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Presumed idiopathic outer lamellar defects of the fovea and chronic solar retinopathy. An OCT and Fundus Autofluorescence study.

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ABSTRACT

AIM: To examine fundus autofluorescence (FAF) findings in eyes with presumed idiopathic outer lamellar defects (OLD) at the fovea and to discuss their pathogenesis.

METHODS: prospective observational case series of five eyes of four patients presenting with OLD at the fovea defined as discrete lesions of 50-100 μ m in size located at the level of the outer retina on biomicroscopy and imaged on optical coherence tomography (OCT) as cylindrical, well demarcated interruption of both hyperreflective bands corresponding to the inner/outer segments junction of photoreceptors and to the complex retinal pigment epithelium-choriocapillaris; none of the enrolled patients had history positive for direct sunbathing, welding-arc or sunbed exposure, whiplash injury, ocular trauma, macular oedema/detachment or evidence of vitreo-macular traction. The corresponding FAF images were evaluated.

RESULTS: in eyes with OLD the neuroretina in the foveal region appeared to be thinner in comparison to fellow, unaffected eyes. FAF revealed well demarcated, hypoautofluorescent areas (corresponding in location to the OLD observed clinically and on OCT), surrounded by an irregular halo of relatively increased autofluorescence in the context of the greater hypoautofluorescent macular region.

CONCLUSION: biomicroscopy, OCT and FAF findings of presumed idiopathic OLD of the fovea strongly resembles those observed in association to chronic solar retinopathy. In association to OCT, FAF might represent an useful technique to detect subtle solar-induced injuries of the retina.

INTRODUCTION

Solar retinopathy[1-5] generally occurs in patients who have viewed an eclipse or have gazed directly at the sun, however it has also been described in patients who have minimally viewed the sun[3] or who have been sunbathing, not sungazing .[3,6] Recently one of the largest series ever published on solar retinopathy[7] reported that up to 49% of patients affected could not remember to have looked at the sun. A reddish, sharply demarcated foveal or juxtafoveal spot located at the level of outer retina, developing approximately two weeks after direct exposure to sunlight, is a well known sign of solar injury. This lesion is permanent and highly suggestive of solar retinopathy[8]. Similar biomicroscopic findings at the fovea have been reported in association to welding arc or sunbed exposure, ocular trauma, initial stages of idiopathic macular holes, persistent retinal defects following successful macular hole repair surgery, whiplash injuries, microaneurisms or hemorrhages, idiopathic parafoveal telangiectasis or grouped under the generic terms of foveal spots[9] or microholes of the fovea centralis. [10,11] Differential diagnosis is usually guided by history along with associated characteristic fluorescein angiography and OCT findings. Fundus autofluorescence (FAF), is a relatively novel, non invasive imaging technique based on the autofluorescent properties of natural occurring retinal fluorophores.[12]

It is generally accepted that the lipofuscin-laden cells of the retinal pigment epithelium (RPE) are the main source of FAF recorded with commercially available confocal scanning laser ophthalmoscopes.[12-14]

The retinal pigment epithelium (RPE) with its high content in melanin is able to reduce photoreceptors exposure to potentially dangerous photons by absorbing and scattering light forward into the choroid. The RPE is particularly exposed to damage when retina's

defense mechanisms against toxic free radicals from light and oxygen are overwhelmed by light levels exceeding those typically encountered in a normal environment.

Subtle RPE abnormalities, overlooked on cursory ophthalmoscopy, can be efficiently imaged using FAF; reduction of the content of lipofuscin, the main fluorophore of the RPE, has been documented following light-induced loss of photoreceptors.[15]

In this study we report OCT and, for the best of our knowledge, the first description of FAF findings in eyes with presumed idiopathic outer lamellar defects (OLD) of the fovea and compare such findings with those reported in eyes with chronic solar retinopathy.

METHODS

A prospective observational case series aimed to investigate FAF findings in eyes with presumed idiopathic OLD at the fovea was designed and carried out at Moorfields Eye Hospital, NHS Trust, between January 2006 and June 2007. The study was performed in accordance to the tenets of the Declaration of Helsinki. Approval from local Institutional Review Board and informed consent from each enrolled subject were obtained.

