(05) Multifocal syphilitic chorioretinitis — a case report

Wieloogniskowe zapalenie siatkówki i naczyniówki w przebiegu kiły – opis przypadku

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Summary:

The aim of the paper is to report a case of a 22-year-old male patient with chronic bilateral uveitis and retinitis secondary to syphilis. Until the diagnosis, the patient had been treated symptomatically with ceftriaxone which resulted in visual acuity improvement. The patient was referred to the Sexually Transmitted Disease Clinic for causal treatment after which he did not continue further ophthalmic monitoring. After a year he contacted the Department again due to vision deterioration and a relapse of retinitis and choroiditis was diagnosed. The patient was referred to the Sexually Transmitted Disease Clinicfor causal treatment which he never received as he did not present there.

Since the beginning of the 21st century the incidence of syphilis has significantly increased. Although it is an infectious disease with potentially permanently debilitating effect e.g. on vision, its treatment is not compulsory in Poland. Infectious etiology and primary syphilis should always be considered in patients with progressive retinitis, choroiditis and vitritis.

Key words: Streszczenie:

syphilis, syphilitic chorioretinitis, uveitis.

Celem pracy jest opis przypadku 22-letniego mężczyzny chorego na przewlekłe obustronne zapalenie błony naczyniowej i siatkówki w przebiegu kiły. Do momentu rozpoznania choroby pacjenta leczono objawowo ceftriaksonem podawanym dożylnie, uzyskując poprawę ostrości wzroku. Pacjent został skierowany do poradni wenerologicznej w celu leczenia przyczynowego, później nie zgłosił się już na kontrolne badanie okulistyczne. Po upływie roku ponownie zgłosił się do kliniki okulistycznej z powodu pogorszenia widzenia, rozpoznano wówczas nawrót zapalenia siatkówki i naczyniówki. Pacjent został skierowany na leczenie przyczynowe do poradni wenerologicznej, leczeniu sie nie poddał.

Od początku XXI wieku obserwuje się znaczący wzrost zakażenia prątkiem kily. Chociaż jest to choroba zakaźna, która może prowadzić do kalectwa (również wzrokowego), w Polsce nie ma przymusu jej leczenia. U chorych na zapalenie siatkówki, naczyniówki i ciała szklistego należy zawsze podejrzewać etiologię zakaźną.

Słowa kluczowe:

kiła, zapalenie siatkówki i naczyniówki, zapalenie błony naczyniowej.

Introduction

Syphilis is a chronic, systemic sexually transmitted disease (STD) caused by *Treponema pallidum*. Its natural course encompasses primary, secondary and tertiary syphilis (1). The incidence of syphilis decreased gradually from 1940 to 2000. However, despite the disease being supposedly eliminated, since the beginning of the 21st century a significant increase in the number of new cases has been observed (2). The estimated annual number of new syphilis cases is 12 million, 90% of which occur in developed countries (2). The infection mostly affects men of homosexual orientation. HIV co-infection, which increases the risk of syphilis 5–7-fold, is considered to be one of the risk factors (1, 2).

Ophthalmic and neurological symptoms of syphilis can vary and largely depend on the disease stage. Vision organ is affected in untreated patients with secondary and tertiary syphilis. The main ophthalmic manifestation of the disease is uveitis which affects 2.5 to 5% of infected individuals (1, 3, 4).

Case report

A 22-year-old homosexual man, who had not received ophthalmic treatment before, was admitted to the Department of Ophthalmology as an emergency due to the decreased visual acuity which persisted for 5 months. At admission: BCVA RE = counting fingers (with full light projection), UCVA LE = 0.05, Sn RE = could not read, Sn LE = 0.75 sc, IOP RE = 8 mm Hg, IOP LE = 8 mmHg. The physical examination did not reveal any abnormalities within the anterior segment. In the vitreous chamber a small-cell exudate was present bilaterally as well as single large floaters. During ophthalmoscopy, yellow lesions were revealed within the posterior pole of the left eye at the level of the retinal pigment epithelium (Fig. 1). In the right eye, a "fluffy" band adjacent to the inferior vascular arcade was revealed (Fig. 2). Numerous small white-yellow spots were present around the retina. Some of them were convex with blur margins, while others were flat and saturated with pigment (Fig. 3a., b). Based on these findings, posterior uveitis was diagnosed. The OCT (Fig. 4a., b)

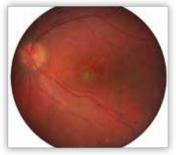


Fig. 1. Left eye – posterior pole. Ryc. 1. Biegun tylny oka lewego.

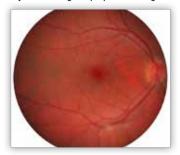


Fig. 2. Right eye – posterior pole. **Rvc. 2.** Biegun tylny oka prawego.

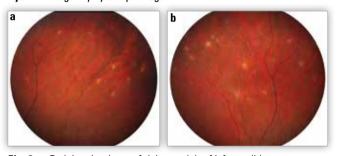
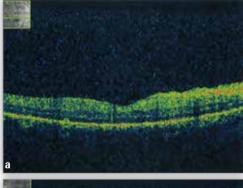


Fig. 3. Peripheral retina – of right eye (a), of left eye (b). Ryc. 3. Obwód siatkówki – oka prawego (a), oka lewego (b).



