CASE REPORT

Unilateral swollen optic disc: do not forget neurosyphilis

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SUMMARY

A 51-year-old man presented with a 3 month history of progressive right visual loss. On examination, the only abnormal findings were a right visual acuity of 4/10 and a swollen right optic disc. The patient had previously undergone MRI that had been reported as being normal and a trial of corticosteroids under the care of the referring ophthalmologist. An extensive battery of blood tests was normal apart from a rapid plasmonic reagin titre of 1:64. ELISA revealed elevated levels of total and immunoglobulin M antibodies against *Treponema pallidum*. Cerebrospinal fluid analysis revealed a mild leucocytosis and mildly elevated protein content. The patient was treated with a 5 week course of penicillin. Three months after starting treatment, his right visual acuity had improved to 8/10 and his right optic disc swelling had resolved.

BACKGROUND

Optic disc swelling is a relatively common condition that can present to different physicians and associate with various entities, some potentially life or sight threatening.¹ Thus, a careful clinical workup is required to initiate the correct treatment.² For diagnosis purposes it is useful to differentiate unilateral from bilateral disc swelling, although a causal overlap exists between the two. Most of the causal entities are ischaemic, inflammatory or infiltrative.³ Although infectious causes are less frequent, they should be kept in mind because they require a rapid specific treatment. Unconventional diagnosis should be considered when the initial evaluation is inconclusive or the manifestations are atypical.

Syphilis is a systemic disease that typically goes through three stages of symptoms (primary, secondary and tertiary) and two asymptomatic stages (early and late latent).⁴ Ophthalmological compromise as a result of neurosyphilis is relatively rare, particularly in the immunocompetent, but is also seen and can be the presenting feature.^{5 6} Because syphilis is re-emerging in developed countries, maintaining the awareness to its variable manifestations may allow an early diagnosis.⁷

CASE PRESENTATION

A 51-year-old man was referred by an ophthalmologist for an internal medicine consultation for aetiological study because of progressive loss of vision in the right eye over a period of 3 months. The patient's ophthalmological evaluation demonstrated a visual acuity of 4/10 in the right eye and generalised oedema of the ipsilateral, without an afferent pupillary defect, biometric changes or

vasculitis/retinitis. No anomalies were observed in the left eye. He had already had systemic corticotherapy, without improvement. He brought previously performed craniocerebral and contrasted ocular nuclear MR showing no changes (no nerve enhancement was found). The patient initially denied any relevant antecedent or additional symptoms, referring only to sexual relations with his spouse. An objective examination showed no changes, beyond ophthalmological signs.

INVESTIGATIONS

The patient's haemogram, biochemistry, C reactive protein and sedimentation rate results were normal. We excluded folic acid and vitamin B₁₂ deficiencies, a serum ACE increase, diabetes mellitus and dyslipidaemia, antineutrophil cytoplasmic antibody-positive vasculitis, antiphospholipid syndrome and thrombophilia. Blood tests for hepatitis B and C, HIV, cytomegalovirus, *Toxoplasma gondii* and *Borrelia burgdorferi* infection were negative. A rapid plasmonic reagin (RPR) test revealed a titre of 1/64. In confirmation of the RPR results, ELISA treponemal test confirmed elevated levels of total and immunoglobulin M antibodies against *Treponema pallidum* in the patient's serum. The patient then admitted to having had an unprotected extramarital sexual relationship 4 years prior, after which he had a single, painless ulcerated penile lesion. At this time, he went to the doctor and was treated with weekly penicillin for 3 weeks. The lesion resolved itself, and the patient was not re-evaluated, nor was his partner informed. He did not notice any later symptoms attributable to secondary syphilis. We performed a lumbar puncture, with a flow of clear cerebrospinal fluid (CSF). CSF analysis revealed a mild leucocytosis (5 mononuclear leucocytes/mm³), a mildly elevated protein content (61 mg protein/dL), a normal glucose value and a non-reactive Venereal Disease Research Laboratory test result.

