Case report

Acute syphilitic posterior placoid chorioretinopathy: An infectious or autoimmune disease?

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ABSTRACT

Purpose: To report a case of acute syphilitic posterior placoid chorioretinopathy (ASPPC) that demonstrated partial resolution with immunosuppressive therapy secondary to a misdiagnosis as Behçet’s disease followed by a relapse which was successfully treated with the appropriate treatment.

Observations: A 34-year-old female patient presented to our service with complaints of decreased vision in the left eye (OS). She initially developed similar symptoms seven months prior to presentation and was diagnosed as Behçet’s disease based on the clinical picture of papillitis, vasculitis and placoid chorioretinitis in the posterior pole of OS. She was started on daily oral prednisone 60 mg and weekly methotrexate 10 mg by her rheumatologist. The patient’s ocular symptoms improved one month prior to presentation with resolution of the placoid lesion but persistence of vasculitis and papillitis. At that time, the dose of the prednisone was decreased to 30 mg which resulted in a relapse of the placoid chorioretinal lesions and worsened visual acuity at the time of presentation to us. Extensive laboratory workup demonstrated positive serology for syphilis. A diagnosis of syphilitic placoid chorioretinitis was made and the patient was treated with intravenous penicillin G for 2 weeks. The vitritis, papillitis, and placoid chorioretinitis resolved along with improvement in vision following the treatment.

Conclusions and importance: Ocular findings in syphilis are heterogeneous and may mimic variety of ocular diseases. ASPPC is a rare ocular manifestation of syphilis and its natural course and underlying pathophysiology is not well understood. However, irrespective of the underlying mechanism of the disease, all patients with ASPPC should receive treatment to prevent recurrence and long-term functional damage.

1. Introduction

Syphilis is a sexually transmitted, systemic infection caused by the Gram-negative spirochete Treponema pallidum. It can affect any organ in the body, including the eyes. Known as the ‘great imitator’, syphilis has a wide variety of ocular presentations such as anterior uveitis, intermediate uveitis, interstitial keratitis, chorioretinitis, retinal vasculitis, retinitis, perineuritis, papillitis, retrobulbar neuritis, optic atrophy, and optic nerve gumma.1,2 Therefore, it is recommended to test every patient with ocular inflammation for syphilis.1,2

Since non-specific treponemal test such as venereal disease research laboratory (VDRL) test and rapid plasma reagin (RPR) test normalize during the latent and tertiary stages, testing for syphilis should include both non-specific and specific treponemal tests such as fluorescent treponemal antibody absorption (FTA-ABS) and the micro-hemagglutination assay for T. pallidum (MHA-TP).3

Acute syphilitic posterior placoid chorioretinopathy (ASPPC) is a rare ocular manifestation of syphilis that was first described by Gass et al.4,5 It is characterized by the presence of one or more yellowish placoid retinal lesions that are typically present in the macula. However, the natural course of ASPPC is not well understood.6

In the index report, we describe a case of ASPPC that demonstrated partial resolution with immunosuppressive therapy secondary to a misdiagnosis as Behçet’s disease followed by a relapse which was successfully treated with the appropriate anti-infectious treatment.

2. Case report

A 34-year-old female patient was admitted to our service with complaints of pain and blurriness of vision in the left eye (OS) for 10
extensive area of chorioretinitis with vitritis and superficial retinal precipitates in OS (Fig. 1). These findings appeared more active compared to those documented in medical records and photographs taken seven months ago when she presented with large yellowish placoid macular lesions with papillitis and vasculitis as seen on fluorescein angiography (FA) (Fig. 2). The retinal lesions resolved a month before the presentation with residual pigment epithelial changes; however, there was persistence of papillitis and vasculitis (Fig. 3). The biomicroscopy and fundus examination of the OD were unremarkable. Various laboratory tests including complete blood count, fasting blood glucose level, blood urea nitrogen, creatinine, HIV antibody, Hepatitis B and C serology, Bartonella IgM/IgG, Toxoplasma IgM/IgG, anti-nuclear antibodies had been performed in the past and were within normal limits. In our clinics, we repeated the laboratory tests with the addition of VDRL and FTA-ABS tests for syphilis as a possible etiology. Both tests for syphilis yielded positive results. The patient also underwent a lumbar puncture and cerebrospinal fluid was negative for VDRL.

Upon the diagnosis of syphilis, she was treated with 4 million units of intravenous penicillin G every 4 hours for 2 weeks. Her steroid therapy was discontinued. After the completion of intravenous penicillin G treatment, her symptoms improved. On ocular examination, BCVA demonstrated improvement to 20/25 in OS. Slit lamp examination demonstrated a decrease in the anterior chamber cells to +0.5 in OS. Dilated fundus examination showed complete resolution of chorioretinitis and improvement of vitreous haze in OS (Fig. 4).

