Spontaneous resolution of acute syphilitic posterior placoid chorioretinitis: reappraisal of the literature and pathogenetic insights

Abstract

Acute syphilitic posterior placoid chorioretinitis (ASPPC) is a rare clinical manifestation of ocular syphilis. Spontaneous resolution of this condition has been reported in a few cases. The aim of this manuscript is to report an additional case and to discuss the possible pathogenesis of this condition by reviewing the current evidence on this subject.

A 45-year-old man presented to us with decreased vision in the right eye secondary to a placoid macular lesion. Fourteen days after presentation, there was a dramatic improvement of the vision, and multimodal retinal imaging showed almost complete spontaneous resolution of the placoid lesion. Syphilis serology turned out positive and a diagnosis of ASPPC was made. The pathogenesis of ASPPC is unclear, and there is contrasting evidence about the role of the cellular immune system. Since this condition may resolve spontaneously before systemic antimicrobial treatment, the presence of a placoid macular lesion should raise a high suspicion of ASPPC in order to make a timely diagnosis and to avoid progression of untreated syphilis.

Keywords: acute syphilitic posterior placoid chorioretinitis, natural course, pathogenesis, retinal imaging, syphilis

Introduction

Syphilis is a sexually transmitted infection caused by the spirochete bacterium *Treponema pallidum* [1]. Syphilis is a re-emerging and rising infection in the developed world. In up to one-quarter of patients with syphilis, ocular involvement manifests at any time during the disease course. Ocular syphilis may precede the diagnosis of systemic disease in up to one-half of cases [2]. Ocular syphilis, known as “the great masquerader”, may affect almost every structure of the eye and has a broad spectrum of presentation, including, among others, interstitial keratitis, optic neuropathy and posterior uveitis, the latter commonly represented by chorioretinitis [3], [4].

In 1988, de Souza et al. [5] reported three young patients with “unilateral central chorioretinitis” as manifestation of ocular syphilis. Two years later, Gass et al. [6] reported six additional similar cases. They concluded that this condition was a separate clinical entity, and coined the term “acute syphilitic posterior placoid chorioretinitis” (ASPPC).

ASPPC is defined by the presence of one or more placoid, yellowish, outer retinal lesions, typically involving the posterior pole and the mid-periphery of the retina near the temporal vascular arcade [6]. ASPPC may have a unilateral or bilateral involvement with a presenting visual acuity ranging from 20/20 to no light perception [7]. The advent of multimodal imaging (MMI) of the retina, especially of spectral domain optical coherence tomography (SD-OCT), has made it possible to report pathognomonic features of ASPPC, which include punctate hyperreflectivity in the choroid, disruption and loss of the ellipsoid zone, nodular irregularity of the retinal pigment epithelium, and transient localized subretinal fluid [8], [9]. Since patients with ASPPC usually receive prompt antimicrobial treatment after serologic results, little is known about the natural course of the disease. To the best of our knowledge, only 5 cases of ASPPC with spontaneous improvement have been reported [10], [11], [12], [13].

We report the natural course and the multimodal retinal imaging features of an additional case, and discuss the pathogenetic implications and the importance of early recognition of this rare clinical entity.

Case presentation

A 45-year-old man with no relevant past medical history presented to the eye casualty service complaining of sudden onset central ‘white ring’ and decreased vision in the right eye (RE) over the past seven days. Best-corrected visual acuity (BCVA) was 6/12 in the right eye and
Figure 1: Fluorescein angiography (FA) and indocyanine green angiography (ICGA) of acute syphilitic posterior placoid chorioretinitis in the right eye at presentation. (a) Early frame of FA shows hypofluorescence (yellow arrowhead) of the placoid lesion which appears hyperfluorescent in the late frames (b), (c), (d) ICGA shows hypocianescence of the placoid lesion (green arrowhead) throughout the whole examination.

Figure 2: Colour fundus photograph (CFP) and fundus autofluorescence (FAF) changes of acute syphilitic posterior placoid chorioretinitis in the right eye over time. (a) CFP shows a yellow placoid lesion (white arrowhead) at the posterior pole which gradually fades 1 week after presentation (b) and 2 weeks after presentation (c). FAF shows increased AF in correspondence of the placoid lesion at presentation (d) with gradual normalisazion of the AF 1 week after presentation (e) and 2 weeks after presentation (f).

