CrossRef]
15. Nishiguchi KM, Ito Y, Terasaki H. Bilateral central retinal artery occlusion and vein occlusion complicated by severe choroidopathy ın systemic lupus erythematosus. Lupus 2013;22:733–5. [CrossRef]
16. Tolba DA, El-Fayoumi DM, Abdelaziz MS, Nabih MH. Fluorescein angiographic findings in patients with active systemic lupus erythematosus. Ocul Immunol Inflamm 2016;20:884–90.
17. Coscas F, Glacet-Bernard A, Miere A, et al. Optical coherence tomography angiography in retinal vein occlusion: Evaluation of superficial and deep capillary plexa. Am J Ophthalmology 2016;161:160–71.e1–2. [CrossRef]
18. Ozturk B, Bozkurt B, Karademir Z, Kerimoglu H. Follow-up of lupus choroidopathy with optical coherence tomography. Lupus 2011;20:1076–8. [CrossRef]
19. Kouprianoff S, Chiquet C, Bouillet L, Romanet JP. Oct follow-up of systemic lupus erythematosus choroidopathy. Ocul Immunol Inflamm 2010;18:113–5. [CrossRef]
20. Farecki ML, Gutfleisch M, Faatz H, et al. Characteristics of Type 1 and 2 CNV in exudative AMD in OCT-angiography. Graefes Arch Clin Exp Ophthalmol 2017;255:913–21. [CrossRef]
21. Takase N, Nozaki M, Kato A, Ozeki H, Yoshida M, Ogura Y. Enlargement of foveal avascular zone in diabetic eyes evaluated by en face optical coherence tomography angiography. Retina 2015;35:2377–83. [CrossRef]
22. Scarinci F, Nesper PL, Fawzi AA. Deep retinal capillary nonperfusion is associated with photoreceptor disruption in diabetic macular ischemia. Am J Ophthalmol 2016;168:129–38. [CrossRef]
23. Cardoso JN, Keane PA, Sim DA, et al. Systematic evaluation of optical coherence tomography angiography in retinal vein occlusion. Am J Ophthalmol 2016;163:93–107. [CrossRef]
24. Sanfilippo CJ, Klufas MA, Sarraf D, Tsui I. Optical coherence tomography angiography of sickle cell maculopathy. Retin Cases Brief Rep 2015;9:360–2. [CrossRef]
25. Rispoli M, Savastano MC, Lumbroso B. Capillary network anomalies in branch retinal vein occlusion on optical coherence tomography angiography. Retina 2015;35:2332–8. [CrossRef]