

A Case of Syphilitic Outer Retinitis Mimicking Acute Zonal Occult Outer Retinopathy

Dear Editor,

Syphilis is a sexually transmitted, systemic infection with increasing prevalence. Ocular syphilis may develop at any stage of the disease process and can involve any ocular structure [1]. Ocular syphilis mimics any type of inflammatory process without pathognomonic manifestations and often leads to misdiagnosis, resulting in permanent vision loss [1,2]. Syphilitic uveitis involving the posterior segment has been reported as several conditions [1]; however, to our knowledge, no previous study describing syphilitic outer retinitis has examined the condition by spectral-domain optical coherence tomography (SD-OCT). Herein, we report a case of syphilitic outer retinopathy, mimicking acute zonal occult outer retinopathy (AZOOR).

A 59-year-old man presented with a 2-week history of blurred vision in the right eye. Visual acuity was 20 / 30 in right eye and 20 / 20 in left eye. Slit-lamp examination and funduscopy were normal (Fig. 1A). Fundus fluorescein angiography showed no abnormality in early and late phases (Fig. 1B). However, fundus autofluorescence imaging showed hyperfluorescence around the macula, while SD-OCT revealed a diffuse disruption of the photoreceptor inner segment-outer segment (IS-OS) junction line in the macula in the right eye (Fig. 1C and 1D). Multifocal electroretinography (ERG) revealed reduced amplitudes in the centermost and inferior retinal areas which corresponded to the area of visual field defect on Humphrey visual field (HVF) perimetry, and to the area of disrupted photoreceptors at the IS-OS junction line on SD-OCT (Fig. 1E and 1F). Full-field ERGs were normal and symmetric. The patient's medical history and laboratory tests were unremarkable; serologic tests for syphilis were not performed.

We initially diagnosed him with AZOOR, based on acute loss of visual acuity, funduscopy, and ERG findings [3]. The patient received 15 mg/day oral prednisolone for 1 month, which was then tapered to 10 mg/day for 2 months. Since vision improved to 20 / 25 and photoreceptor disruption was partially restored after treatment (Fig. 1G), we

added oral methotrexate (15 mg/wk). However, at 5 months, vision decreased to 20 / 30, vitreous opacity developed, and photoreceptor disruption progressed on SD-OCT (Fig. 1H). At 6 months, the photoreceptor disruption worsened and his vision deteriorated to 20 / 50 (Fig. 1I). Complete serologic tests were performed, which diagnosed him as active syphilis (both the Venereal Disease Research Laboratory assay and fluorescent treponemal antibody-absorption test were positive). We diagnosed him with syphilitic outer retinopathy in right eye and discontinued immunosuppressant therapy and intramuscularly administered 2.4 million units of benzathine penicillin G three times per week. One month later, vision had improved to 20 / 30, and the disruption of photoreceptor on SD-OCT was partially restored (Fig. 1J). At 3 months after antibiotics therapy, vision had improved to 20 / 20 and SD-OCT showed complete restoration of photoreceptor function (Fig. 1K). Multifocal ERG and HVF revealed complete normalization (Fig. 1L and 1M).

AZOOR is a rare disease of unknown etiology and is characterized by focal degeneration. While associated with both ocular and systemic diseases [3,4], syphilis has never been reported as mimicking AZOOR in Korea. Given the clinical presentation and the lack of syphilis as differential diagnosis, we diagnosed AZOOR without exploring other possible causes. Based on this case, we suggest that syphilitic retinitis should be considered as a differential diagnosis for AZOOR, and that meticulous serologic tests should be performed prior to a diagnosis of AZOOR. In this case, SD-OCT was helpful in the evaluation and monitoring of photoreceptor damage, and the results were correlated with visual function.

Although syphilitic retinitis has increased with an increasing global incidence of syphilis [5], its pathogenesis remains controversy [2,4]. However, given the response to antibiotics therapy, our case substantiates that syphilitic retinitis occurred via direct invasion of spirochetes into the lesion, rather than by an autoimmune reaction as a result of molecular mimicry. Photoreceptor disruption completely recovered with antibiotic therapy; timely administration of systemic antibiotics is thus warranted for rescuing vision in eyes with syphilitic retinitis. Furthermore, SD-OCT accurately reflected the status of photoreceptor disruption at diagnosis, as well as efficacy of therapeutic intervention.

Acute syphilitic posterior placoid chorioretinitis should be also suspected as a differential diagnosis. In our case, however, we ruled out acute syphilitic posterior placoid chorioretinitis based on normal fluorescein angiography results.

