Early presentation of sympathetic ophthalmia in optical coherence tomography studies: A case report

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Sympathetic ophthalmia (SO) is often diagnosed when an inflammatory process appears to be advanced. Herein, the authors present the prospective optical coherence tomography (OCT) study of the onset of SO in the sympathizing eye. Prior to any signs of uveitis, we noted the mild disintegration of the retinal pigment epithelium (RPE) layer, the interdigitation zone (IZ), and the ellipsoid zone (EZ). The complete disruption of IZ and EZ was seen 12 weeks later. After 14 weeks, the uveal inflammation was present, and OCT imaging disclosed the formation of nodule-like lesions between the Bruch's membrane and the RPE layer. The histopathological evaluation of the enucleated exciting eye confirmed the diagnosis of SO.

Key words: Dalen-Fuchs nodules, granulomatous uveitis, ocular trauma, optical coherence tomography, sympathetic ophthalmia

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Sympathetic ophthalmia (SO) is a bilateral, nonnecrotizing granulomatous inflammation of the uveal tract preceded by surgical or traumatic injury of one eye. Patients present with anterior uveitis, vitritis, choroiditis with multiple yellow-white lesions, papillitis, macular edema, and serous retinal detachment. The onset of SO is recognized by the blurring of vision and signs of the inflammatory process.[1,2] Apart from physical examination, several authors reported the optical coherence tomography (OCT) findings in patients with SO and found it useful for monitoring the course of the disease.[3-7]

Herein, we report a patient with SO who underwent systematic ophthalmic examination since the day of the injury of one eye. By SD-OCT imaging, we visualized early changes at

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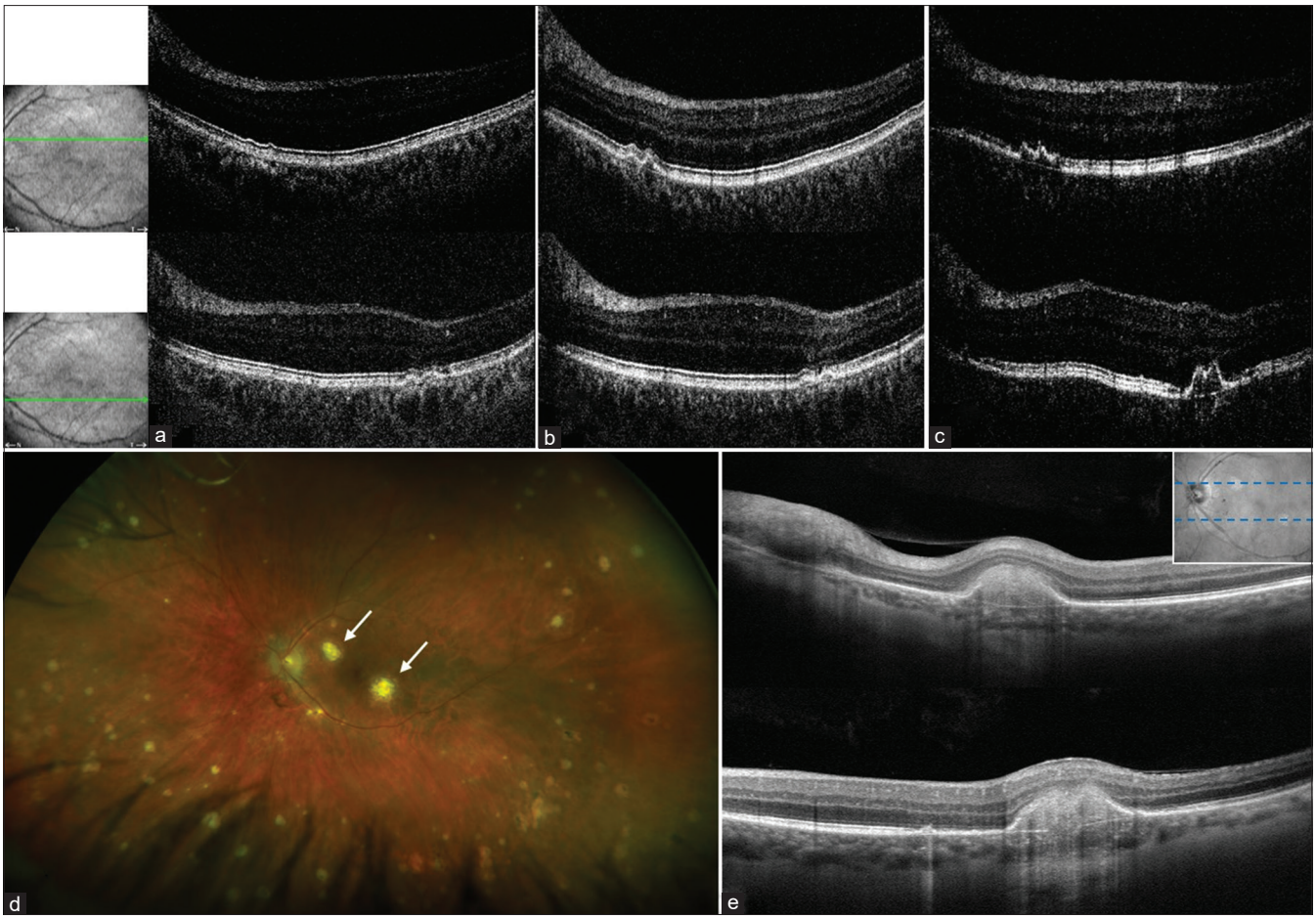

