

Neuroretinitis as presenting and the only presentation of Lyme disease: Diagnosis and management

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We present a case of neuroretinitis as presenting and the only presentation of Lyme disease in a 25-year-old female who visited hilly areas in the Himalayas of North India. She presented with right eye sudden and painless blurring of vision. Her vision at presentation was 20/60. She had fundus examination; fundus fluorescein angiography (FFA) and optical coherence tomography (OCT) imaging showed classical features of neuroretinitis. No other organ was involved. Oral steroids were prescribed and relevant investigations sent for noninfective and infective causes. Worsened visual acuity (VA) to hand movement and positive IgM titers for *Borrelia burgdorferi* led to the diagnosis of Lyme disease-associated neuroretinitis. Treatment with oral doxycycline plus oral steroids for 4 weeks revealed VA of 20/20 and resolution of fundus and OCT changes. Neuroretinitis as presenting and the only presentation of Lyme disease will be discussed with serial fundus, FFA, and OCT pictures.

Key words: *Borrelia burgdorferi*, doxycycline, Lyme disease, neuroretinitis, tick bite

Lyme disease occurs due to *Borrelia burgdorferi*, a spirochete, and is transmitted by the bite of its vector *Ixodes* tick. It occurs

in three stages: primary, secondary, and tertiary. Ocular involvement can occur in any stage. Any part of the eye can be involved. No large published series is available for neuroretinitis as the first and only presentation of Lyme disease.

Case Report

A 25-year-old female presented with right eye sudden and painless blurring of vision for 4 weeks, who visited a hilly area in the Himalayas, North India. No history of headache, fever, stiff neck, malaise, nausea, redness, watering of eyes, floaters, and trauma was reported. Family and treatment history were negative. On examination, her visual acuity (VA) in the right eye was 20/60 (unaided) and 20/20 in the left eye. Anterior chamber was normal, relative afferent pupil defect was present, and fundus examination showed signs of papillitis, superior temporal retinitis contiguous with disc, vasculitis involving large vessels near the disc [Fig. 1]. Examination with +78 D revealed vitreous cells in the vicinity of neuroretinitis. Intraocular pressure was 14 mmHg in both eyes. Fundus fluorescein angiography (FFA) and optical coherence tomography (OCT) confirmed the diagnosis of neuroretinitis [Fig. 2a and b]. Left eye examination was normal. Preauricular and submandibular lymphadenopathy was absent. Dermatologic and systemic examinations were normal. The patient was empirically started on oral steroids (prednisone 40 mg/day [1 mg/kg]).

Rheumatoid factor, Cytoplasmic Antineutrophil Cytoplasmic Antibodies (c-ANCA), Perinuclear Anti-Neutrophil Cytoplasmic Antibodies (p-ANCA) (anti-myeloperoxidase) antibodies, antinuclear antibody, anti-double-standard antibody, angiotensin converting enzyme, human leukocyte antigen B-27, rapid plasma reagin and fluorescent treponemal antibody test, *Treponema pallidum* immobilization and *T. pallidum* hemagglutination assay for syphilis, toxoplasma titer, HIV ELISA were within limits. Lyme disease antibodies, IgG and IgM serum were 0.3 (<0.90) and 9.1 (<0.90). Mantoux test (5 mm × 5 mm), chest X-ray, magnetic resonance imaging head, and echocardiography revealed no abnormality.

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On follow-up examination after 1 week, her VA dropped to hand movement close to face and signs of fluid retention were present. Fundus examination showed exaggeration of extent of all signs [Fig. 3a]. OCT showed neurosensory detachment [Fig. 3b]. Considering the worsening of signs, symptoms, and positive IgM titer for *Borrelia burgdorferi*, serum Western blot analysis test confirmed Lyme disease. Diagnosis of neuroretinitis associated with Lyme disease was made. Oral doxycycline 100 mg was administered twice daily for 6 weeks and prednisone was tapered over a period of 4 weeks. Four weeks follow-up showed VA of 20/20 right eye. Fundus and OCT showed settling of neurosensory detachment and reduction of intraretinal fluid [Fig. 4a and b]. IgM titer was 1.2 (<0.90). The patient was advised strict follow-up for a longer duration. One-year follow-up showed no recurrence.

