

Ocular involvement in Behçet's Disease: relevance of new diagnostic tools

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Key message: SD-OCT and OCTA can valuably recognize early retinal ischemic changes in patients with Behçet's disease.

Dear Editor,

Ocular involvement in Behçet's Disease (BD) is characterized by a relapsing non-granulomatous uveitis that may affect both the anterior and the posterior eye segment, often causing a decrease of visual acuity that may progress to blindness [1]. This particularly takes place when the recurrent inflammatory flares of the disease translate to damage of the posterior eye segment, leading to a permanent injury of the retinal vasculature [1]. The early identification of disease activity is still challenging, although its prompt recognition can decrease the risk of poor visual outcome [2]. Spectral domain optical coherence tomography (SD-OCT) and optical coherence tomography angiography (OCTA) could complement the information given by the standard fluorescein angiography (FA).

Herein, we report the case of a 39-year-old Caucasian man with BD, who came to our department for his annual evaluation. The patient gave written informed consent for this case report. Seven years ago, he was diagnosed with BD based on the recurrence of oral and genital ulcers, skin lesions and a positive pathergy test. He also had an anterior right uveitis, and an asymmetrical polyarthritis, currently deserving only a low-dose of methyl-prednisolone (4 mg/day). His medical history was otherwise unremarkable.

At current presentation, he reported that two episodes of transient bilateral scotomas had occurred 3 months before. His best-corrected visual acuity was 20/20 in the right eye (OD) and 20/25 in the left eye (OS). Both fundus examination and fluorescein angiography (FA) were normal. However, SD-OCT revealed a bilateral limited area of severe thinning of the retinal inner nuclear layer, determining a partial collapse of the overlying retinal structures (fig. 1 panel A, B). A complementary information was provided by OCTA, which bilaterally showed an isolated ischemia of the deep capillary plexus corresponding to the atrophic lesions detected by SD-OCT (fig 1 panel C, D). These findings, documenting subtle ischemic changes, were consistent with the chronic evolution of a paracentral acute middle maculopathy. Therefore, azathioprine 100 mg per day (1.5 mg/kg/die) was added to the current therapy, and we planned a tighter ocular follow-up aimed to identify further ocular disease progression.

The retinal capillary network is a multilayer structure, composed of the superficial, intermediate and deep capillary plexuses [3]. FA, the current reference method for the assessment of posterior ocular

involvement in BD, only allows a two-dimensional study of the superficial retinal circulation, and frequently, the damage of retinal vessels limits a reliable assessment of disease activity [3]. SDOCT better defines the morphology of the retina, whereas OCTA can separately evaluate its superficial and deep circulation, simulating angiographic images [2,4]. Through these techniques, we identified the ocular involvement of our patient, whose deep retinal capillary plexus was severely affected. Indeed, paracentral acute middle maculopathy, whose clinical course might be asymptomatic or characterized by transient or persistent paracentral scotomas, is caused by a focal ischemia of the deep capillary plexus [5]. This disorder may be detected in eyes with compromised retinal arterial circulation in the absence of FA abnormalities as an isolated phenomenon or as expression of an underlying systemic disease, as occurred in our patient [4,5]. Actually, the posterior eye involvement is a prognostic factor of poor visual prognosis, especially in young men. In our patient, we added a dose of azathioprine apt to allow a better control of the disease, as our findings were expression of a past ocular flare, likely responsible of scotomas. In the subsequent six months, the patient did not experience further episodes of scotomas.

SD-OCT and OCTA, by yielding an in-depth investigation of retinal vascular pathologies, could allow us to identify unrecognized ocular BD cases. In fact, our SD-OCT and OCTA findings are in line with those of a recent study, reporting that patients with diagnosis of BD and without significant FA perfusion abnormalities did have a significant damage of the deep capillary plexus [6]. It is conceivable that the ischemia of the deep capillary plexus, situated in a watershed region of oxygen supply, may be an early expression of BD. This could also account for the progressive vision loss observed in some patients with BD and negative FA findings, and could possibly be useful screening tools in this condition [7]. Therefore, methods apt to more accurately explore retinal circulation may evidence earlier stages of the disease.

Overall, the finding we obtained by SD-OCT and OCTA would suggest that the prevalence of ocular involvement might be higher than expected in BD. This may have implications for treatment, such as a lower threshold for starting a therapy. In summary, the present case outlines that SD-OCT and OCTA could be valuably used in the evaluation and monitoring of BD patients. These techniques may provide a more comprehensive assessment of retinal inflammatory involvement, revealing subtle ischemic changes which precede clinically overt ocular BD.

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Figure 1. SD-OCT and OCTA features of paracentral acute middle maculopathy. SD-OCT scan of the right (A) and left (B) eye reveals thinning of the inner nuclear layer (top arrows) and irregularity of the outer plexiform layer (bottom interrupted arrow). OCTA scan shows the corresponding area of not perfused deep capillary plexus (circle) in the right (C) and left (D) eye.

SD-OCT: Spectral domain optical coherence tomography; OCTA: optical coherence tomography angiography.

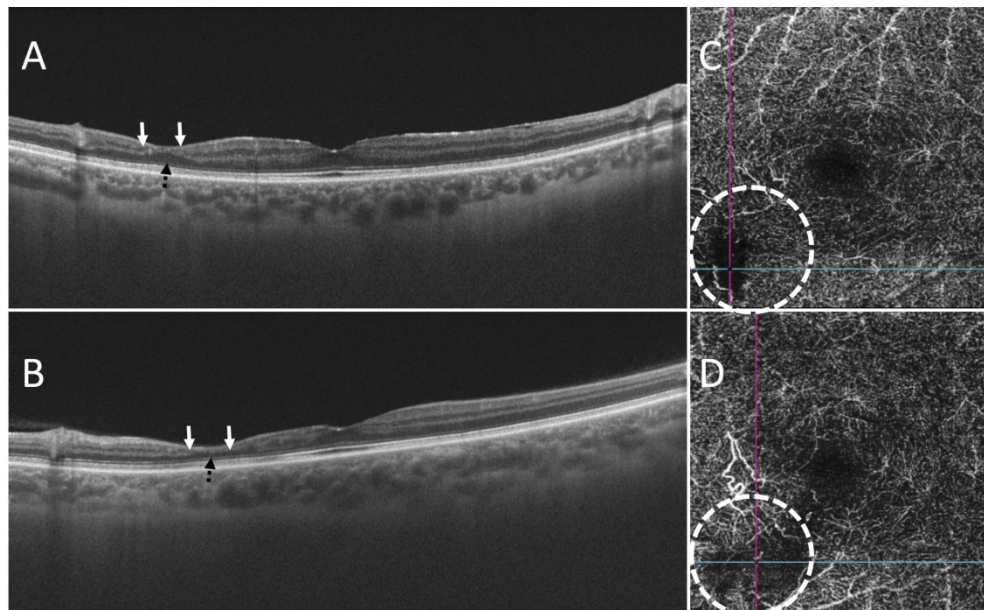


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