Figure 1: The optical coherence tomography (OCT) scans of the left eye showing the progression of two retinal lesions in time. (a) It demonstrates the areas of retinal pigment epithelium (RPE) elevation and mild disintegration of interdigitation zone (IZ) and the ellipsoid zone (EZ) (23 h after right eye trauma) with further RPE elevation and disruption of IZ and EZ 12 weeks later (b). (c) OCT discloses the thickened retina and forming of the nodule-like lesion between Bruch's membrane and the RPE layer (14th week). (d) Fundus photo shows yellow Dalen-Fuchs nodules (white arrows), which correspond with lesions between Bruch's membrane and the RPE layer on OCT scans (e) 6 months later

the level of retinal pigment epithelium (RPE), the interdigitation zone (IZ), and the ellipsoid zone (EZ), while the patient had no visual disturbances and signs of inflammation.

Case Report

A 62-year-old Caucasian male presented with a penetrating injury to his right eye (RE) caused by a metallic splinter a day before. Best-corrected visual acuity (BCVA) was light perception and 20/20 in his RE and left eye (LE), respectively. The slit-lamp examination disclosed a previously sutured corneal wound and a traumatic cataract in the injured eye. The anterior segment and fundus of the fellow eye were unremarkable; however, OCT scans showed a mild focal disintegration of RPE, IZ, and EZ in the macula [Fig. 1a]. The patient underwent pars plana vitrectomy of the RE with the removal of IOFB, cataract phacofragmentation, and intraocular injection of antibiotics. A giant peripapillary retinal tear hindered the attachment intraoperatively.

Twelve weeks postoperatively, the patient developed a painful phthisis bulbi with no light perception in RE [Fig. 2a]. Although BCVA in LE was still 20/20 with no abnormalities in ophthalmoscopy, the OCT imaging revealed previously observed macular lesions [Fig. 1b]. Two weeks later, the patient complained of blurry vision and photophobia—the visual acuity diminished to 20/25. Slit-lamp examination of the LE disclosed mutton-fat keratic precipitates [Fig. 3], posterior synechiae (2–4 and 8–9 o'clock meridians), vitritis (1+),[8] hyperemic optic disc, and multiple yellowish subretinal lesions [Fig. 2c]. The intraocular pressure (IOP) rose from 18 mmHg to 25 mmHg. OCT disclosed the formation of nodule-like lesions between Bruch's membrane and the RPE layer and thickened retina [Fig. 1c]. Fluorescein angiography revealed multiple hypofluorescent foci in the early venous phase followed by late staining, and the optic nerve head demonstrated leakage in the late phase of the angiogram [Fig. 2d and e]. The areas of early blocked fluorescence corresponded to the lesions observed on OCT images. The systemic workup was negative.

Based on the history and clinical findings, we made a diagnosis of SO. The patient was started on topical dexamethasone 0.1% every hour, tropicamide 1% t.i.d., dorzolamide 2% b.i.d., timolol 0.5% b.i.d., and oral prednisone 80 mg daily along with proton pump inhibitor (patient refused

