

# Akutni sifilitični posteriorni plakoidni horioretinitis: prikaz primera

## Acute syphilitic posterior placoid chorioretinitis: a case report

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sifilis, horioretinitis, fluoresceinska, indocianin green angiografija

**Key words:**

syphilis, chorioretinitis, fluorescein, indocyanine green angiography

**Članek prispel / Received**

21.9.2013

**Članek sprejet / Accepted**

3.11.2014

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**Izvleček**

**Namen:** Predstaviti primer akutnega sifilitičnega posteriornega plakoidnega horioretinitisa (ASPPC), enega od možnih očesnih prezentacij sifilisa, in opozoriti na ponovni porast sifilisa v svetu.

**Metode:** Prikaz klinične slike, izvidov fluoresceinske in indocianin–green angiografije, slik autofluorescence in optične koherenčne tomografije ter poteka obravnave bolnika.

**Rezultati:** 39–letni bolnik se je predstavil kot sicer zdrav moški s 3–dnevno anamnezo meglene vida desnega očesa. Ugotovljeni so bili znaki posteriornega uveitisa. Klinična slika in izvidi slikovnih diagnostičnih preiskav so ustrezali opisu ASPPC, pozitivni serološki test za sifilis pa je potrdil diagnozo. Šele potem, ko je bilo predvideno dodatno testiranje na okužbo s humanim virusom imunske pomanjkljivosti (HIV), je bolnik razkril anamnezo že znane in z antiretrovirusno

**Abstract**

**Purpose:** To present a case of acute syphilitic posterior placoid chorioretinitis (ASPPC), one of the possible ocular manifestations of syphilis, and alert clinicians to the reemergence of syphilis worldwide.

**Methods:** Clinical findings, fluorescein angiography, indocyanine green angiography, autofluorescence imaging, time domain optical coherence tomography features, and management of the patient are presented.

**Results:** A 39–year–old male presented as an otherwise healthy man with a 3–day history of blurred vision in his right eye. Clinical examination revealed signs of posterior uveitis. Clinical features and additional imaging findings were consistent with the description of ASPPC. Syphilis was confirmed with a positive serological test for syphilis. Additional human immunodeficiency virus (HIV) testing was intended and the patient only

terapijo zdravljene okužbe s HIV. Pregled likvorja je pokazal prisotnost neurosifilisa. Dva tedna je prejemal penicilin G intravensko, po čemer se je vidna ostrina normalizirala.

**Zaključki:** Prizadetost oči je lahko tista, zaradi katere bolnik s sifilisom najprej poišče zdravniško pomoč. Ozaveščenost zdravnikov o možnih številnih prezentacijah sifilisa je pomembna za takojšnjo prepoznavo in zgodnje zdravljenje te sicer ozdravljive bolezni, ki pa nezdravljena vodi v hude okvare zdravja in povečuje umrljivost.

then disclosed his medical history of being HIV positive and receiving antiretroviral therapy. Cerebrospinal fluid examination revealed neurosyphilis. He was treated with a 2-week course of intravenous penicillin and his visual acuity recovered fully.

**Conclusions:** Ocular findings may be the presenting feature of syphilis. Physicians' awareness of the variety of clinical presentations of syphilis is important for prompt recognition and early management of this curable disease, which can cause substantial morbidity and increased mortality if left untreated.

## INTRODUCTION

Syphilis represents an important and increasingly prevalent public health concern worldwide (1). According to recent data, an overall rate of 4.9 new cases per 100 000 population was reported in 2011. Compared with 2010, some countries (including Slovenia) reported increases of 20% or more, and 42% of all cases were reported in men who have sex with men (2). A missed diagnosis of syphilis can have serious consequences for the patient. Clinicians therefore must be familiar with its possible clinical manifestations. It can affect almost every organ system, and the eye is no exception (3, 4). Nearly all of the eye structures may be affected and multiple ocular manifestations can occur at any stage of the disease (3, 4). Acute syphilitic posterior placoid chorioretinitis (ASPPC), named by Gass and colleagues in 1990, is a distinctive manifestation of ocular syphilis (5). According to the above authors, the ophthalmoscopic and angiographic appearance of the lesions is sufficiently characteristic to suggest a diagnosis of syphilis. The condition is characterized by unilateral or bilateral, single or multiple, large nonelevated, placoid, pale yellowish, ill-defined, outer retinal and choroidal lesions that often are confluent in the posterior pole and mid periphery of the fundus. The lesions tend to fade centrally and often there is clumping of the retinal pigment epithelium (RPE) in a small leopard-spot pattern (5, 6).

