Case Report

Unilateral focal lesions in the macula as an early presentation of syphilis

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Abstract: A 55-year-old male complained of right eye blurry vision for 3 days. His best-corrected visual acuity (BCVA) was 0.2 for the right eye and 1.0 for the left eye. Anterior segment and vitreous body examinations of both eyes were normal. Yellowish-white focal lesions in the macula of the right eye were observed and subtly changes of lesions were found along the superotemporal and inferotemporal arcades in the macula two days later. Fluorescein fundus angiography (FFA) revealed slight fluorescent leakage from the lesions in the macula of the right eye, and segmental venous leakage and optic disc hyperfluorescence were observed in both eyes. Indocyanine green angiography (ICGA) demonstrated that the lesions in the macula of the right eye had hypofluorescence at a late stage and spectral domain optical coherence tomography (SD-OCT) imaging of the macula showed focal impairment of the inner segment and outer segment (IS/OS). The blood investigation indicated a positive treponema pallidum hemagglutination assay (TPPA) and a rapid plasma reagin test (RPR) of 1:32. After antisyphilitica treatment for 6 weeks, the yellowish-white lesions had vanished and the BCVA was 1.2 followed by restoration of the IS/OS for the right eye, with an RPR of 1:4. In conclusion, ophthalmologists should alert unilateral focal lesions in the macula may be the first sign of syphilis. Prompt treatment is highly effective in resolving vision.

Keywords: Macula; syphilis; fluorescein fundus angiography; indocyanine green angiography; chorioretinitis; posterior uveitis

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Introduction

Syphilis is a bacterial infection caused by the spirochete Treponema pallidum (1). The first signs and symptoms of syphilis may be ocular, which can occur at any stage of syphilis (2). Manifestations of ocular syphilis can include interstitial keratitis; posterior uveitis; retinal vasculitis; chorioretinitis; and retinitis (3,4). Chorioretinitis is a common clinical presentation of syphilis and a cause of painless severe vision loss (5,6). Acute syphilitic posterior placoid chorioretinitis (ASPPC) characterized by large, yellow-white geographic lesions involving the macula is considered to be a distinct clinical presentation of ocular syphilis (7-9). However, focal lesions in the macula usually lead to a diagnostic challenge. Herein, we reported a case of unilateral focal lesions in the macula in a patient who presented with clinical and laboratory findings consistent with a diagnosis of syphilis.

Case presentation

A 55-year-old male presented with acute painless unilateral blurred vision for 3 days without discharge, photophobia and photopsia. The patient was generally healthy and had no chronic medical conditions. There was no previous history of eye diseases, trauma and surgery. The best-corrected visual acuity (BCVA) was 0.2 for the right eye and 1.0 for the left eye. Intraocular pressure was 16 mmHg in both eyes. The corneas were clear and the pupils were equally