6/6 in the left eye (LE). Intraocular pressure was 14 mmHg in both eyes. Examination of the RE showed no cells in the right anterior chamber and 1+ vitreous cells; fundus examination revealed a yellow placoid lesion involving the macular area with no signs of vasculitis or retinal necrosis. Examination of the LE was unremarkable. MMI of the retina including colour fundus photograph, fundus autofluorescence, SD-OCT, fluorescein angiography and indocyanine green angiography are presented in Figure 1, Figure 2, and Figure 3.

The medical history was carefully reviewed; the patient admitted to be addicted to poppers and cocaine, and reported promiscuous homosexual activity over the last months. He denied intravenous drug use and any systemic symptoms such as headache, skin rash, nausea, weight loss, cough, or night sweats. A complete laboratory work-up was ordered, including TB QuantiFERON-TB testing.
Figure 3: (a) Spectral domain optical coherence tomography (SD-OCT) scan of the right eye at presentation shows disruption of the ellipsoid zone (white asterisks), nodular thickening of the retinal pigment epithelium (yellow arrowheads) and punctate hyperreflectivity in the inner choroid (white arrows). SD-OCT scan 1 week after presentation (b) and 2 weeks after presentation (c) show gradual recovery of the ellipsoid zone and retinal pigment epithelium.

syphilis serology and human immunodeficiency virus (HIV) antibodies. Seven days after presentation, the patient reported spontaneous improvement in the vision of the RE, and BCVA improved to 6/9 in the RE and was stable in the LE. Full blood count, liver function test, kidney function, angiotensin-converting enzyme level were within normal range, HIV antibodies were negative. However, results for QuantiFERON-TB testing and syphilis had not been available yet. MMI revealed spontaneous improvement of the placoid lesion (Figure 2, Figure 3).

Two weeks after presentation, BCVA further improved to 6/6 in the RE and MMI showed signs of early resolution of the placoid lesion. Laboratory results returned negative for QuantiFERON-TB testing, and positive for venereal disease research laboratory test and fluorescent treponemal antibody testing. Therefore, a definite diagnosis of ASPPC was made, and the patient was promptly referred to the Infectious Disease Department for systemic treatment with penicillin.

Discussion

ASPPC is a rare clinical manifestation of ocular syphilis. Although the pathophysiology of ASPPC is not completely understood, timing and characteristics of SD-OCT findings may be the reflection of the sequence of disease events [9]. It has been suggested that circulating T. pallidum organisms may enter the choroidal circulation, giving the choroidal hyperreflective pinpoint lesions seen on SD-OCT; subsequent access to the outer retina may give a variable amount of subretinal fluid and impaired photoreceptor function expressed by disruption of EZ seen on SD-OCT [9]. However, the role played by the cellular immune system in the pathogenesis of this condition remains controversial. While it was initially suggested that ASPPC is secondary to immunocompromised status such as in HIV-positive patients [5], [6], [14], it was later described in both immunocompetent and immunocompromised individuals [7], [9], [15].

Of note, no differences have been found in terms of clinical characteristics and long-term visual outcome in HIV-positive versus HIV-negative patients with ASPPC [7]. To the best of our knowledge, spontaneous resolution of ASPPC before initiation of systemic antimicrobial treatment has been reported in 5 cases [10], [11], [12], [13]. The first cases were described in 2015 by Ji et al. [10], who reported two HIV-negative patients with ASPPC which spontaneously improved 10 days (for the first case) and 3 weeks (for the second case) after presentation. In the same year, Aranda et al. [11] reported an HIV-positive patient on anti-retroviral therapy for 4 years and CD4+
be the manifestation of different pathogenetic pathways. There is contrasting evidence about the role of the im-

Conclusions

There is contrasting evidence about the role of the immune system in the pathogenesis of ASPPC which may be the manifestation of different pathogenetic pathways that ultimately lead to an inflammatory response driven by the presence of the spirochete. Since this condition may resolve spontaneously before antimicrobial treatment, the presence of a placoid macular lesion should raise a high suspicion of ASPPC, as the ophthalmologist may be the first to diagnose syphilis in the patient. Indeed, timely diagnosis and antimicrobial treatment are essential to preventing the progression of syphilis which may include irreversible visual damage [21].

Notes

Competing interests

The authors declare that they have no competing interests.

Informed consent

We obtained written informed consent from the patient for publishing the information and images.

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Corresponding author:
Giuseppe Casalino, MD, FEBO
Oftalmico Hospital, ASST Fatebenefratelli Sacco, Piazzale Principessa G. Clotilde 3, 20121 Milano, Italy
giuseppe.casalino@gmail.com

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