CASE REPORT

Fundus Autofluorescence Features of Optic Disc Pit Related Maculopathy

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ABSTRACT

Fundus autoflorescence (FAF) is a new investigational tool used to identify lipofuscin distribution in the retinal pigment epithelium (RPE) cell monolayer. It has recently been used to analyze age-related macular degeneration, central serous chorioretinopathy, retina telangiectasia and diffuse and macula retina dystrophies. Our study reports its use to evaluate two different patterns of presentation in optic disc pit related maculopathy, and findings suggest that FAF as an investigational tool can give valuable information for evaluation of the RPE – photoreceptor complex and the metabolic condition of RPE in this disorder.

Keywords: Fundus autofluorescence, maculopathy, optic disc pit

INTRODUCTION

Fundus autoflorescence (FAF) is a new investigational tool used to identify lipofuscin distribution in the retinal pigment epithelium (RPE) cell monolayer.[1] Lipofuscin (LF) accumulation occurs because RPE cells have no mechanism for degrading or transporting LF granules that come from phagocytosis of photoreceptor outer segments into the extracellular space.[1] Excessive LF accumulation is a final common pathway in the pathology of many hereditary retina disorders as well as age-related macular degeneration (ARMD).[2] Several studies define FAF features of ARMD, central serous chorioretinopathy, retina telangiectasia and diffuse and macula retina dystrophies.[1,3-8] These features give useful clinical and prognostic information, making FAF a desired day-to-day clinical tool.

Fundus autoflorescence signals can be detected using 3 different systems, the Delori's fundus spectrophotometer, modified Fundus Camera and the confocal scanning laser ophthalmoscope.[1] These systems are able to bypass anterior segment autoflorescence structures to produce single high-contrast monochromatic images of the retina FAF characteristics. In a normal FAF image of the retina, the optic nerve head is dark (black) due to the absence of RPE (and hence no lipofuscin). The blood vessels also appear dark due to absorption of the light by blood. The FAF signal is reduced at the fovea owing to absorption by luteal pigments such as lutein and zeaxanthin in the neurosensory retina and in the parafoveal area, the signal tends to be higher even as its intensity is decreased relative to peripheral retinal areas.[1]

Optic disc pits were first described in 1882 by Wiethe as small gray-white oval depressions found at the optic nerve head.[9] Associated maculopathy is reported in between 25% and 75% of patients[10] and may be a cause of significant visual impairment. Better understanding of the structural changes occurring in the macula is now possible with the advent of the optical coherence tomography (OCT) scan.[11] For evaluation of the RPE – photoreceptor complex and the metabolic condition of RPE, however, OCT and FAF photography need to be used together. The main source of FAF is lipofuscin pigment that is a metabolic substrate generated after phagocytosis of the outer segments of photoreceptors by RPE in older age groups.[12,13]

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The density of autofluorescence is directly proportional to the amount of lipofuscin. In the condition of physical detachment between RPE and the outer segments of the retina, there may be more autofluorescence related with the precipitation of these structures on the outer retinal surface and subretinal space, because of the deterioration of phagocytosis of the outer segments of photoreceptors.[14]

This study uses the modified Zeiss P450 fundus camera pictures to describe the FAF characteristics of optic disc pit related maculopathy in two patients with different presentations. Fundus cameras utilize high sensitivity monochrome (black and white) digital sensors with different wavelength combinations than used for angiography. Monochrome sensors are more light sensitive than their color counterparts and have all pixels available for exposure by the relatively limited band of wavelengths generated by autofluorescence. The digital fundus camera technique first described by Spaide and Klancnik,[8] employs an excitation filter centered at 580 nm and a barrier filter centered at 695 nm. These wavelengths are shifted toward the red end of the spectrum to avoid unwanted short-wavelength autofluorescence from the crystalline lens. The FAF excitation wavelength of 580 causes minimal excitation of fluorescein and the barrier filter centered at 680 nm blocks the emission peak of fluorescein (520 nm), giving a high-quality fundus image with minimal interference from other autofluorescent anterior segment structures.