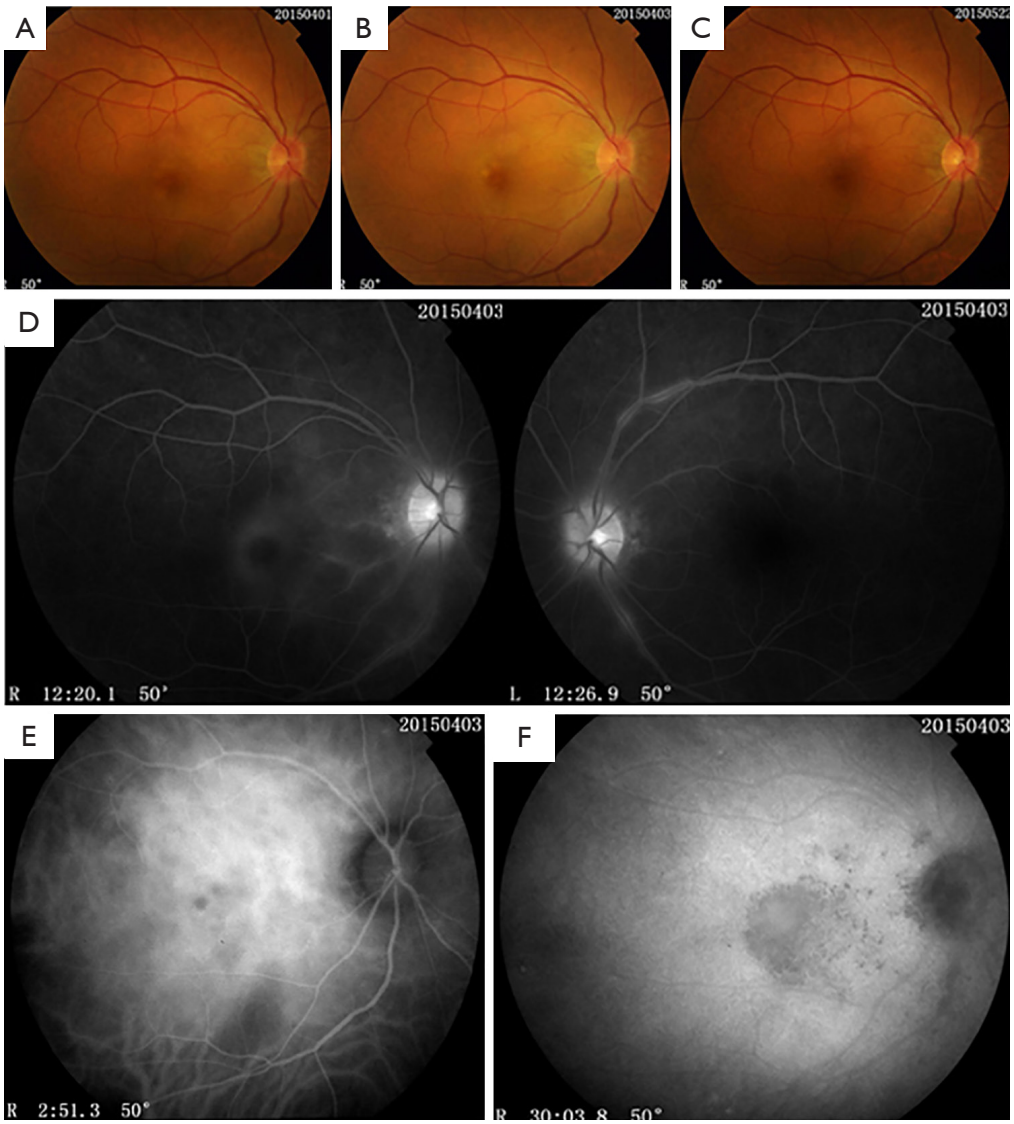

Figure 1 Eye examinations of the patient. (A) Fundus color photograph showed yellowish-white focal lesions of the right eye in the macula at presentation; (B) showed subtly changes along the superotemporal and inferotemporal arcades in the macula two days after presentation; (C) showed lesions vanished after treatment; (D) fluorescein fundus angiography (FFA) demonstrated slight fluorescent leakage in the macula of the right eye, and segmental venous leakage and optic disc hyperfluorescence were observed for both eyes; (E) indocyanine green angiography (ICGA) indicated that the lesions in the macula of the right eye had spotty hypofluorescence at early stage; (F) the hypofluorescence was significant at late stage .

round in both eyes. The anterior segment of both eyes was normal without evidence of relative afferent papillary defect in either eye. There were no inflammatory cells in the vitreous of both eyes. In the macula, yellowish-white focal lesions of the right eye were observed (*Figure 1A*). Two days later, subtly changes of lesions were found along the superotemporal and inferotemporal arcades in the macula (*Figure 1B*) and BCVA was 0.12, while the posterior of the left eye was normal. Fluorescein fundus angiography (FFA) demonstrated (*Figure 1*) slight fluorescent leakage in the macula of the right eye, and segmental venous leakage and optic disc hyperfluorescence were observed in both eyes (*Figure 1D*). Indocyanine green angiography (ICGA) indicated that the lesions in the macula of the right eye had spotty hypofluorescence at early stage and the hypofluorescence

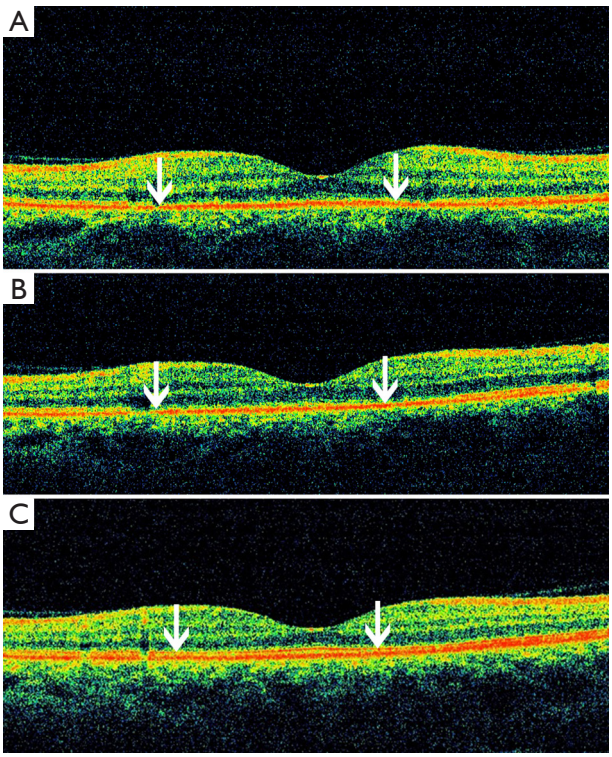

Figure 2 Spectral domain optical coherence tomography (SD-OCT) for the case. (A) The impairment (white arrows) of the inner segment and outer segment (IS/OS) before treatment; (B) the impairment (white arrows) of the IS/OS persisted two weeks after treatment; (C) restoration (white arrows) of the IS/OS six weeks after treatment.

was significant at late stage (*Figure 1E,F*). Spectral domain optical coherence tomography (SD-OCT) imaging of the macula for the right eye showed focal impairment of the inner segment and outer segment (IS/OS) (*Figure 2A*), while OCT imaging of the macula for the left eye was normal.