3. Discussion

Syphilis is a sexually transmitted disease caused by the spirochete Treponema pallidum. It is colloquially known as ‘the great imitator’ for its ability to mimic many diseases due to its extensive range of clinical manifestations. Therefore, a high index of clinical suspicion and a low threshold for serological testing can help with early detection and treatment of the disease, allowing us to reverse retinal changes and prevent permanent damage. Posterior segment manifestations of ocular syphilis can include, among others, superficial retinal precipitates, exudative retinal detachment, acute syphilitic placoid posterior chorioretinopathy, papillitis, vasculitis, neuroretinitis, chorioretinitis, and retinitis.

ASPPC is a rare ocular manifestation of syphilis and its natural course is not well known. It is characterized by the development of a placoid macular deposit in the outer retina. Furthermore, the ocular disease can be accompanied by central nervous system and/or mucocutaneous abnormalities, which can mimic other systemic diseases and complicate the diagnosis.

The underlying pathophysiology of ASPPC is not very well understood. Similar to our report, it has been documented in immunocompetent patients. Franco et al. and Ji et al. reported findings of ASPPC in immunocompetent patients with spontaneous resolution without receiving any therapy. On the other hand, our patient was initially treated with immunosuppressive therapy which is often not a suggested therapy for syphilis. Surprisingly, in contrast to the general infectious behavior of worsening with immunosuppression, our case demonstrated improvement in the visual and anatomical outcome as the placoid chorioretinal lesion resolved after steroid therapy. The vasculitis and papillitis, however, persisted. Such treatment outcomes underscore the thought that maybe the underlying mechanism leading to the chorioretinal lesions seen in ASPPC is different than the vasculitis and papillitis seen in syphilitic uveitis which has been thought previously. Additionally, unlike other reports of ASPPC, there was a

increase in leakage in the area of the lesion along with papillitis and vasculitis. (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)
As Souza et al. demonstrated an increase in the level of anti-beta 2 glycoprotein antibodies in a subject with ASPPC. They suggested that these antibodies can potentially lead to choroidal thrombosis and impairment of ocular syphilis, which further highlights the importance of local immunomodulation in the pathogenesis of the disease.

On the other hand, some of the studies have suggested immunosuppression as a major stimulator of ASPPC. Aranda et al. documented a case with ASPPC in an HIV patient which resolved spontaneously as the immune system recovered following anti-retroviral therapy. Zamani et al. reported another case of ASPPC which presented secondary to immunosuppression following corticosteroid therapy. Furthermore, Erol et al. and Song et al. published cases of ASPPC following local intravitreal triamcinolone injections suggesting a role of local immunomodulation in the pathogenesis of the disease. In contrast to the findings of these reports, our patient received corticosteroid and methotrexate for 7 months and demonstrated no worsening of the disease.

Based on these contrasting evidences published in the literature and what we noted in our patient, we suggest that the ASPPC lesions may be a single clinical manifestation of two different underlying pathophysiological pathways. These lesions can either result from direct attack by the spirochete secondary to reactivation from immunosuppression or as a consequence of indirect immune-mediated hypersensitivity.

Despite the complex underlying mechanisms leading to ASPPC, there should be a low threshold to test for syphilis in patients presenting with chorioretinitis. It is important to recognize that the chorioretinal lesions of ASPPC may appear similar to other ocular autoimmune diseases as was seen in our case which was initially misinterpreted and treated as Behçet's disease. Furthermore, all the patients with positive serology for syphilis, even with spontaneous resolution of the chorioretinal lesions, should receive the appropriate anti-infectious treatment for the underlying infection. Otherwise, the ocular syphilis can lead to recurrence of ASPPC as we noted or can lead to a more permanent functional damage via other syphilitic manifestations in the eye as was seen in eyes in which the diagnosis was delayed.

4. Conclusion

ASPPC is a rare ocular manifestation of syphilis and its natural course is not well understood. However, it may have both autoimmune as well as infectious basis in its pathophysiology. Irrespective of the pathogenesis, all patients with ASPPC should receive anti-infectious treatment to treat the underlying disease to avoid recurrence of the disease and permanent functional damage.

Patient consent

Consent to publish this case report has been obtained from the patient in writing.

Conflicts of interest

QDN is a recipient of a Physician Scientist Award from Research to Prevent Blindness, New York, NY, and serves on the Scientific Advisory Board for AbbVie, Bayer, Genentech, Regeneron, and Santen, among others. QDN also chaired the Steering Committee for the RISE and RIDE studies and was on the Steering Committee for the VISTA Study, and other studies sponsored by Genentech and Regeneron.

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Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.ajoc.2019.03.002.

Abbreviations

ASPPC Acute Syphilitic Posterior Placoid Chorioretinopathy
BCVA Best Corrected Visual Acuity
FA Fluorescein Angiography
FTA-ABS Fluorescent Treponemal Antibody Absorption
MHA-TP Micro-Hemagglutination Assay for T. pallidum
OD Right Eye
OS Left Eye
RPR Rapid Plasma Reagin
VDRL Venereal Disease Research Laboratory

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