Neurosensory Misa“Lyme”ment:

Functional vision assessment and rehabilitation in neurological Lyme disease

Jennifer Zhang, OD

Summary

Lyme disease is a complex systemic inflammatory condition that can cause significant neurological effects. Patients with neurological Lyme disease often have visual symptoms such as blurry vision, fatigue, double vision, and are overwhelmed by visually busy environments. They also suffer from sensory processing difficulties such as disequilibrium, compromised spatial orientation, and memory problems. Systemic antibiotics are the mainstay of treatment for Lyme disease, but are well documented to be ineffective against neurological Lyme. Thus, functional management of the disease should be considered to improve patients' quality of life. This paper describes two cases of patients with neurological Lyme disease, addresses important diagnostic considerations, and discusses rehabilitation strategies through vision therapy with an emphasis on sensory processing, visual-vestibular integration, and localization.

Introduction

Lyme disease is a complex systemic infection caused by the bacterium *Borrelia burgdorferi*. According to the CDC, 30,000 cases of Lyme disease are reported in the USA per year. However, the condition is likely underdiagnosed; the estimated yearly case rate is estimated to be 476,000 per year. Early signs and symptoms of the disease include fever, chills, and flu-like symptoms, as well as the classic "bullseye" rash. However, the nervous system is affected in 15% of patients (Halperin 2015), and in such cases, can lead to vision problems such as blur, visual fatigue, double vision, headaches, light sensitivity, and losing place while reading. It also affects higher order visual processing in the brain, and can cause more widespread sensory processing deficits (Padula Institute of Vision).

Currently, the common treatment for Lyme disease is high-dose systemic antibiotics, (Halperin 2015). However, such treatment poses substantial risks and has been widely documented not to be effective in patients with "persistent fatigue and cognitive symptoms" (Halperin 2015, Klemper et al 2001, Klemper et al 2013), which are consequences of disruptions to visual and other sensory processing mechanisms within the brain. Patients who suffer from such functional vision deficits present with symptoms that range from uncomfortable to debilitating. Yet, functional vision rehabilitation is not a mainstay of neurological Lyme disease treatment, despite the significant limitations that the disease imposes on the patients' activities of daily living.

This paper presents two cases of patients with neurological Lyme disease, describes visual and visual-vestibular effects of the condition, and discusses rehabilitation strategies through vision therapy with an emphasis on sensory processing, visual-vestibular integration, and localization.

Case 1: Patient EE

Case History

Patient EE is a 47-year-old female who presented for a comprehensive eye exam with complaints of her right eye vision feeling "off", or "not right" constantly for several months. She also notes a vague "fog in the middle of [her] right eye" that worsens when looking at headlights. The symptoms are associated with a general sense of disorientation and "not knowing where [she] is in space". At her last exam 4 years ago, the patient was found to have a low

refractive error with presbyopia and was prescribed glasses for near. The patient habitually does not wear correction for distance or near. The patient's medical history is remarkable for neurological Lyme disease, diagnosed 1.5 years ago. She received ceftriaxone treatment for 6 months before discontinuing due to liver and kidney toxicity. She also reports a history of exposure to an environmental mold that caused significant levels of mycotoxin in her blood.

Given her history of Lyme disease, the patient was given a concussion symptom screening checklist to futher assess her functional symptoms. The checklist yielded a score of 40 and revealed significant near vision symptoms, including blur at near, closing or covering an eye when reading, declining reading comprehension over time, difficulty with visual work at the end of the day, and dizziness/nausea after 30-45 minutes of near work. The checklist also revealed symptoms consistent with sensory and vestibular maladaptations: light and sound sensitivity, poor ability to judge distances, poor eye-hand coordination, clumsiness, motion sickness, and a history of vertigo and disorientation.