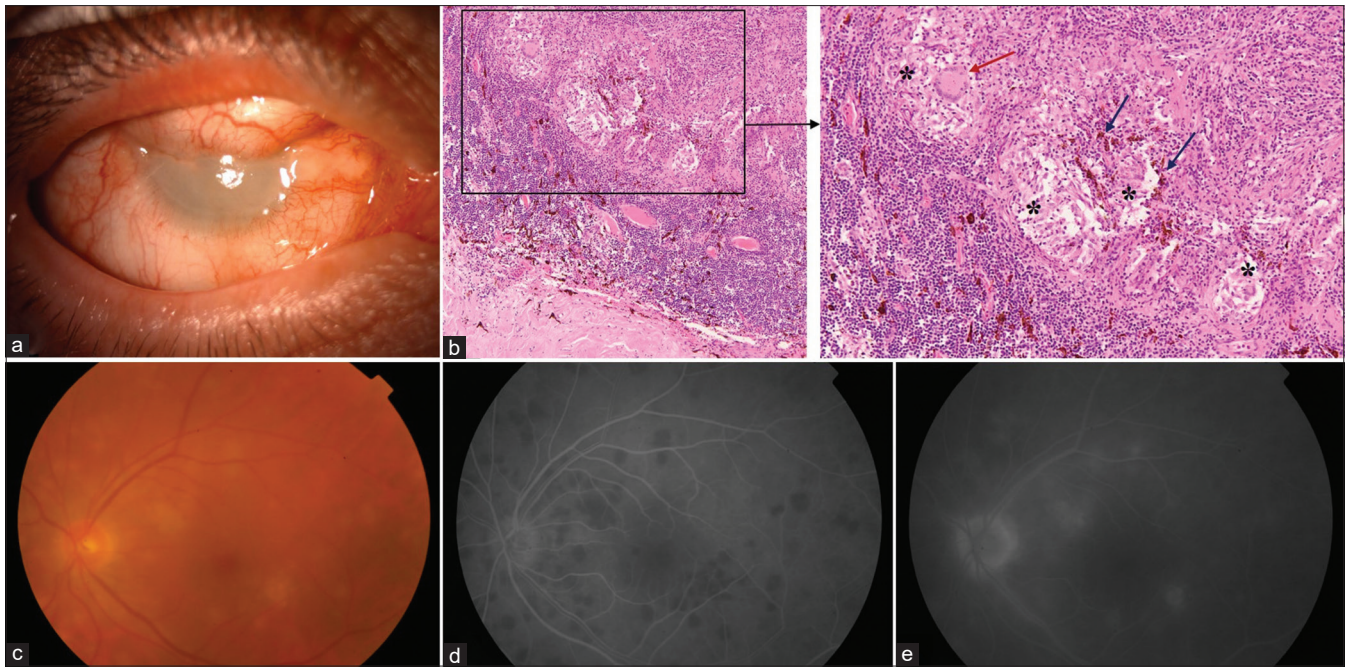

Figure 2: The acute phase of sympathetic ophthalmia. (a) The exciting eye 12 weeks after trauma—phthisis bulbi with no light perception. (b) The enucleated eye shows the thickened choroid with lymphocytes and granulomas. Please note the Dalen-Fuchs nodules (asterisk), Langhans' giant cell (red arrow), and epithelioid cells containing melanin granules (blue arrow); H and E stain; 100 × mag. (c) Fundus photo of the left eye shows hyperemic optic disc and multiple yellow lesions. FA reveals focal areas of hypofluorescence in the early venous phase (d) and late staining of lesions observed on OCT images (e)

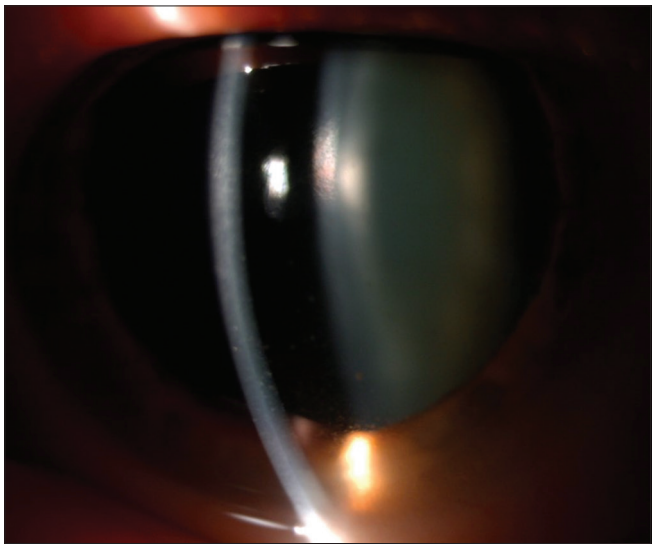

Figure 3: The acute phase of sympathetic ophthalmia. Mutton-fat keratic precipitates forming an Arlt's triangle in the left eye (indirect diffuse illumination, yellow field)

intravenous corticosteroids administration), calcium and vitamin D supplementation. The exciting RE was enucleated. The histopathology showed lymphocytic infiltration of the thickened choroid with Dalen-Fuchs nodules, Langhans' giant cells, and epithelioid cells [Fig. 2b]. Corticosteroids were slowly tapered, and we added cyclosporin A (5.0 mg/kg/day) and adalimumab (initially 80 mg followed by 40 mg subcutaneously every 2 weeks). The observed macular lesions developed into yellow Dalen-Fuchs nodules [Fig. 1d and e]. After a 2-year follow-up, the BCVA was stable at 20/20, and Dalen-Fuchs nodules have regressed [Fig. 4].

