CASE REPORT
DOI: 10.5603/OJ.2021.0027

Swept-source optical coherence tomography angiography findings in a patient with Behçet's disease: a case report

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ABSTRACT

We present the clinical case of a 28-year-old female patient with a diagnosis of Behçet's disease and unilateral relapsing ocular involvement in systemic management with infliximab. Optical coherence tomography angiography with swept-source technology (DRI OCT Triton Plus™, Topcon Corporation, Tokyo, Japan) revealed asymmetric compromise between both eyes. Decreased foveal vascular density, asymmetrical foveal avascular zone enlargement at superficial and deep capillary plexuses, and focal areas of hypoperfusion at the deep capillary plexus around the macula with corresponding hyporreflective lesions in the *en face* slab in the left eye were noted.
Swept-source OCT angiography (SS-OCTA) helped identify early asymmetrical microvascular structural alterations compatible with macular ischemia in a patient with systemic manifestations and unilateral ocular compromise.

KEY WORDS: Behçet's disease; optical coherence tomography angiography; uveitis; vasculitis; retinal vasculature

Ophthalmol J 2021; Vol. 6, 193–198

INTRODUCTION

Behçet's disease (BD), also known as "silk road disease" [1], was first described in 1937 [2]. This multisystemic rheumatologic disease is characterized by necrotizing, chronic, and occlusive vasculitis, of unknown cause, which affects arteries and veins, with recurrent self-limited episodes [1–3]. Classically it presents with a triad of oral aphthous ulcers, genital ulcers, and uveitis. Nevertheless, it can also compromise other systems and organs less frequently, such as the central nervous system, cardiovascular, skin, gastrointestinal, renal, pulmonary, and musculoskeletal systems [1–4].

The prevalence of BD varies with demographic profiles and is determined by environmental and genetic factors [2]. Turkey has the highest prevalence of BD, with 420 per 10,000 inhabitants [1,5]. Usually, the disease presents between 20-40 years of age, and both genders are equally affected [1, 6].

Ocular symptoms occur in 40-70% of the cases [7–9] with different clinical manifestations such as anterior non-granulomatous uveitis with or without hypopyon, panuveitis, retinitis, retinal vasculitis, venous or arterial retinal occlusion, cystoid macular edema, and papillitis [7–11]. Potential irreversible visual-threatening complications are also possible,

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such as retinal detachment, secondary glaucoma, and optic atrophy [6].

Retinal vasculitis is the most common cause of visual loss [7, 8, 10]. Fluorescein angiography (FA) has become the gold standard for evaluating and following this vascular disease [4,8,11]. However, FA presents some limitations. The procedure is invasive and uses dye which can cause adverse effects such as nausea and anaphylaxis. Additionally, FA provides limited information on the capillary structure of both superficial and deep plexuses separately [4, 8].

Swept-source optical coherence tomography angiography (SS-OCTA) is a novel non-invasive imaging technique that provides highly detailed cross-sectioned structural imaging of the retinal capillary anatomy separating the superficial capillary plexus (SCP), deep capillary plexus (DCP), outer retina, and choriocapillaris [4, 8, 10, 12, 13]. It works through contrast blood movement images generating instantaneous angiographic-like pictures based on different sequential slices of optical coherence tomography (OCT) taken in the same cross-section (B-scan) and delivers 3-dimensional information about the structures of retinal vasculature to construct a map of blood flow [14, 15].

We present SS-OCTA findings of a patient with BD and unilateral ocular manifestations.

To the best of our knowledge, this is the first case that describes SS-OCTA alterations in a patient with BD in South America.

CASE PRESENTATION

We present a case of a 28-year-old female patient with a past medical history of celiac, Sjögren's syndrome, and relapsing recurrent oral aphthous ulcers, joint pain, folliculitis, multiform erythema, deep venous thrombosis, and anterior uveitis in her left eye (OS). HLA-B51 antigen testing was positive, as well as the pathergy test. She was diagnosed with BD by a rheumatologist and uveitis specialist.

Best-corrected visual acuity (BCVA) was 20/20 in the right eye (OD) and 20/25 in the OS. Anterior segment was unremarkable in both eyes (OU). Fundus eye examination revealed no alterations in OD and focal points of retinal pigment epithelium (RPE) atrophy within the fovea and at the end of temporal vascular arcades in OS (Fig. 1A).

FA in the OD was normal. The OS demonstrated hyperfluorescence and vascular leakage around the optic nerve, besides focal punctiform transmission defects within the foveal avascular zone (FAZ) and between temporal vascular arcades compatible with focal RPE atrophy (Fig. 1B).

SS-OCT and SS-OCTA images were taken with DRI OCT Triton Plus™ (Topcon Corporation, Tokyo, Japan).