Systemic general examination and the peripheral blood showed normal. Chest X-ray and MRI of the brain were normal. Laboratory examinations showed human immunodeficiency virus (HIV) antibodies, cytomegalovirus IgM, toxoplasma IgM antibodies, and herpes simplex virus IgM antibodies were negative. C-reactive protein (CRP) and erythrocyte sedimentation rate were normal. All other autoimmune screening tests were negative. Serum glucose level was 5.9 mmol. The blood investigation indicated the peripheral blood a positive treponema pallidum hemagglutination assay (TPPA) and a rapid plasma reagin test (RPR) of 1:32. Lumbar puncture was recommended to evaluate for CSF antibodies, but the patient declined this invasive test. After the diagnosis was confirmed to be ocular syphilis, the patient was recommended a treatment of benzathine penicillin intramuscular injection of 120 million units bilaterally once a week. Two weeks after initial treatment, visual improvement to 0.2, while OCT changes persisted in right eye (*Figure 2B*). After antisyphilitic treatment for 6 weeks, the yellowish-white lesions had vanished (*Figure 1C*) and the BCVA was 1.2 followed by restoration of the IS/OS for the right eye (*Figure 2C*), with an RPR of 1:4.

Discussion

The ocular presentation of syphilis is proved to be very different among affected patients, which makes the diagnose more challenging. There are certain features considered to be characteristic of syphilitic uveitis. Ground glass retinal opacification associated with retinal vasculitis is considered to be characteristic for syphilitic uveitis (2) and another distinctive feature is ASPPC characterized by large, yellow-white geographic lesions involving the macula (7). In syphilitic posterior uveitis, inflammatory changes in the choroid, as well as in the retina, are common. Chorioretinitis is the most common posterior segment involvement of ocular syphilis (10). Baglivo *et al.* postulated that lesion of ASPPC was focused on choroid (11). RPE impairment and vasculitis were secondary (12). In this case, unilateral focal lesions in the macula was first found. Then, subtly changes of focal lesions were observed along the superotemporal and inferotemporal arcades in the macula 2 days after presentation. We also observed the changes of retinal vasculitis and optic disc dyeing for both eyes. It is not sure whether macular lesions of this case was a manifestation of posterior uveitis or an early stage of the discoid lesions. More cases need to observe to find special features and further evolution of affecting the macular and optic disk with syphili.

Focal lesions in the macula usually give rise to a diagnostic difficulty. In up to 40% of reported cases, posterior uveitis (choroiditis, retinitis, chorioretinitis, retina vasculitis) is more commonly associated with an infectious cause with pathogens that include syphilis, toxoplasmosis, and cytomegalovirus (13). Other differential diagnoses include central serous retinopathy, viral retinitis, and punctate inner choroidopathy. For this case, according to clinical and laboratory findings, the macular lesions of syphilis could be confirmed. The macular was easy to be affected that was probably because of the redundant circulation of macular.

In this case, ICGA demonstrated that the lesions of

the macula had significant hypofluorescence at a late stage. Some authors postulated that hypofluorescence resulted from deposition of degraded material from the RPE and photoreceptor segments (14). We considered the hypofluorescence may be because of inflammation infiltration, or choroid capillary blockage or both effects. ICGA could be used to investigate lesions and supply information for diagnosis. According to SD-OCT imaging, we observed the impairment of IS/OS without thickening and hyperreflective nodularity of RPE in the area corresponding to the fundus lesion , which were not consistent with the findings of ASPPC (8). OCT can provide more information about the pathophysiology of the disease. For this patient, vision improved to 1.2 after treatment followed by restoration of the IS/OS. So OCT could be a noninvasive method for follow-up.

In conclusion, unilateral focal lesions in the macula may be the first sign of syghilis. Physicians should be aware that syphilis serology should be routinely done in every case of uveitis that requires investigation. A correct diagnosis is important to prompt treatment. Prompt treatment can result in good visual recovery for syphilis and can prevent potential irreversible complications.

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None.

Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

Informed Consent: Written informed consent was obtained from the patient for publication of this manuscript and any accompanying images.

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