CASE REPORTS

Case 1

A 19-year-old student presented with 10-month history of poor vision in the left eye noticed during a routine eye examination. Medical history and systemic review of symptoms were not contributory. Examination revealed visual acuities of 6/6 in the right eye (OD) and 6/36 in the left (OS) with no improvement on refraction. Anterior segment findings were normal with round and reactive pupils in both eyes (OU). Intraocular pressures were 11 mmHg OD and 12 mmHg OS by applanation tonometry. Both pupils were round and reactive to light.

Fundoscopy revealed flat retinae with cup to disc ratio of 0.4 OU, a temporal optic disc pit at 3 O'clock with associated maculopathy OS. These findings were confirmed on OCT and fundus fluorescein angiography (FFA). A small foveal area of <500 microns of hyper autofluorescence is seen on FAF imaging, indicative a localized schisis cavity OS [Figure 1].

Case 2

A 29-year-old gravida 3 para 2 presented with 2 months history of bumping into rough areas of the road

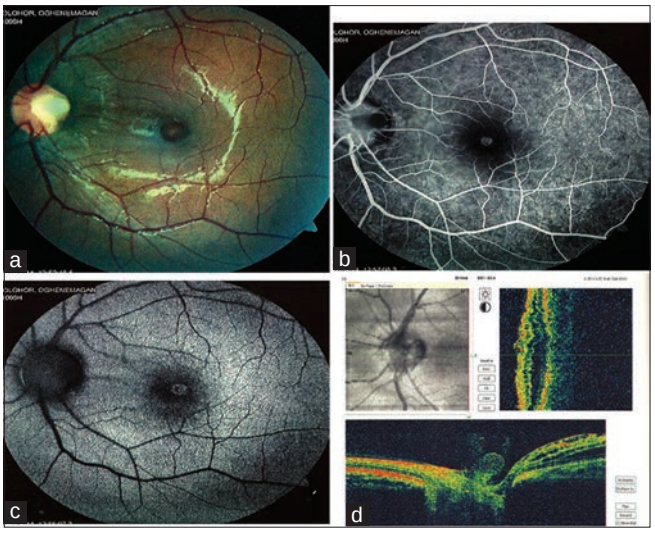

Figure 1: Fundus autofluorescence, fluorescein angiography and optical coherence tomographic features of optic disc related maculopathy in a 19-year-old student. (a) Fundus picture shows an optic disc pit with associated maculopathy. (b) The central hyperfluorescence is seen on fluorescein Angiography. (c) Hyperautofluorescence is seen in the small schisis cavity in the macula. (d) Optical coherence Tomography confirms a schisis cavity within the macula

while driving. She is not a known glaucoma patient or spectacle wearer. Examination revealed visual acuities of 6/9 OD best corrected to 6/6 and 6/36 OS best corrected to 6/24. Anterior segment findings were normal with intraocular pressures of 10 mmHg by applanation tonometry and round pupils reactive to light OU.

Fundoscopy revealed flat retinae with cup to disc ratio of 0.4 OU, an inferotemporal optic disc pit at 4 o'clock with associated maculopathy OS. These findings were confirmed on fundus photography and OCT. FFA was not done because the patient was pregnant. A large area of about 2 disc diameters (3000 microns) of hypo autofluorescence with multiple yellowish subretina deposits are seen on FAF imaging [Figure 2].