Visual and Ocular Examination

Table 1 provides a summary of the patient's visual exam and sensorimotor evaluation:

Test	**Finding**	**Subjective response**
Entering distance acuity (sc)	OD: 20/20-1 OS: 20/25 OU: 20/20-1	Slow, strenuous
Subjective refration (distance)	OD: -0.50 sph (20/20) OS: -0.50 -0.75 x045 (20/20)	Feels like "eyes are being squeezed"
Binocular balance	OD: pl sph OS: pl sph	"Blurry but calmed"
Subjective refraction (near)	OD: +0.75 sph (20/20) OS: +0.75 sph (20/20) OU: 20/20	OD: "hazy" OS: "feels like eyes are squeezing"
Cover test/Maddox Rod	D: ortho N: 6^ XP, 1^ R hyper	
Von Graefe	D: 4^ XP N: 7^ XP, 3^ R hyper	
NPC (break/recovery)	3"/4"	Significant visual discomfort
Smooth vergences (distance)	BO: x/6/2 BI: x/6/4	
Smooth vergences (near)	BO: x/20/2 BI: 16/20/16	
NRA/PRA	+2.00/-2.00	Significant strain, "squeezing feeling"
Stereopsis (Randot 1)	125 arcsec	

Table 1: Patient EE Visual Exam Findings

Clinical Management – Vision Therapy

The goal of vision therapy in this case is to help the patient gain an understanding of her visual world and the status of her visual systems, with future goals to gain more control of her visual system and to integrate vestibular and other sensory systems. At the time of writing, the patient is in the initial part of her vision therapy course, in which an emphasis is being placed on learning basic accommodative, vergence, and ocular motility skills, as well as the associated tone. Visual-spatial exercises are also being used to encourage spatial processing. As the patient progresses in her treatment, it is expected that exercises involving mobility, balance, and head/eye movement will be incorporated into her program to emphasize central-peripheral integration and vestibular integration. Examples of such activities include infinity walks, walking rail exercises, balance board exercises, vestibulo-ocular reflex exercises, and navigating through space while holding vectograms or otherwise engaging central vision.

Case 2: Patient DL

Case History

Patient DL is a 37 year old Caucasian female who was referred from Wilmer Eye Institute at Johns Hopkins Medicine for binocular vision/sensorimotor assessment. The patient's chief complaints included distance blur especially in the left eye, headaches, painful and tired eyes, covering an eye, and poor reading comprehension. She also reports visual and auditory hypersensitivity, "misaligned vision", and an uncertainty of spatial relationships that she describes as an "out-of-body-experience". She also reports that she has "lost confidence in [her] eyes" and that her eyes make her feel "vulnerable". The symptoms have persisted for the past six years with no effective treatment.

Her concussion symptom checklist revealed an avoidance of near work, poor reading attention and comprehension, and increased difficulty with visual work at the end of the day; as well as sensory processing symptoms including poor ability to judge distances, being clumsy or accident prone, motion sickness, and poor memory.

Visual and Ocular Examination

Table 2 summarizes pertinent findings from the patient's visual exam per records from Wilmer Eye Institute and sensorimotor testing.

Clinical Management – Vision Therapy

Patient DL reported that her goals for vision therapy were to "have better spatial awareness of both my body in space and my relationship to other people/objects", to "feel less out-of-body", and to "gain back confidence I have lost at work because navigating visual stimuli is so challenging for me". Thus, her therapy was tailored to focus on binocular stability, localization, and spatial awareness. She was given vectograms and similar activities with increasing accommodative and vergence demand at near to strengthen binocularity at near and develop stronger central stereopsis; she also worked on exercises that incorporated movement and changing visual enviroments to engage central-peripheral integration and vestibular integration.

After three months of in-office therapy and home exercises, the patient was re-evaluated. She demonstrated improved divergence and convergence ranges, improved NPC (from a break point of 12" to convergence to the nose), and improved stereopsis (from 200 arcsec to 25 arcsec). The patient also reported that she was better able to read road signs while driving and thus felt more confident while driving, and was able to complete vision therapy tasks and handle visual demands more easily and with more confidence. At the time of writing, the patient is continuing with her vision therapy course. She is also seeking systemic Lyme treatment.