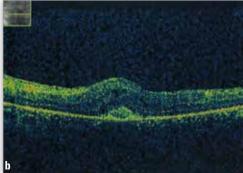
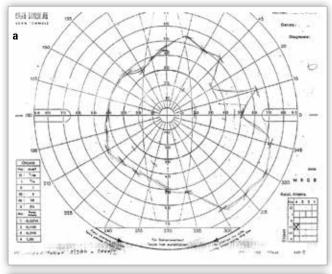


Fig. 4. OCT – of right eye (a), of left eye (b).

Ryc. 4. Badanie OCT – oka prawego (a), oka lewego (b).



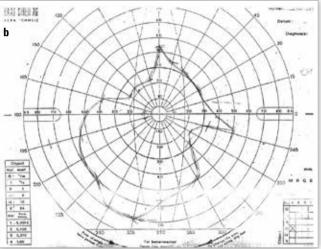


Fig. 5. Kinetic perimetry – of right eye (a), of left eye (b).Ryc. 5. Kinetyczne badanie pola widzenia – oka prawego (a), oka lewego (b).

and the kinetic perimetry (Fig. 5a., b) were performed. During fluorescein angiography a leakage and a contrast medium concentration in the peripapillary area, within the choroid and retina were observed as well as the leakage from retinal vessels (Fig. 6a.— f.). The patient was admitted to the Department of Ophthalmology where he received symptomatic intravenous treatment with ceftriaxone 1.0 g twice a day and hydrocortisone iontophoresis, until the underlying cause of ocular lesions was determined.

During hospitalization the patient was tested for HIV, CMV, Toxoplasma gondii, Borrelia, sarcoidosis and syphilis. HIV screening test was negative. Neither CMV Toxoplasma infection was confirmed. However, positive laboratory test results were obtained in all assays specific for syphilis: USR (+++); VDRL – extremely positive (+++, titre 1/128), FTA – positive (titre 1/32000), FTA-ABS – positive. The patient did not have other symptoms of secondary syphilis. A positive antibody titre for borreliosis was also observed: IgM - 56.02 RU/mI (positive result), IgG - 18.32 RU/mI (questionable result). The infectious disease specialist was ruled out this result as false positive, due to Treponema pallidum infection.

After a two-week treatment, a significant improvement in visual acuity was achieved with a partial resolution

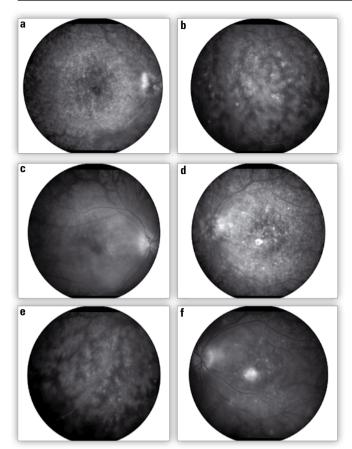


Fig. 6. Fluorescein angiography: of right eye (56 sec) (a), of right eye – peripheral retina (2.42 min) (b), of right eye (11.35 min) (c), of left eye (1.08 min) (d), of left eye – peripheral retina (4.18 min) (e), of left eye (12.12 min) (f).

Ryc. 6. Badanie angiografii fluoresceinowej: oko prawe (56. sekunda) (a), oko prawe – obwód siatkówki (2,42 min) (b), oko prawe (11,35 min) (c), oko lewe (1,08 min) (d), oko lewe – obwód siatkówki (4,18 min) (e), oko lewe (12,12 min) (f).

of the exudate in the vitreous cavity. At discharge, the visual acuity was: BCVA RE =0.8-0.25 Dsph, BCVA LE =0.2-0.25 Dsph, UCVA RE (Snellen) =0.5, UCVA LE (Snellen) =0.5, IOP RE -12 mmHg, IOP LE -12 mmHg. The patient was subsequently referred to the Department of Dermatology and Sexually Transmitted Diseases for causal treatment. The therapy was continued for a month, after which the patient failed to arrive for the scheduled ophthalmic check-up. After a year he was re-admitted to our Department as an emergency with vision deterioration. A relapse of retinitis and choroiditis was diagnosed. The patient was referred to the Department of Dermatology and Sexually Transmitted Diseases in Warsaw for causal treatment, which he never received as he did not present there.

Discussion

In the pre-antibiotics era, syphilis was the main cause of intraocular inflammations (5). Nowadays, it is rare for syphilis to affect the eyes and the disease is known as "the great imitator". There are no specific symptoms specific. Throughout the natural course of syphilis, all ocular structures can be involved, thus manifesting as chorioretinitis, panuveitis, and iry-docyclitis with corneal precipitates, vitritis and retinal vasculitis. In infected patients optic disc oedema can also develop as well as exudative retinal detachment or necrotising retinitis (4).