OCT B-scan showed a normal foveal retinal thickness (FRT) without pathologic findings (Fig. 2).

Full-thickness 6.0 × 6.0 mm angiogram images were used to analyze vascular changes. Structural analysis evidenced asymmetric values of foveal vascular density (VD) between OD and OS, with 21.75% and 16.91%, respectively (Fig. 3), and multiple focal areas of VD loss at the level of DCP in the macular region of OS (Fig. 4A) that correlates with hyporreflective spots in *en face* slabs (Fig. 4B). Additionally, it showed marked asymmetry at FAZ in SCP of OD 169,805 μm^2 and

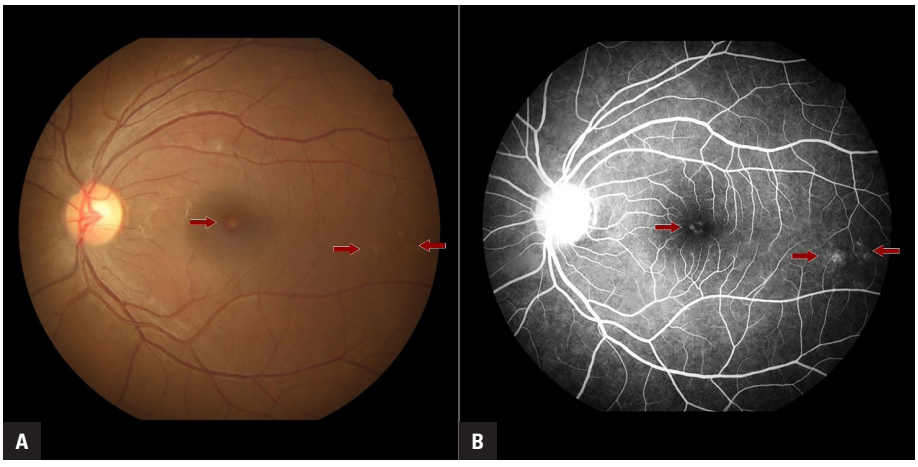

FIGURE 1. Images of left eye. **A.** Focal points of retinal pigment epithelium atrophy (arrows). **B.** Disc hyperfluorescence. Focal transmission defects within foveal avascular zone and between temporal vascular vessels (arrows)

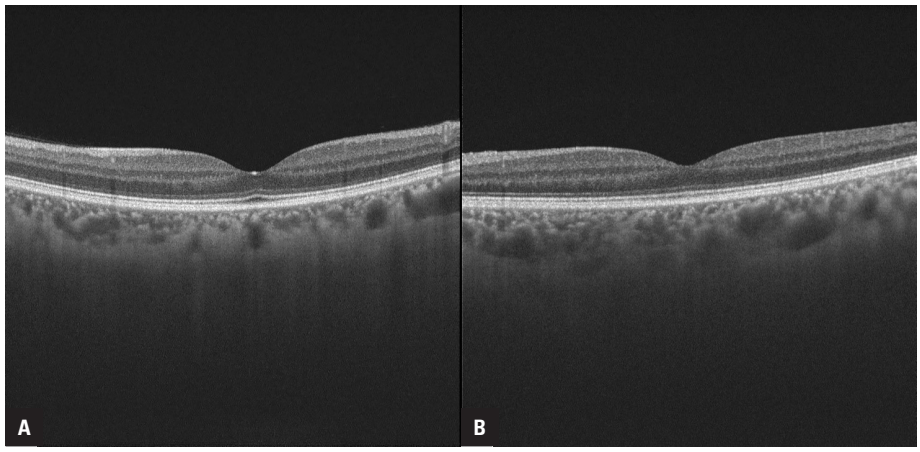

FIGURE 2. Normal optical coherence tomography in both eyes

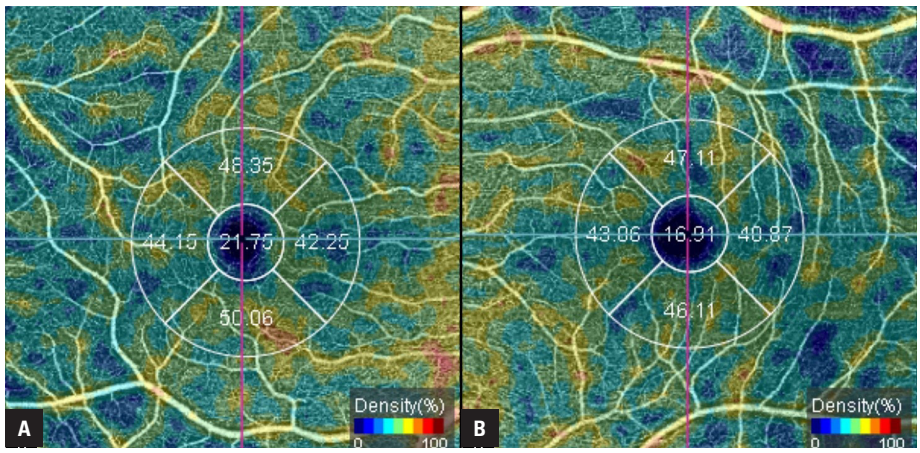

FIGURE 3. Asymmetric values of foveal vascular density. **A.** Right eye: 21.75%. **B.** Left eye: 16.91%