DIFFERENTIAL DIAGNOSIS

Unilateral swollen disc associates, most commonly, with anterior ischaemic optic neuropathy (AION), particularly non-arteritic AION, optic neuritis and orbital compressive lesions. The normal brain images excluded intracranial lesions. Non-arteritic AION is usually seen in elderly individuals with atherosclerotic risk factors or in use of sildenafil and arteritic AION is generally associated with giant cell arteritis. In this condition, vision loss is generally sudden and altitudinal defects are a classic finding. Our patient did not present relevant personal or toxicological antecedents, clinical or

To cite: Porto L, Capelo J, Carragoso A. *BMJ Case Rep* Published online: [please include Day Month Year] doi:10.1136/bcr-2013-008891

analytical evidence of giant cell arteritis, or altitudinal defects. Retinal vein occlusion and papillophlebitis also cause unilateral vision loss and disc swelling but vessel enlargement and retinal haemorrhage are also described. Leber's hereditary optic neuropathy could also be considered but as an improbable cause because of the patient's age and the absence of familiar history. So, optic neuritis remained as the most probable diagnosis. Optic neuritis can be secondary to demyelinating lesions or, less frequently, to autoimmune and infectious diseases, like syphilis.

TREATMENT

We started treatment with 24 million units of intravenous penicillin G per day for 14 days and 2.4 million units of intramuscular penicillin benzathine per week for an additional 3 weeks.

OUTCOME AND FOLLOW-UP

Three months after the treatment started, there was a significant improvement in the patient's vision, with the visual acuity of his right eye reaching 8/10 and an absence of papillary oedema. Furthermore, his RPR titre was then 1/16.

DISCUSSION

In this case, the primary infection was identified as a probable sexual contact followed by a compatible ulcerated penile lesion and associated medical therapy. The patient denied any symptoms/signs attributable to secondary syphilis. In fact, secondary manifestations can be so subtle that 60% of patients do not acknowledge them.⁸ Neurosyphilis is diagnosed based on a combination of signs and symptoms, as well as laboratory studies of blood and CSF.⁴⁻⁹ In this case, the non-treponemal and treponemal tests by enzymatic immunoassay were both positive. Analysis of the CSF, obligated because of the positive blood tests and ophthalmological symptoms, showed high mononuclear cellularity and a high protein concentration of the CSF, with non-reactive VDRL serology. In neurosyphilis with isolated cranial nerve involvement, CSF cellularity may be normal or elevated, and the CSF-VDRL in neurosyphilis, particularly of the eye, may be negative.⁴ Given the clinical observations, ophthalmological findings, positive treponemal blood test result, considerable titre level of the non-treponemal test and cytochemical changes in the fluid in this case, we were able to affirm a diagnosis of optical neuritis and tertiary syphilis and proceeded with a neurosyphilis treatment.⁴ Post-treatment clinical improvement and reduction of blood titres to normal levels further confirmed the diagnosis.

Syphilis can compromise nearly all of the ocular structures. Syphilitic optical neuritis, particularly in an immunocompetent person, is rare and does not have any distinctive characteristics. The cases presented in the literature describe syphilitic optic neuritis in secondary and tertiary syphilis, immunocompetent and immunocompromised patients and unilateral and bilateral affections.^{2-5, 10-12} Based on the location of the optic nerve involvement, we found cases of papillitis with swollen disc, perineuritis and neuroretinitis.^{2-5, 10-12} Regarding the ophthalmological findings, an optic neuropathy usually, but not always, associates with a relative afferent pupillary defect. In other cases

of syphilitic optic neuritis the afferent pupillary defect was also absent.¹⁰⁻¹¹

We still need to discuss how the patient developed this clinical picture after receiving appropriate treatment for primary syphilis. One hypothesis is reinfection, since his partner was not treated, although the patient did not describe a new primary syphilis lesion after the first infection. Another hypothesis is therapeutic failure. Indeed, 10% of patients with early syphilis require retreatment.⁴⁻⁸ We can only speculate about what happened in this case.

Learning points

- ▶ Optic disc swelling is a relatively common condition with numerous causes.
- ▶ A careful clinical work up is required to establish the correct diagnosis and treatment.
- ▶ Syphilitic optic neuritis, although uncommon, should be considered if a patient presents with visual loss and optic disc swelling.
- ▶ In neurosyphilis, particularly of the eye, cerebrospinal fluid (CSF) cellularity may be normal or elevated and the CSF-Venereal Disease Research Laboratory may be negative.

Contributors All the authors contributed to conception and design, or analysis and interpretation of data, drafting the article or revising it critically for important intellectual content and final approval of the version to be published.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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