## CASE REPORT

A 39-year-old male presented with a 3-day history of blurred vision in his right eye (RE). At presentation, visual acuity was 0.63 in the RE and 1.0 in the left eye (LE). Clinical examination revealed mild bilateral vitritis and initially discrete yellowish discoloration in the macula of the RE, which over the following 2 days progressed to ill-defined, confluent, yellow-white, subretinal, nonelevated lesions at the posterior pole (Figure 1). The visual acuity of the RE deteriorated to 0.2, but then spontaneously improved to 0.63 in the next 3 days. The anterior segments were quiet, and the posterior segment of the LE was unremarkable.

Figures 2-5 present time-domain optical coherence tomography (TD-OCT), autofluorescence imaging (AF), fluorescein angiography (FA) and indocyanine-green angiography (ICGA) findings in the RE.

The results of a systemic workup for posterior uveitis (complete blood count, erythrocyte sedimentation rate, C-reactive protein, electrolytes, creatinine, blood urea nitrogen, urinalysis, liver enzymes, antinuclear antibody and antineutrophil cytoplasmic antibody testing, Mantoux test, chest X-ray, angiotensin-converting enzyme, serologic screening for toxoplasmosis and Lyme borreliosis) were all normal, except for a positive treponemal serologic test



**Figure 1.** Color photography of the fundus of the RE showing ill-defined, confluent, yellow-white, nonelevated subretinal lesions at the posterior pole.

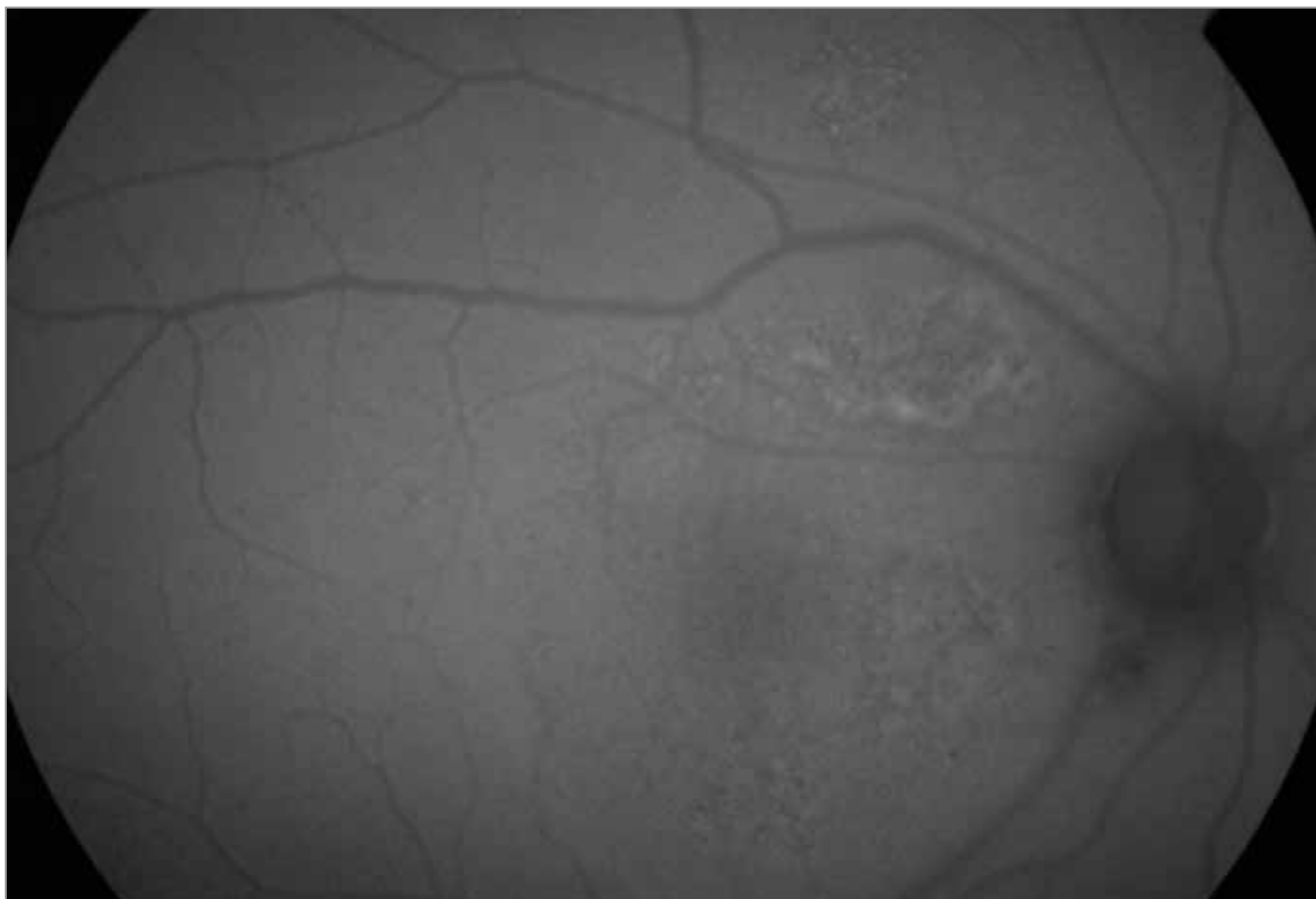
for syphilis (enzyme immunoassay). Additional HIV testing was mandatory. The patient only then disclosed his medical history of being HIV positive and having received antiretroviral therapy for the last 2 years. Lumbar puncture was performed by the infectious disease service; analysis of cerebrospinal fluid revealed neurosyphilis. The patient was treated with a 2-week course of intravenous penicillin G (18 million units/day for 14 days). One month later his visual acuity had recovered fully, although some paracentral visual field defects remained. The vitritis and posterior segment inflammation resolved, leaving mild pigment mottling in the macula of the RE.