An unusual manifestation of chorioretinitis is a formation of yellow lesions within the posterior pole on the level of retinal pigment epithelium (posterior placoid chorioretinitis) (6). The disease can also cause the intraocular pressure elevation (ocular hypertension or secondary uveitic glaucoma), Argyll--Robertson pupil or cranial nerve (including oculomotor nerve) palsy (1). The above symptoms develop in patients with secondary and tertiary syphilis. Barile reported that the most common ocular manifestation of syphilis was iridocyclitis followed by panuveitis, posterior uveitis and keratouveitis (7). Hong described 8 syphilitic patients. He diagnosed panuveitis in 11 eyes, anterior uveitis in 2 eyes and posterior uveitis in 1 eye (8). Villanueva presented 20 patients with syphilitic posterior uveitis – 15 of them had acute or chronic chorioretinitis, panuveitis was diagnosed in 3, retinal vasculitis in 2 and pseudoretinitis pigmentosa in one patient (5). The International Uveitis Study Group introduced a classification of syphilitic posterior uveitis to organize and show the spectrum of syphilic-induced ocular lesions. This classification is shown in Table I (5,9). These lesions are usually bilateral although often asymmetric (9).

Characteristic/ Właściwość	Classification/ Klasyfikacja
Onset/ Początek	Acute (≤ 3 months)/ 0stry ≤ 3 miesiące
	Chronic (> 3 months)/ Przewlekły > 3 miesiące
Extent/ Zasięg	Focal/ Ogniskowy
	Multifocal/ Wieloogniskowy
	Diffuse/ Rozsiany
Pattern/ Typ	Chorioretinitis/ Zapalenie naczyniówki (pierwotne) i siatkówki (wtórne)
	Retinochoroiditis/ Zapalenie siatkówki (pierwotne) i naczyniówki (wtórne)
	Neuroretinitis/ Zapalenie nerwu wzrokowego i otaczającej go siatkówki
	Panuveitis/ Zapalenie całej błony naczyniowej
	Posterior placoid chorioretinitis/ Tylne płaskie zapa- lenie naczyniówki i siatkówki na poziomie nabłonka barwnikowego siatkówki i neurosensorycznej siatkówki
	Peripheral necrotizing retinitis/ Obwodowe martwi- cze zapalenie siatkówki
	Primary retinal vasculitis/ Zapalenie naczyń siatkówki

Tab. I. Classification of syphilitic posterior uveitis.

Tab. I. Klasyfikacja tylnego zapalenia błony naczyniowej na tle kiły.

Approximately 50% of patients with acute syphilitic uveitis are infected with HIV (9). In HIV-positive patients syphilis is the most common intraocular bacterial infection (1). Considering the above, all patients with eyes affected by syphilis should be screened for HIV. It is also advisable to examine the patient's sexual partner(s) for syphilis and HIV.

As syphilitic uveitis and neuroretinitis are classified as neurosyphilis, cerebrospinal fluid (CSF) analysis should be perfor-

med in such patients. The ENT assessment should also be performed due to possible hearing loss (10). Such management is based on an assumption that if the eyes are involved, the entire nervous system may be involved as well (10).

The described patient was not infected with HIV. His sexual partner was not infected with syphilis. The patient initially received symptomatic treatment, due to time needed for obtaining the results of blood tests for syphilis. This treatment significantly improved his visual acuity. The results of all syphilis diagnostic tests: USR, VDRL, FTA and FTA-ABS were positive. Serology testing for borreliosis was also positive, although it was considered false positive due to syphilis infection. The patient was referred to the Department of Dermatology and Sexually Transmitted Diseases for causal treatment. Recommended therapy in such cases includes the use of crystalline penicillin G (agueous solution) at the daily dose of 18-24 million IU administered as a continuous infusion or divided into smaller doses administered at 4-hour intervals. The treatment should be continued for 10-14 days (10). After the causal treatment, the patient was lost to follow-up at the Department of Dermatology and Sexually Transmitted Diseases.

Patients with syphilis require periodic clinical assessment and serological testing (11).

- In early syphilis: clinical examinations and serological tests (non-specific tests) should be carried out in the 1st, 2nd, 3rd, 6th and 12th month. In HIV-infected patients check-ups should be performed in the 1st, 2nd, 3rd, 6th, 9th, 12th, 18th and 24th month and can be finished with cerebrospinal fluid examination.
- After syphilis treatment completion, the titre in non-specific tests should decrease four-fold within 6 months and in patients with HIV co-infection – within a year.
- Cerebrospinal fluid analysis should be performed 1–2 years following treatment completion of neurosyphilis. It should be noted that ocular syphilis is also classified as neurosyphilis.
- The sexual partner(s) of syphilis patients should also be examined.

A four-fold non-specific tests titre increase indicates re-infection or reactivation of the infection. Specific syphilis tests can remain positive even after a successful therapy.

Although syphilis is an infectious disease with a potential permanent debilitating effect on e.g. vision, its treatment is not compulsory in Poland.

Conclusions

An increase incidence of syphilis infections has been observed since the beginning of the 21st century. Therefore infectious etiology and primary syphilis should always be considered in patients with progressive retinitis, choroiditis and vitritis.

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