Test	Finding
Entering distance acuity (cc)	OD: 20/20 OS: 20/20 OU: 20/20
Subjective refraction/ Binocular Balance (distance)	OD: -4.00 sph (20/20) OS: -4.00 sph (20/20)
Subjective refraction (near)	OD: pl sph (20/20) OS: pl sph (20/20) OU: 20/20
Cover test/Maddox Rod	D: 1-2^ XP N: 12-14^ IAXT
Von Graefe	D: 3^ EP, 2^ R hyper N: 5^ XP, 1^ R hyper
NPC (break/recovery)	12"/ 18"
Smooth vergences (distance)	BO: 6/20/8 BI: x/6/-2
Smooth vergences (near)	BO: 12/30/6 BI: x/28/6
NRA/PRA	+0.75/-3.00
Stereopsis (Randot 1)	200 arcsec

Table 2: Patient DL Visual Exam Findings

Discussion

Lyme disease as an ABI

Lyme disease is a complex systemic infection caused by the bacterium *Borrelia burgdorferi* and is transmitted to humans through the bite of infected blacklegged ticks (Neuro-Optometric Rehabilitation Association, Padula Institute of Vision). The bacterium causes widespread effects, including the characteristic bullseye rash, fever and flu-like symptoms, and many ocular conditions. In the eye itself, it manifests as conjunctivitis, uveitis, choroiditis, optic neuritis, and vasculitis (Kauffman and Wormser 1990). Currently, the common treatment for Lyme disease is high-dose systemic antibiotics, such as penicillin, ceftriaxone (and related antibiotics, cefuroxime axetil, cefotaxime), and doxycyline (Halperin 2015). However, antibiotic treatment has been widely documented not to be effective in patients who present with the chronic fatigue and cognitive symptoms suggestive of a more widespread neurological effect of the disease (Halperin 2015).

Neurological complications of Lyme disease occur in up to 15% of cases, where inflammation of the brain causes cranial nerve palsies, optic atrophy, and a host of vision problems, including blurry vision, diplopia (double vision), asthenopia, visual processing deficits, and visual-vestibular deficits (Padula Institute of Vision). Whether caused by these vision compromises themselves or by the compensatory mechanisms developed by the brain to rectify them, the visual system undertakes excessive strain that leads to fatigue, discomfort, and difficulties with higher-order visual processing mechanisms (Neuro-Optometric Rehabilitation Association). Although Lyme disease is often not viewed as an acquired brain injury (ABI) in mainstay treatment, it undoubtedly fits the criteria. Acquired brain injury (ABI) is a term that encompasses conditions that appear suddenly

and result in neurological dysfunction (Suchoff, Ciuffreda, and Kapoor 2001).

ABI effects on visual and visual-vestibular processing

The most frequent visual dysfunctions expected in the ABI population are 1) visual field losses, 2) eye movement dysfunctions, 3) ocular muscle disfunctions (including strabismus), 4) binocular dysfunctions (including convergence insufficiency, exophorias, vertical phorias, and fusional instability), 5) accommodative dysfunctions, 6) perceptual dysfunctions, and 7) visually-involved vestibular dysfunctions (Suchoff, Ciuffreda, and Kapoor 2001). The patients described in both cases were affected by dysfunctions in multiple of the above categories: both had significant convergence insufficiency, difficulty sustaining accommodation or poor accommodative flexibility, horizontal and vertical misalignment, and both showed signs of significant visual stress. All of these sequelae are functional in nature; thus, the role of neuro-optometry and visual rehabilitation is paramount, in both the diagnosis and treatment of these conditions.