## DISCUSSION

Our patient's only complaint was blurring of vision in his RE. He had no systemic symptoms or signs of

syphilis, such as mucocutaneous lesions. The clinical and angiographic findings, findings on literature review and positive syphilis serology were consistent with the description of ASPPC. The typical large placoid yellowish lesion has been postulated to be the result of an active inflammatory reaction at the level of the choriocapillaris-RPE-photoreceptor complex (5,6).

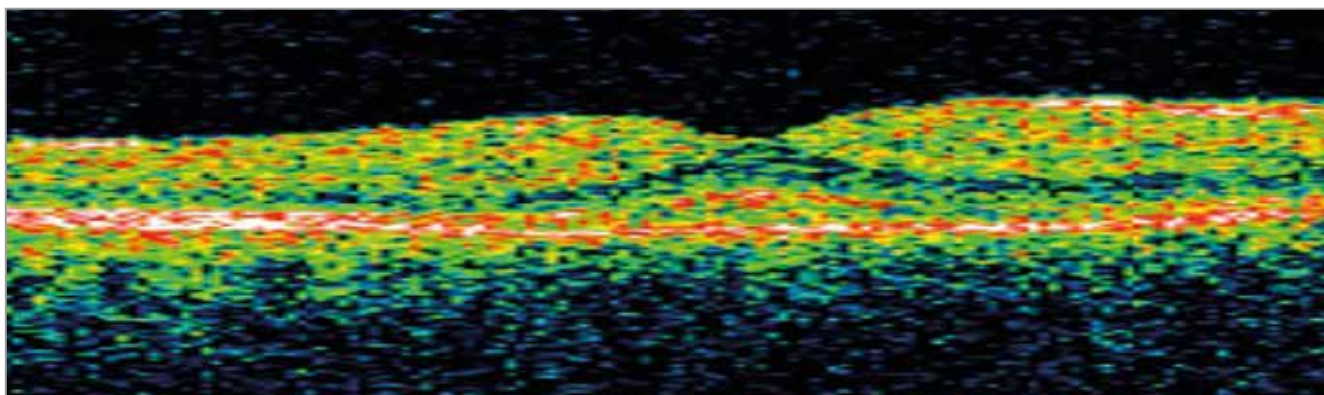
The FA and ICGA findings were similar to those described for other reported cases (5–7). The sometimes observed optic disc edema, retinal haemorrhages, and late staining of the optic disc and retinal veins were not present in this patient, and neither were late hypofluorescent peripheral spots in ICGA, reported by Eandi CM et al. (6–8). The AF imaging revealed irregular hyperautofluorescent patches with granular hypoautofluorescence, in addition to



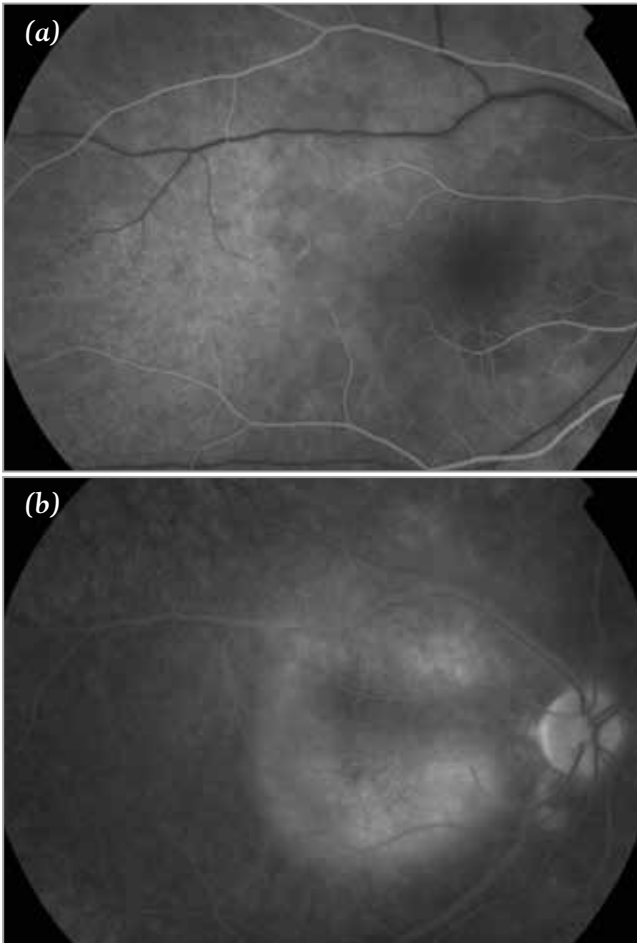
**Figure 2.** Autofluorescence image with slightly increased autofluorescence in the area of the lesion and some irregular hyperautofluorescent patches with granular hypoautofluorescence.