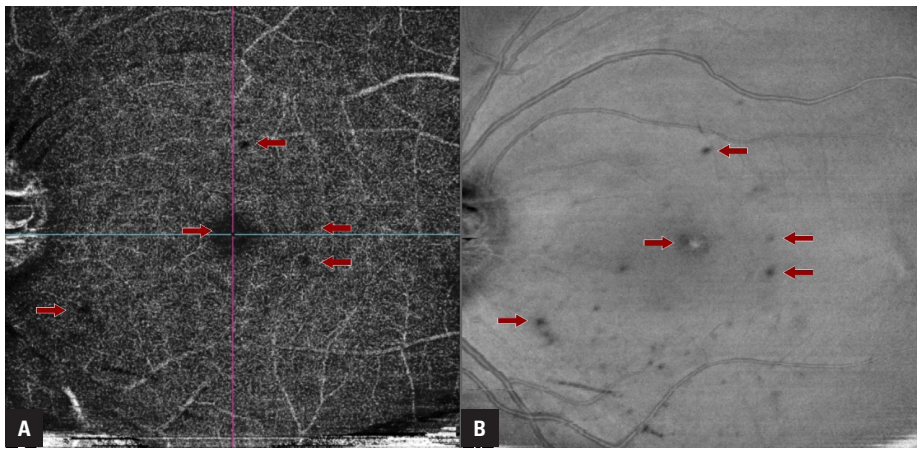

FIGURE 4. Images of left eye. **A.** Focal areas of vascular density loss at deep capillary plexus (arrows). **B.** Focal hyporreflective spots in *en face* that correlates with figure 4A (arrows)

OS 281,602 μm^2 (Fig. 5) and DCP with OD 228,867 μm^2 and OS 357,891 μm^2 (Fig. 6).

At present, the patient remains on systemic infliximab therapy.

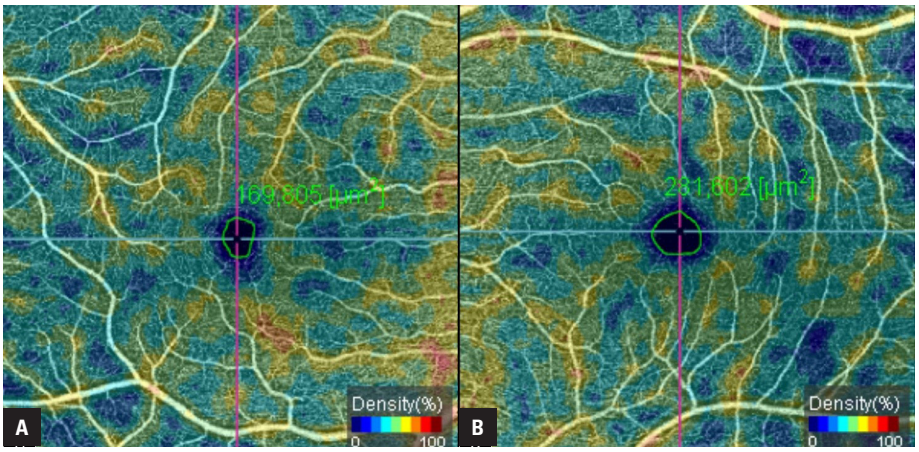

FIGURE 5. Size of foveal avascular zones in superficial capillary plexus. **A.** Right eye: 169.805 m^2. **B.** Left eye: 281.602 m^2

