RETROBULBAR OPTIC NEURITIS AND CYSTOID MACULAR EDEMA AS A FIRST MANIFESTATION OF LYME DISEASE

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ABSTRACT

Lyme disease is a multi-system disorder caused by the spirochete *Borrelia burgdorferi*. Eye manifestations are a rare involvement. We report two cases of patients who developed a retrobulbar optic neuritis and a cystoid macular edema as a sole clinical presenting in Lyme disease.

A 42-year-old female presented with left eye decreased visual acuity and painful ocular movement. Dilated funduscopy and neurological examination were normal. A diagnosis of left eye retrobulbar optic neuritis was made. The brain and spinal cord MRI showed typical lesions characteristic of MS. Serological tests for Lyme disease revealed positive results for IgM ELISA and Western blot. The possible serologically proven Lyme disease was diagnosed. Visual acuity returned to normal following a course of high-dose steroids and intravenous antibiotic, after by oral antibiotic. A 28-year-old male presented with left eye blurred vision and intermittent blurring vision in his right eye. Dilated funduscopy revealed bilateral cystoid macular edema (left more than right). He was positive for Lyme serology, IgM ELISA and Western blot, and intravenous antibiotic therapy was commenced, followed by oral antibiotic. Visual acuity was restored and funduscopy was normal.

In summary, although Lyme disease is an uncommon cause of these neuro-ophthalmic complications, our two patients highlight the importance of considering this disorder as a differential diagnosis and to initiate an early adequate therapy.

Keywords: *Lyme disease, retrobulbar optic neuritis, cystoid macular edema*

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INTRODUCTION

Lyme disease, first identified in 1975 in Lyme, Connecticut, USA is caused by *Borrelia burgdorferi*. This tick-borne spirochetal disorder progresses in three stages and can cause symptoms in many body systems, including the eyes (1,2). Ocular problems are infrequent involvements of Lyme disease that can appear at any stage of the disease and can take many different forms. Conjunctivitis is the most common ocular finding in stage I and II (3-6). Ophthalmic complications can be associated with neurologic in-

volvement, manifesting as ocular motility problems, optic neuritis, papilledema, and other possible ocular signs including uveitis, cystoid macular edema, scleritis, exudative retinal detachments (7-15). In the late stages of the Lyme borreliosis bilateral interstitial keratitis and symblepharon formation are described as characteristic findings. Enzyme linked immunosorbent assay (ELISA) test is used to detect antibody titers for the diagnosis of Lyme disease. Western blot is a qualitative test that is generally more sensitive and specific than ELISA. Although no test is 100% accurate, Western blot test is helpful in differentiating a false positive result that can occur in patients with syphilis, tuberculosis, sarcoidosis, autoimmune disease (MS) or other neurologic disorders (16,17). We report two cases of patients who developed a retrobulbar optic neuritis and a cystoid macular edema as a sole clinical presenting in Lyme disease.

CASE REPORT I

A 42-year-old female (DDR) was admitted to the Neuro-ophthalmology section of the Department of Neurology because of progressive decrease of visual acuity in the left eye and painful ocular movement during the last week. Neuro-ophthalmic examination revealed decreased visual acuity of 5/500 OS and 5/5 OD, left eye relative afferent pupillary defect and Ishihara testing with significant decrease in color vision. Dilated funduscopy was normal. A diagnosis of left eye retrobulbar optic neuritis was made. Neurological examination was normal. Standard hematological investigations, thyroid function tests, antinuclear antibodies, and syphilis serology were negative. Lumbar puncture was not performed. Cerebral and spinal cord Magnetic Resonance Imaging (MRI) showed typical lesions characteristic of MS. Serological examination of Neuromyelitis optica (NMO) – IgG/antiAQP4 antibody was negative. Intravenous high-dose Methylprednisolone was initiated for five days. After five days of treatment the visual acuity had improved to 5/6.3 OS. Further serological tests for Lyme disease showed a raised IgM ELISA titer and a positive result for *Borrelia burgdorferi* in the Western blot. The possible serologically proven Lyme disease was made in the absence of tick bites history and erythema migrans. A course of intravenous Medaxone and oral Cefzil was administered.

One month later visual acuity (5/5 OD and OS), pupillary light reaction, and color vision were normal.

CASE REPORT II

A 28-year-old male (VSI) presented to the Neuro-ophthalmologic section of the Department of Neurology with left eye blurred vision and intermittent blurring vision in his right eye for the last ten days. His medical history included a tick bite one year before but had never identified any rash. Neuro-ophthalmologic examination showed visual acuity of 5/6.3 OD and 5/50 OS. Dilated funduscopy revealed bilateral cystoid macular edema (left more than right), confirmed by fluorescein angiogram (FA) and OCT. Brain MRI was normal. Initial blood tests, including autoantibodies and syphilis serology were also negative. Serological tests for Lyme disease revealed a positive result for IgM ELISA and IgM Western blot. A diagnosis of Lyme disease was made. Antibiotic therapy with Medaxone (3.0 g daily) was commenced and oral Cefzil was administered. The patient responded subsequently to this antibiotic therapy. Six months after the treatment visual acuity was restored to 5/5 OD and OS, funduscopy was without macular edema.

DISCUSSION

Lyme disease is a multi-system disorder caused by the spirochete *Borrelia burgdorferi*, and has three clinical stages. Eye manifestations of Lyme disease are rare involvement (1,2,4). Neuro-ophthalmological complications are most frequently seen in the late phases of the disorder, but some patients may not present all stages. As the serological tests lack sensitivity and specificity the etiological diagnosis of an isolated ocular manifestation as the presenting sign of Lyme borreliose must be based on medical history, clinical findings, and positive serological testing (5,6,10,16,17). Retrobulbar optic neuritis as a sole sign of Lyme disease is rare, but it is most often caused by demyelinating disease. In our first case the patient with retrobulbar neuritis was with MRI typical lesions of MS, but with ELISA and Western blot positive results for *Borrelia burgdorferi*. In the recent literature the researchers discuss that patients with MS-like ocular symptoms probably had these symptoms due to MS and had also been exposed to the *Borrelia* bacterium. Based on the long-term follow-

up of cases of isolated optic neuritis labeled as Lyme disease, because of the positive serology for *Borrelia*, there is an evidence for a later development of demyelination syndromes (8,9,10,13,14,15). The second patient suffered from serologically proven Lyme disease with isolated cystoid macular edema as a first manifestation.

Current treatment protocols for Lyme disease include the use of oral, intramuscular, and intravenous antibiotic medications. A treatment protocol for severe neuro-ophthalmic or posterior ocular segment involvement in Lyme disease has not been established. In these cases therapeutic strategy should be based on the association of antibiotics and corticosteroids. A new course of therapy may be recommended to patients with relapsing ocular inflammation (18).

CONCLUSION

In this report we present two patients with retrobulbar optic neuritis and cystoid macular edema as a first manifestation of Lyme disease. In summary, although Lyme disease is an uncommon cause of these neuro-ophthalmic complications, our cases highlight the importance of this spirochetal disorder as a differential diagnosis and the necessity to initiate an early antibiotic therapy.

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