The diagnosis was based on biomicroscopic identification of small, reddish, sharply demarcated outer lamellar defects at the fovea using a fundus contact lens (Goldmann

three mirror lens, Ocular Instruments, Bellevue, Washington) and on third-generation OCT evaluation (Stratus Tomographer, Model 3000, Carl Zeiss Ophthalmic Systems Inc., Humphrey Division, Dublin, California). OCT examination was performed using six linear 6.00 mm scans oriented at intervals of 30 degrees and centered on the fixation point. To verify the accuracy of the OCT findings, the fundus area encompassing the lesions was additionally scanned using several vertical and horizontal scans of 5.00 mm in length. Retinal thickness was measured manually, using software callipers, at the foveal centre. OCT findings were considered consistent with diagnosis of OLD in presence of well demarcated interruption involving the hyperreflective bands corresponding to the inner/outer segments junction of photoreceptors and to the retinal pigment epithelium-choriocapillaris complex (Fig.1,2); eyes with concomitant macular abnormalities detectable on OCT such as fine foveal epiretinal membrane or vitreomacular traction were excluded. Patients with evidence of other fundus disease, such as hereditary maculopathy, diabetes and other retinal vascular diseases, intraocular infection or inflammation, intraocular tumors, myopia, previous macular oedema, previous surgery for macular hole or retinal detachment, ocular trauma and whiplash injury were also excluded from the study.

Particular attention was paid in recording information about history of direct sungazing, welding arc and sunbed exposure, [16] or assumption in the past of compounds with photosensitizing properties. [17, 18]

All the participants received a detailed ophthalmologic examination including best-corrected visual acuity on Snellen chart, slit-lamp biomicroscopy, indirect fundus examination, fundus photography, OCT 3 STRATUS and FAF evaluation at first visit and in occasion of follow-up appointments. For color fundus pictures, a high-resolution (1024 x 1024) fundus camera system (TRC-50IA/IMAGEnet 2000; Topcon, Tokyo, Japan) was used. FAF was recorded using a confocal scanning laser ophthalmoscope [Heidelberg Retina Angiograph, HRA, Heidelberg Engineering, Germany]. An argon

blue laser (488 nm) was used for excitation, emitted light detected above 500 nm (barrier filter). Before recording FAF images, individual refraction was corrected by varying the optics of the instrument in the reflection mode. From each individual eye at least 16 FAF images were obtained by using the frame grabber (10.2 images per second). In order to amplify the FAF signal, the images were aligned and a mean image was calculated after automated detection and correction of eye movements using image analysis software. The digital images were saved on disk for further analysis and processing.

RESULTS

Five eyes of four patients with diagnosis of presumed idiopathic OLD at the fovea were identified and enrolled in the study. No patient reported history of direct sungazing, welding arc and sunbed exposure, or assumption in the past of compounds with photosensitizing properties.

Mean follow-up (\pm SD) was 10.2 (\pm 2.2) months. All four patients were females. Mean (\pm SD) age was 52,5 (\pm 4,4) years.

Characteristics of the examined eyes are summarized in the Table.

pt/age/ gender	eye affected (OLD at the fovea on biomicroscopy and OCT)	VA	central foveal thickness on OCT
1. 47/F	BE	6/6 RE 6/6 LE	RE: 118 μm LE: 127 μm
2. 51/F	RE	6/9 RE 6/6 LE	RE: 125 μm LE: 182 μm
3. 55/F	RE	6/9 RE 6/6 LE	RE: 112 μm LE: 194 μm
4. 57/F	LE	6/9 RE 6/12 LE	RE: 175 μm LE: 154 μm

Table. Summary of the characteristics of the sample.

Abbreviations: pt: patient; F: female; OLD: outer lamellar defect; OCT: optical coherence tomography; BE: both eyes; VA: visual acuity; RE: right eye; LE: left eye.

Mild or moderate nuclear sclerosis was present in all but one eye, which had previously undergone uncomplicated phacoemulsification with posterior chamber intraocular lens implantation. One patient presented foveal outer lamellar defects in BE, the other three had unilateral lesions. In the five eyes with outer lamellar defects, best-corrected visual acuity was 6/12 in one, 6/9 in two and 6/6 in two eyes; visual acuity in the three fellow eyes of the patients with unilateral defect was 6/6. On biomicroscopic examination none of the patients had a complete posterior vitreous detachment (PVD). None of the patients demonstrated any retinal haemorrhage, lipid or exudate, full thickness macular hole, or operculum. In one patient drusen-like deposits were present at the macula in the eye with outer lamellar defect (Fig 2).