Discussion

Lyme disease was first described in the USA in 1977 by Steere *et al.*^[1] It is characterized by prominent dermatological, rheumatological, and cardiac manifestations. The infectious agent of Lyme disease is *B. burgdorferi*, a spirochete, isolated

successfully from blood, synovial fluid, spinal fluid, retina, and vitreous of the eye.^[2] It is transmitted by the vector *Ixodes* tick bite and occurs in three chronological stages; primary, secondary, and tertiary [Table 1]. Over 50% of untreated, treated initially, and on treatment, patients may develop stage 3 Lyme disease. Hence, long follow-up is recommended for early diagnosis of cardiac, neurological, and arthritic involvement.^[3,4] The Centers for Disease Control and Prevention made Lyme disease a reportable disease in 1982.

Individual case reports with positive Lyme serology have reported papillitis,^[5] anterior ischemic optic neuropathy, optic neuritis, and neuroretinitis;^[6] false positives have been up to 10% among residents of endemic areas without any apparent signs of infection.

The current definition of Lyme disease includes a patient who develops erythema migrans (EM) within 30 days of exposure in an endemic area or endemic area exposure, without EM, but with signs involving one organ system and a positive laboratory test or no history of exposure, but with EM as well as involvement of two organ systems or no exposure

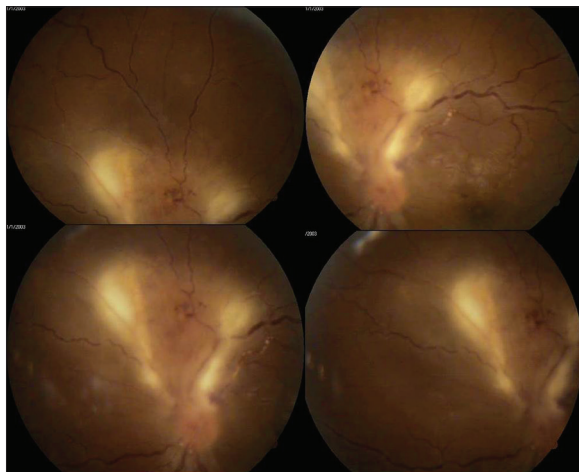


Figure 1: Fundus photograph showing papillitis, neuroretinitis, vasculitis

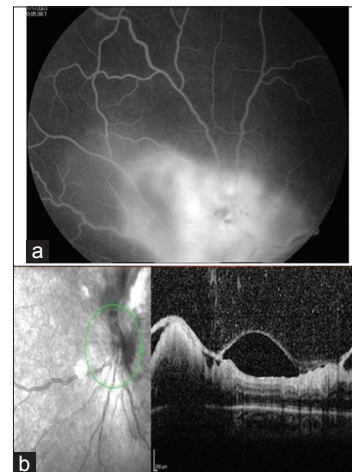


Figure 2: (a) Fundus fluorescein angiography showing active vasculitis. (b) Optical coherence tomography showing vitritis

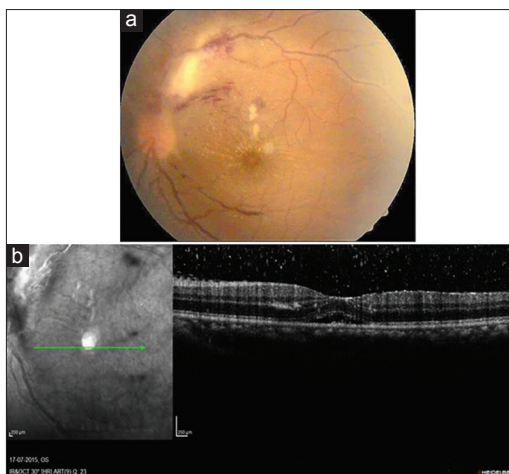


Figure 3: (a) Fundus photograph showing exaggeration. (b) Macular optical coherence tomography showing neurosensory detachment and vitritis