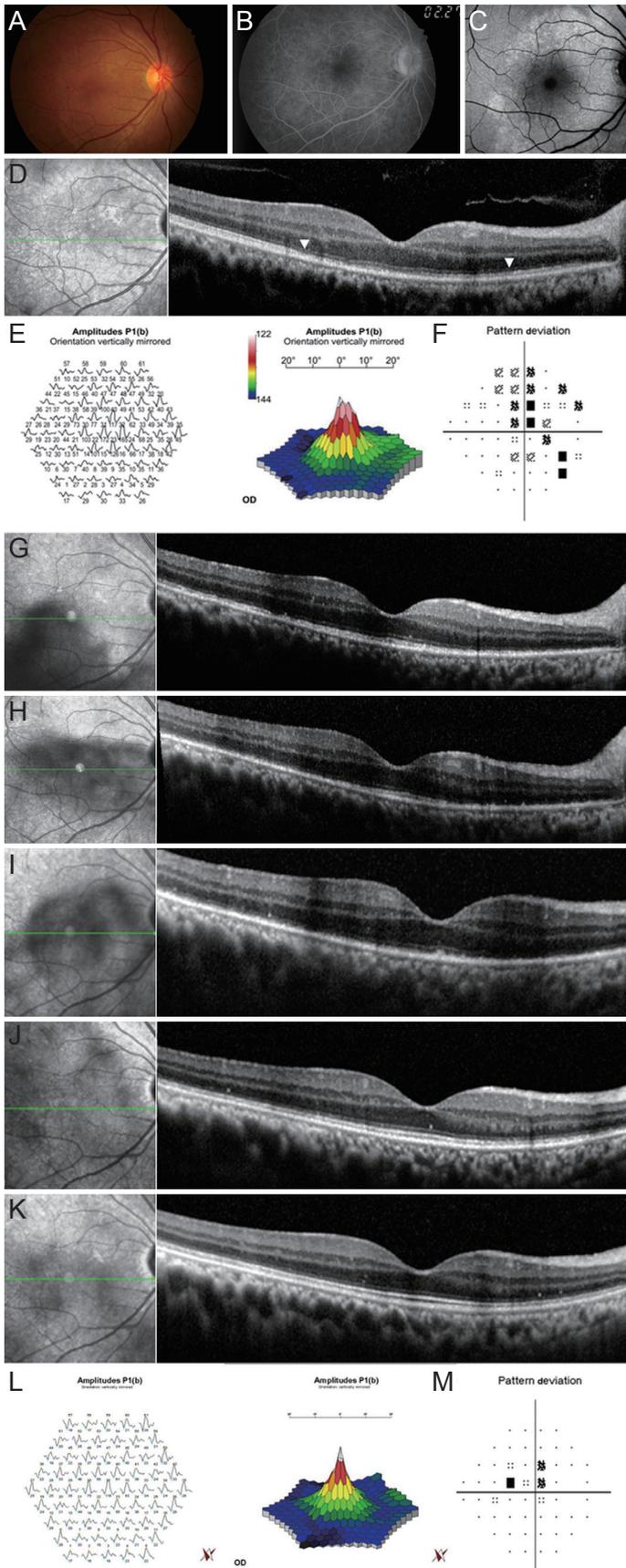


Fig. 1. Summary of clinical and diagnostic features. At initial visit, color fundus photography (A) and fundus fluorescein angiography (A) showed normal appearance. However, fundus autofluorescence (FAF) imaging (C) and spectral-domain optical coherence tomography (SD-OCT) (D) showed hyperfluorescence and diffuse disruption of the photoreceptor inner segment-outer segment (IS-OS) junction line (white arrowheads indicate the border of the irregular photoreceptor IS-OS junction area) in the centermost area and inferior retinal areas of macula corresponding to the area of reduced amplitudes in the multifocal electroretinography (ERG) (E). Humphrey visual field (HVF) perimetry showed that the visual field defects corresponded to SD-OCT, FAF imaging, and multifocal ERG (F). Although the photoreceptor disruption was partially restored at 3 months after treatment (G), the disruption became aggravated gradually thereafter (H), at 5 months after treatment (I), at 6 months after treatment. (J) One month after antimicrobial treatment with penicillin, restoration of the photoreceptor IS-OS junction occurred, and vision was restored to 20 / 30 in right eye. Three months after antimicrobial therapy, vision improved to 20 / 20 in right eye and SD-OCT revealed complete restoration of the photoreceptor IS-OS junction (K). Multifocal ERG (L) and HVF (M) showed nearly complete normalization of visual function.

In conclusion, clinical suspicion of syphilitic outer retinitis in patients presenting as AZOOR is crucial for early diagnosis and treatment. Photoreceptor evaluation using SD-OCT can be useful for diagnosis and monitoring response to treatment.

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Conflict of Interest

No potential conflict of interest relevant to this article was reported.

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Bilateral Free Floating Vitreous Cysts with Posterior Embryotoxon

Dear Editor,

A 9-year-old female child presented with diminution of vision in both the eyes and deviation of eyes since birth. The corrected visual acuity was 1 / 60 in both eyes. There was no history of any systemic disorder or trauma. Hirschberg's test showed 20 degrees of exotropia. The prism bar covered test showed 40 prism diopter exotropia. Slit lamp examination revealed an unremarkable anterior segment with no signs of inflammation except for the presence of posterior embryotoxon in both eyes. A spherical, translucent, smooth cyst covered with a brown pigment was noted in posterior vitreous, moving freely with the movements of the eyeball in both the eyes (Fig. 1). The intraocular pressure was 16 and 15 mmHg in the right and left eye respectively.

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Indirect ophthalmoscopy and biomicroscopy with 90D lens revealed a healed chorioretinitis patch in both the eyes. In the right eye, a single oval cyst was identified floating freely in the vitreous, partially masking the underlying retinal vasculature. B-scan ultrasound revealed an echogenic, round-shaped cyst measuring 2 × 1.5 mm, free from surrounding vitreous strands or retina, localized at the posterior vitreous in the right eye, and four similar miniature cysts were found in the left eye. Color doppler revealed absence of vessels passing through the vitreous of both of the globes and within the cysts. There were no signs of retinitis pigmentosa.

Indirect hemagglutinin tests for echinococcus and cysticercosis were negative. Eosinophilia was absent on peripheral blood smear. Based on these findings the patient was diagnosed with bilateral free floating vitreous cysts with posterior embryotoxon.

The first description of a vitreous cyst was by Tansley in 1899. These are classified as congenital and acquired [1]. Congenital cysts are associated with residues of the hyaloid vascular system and are occasionally seen in normal eyes.