OCT 3 STRATUS and FAF were performed for both eyes of all patients. In eyes diagnosed with outer lamellar defects at the fovea, OCT demonstrated discrete abnormalities in shape of focal interruption at the level of the hyperelective bands corresponding respectively to the junction between inner and outer photoreceptor segments and to the retinal pigment epithelium-choriocapillaris complex. In addition, in the affected eyes, the neuroretina at the fovea appeared to be substantially thinner in comparison to the fellow unaffected eyes (centrofoveal mean thickness: 127.2 μm and 183.7 μm in the affected and unaffected eyes respectively). A preserved foveal contour and a normal vitreoretinal interface with no evidence of vitreo-macular traction was noted in all cases. FAF revealed well demarcated, hypoautofluorescent areas (corresponding in location to the OLD observed clinically and to the defects of the outer retina/RPE layers detected by OCT) surrounded by an irregular halo of relatively increased autofluorescence in the context of the greater hypoautofluorescent macular region (Fig. 1,2). FAF images of the fellow, unaffected eyes were consistent with physiologic phenotypes previously described in the literature.[12,14] Biomicroscopic,

OCT and FAF features of the OLD didn't change over the course of the follow-up period.

DISCUSSION

Solar retinopathy refers to maculopathy that occurs following exposure to solar radiation. [1-4] There is a wide spectrum of solar-induced injuries and great individual variation in susceptibility for developing such lesions.[5] Prolonged staring at the sun may cause only minimal abnormalities[5] whereas minimal exposure (e.g. during sunbathing) may induce macular lesions. [3] Many factors including characteristics of the light (wavelength, dose rate), ocular pigmentation, clarity of the ocular media and body temperature (influenced by climate, exercise or infection) along with unusual environmental conditions such as highly reflective surroundings and reduced atmospheric ozone[6] are believed to influence the final injury on the retinal cells. Most cases are bilateral but often asymmetry between the two eyes is observed. Unilateral cases of solar retinopathy have been documented and are not uncommon. Ophthalmoscopic examination in acute phases may reveal a small yellowish spot in the centre of the foveal region surrounded by an area of faint grey, granular pigmentation. [8] This spot usually fades gradually within few weeks and assumes a reddish, sharply circumscribed appearance at the fovea, which Gass[8] described as pathognomonic of solar retinopathy.

Recently, N. Rai et al.[7] have described clinical findings of 319 patients presenting with retinal lesions in the fovea highly suggestive of solar retinopathy, underscoring that only 51% of the examined patients could report history of sun-gazing in the past.

Therefore, although diagnosis may be straightforward in patients with acute loss of vision following prolonged sun exposure, patients who present months to years later may present more of a diagnostic challenge.

Histopathological studies[19,20] have demonstrated that the RPE and outer segments of the photoreceptor layer are most susceptible to solar damage and that the outer lamellar defects visible a few weeks after the insult are probably caused by focal loss of the photoreceptors, which occurs maximally few days after exposure. [21] OCT findings of chronic solar retinopathy seem to correlate with histopathology observations; Garg et al.[22] described small, hyporeflective space in the outer retinal and retinal pigment epithelium (RPE) layers and thinning of the overlying fovea in a series of four patients with documented history of chronic solar retinopathy. Similar lesions involving both the outer retina and the inner retinal pigment epithelium had been previously described by Huang et al.[23] in three cases of photic maculopathy.

In our study, eyes with biomicroscopic and OCT findings consistent with diagnosis of OLD, strongly resembling chronic solar retinopathy, were further analyzed using FAF. FAF imaging of the ocular fundus depends both on stimulated emission of light from molecules, chiefly lipofuscin, in the RPE cells and on light attenuating factors such as macular pigment, cone and RPE pigment in the fovea.[12-14]

In the eyes with OLD at the fovea we observed a characteristic autofluorescence pattern consisting of small areas of decreased autofluorescence, corresponding in location to the OLD, surrounded by a ring of faint increased autofluorescence in the context of the relatively hypoautofluorescent macular area.

To the best of our knowledge this pattern has not been associated with other maculopathies and doesn't fit any of the physiologic FAF phenotypes previously described in the literature.[12-14]

Interestingly, similar FAF findings (i.e. small focal areas of decreased autofluorescence corresponding to the OLD visible on biomicroscopy) have been reported in association

to documented cases of solar retinopathy (Dr. L.A. Yannuzzi, lecture on Fundus Autofluorescence, 14 April 2005, Fermo, Italy and Prof A. Bird, personal communication, 2007).

