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Case Report

Ophthalmology Section

Bilateral Chorioretinitis as Syphilis Presentation: Multimodal Characterization and Therapy Response

MÓNICA MIRANDA LOUREIRO[1], PAULA ALEXANDRA SEPÚLVEDA[2]

ABSTRACT

Ocular involvement of syphilis may occur in any stage of infection. We report a case of bilateral syphilitic chorioretinitis, in an immunocompetent patient, as the presenting feature of a tertiary syphilis we performed a multimodal characterization through Fundus Autofluorescence (FAF), Fluorescein Angiography (FA) and Spectral-Domain Optical Coherence Tomography (SD-OCT).

Keywords: Fluorescein angiography, Neurosyphilis, Ocular syphilis, Optical coherence tomography

CASE REPORT

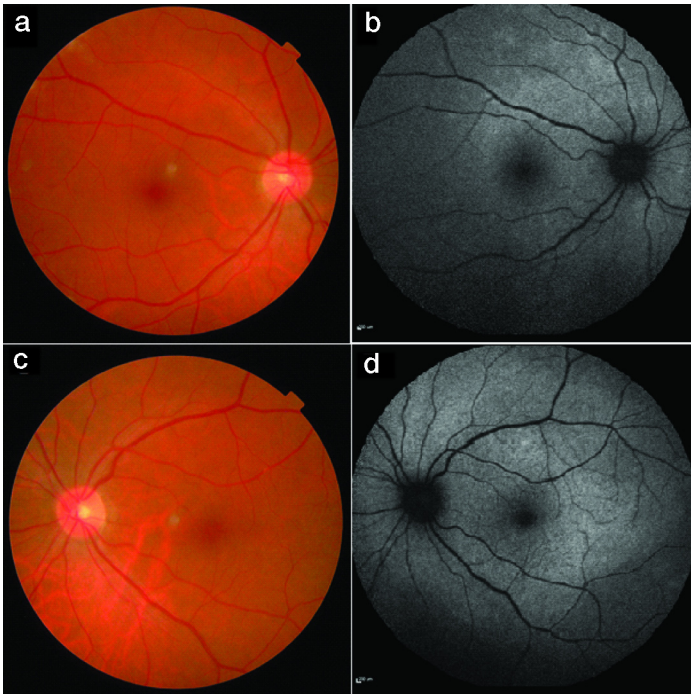
A 47-year-old caucasian man presented bilateral and progressive vision decline since two weeks. His medical and family history was unremarkable; he was not taking any medication and he denied other symptoms. He reported a history of multiple sexual partners in the past few years, with inconsistent use of barrier protection.

On the ophthalmic examination, his Best-Corrected Visual Acuity (BCVA) was of 20/25 in the right eye and 20/63 in the left eye. The anterior segment, the tonometry and the pupillary reflexes were normal.

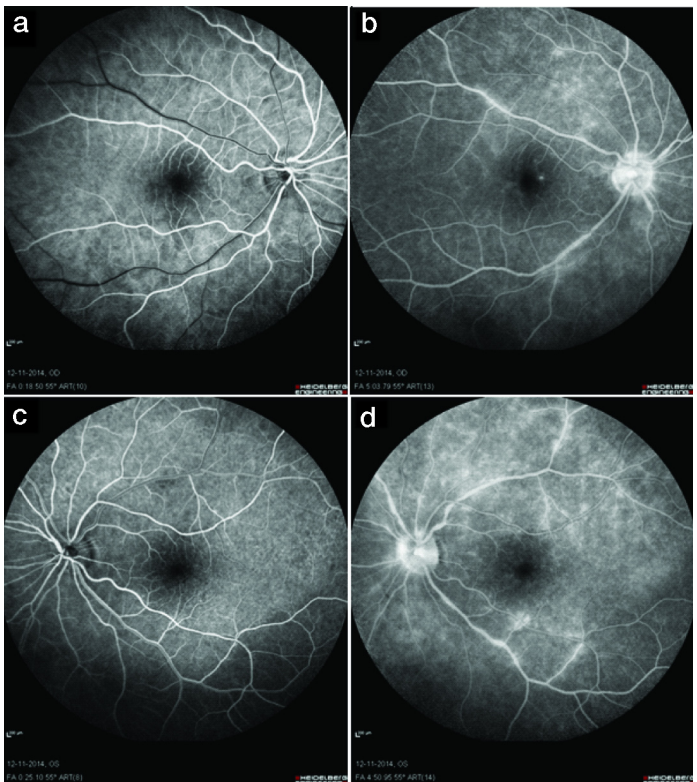
Fundus examination showed discrete pigmented changes involving the macula and the Fundus Autofluorescence (FAF) evidenced a mottled pattern of hyperautofluorescence in the corresponding area [Table/Fig-1]. The Fluorescein Angiography (FA) presented progressive hyperfluorescence of the affected area, which extended beyond the temporal vascular arcades; as well as, leakage from the retinal vasculature and the optic disc [Table/Fig-2]. The Spectral-Domain Optical Coherence Tomography (SD-OCT) revealed a grainy aspect of the Retinal Pigment Epitheliun (RPE) and absence of the outer segments of photoreceptors [Table/Fig-3], especially in the left eye.