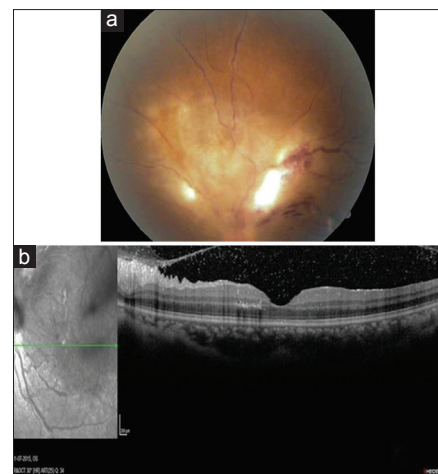


Figure 4: (a) Fundus photograph showing resolving lesions. (b) Optical coherence tomography showing absence of neurosensory detachment and a normal foveal contour

Table 1: Stages of Lyme disease - major manifestations

Stage	Ophthalmic	Neurologic
Stage 1	Conjunctivitis, photophobia	Headache, stiff neck, nausea
Stage 2	Iridocyclitis, uveitis, vitritis, pars planitis, diplegia, painful radiculopathy, choroiditis, macular edema, optic disc edema, optic disc pallor (AION, optic neuritis, neuroretinitis)	Meningitis with papilledema, painful radiculopathy, encephalitis
Stage 3	Stromal keratitis, episcleritis, orbital myositis	Fatigue, encephalomyelitis, demyelination (MS-like, anorexia nervosa, ALS)

AION: Anterior ischemic optic neuropathy, ALS: Amyotrophic lateral sclerosis, MS: Multiple sclerosis

in an endemic area, but with EM and a positive serology.^[7] Our patient was diagnosed as Lyme disease based on history of exposure, involvement of one organ, i.e., eye and positive serology for *B. burgdorferi*. Classical presentations of three chronological stages of ophthalmic Lyme disease were not present. Vitreous, retina, and optic nerve of one eye were involved.

Neuroretinitis is a focal inflammation of the optic nerve and peripapillary retina or macula of either infectious or idiopathic etiology. Infectious causes are Cat scratch disease, syphilis, Lyme disease, toxoplasmosis, mumps, salmonellosis, tuberculosis, and histoplasmosis. Cat scratch disease is the most common cause of neuroretinitis. Diagnosis for neuroretinitis in Lyme disease is established by exclusion. Babu and Murthy^[8] reported a similar case in South India. Although rare in India, we need to consider Lyme disease in the differential diagnosis of neuroretinitis, especially if the patient hails from a forested area. Our case had an exposure in the Himalayan region of North India.

The role of steroids has been controversial in infective neuroretinitis. In our case, oral steroids started before the serological confirmation did worsen the signs. Babu and Murthy^[8] and Vanya *et al.*^[9] have reported resolution of neuroretinitis with a combination of doxycycline and corticosteroids. Treatment with prednisone and doxycycline resolved neuroretinitis in our case. There is no recurrence within 1-year follow-up of our case. Long-term use of antibiotics is successful in preventing disease progression to tertiary stage. The same was also suggested by Steere *et al.*^[10]

Conclusion

We conclude that neuroretinitis can be the presenting and the only presentation of Lyme disease, hence should be considered in the differential diagnosis of neuroretinitis. Documentation with fundus photography, FFA, and OCT for response to treatment is recommended. Doxycycline and corticosteroids

should be started at the earliest as it is a treatable disease. Long-term follow-up is important.

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Conflicts of interest

There are no conflicts of interest.

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