Unusual Fundus Autofluorescence Image in Acute Syphilitic Posterior Placoid Chorioretinitis Associated with Visual Field Defect

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Abstract

Acute syphilitic posterior placoid chorioretinitis (ASPPC) is an uncommon but very characteristic ocular syphilitic manifestation. ASPPC presents itself as an oval or circular lesion at the posterior pole, corresponding to outer retinal and choroidal inflammation. Here we describe a case of a 42-year-old man diagnosed with ASPPC, presenting an unusual fundus autofluorescence (FAF) pattern. A mottling aspect of hypo and hyperautofluorescence was observed, different from the typical homogeneous hyperautofluorescence described in the literature. Six months after treatment, a new FAF revealed shrinking, but not disappearing, of the hyperfluorescent spots, and a visual field test performed elucidated a defect, which corresponded to the ASPPC lesion.

Keywords: Fundus Autofluorescence; Syphilis; Acute Syphilitic Posterior Placoid Chorioretinitis; Visual Field Defect

Introduction

Acquired syphilis is a chronic sexually transmitted disease caused by the spirochete Treponema pallidum. Ocular syphilis is a relatively uncommon manifestation, believed to occur most often during the secondary or latent stages of infection and affecting either the anterior or the posterior segment [1]. Acute syphilitic posterior placoid chorioretinitis (ASPPC) is one of the distinctive clinical patterns of ocular syphilis that assist to its rapid diagnosis [2], as the disease is known as “the great imitator” and, for that, is a differential diagnosis for any uveitis. ASPPC is usually unilateral and presents as an oval or circular, placoid, yellowish lesion on the posterior pole, corresponding to outer retina and inner choroidal inflammation [2-4]. Multiple lesions were also described. Fluorescein angiography (FA), indocyanine green angiography (ICGA), optical coherence tomography (OCT) and fundus autofluorescence (FAF) are some of the ancillary exams that help characterizing this entity. Early-phase FA shows either hypofluorescence or faint hyperfluorescence, often with scattered hypofluorescent spots in the area corresponding to the yellowish opacification, a pattern referred to by some as leopard spotting [3]. Mid- and late-phase FA typically shows progressive hyperfluorescence. Early-phase ICGA reveals hypofluorescent areas corresponding to the lesion, changes that persist into later phases of the study. In some patients, late ICGA hyperfluorescence in affected areas has also been described [4]. Subretinal fluid, thickening of the neurosensory retina and RPE-choriocapillaris complex, ELM and IS/OS bands disruption are some of the Sd-OCT findings already reported [1,2].

Here we present an unusual FAF pattern in ASPPC with a correspondent visual field impairment.

Case Report

A 42-year-old man referred a 1-month history of photopsia on his right eye (RE) and sudden vision loss over the following 10 days. His best-corrected visual acuity (BCVA) was 20/50 on that eye. Fundoscopy disclosed an oval yellowish lesion at the posterior pole of the RE (Figure 1). Fluorescein angiography revealed initial hypofluorescence, with late diffuse leakage. OCT showed RPE and outer retina irregularities at the lesion area (Figure 2). FAF presented a mottling aspect of hypo and hyperautofluorescence (Figure 3). Serum workup
was positive for syphilis, and the patient received intravenous crystalline penicillin for 14 days. HIV tests were negative. BCVA improved to 20/20 after less than a month, but the patient remained with a superior RE visual field defect (Figure 4) that corresponded to the inferior FAF changes. Six months later, a new FAF revealed shrinking, but not disappearance of the hyper and hypoautofluorescent spots (Figure 5).

**Figure 1:** Fundus photograph showing a yellowish oval lesion at the posterior pole affecting the outer retina, with the classic aspect of ASPPC.

**Figure 2:** OCT reveals RPE and outer retina irregularity presented only at the lesion area; neurosensory retina remains intact.

**Figure 3:** FAF presents as a mottling area of hypo and hyperautofluorescence corresponding to the lesion, which contrasts to the most typical presentation seen in ASPPC (hyperautofluorescent area).

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**Figure 4:** Superior visual field defect observed at the 24-2 VF Humphrey, which corresponds to the inferior FAF defect.

**Figure 5:** FAF image 6 months after treatment; note a reduction in the defect compared to the previous exam.
Discussion

The typical large placoid pale yellowish lesion of ASPPC has been postulated to be the result of an active inflammatory reaction at the level of the choriocapillaris–pigment epithelial–retinal photoreceptor complex. Although histopathologic analysis is unavailable, ophthalmoscopic, angiographic, and fundus autofluorescence findings support this hypothesis [1].

The clinical and angiographic features of ASPPC are quite distinctive. Few other conditions produce inflammation so evenly distributed over such a uniquely circular or oval area of the outer retina and inner choroid [1]. Imaging studies not only contribute to diagnosis, but can also be helpful in distinguishing placoid syphilis from other entities, such as sarcoidosis, Vogt–Koyanagi–Harada, posterior scleritis, viral retinitis, and lymphoma [2]. Even a prognostic value can be attributed to ancillary exams, as a study showed that the presence of vascular staining in fluorescein angiography of placoid patients was correlated to improvement in visual acuity after treatment – proving the reversible nature of this type of ocular inflammatory reaction in the context of ocular syphilis [5].

Fundus autofluorescence of ASPPC was first described in 1999 [6]. The active placoid lesion was hyperautofluorescent and normalized with treatment. This pattern of FAF is suggestive of lipofuscin accumulation at the level of the RPE–choriocapillaris complex, and is the most typical presentation of placoid syphilis [1, 7, 8]. Thus, the mottling aspect of hypo and hyperautofluorescence observed in our subject is not a common feature, but has already been described once [9].

Visual field defects are shown in ASPPC [10], although their correspondence with FAF lesions has not been reported so far. Evidence suggests that ASPPC is caused by deposition of soluble immune complexes at the RPE–choriocapillaris [4]. This deposition may lead to variable RPE damage, causing different FAF patterns, and some of these damages may be permanent, even after correct syphilis treatment. The hypoautofluorescent areas observed in our patient, even 6 months after treatment, may suggest a permanent damage of RPE cells, which could cause photoreceptor impairment, ultimately explaining the related VF defect and the complain of superior visual scotoma.

Despite VF defects, visual acuity recovery is common after treatment of ASPPC, as we observed in our subject (final BCVA 20/20). A report of 16 new cases and a review of 44 previously reported ASPPC cases (most of them male, middle-aged, and one-third HIV positive) noted vision improvement in most eyes regardless of the patient’s HIV status, except three eyes previously treated with intravitreal triamcinolone acetonide that developed RPE changes in the macula [1].

Conclusion

Here we described the case of 42-year-old man with typical clinical features of ASPPC in the RE: an oval yellowish lesion at the posterior pole, correspondent to late diffuse leakage on FA and to RPE and outer retina irregularities on OCT. However, his FAF image with mottling areas of hypo and hyperautofluorescence, was different from the classic pattern described on the literature (homogeneous hyperautofluorescence at the placoid area). The lesion did not disappear even 6 months after correct treatment, so the possibility of persistent RPE and photoreceptor damage should be kept in mind. In our case, it translated as a complain of superior scotoma, confirmed by VF testing. Besides aiding on the correct diagnosis, knowing this unusual FAF presentation also has a prognostic value, as hypoautofluorescent areas suggest permanent damage.

In this time of alarming increase in incidence of syphilis in developed and developing countries, the ophthalmologist plays an important role. Despite of supporting the placoid syphilis diagnosis, ancillary exams like FAF, FA and OCT can also be useful in following response to antibiotic therapy [11].

Bibliography

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