Screening blood tests were requested and the VDRL (Venereal Disease Research Laboratory) returned reactive, followed by positive treponemal-specific tests (FTA-ABS and TPHA), confirming active infection. The patient was HIV negative and his HIV status remained negative three months following initial presentation. All other infectious and autoimmune screening tests were negative. A lumbar puncture was also performed and the cerebrospinal fluid cell count was 23 white blood cells per microliter with 96% of mononuclear cells, the glucose level was 67 mg/dl and the protein level was 37 mg/dL, VDRL was reactive and TPHA was positive. The cranial computerized tomography performed was normal.

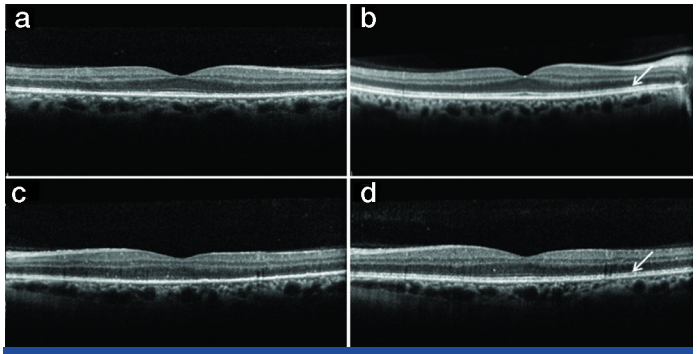
Therefore, he was diagnosed with Acute Syphilitic Posterior Placoid Chorioretinitis (ASPPC) and neurosyphilis and prompt



[Table/Fig-1]: Fundus photograph and autofluorescence of the eyes.
Color fundus photograph of the right (a) and left eye (c) showing subtle pigmented changes, better evidenced in the respective autofluorescence images of the right (b) and left eye (d) as a mottled pattern of hyperautofluorescence.



[Table/Fig-2]: Fluorescein angiography of the eyes.
Early hyperfluorescence corresponding to the area of affected retina in the right (a) and left eye (c). Progressive hyperfluorescence, with perivascular (vasculitis) and disc (papillitis) leakage in the late frames of the right eye (b) and left eye (d).



[Table/Fig-3]: Spectral domain-optical coherence tomography of the eyes, before and after treatment.
Absence of the outer segments of photoreceptors and grainy aspect of the retinal pigment epithelium before treatment of the right (a) and left eye (b). Restitution of the outer segments of photoreceptors layer (arrow) and normalization of the retinal pigment epithelium after the treatment of the right (c) and left eye (d).

treatment with intravenous penicillin G 24 million units every day for two weeks was instituted.

At the first month of follow-up, there was a fantastic visual acuity recovery with a BCVA of each eye of 20/20, coupled with restoration of the outer segments of photoreceptors and normalization of the retinal pigment epithelium in the SD-OCT [Table/Fig-3].

DISCUSSION

Syphilis, known as the great imitator, has a myriad of manifestations [1]. Syphilitic uveitis is a rare cause of uveitis (1.6- 4.5% of cases), occurring in up to 5% of cases who have progressed to tertiary syphilis [2]. The ASPPC, described by Gass, is an uncommon presentation that is probably related with an inflammatory reaction to the *Treponema pallidum* that affects the RPE [3].

Although challenging to diagnose because of the similarities with other clinical entities, this patient presented characteristic findings, such as mottled pattern of hyperautofluorescence in the FAF [4,5], hyperfluorescence of the lesion in the FA [3,6,7] and grainy aspect of RPE with segmental loss of the outer segments of photoreceptors in the OCT [4,8]. A recent study also evidenced the presence of a small amount of fluid under the fovea in the first days that resolves in a week [8]. Though this finding was not observed in our case, the patient reported the onset of symptoms for more than a week, so the fluid might already be gone.

Uncommon characteristics were the extension of the lesion beyond the temporal vascular arcades, the association with vasculitis and papillitis, and the bilateral ocular involvement in a patient who showed immunocompetence [9].

Since the optic nerve and retina are considered extensions of the central nervous system, the patient was submitted to lumbar puncture, which allowed the detection of syphilitic neurological involvement and the prompt treatment with recommended high-dose of intravenous penicillin G.

After the treatment, the visual acuity and the retinal appearance returned to normal. This fact is curious and showed the reversibility of the macular lesion, suggesting that the absence of the outer segments of photoreceptors is not an anatomical irreversible damage and instead, may correspond to structural disorganization that determines a decrease of the reflectivity of the line. Probably, the microbial eradication allows the metabolic functions of the RPE to resume their normal activities, restoring the optical properties of the layer of the photoreceptors.

CONCLUSION

It is essential to recognize the multimodal characteristics of the ASPPC and subsequently perform analysis of cerebrospinal fluid in all patients. This is the key to diagnose a neurosyphilis in the absence of other manifestations, allowing the treatment with excellent outcomes.

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PARTICULARS OF CONTRIBUTORS:
1. Medical Doctor, Department of Ophthalmology, Centro Hospitalar Vila Nova de Gaia/Espinho, Portugal. Institute of Biomedical Sciences Abel Salazar, Oporto University, Portugal.
2. Medical Doctor, Department of Ophthalmology, Centro Hospitalar Vila Nova de Gaia/Espinho, Portugal.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Mónica Miranda Loureiro,
R. Dr. Miguel Assunção Lopes, nº 15, 3ºB. 4400-698 Vila Nova de Gaia, Porto, Potugal.
E-mail: monicamloureiro@gmail.com

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