DISCUSSION

Following Lincoff *et al*.[15] description in 1988 of the pathology of macula changes associated with optic disc pits, different surgical techniques have been described, aiming at relieving the tangential traction and enhancing adhesions between the RPE and the neurosensory retina at the edge of the disc pit.[16] Inducing a posterior vitreous detachment after pars plana vitrectomy, with or without fluid gas exchange or laser photocoagulation to the edge of the optic disc pit, is well-described with encouraging visual outcomes.[17-19]

However, despite reported success, long-term outcomes are still dependent partly on such factors as duration of the maculopathy, achievement of anatomic

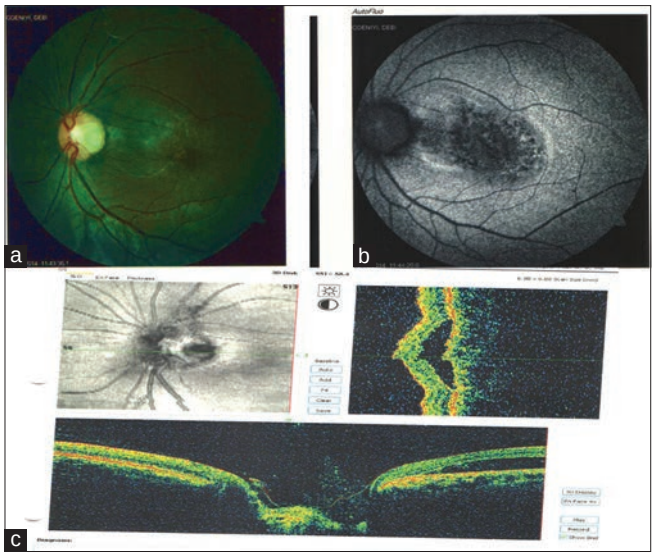

Figure 2: Fundus autofluorescence and optical coherence tomographic features of optic disc related maculopathy in a 29-year-old patient. (a) Fundus picture shows an optic disc pit with associated maculopathy. (b) Hypoautofluorescence is seen in large schisis cavities in the macula. (c) Optical coherence tomography confirms a schisis cavity within the macula

success and functional recovery of photoreceptors that may take up to a year to occur.[20] FAF imaging defines two different patterns in our patients. Case 1 has a focal area of hyper autofluorescence on FAF and hyperfluorescence on FFA. The size is about 100 microns and corresponds to a localized schisis cavity seen on OCT. The hyper autofluorescence appearance is due to lipofuscin accumulation, likely because the macula pathology is of recent onset. The long standing pathology in Case 2 appears hypoautofluorescent from failure of lipofuscin accumulation. There are multiple yellowish subretina deposits and a much larger schisis cavity on OCT. Accumulation of intensely autofluorescent subretinal deposits in the area of a serous retinal detachment has been reported in the FAF images of eyes with optic disc pit maculopathy.[21] Subretina deposits might have accumulated because, the serous detachment separated the photoreceptor outer segments from the underlying RPE and potentially, inhibited proper phagocytosis of shed outer segments. Accumulation of the shed outer segments would allow accrual of fluorophores under the outer retinal surface. Alternate explanations for the formation of the subretinal precipitates would be that they represent depositions of fibrin or lipoprotein.[22,23] Deposition of subretinal material has been described in another similar retinopathy, central serous retinopathy (CSR),[24] but in Case 2 above, there is an associated optic disc pit, with the OCT revealing a continuation between the pit and the serous detachment. This distinguishes it from a CSR.

The autofluorescent appearances of these schisis cavities seem to be related to the height and the duration of the lesion. In a recent case series, reported cases had large areas of hypo autofluorescence on FAF, these areas gradually changed to normal and then hyper autofluorescence as the schisis cavities reduced in height and gradually closed up after surgery.[25] FAF changes thus provide useful information about improvements in the function of the RPE – photoreceptor complex, and help to monitor return in function of the RPE postoperatively or rarely when closure occurs spontaneously. However, FAF photography needs to be used together with OCT images, to obtain more valuable information. Such information is important for counseling, in order for patients to have realistic expectations for outcomes. Further studies need to be done to elucidate the clinical importance of hypo and hyperautofluorescent appearances of maculopathy associated with optic disc pits.

CONCLUSION

Fundus autofluorescence as an investigational tool can give valuable information for evaluation of the RPE – photoreceptor complex and the metabolic condition of RPE in maculopathy associated with optic disc pits.

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