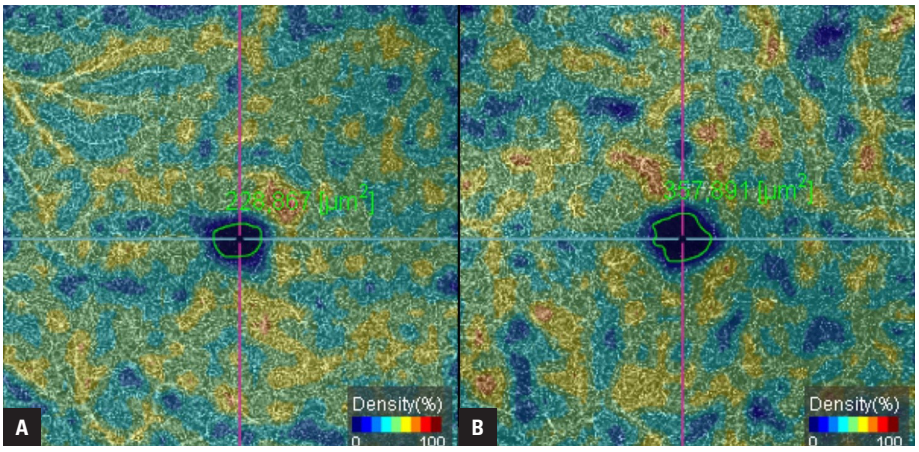

FIGURE 6. Size of foveal avascular zones in deep capillary plexus. **A.** Right eye: 228.867 m^2. **B.** Left eye: 357.891 m^2

DISCUSSION

Behçet´s disease is a systemic vasculitis that affects veins as well as arteries. Ophthalmological manifestations are present in about 20% of cases. It can affect many structures, and retinal vasculitis is the most common in the posterior segment. Generally, it manifests 2-3 years after onset [9].

FA is considered the main diagnostic exam in these cases to show zones with no-perfusion [4, 7, 8, 13]. In our case, FA was also helpful to identify transmission defects compatible with RPE atrophy and active inflammation around the optic nerve in late stages in the OS. However, this imaging modality did not allow an accurate measurement of the FAZ and made it challenging to identify small areas of non-perfusion in different levels, especially in cases where leakage of dyes is present [4, 8].

Retinal pigmentary epithelium atrophy zones were associated with VD focal loss in SS-OCTA, and it also evidenced FAZ enlargement with foveal VD loss in both plexuses of the affected eye. These results are similar to those reported in the literature [4,7–10, 12, 13].

As in our report, Somkijrungroj [4], Khairallah [7], and Emre [8] observed that both plexuses were altered. Nevertheless, DCP is affected more often with more significant non-perfusion areas compared to SCP in the early stages of the disease, with decreased capillary density and FAZ enlargement in the eyes of patients with active BD. This is probably due to the fact that the DCP is a watershed zone between inner and outer retinal circulation, and its capillaries have no direct connection with arterioles and could be more vulnerable to an ischemic process [4, 7–9].

It is interesting to consider that few reports have shown retinal microvasculature alterations in patients with BD diagnosis. Still, with no evident

ocular affection [9, 12, 13], this could indicate that posterior segment vessels are affected simultaneously with other organs. This could indicate that posterior segment vessels are affected simultaneously with other organs because its manifestations are the result of a chronic progressive process, being its expression unpredictable at ocular level.

Other studies have demonstrated that vascular supply in the posterior pole tends to be spared until the late stages of the disease with generalized vascular occlusion [6]. However, this theory would not apply in our case: the posterior pole in the OS had a marked FAZ enlargement in both superficial and deep capillary plexuses accompanied by decreased FVD compared to the OD. This was evidence that ocular compromise could be asymmetric and independent of the stage of the disease. Macular ischemia, the major cause of permanent visual loss in retinal vascular diseases, was undetectable in FA [4, 8, 11].

Foveal retinal thickness was normal in OU, demonstrating that vascular changes in our patient may have not affected retinal structures at this stage but may have an effect on visual acuity, hence the patient´s decreased BCVA. In contrast to our case, some authors have previously identified retinal microvascular changes without altered visual acuity [9, 12, 13].

At present, there is no established consensus regarding normal FAZ size, so we determined there was enlargement by comparison to the contralateral non-affected eye.

CONCLUSION

Swept-source optical coherence tomography angiography is a novel imaging modality that does not require invasive dye injection to identify alterations in the retinal vasculature. It was useful to detect unilateral hypoperfusion areas, FAZ enlargement at SCP and DCP, and decrease FVD in early stages compatible with macular ischemia, secondary to BD chronic damage.

This imaging modality could be useful in monitoring the progression of visual loss and guide early systemic treatment. However, we consider that large studies are necessary to define the value of OCT-A in the management of BD.

Conflict of interest

The authors declare that they have no conflict of interest.

Funding

None declared.

Informed consent and human and animal rights statements

Informed consent has been obtained from all individuals included in this study.

Authorization for the use of human subjects and ethical approval

The research related to human use complies with all the relevant national regulations and institutional policies, according to the tenets of the Helsinki Declaration, and has been approved by the Ethics Committee of Grupo Oftalmológico Abdala-Figuerola AF, Barranquilla, Colombia.

Acknowledgements

None declared.

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