Discussion

We present herein the SD-OCT scan series of Dalen-Fuchs nodules formation in the sympathizing eye. The initial OCT scan of the LE was taken 23 h after the trauma of the exciting eye. Our attention was focused on two areas of RPE elevation and disintegration of IZ and EZ. We were performing the ophthalmic examination, including OCT imaging, every 2–3 weeks. Twelve weeks after eye trauma, we noted the progression of monitored lesions, although there were no inflammation and IOP elevation. Clinical signs of panuveitis occurred 2 weeks later when the structures growing beneath the RPE formed well visible yellow foci. Finally, they evolved into Dalen-Fuchs nodules. Due to the development of a painful blind eye, the exciting RE was enucleated. Histopathologic findings revealed that the inflammatory process was more marked in the exciting eye than in the sympathizing eye. Eye involvement does not have to be symmetrical. Aziz *et al*.[9] reported that the clinical course of SO might have a minimal correlation with the histopathologic findings, mainly in cases with corticosteroid therapy before enucleation. Our patient was on corticosteroids for 4 days only before enucleation. Therefore histopathologic characteristics of SO were prominent. Reynard *et al*.[10] described morphological variations of Dalen-Fuchs nodules and distinguished three types of them. The first type consisted of focal hyperplasia and aggregation of RPE cells, the second type included epithelioid cells and lymphocytes underlying an intact dome of RPE, and the third type was characterized by degeneration of the RPE and disorganization of the nodule. All three types may appear

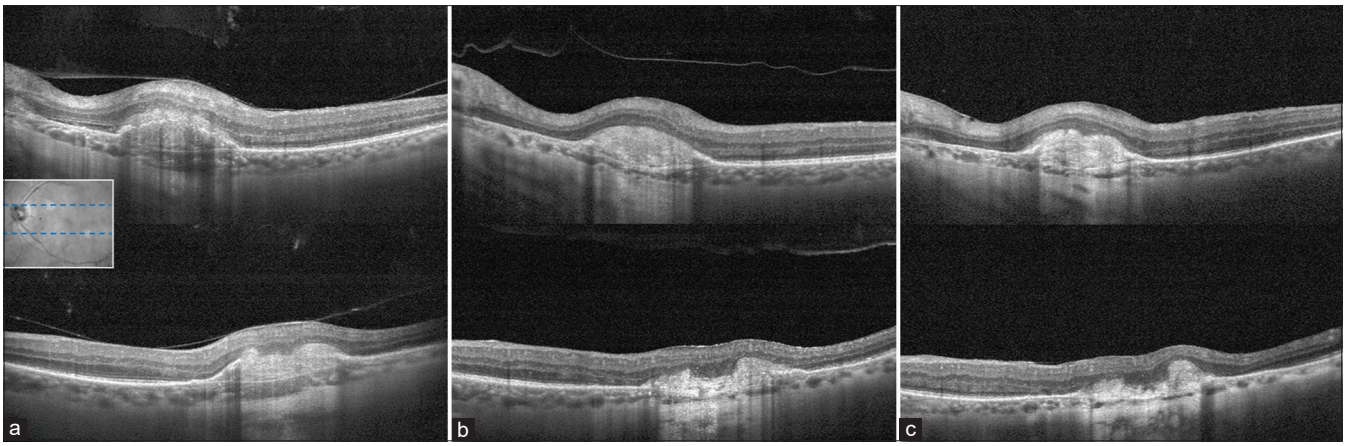

Figure 4: The regression of Dalen-Fuchs nodules. The OCT scans demonstrate the regression of the same nodules after 1 year (a), 1.5 years (b), and 2 years (c) of treatment. Please note the reduced thickness and increased hyperreflectivity of the nodules

in the same eye, not depending on the advancement of the inflammatory process. Due to total retinal detachment and loss of anatomic compartments, we were not able to assign the type of Dalen-Fuchs nodules in the histopathological specimen of the enucleated eye explicitly.

So far, no prospective observation of developing SO was published. There are several reported cases of SO, where the course of the disease was monitored with OCT and enhanced depth imaging OCT. It was shown that in the acute phase of SO, the choroidal thickening was more than 500 µm with a subsequent reduction of about 200 µm during treatment.[3,5] Gupta *et al*.[6] reported the reversible changes in the photoreceptor layer and the resolution of subretinal fluid in cases with prompt systemic corticosteroid therapy. Thus, early treatment is crucial to preserve vision. In OCT imaging, the Dalen-Fuchs nodules presented as hyper-reflective, sub-RPE lesions associated with the disruption of the ellipsoid zone and RPE.[11,12] In those studies, the lesions were visualized in the course of the disease. In our patient, we depicted the disruption of RPE, IZ, and EZ long before the signs of inflammation.

Conclusion

Our case illustrates the possibility to detect the developing SO before typical clinical findings occur. The small choroidal lesions can be initially overlooked, especially in patients with no symptoms, insidious onset, or recurrence of the disease. We recommend the frequent OCT imaging of preferably both eyes. We believe that the careful OCT evaluation may expedite early treatment when indicated.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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