the previously described diffuse hyperautofluorescence in the area of the lesion (6). The TD-OCT showed hyperreflective thickening at the level of the RPE-choriocapillaris complex, without signs of sub-

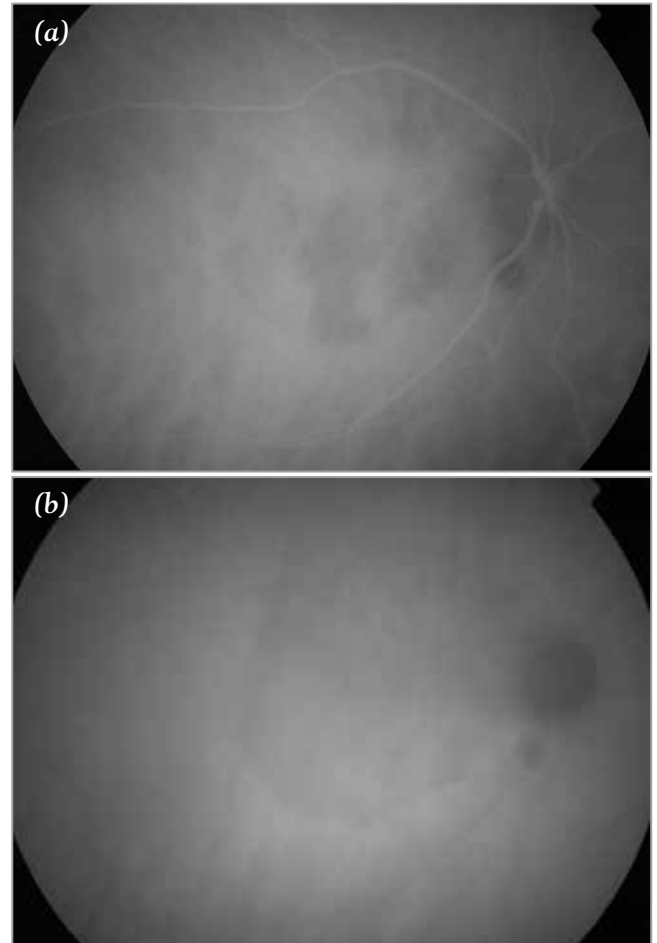
retinal fluid, which has been reported in some cases (9). The changes resolved spontaneously a few days after presentation. Interestingly, the visual acuity also partly improved spontaneously after an initial



**Figure 3.** Time domain optical coherence tomography of the RE showing hyperreflective thickening at the RPE-choriocapillaris complex. The changes resolved spontaneously less than a week after presentation.



**Figure 4.** (a) Early fluorescein angiogram showing irregular confluent hypofluorescent lesions, and (b) progressive hyperfluorescence with leakage and staining of the lesion in the late frame.



**Figure 5.** Indocyanine green angiography revealing (a) early and (b) late hypofluorescence in the area of the lesion.

deterioration but the clinical course of the disease is variable. In some cases it resolves spontaneously, whereas in others widespread areas of chorioretinal atrophy and loss of retinal function occur (6). Prompt and adequate therapy usually results in good visual recovery, as in our patient, who also had HIV coinfection and asymptomatic neurosyphilis (5, 4, 7). The recommended standard therapy for ocular syphilis is the neurosyphilis treatment regimen: 18 to 24 million units of aqueous penicillin G intravenously per day for 10 to 14 days, regardless of the immune status of the patient (3, 4, 10, 11).

To conclude, our aim in presenting this case was to alert clinicians to the reemergence of syphilis and to highlight one of its possible ocular manifestations with a distinctive clinical and angiographic pattern which can assist prompt diagnosis. For definitive diagnosis, laboratory serological confirmation is required. Nevertheless, syphilis should always be considered in patients with unexplained ocular inflammation because its ocular manifestations in general are nonspecific. Therefore, a high level of suspicion is crucial. All patients with ocular syphilis should also be tested for neurosyphilis and HIV coinfection.

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