In case 1, the subjective responses from the patient throughout testing indicated significant visual stress and discomfort, binocular instability, and more visual fatigue than expected. Even though the patient's visual acuity was 20/20 and her binocular testing objectively did not yield extreme deficits, she was unable to have comfortable vision.

Both patients exhibited vertical phorias during testing, although neither reported vertical diplopia and both were able to achieve stereoscopic fusion, even at initial testing. Vertical deviations are highly disruptive to binocular vision, and likely contribute significantly to binocular dysfunction. The resulting implications are twofold: firstly, binocular dysfunction places an enormous strain on the visual processing system. Literature supports that neurological processing and attention are limited resources, and when processing mechanisms are required in excess to maintain fusion, less capacity is available for other cognitive functions such as reading comprehension, retention and memory, and sustained visual work (Leslie 2001, Gallaway and Boas 2007, Garzia 1989). The goal of rehabilitation is to redevelop the automatic nature of binocular vision, such that the pathways themselves may operate with minimal attention, and allow attention to be available for higher processing (Leslie 2001).

Secondly, binocular dysfunction reduces fixation speed and visual stability (Gallaway and Boas 2007, Laukannen 1995, Morad 2002). Visual information is not received as quickly or accurately as expected is consistent with poor reading ability and feeling overwhelmed in busy visual environments. To examine the scope of these effects, it is important to consider that the visual system does not operate in isolation—rather, it is intimately connected to other sensory systems, including, importantly, the vestibular system. The vestibular sense is often recognized as the corporal sense that is most responsible for the feeling of being "grounded"; it allows the body to sense its orientation in space; it is the most organizing sense (Kranowitz 1998). A significant amount of input for the vestibular system is derived from vision, and the two systems are intimately connected. The insertion of the extraocular muscles on the eyeballs geometrically mirror the positions of the semicircular canals within the inner ear; thus, visual system disruption as seen in ABI is often concurrent with visual-vestibular misalignment (Rosen, Cohen, and Trebing 2001). An example of a visual-vestibular misalignment is visual motion sensitivity (VMS), which is a condition often seen after brain injuries that is characterized by dizziness, loss of balance,

headaches, and visual strain in "busy" environments (Tannen and Cohen 2018). Visual processing takes into account both central and peripheral information: the former accounts for stable fixation and acuity; the latter provides spatial cues and plan eye movements. When these pathways are unstable, information regarding the location and spatial relationships of objects is inconsistent or erroneous, which causes patients to be disoriented or uncomfortable in their spatial surroundings. In both of the above cases, the patients experienced discomfort related to spatial processing. Patient DL described her vision as being "misaligned", uncertain how far she was from certain objects, and how her vision left her feeling "vulnerable". Patient EE reported feeling disoriented and did not know where she was in space. Therefore, emphasizing spatial relationships, localization, movement, and balance-type activities in the patients' vision therapy programs strengthens the realignment of the visual-vestibular system and promotes sensory integration.

Other Management Considerations

Both objective findings and subjective responses are pertinent to the comprehensive evaluation of a patient with neurological Lyme disease. Together, they reveal the amount and quality of visual stress and discomfort experienced by the patient. Patients often find their symptoms difficult to describe, describe their symptoms vaguely, or become frustrated or defeated due to their visual experiences. A clinician's empathy towards the affective responses of the patient may be affirming and motivating in the rehabilitation process. Additionally, establishing motivational and realistic goals for therapy aids in managing patient expectations. In the above cases, emphasis was placed on improving binocular stability to improve the quality of daily living activities.

Conclusion

Neurological Lyme disease is a poorly understood and complex condition, one that causes debilitating visual dysfunction and is largely unable to be managed with traditional antibiotic Lyme treatments. Effectively classifying neurological Lyme disease under the umbrella term of acquired brain injury, and addressing the condition from a functional visual perspective, allows for the investigation of visual and sensory dysfunctions caused by the condition. Vision therapy is an effective management strategy to decrease binocular instability and improve sensory integration.

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