The well demarcated hypoautofluorescent areas corresponding to the OLD visible on funduscopy and on OCT could be secondary to lack of RPE cells or more likely to dead photoreceptors with consequent reduced accumulation of lipofuscin in the underlying RPE cells as it has been demonstrated in previous studies evaluating the effect on FAF of light-induced damage to the retina.[15]

Conversely, the halo of relatively increased signal in the context of macular hypofluorescence might be due to lipofuscin accumulation secondary to disturbances occurring at the level of photoreceptors-RPE interface. We didn't observe any obvious alterations of the layers corresponding to the junction between outer and inner photoreceptor segments and to the RPE-choriocapillaris complex in the juxtafoveal areas on OCT 3 images, but it is possible that new generation Spectral-domain OCTs, with their improved image resolution and sensitivity, may reveal subtle changes undiscovered by time-domain devices.

Another hypothesis could be that the increased signal is secondary to alterations in the distribution or to reduction of the luteal pigment in the perifoveal region. It has been demonstrated that any defect involving the foveal tissue, including lamellar macular holes and macular pseudoholes[24] , may alter the degree of foveal FAF by decreasing the amount of masking macular pigment, and therefore increasing the foveal FAF. Studies revealing alterations affecting optical density measurement and lateral distribution of macular pigment in eyes with features similar to those reported in this paper, might validate such hypothesis.

In our series a substantial thinning of the foveal region was observed in eyes with foveal OLD in comparison to the fellow, unaffected eyes with a resulting area of relatively increased autofluorescent signal. It remains uncertain if such retinal thinning is

secondary to the damage inflicted to the retina by the light or if it represents just an anatomical peculiarity making these eyes more susceptible than contralateral to develop light-induced injuries.

Depletion of macular pigment leading to an increased FAF signal has been described in eyes with idiopathic macular telangiectasis[25], where an impaired trafficking and/or storage of lutein and zeaxanthin has been proposed to play a role; a similar deficiency could in theory make some eyes more prone to develop solar retinopathy.

We acknowledge that the present study presents several limitations: the sample of patients analysed is small; all the patients are females whereas solar maculopathy equally affects both genders; furthermore, although the series presented shares clinical and OCT features commonly seen in association to solar retinopathy, no patient reported history of solar exposure. Consequently, we cannot exclude that the macular changes herein described, are actually secondary to undefined forms of macular dystrophies or to resolved, subtle chorioretinal inflammatory disease (for example acute retinal pigment epitheliitis or acute macular neuroretinitis). Self-limiting forms of central serous chorioretinopathy might be considered in the differential diagnosis too. Another possibility is that the OLD were caused by a vitreous-retinal interface syndrome; however, none of the eyes presented in this series had evidence of vitreous-macular traction; the stability of symptoms and funduscopic appearance of the lesions during the follow-up, makes, in our opinion, an interface syndrome for the pathogenesis of these OLD, at least unlikely.

In conclusion our findings suggest that presumed idiopathic OLD at the fovea might be secondary to chronic solar retinopathy. Further studies evaluating larger number of patients affected by this relatively uncommon maculopathy are warranted.

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COMPETING INTEREST

None declared.

FIGURE LEGENDS

Figure 1. (A and B) Case 1, color fundus photograph showing small, reddish, sharply demarcated spots at the fovea in both eyes (BE). (C and D) Optical coherence tomography reveals interruption of the thin hyperreflective band corresponding to the junction between inner and outer segment of photoreceptors and of the inner portion of the hyperreflective band corresponding to the retinal pigment epithelium/choriocapillaris complex (arrows); note the partial posterior vitreous detachment not involving the fovea in the right eye. (E and F) Fundus autofluorescence shows well demarcated, hypoautofluorescent areas corresponding in location to the outer lamellar defect observed clinically surrounded by an irregular halo of hyperautofluorescence in the context of the greater hypofluorescent area of the macular region of BE.

Figure 2. (A) Case 3, color fundus photograph showing a small red lesion at the fovea and surrounding drusen-like deposits in the right macula. (B) Funduscopy in the left eye (LE) is unremarkable; (C) optical coherence tomography (OCT) in the right eye reveals interruption of the thin hyperreflective band corresponding to the junction between inner and outer segment of photoreceptors and of the inner portion of the hyperreflective band corresponding to the retinal pigment epithelium/choriocapillaris complex (arrow). (D) OCT shows a normal retinal structure in the LE; (E) fundus autofluorescence (FAF) of the RE shows a well-demarcated, hypoautofluorescent lesion corresponding in location to the outer lamellar defect observed clinically; the drusen-like lesions seen ophthalmoscopically are hyperautofluorescent. (F) FAF findings in the LE appear

consistent with type II phenotype of distribution of macular pigment in the foveal and perifoveal area as described by Trieschmann M. et al. [14]

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