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CLINICAL VIGNETTE

A Man Dizzy with Lyme

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Introduction

This case involves a patient with Lyme neuroborreliosis in the form of cerebral vasculitis who was noted to have increasing dizziness/vertigo, right sided pulsatile tinnitus with eventual progression to bilateral tinnitus and autophony. Despite aggressive Lyme directed therapy, he continued to decline clinically. Upon further investigation, he was diagnosed with superimposed diagnosis of bilateral superior semicircular canal dehiscence (SSCD) syndrome.

History of Present Illness

A 66-year-old male presented in 2015 with progressive weakness, fatigue, body aches, and memory impairment. After an extensive workup and the input of multiple consultants, he was diagnosed with chronic Lyme disease with central nervous system (CNS) involvement. Brain nuclear MRI spectroscopy confirmed the diagnosis of cerebral vasculitis. Since the Lyme disease diagnosis, he has been on and continues to be treated with multiple types of antibiotics. The Lyme disease remained clinically active with an only partial response to antibiotic therapy.

In 2016, the patient developed new right-sided tinnitus which then deteriorated into bilateral tinnitus. During the subsequent year, his symptoms progressed to right-sided pulsatile tinnitus, aural fullness, mild to moderate autophony, gait unsteadiness, dizziness and abnormal sound perception in the right ear.

At first, the diagnosis remained elusive as some of the new symptoms were attributed to the neurologic/sensorineural complications of the Lyme disease. However, given the progressive symptoms, he was further evaluated. In 2017, the patient underwent extensive testing including high-resolution CT of the temporal bones. The temporal bone and the brain CT showed no evidence of intracranial bleeding, mass or territorial infarction. They did suggest bilateral superior canal dehiscence and he was referred to head and neck surgery. He underwent successful right sided intra-temporal craniotomy and repair of skull base defect. The corrective surgery resulted in a partial improvement of the SSCD related symptoms.

His right-sided tinnitus resolved but he continued somewhat symptomatic with dizziness, left-sided tinnitus and gait unsteadiness and is being considered for a left-sided skull base repair. The Lyme disease related symptoms continued unabated.

Discussion

To date, there are no documented associations between Lyme disease, more specifically neuroborreliosis and SSCD. Late Lyme disease due to tick transmitted spirochete *Borrelia burgdorferi* is usually multifaceted and clinically very challenging. This disease may mimic rheumatologic, neurologic, ophthalmologic, cardiac, dermatologic, psychiatric and hematologic conditions.¹⁻⁴ On the other hand, the SSCD is a newly defined rare entity which was first reported by Lloyd Minor and colleagues in 1998.^{5,6} The SSCD is caused by the dehiscence of the superior semicircular canal which creates a “third window”.⁵ Please refer to Figure 1. The prevalence of SSCD is poorly defined in the literature.⁶

Figure 1.



The creation of the “third window” seen SSCD explains the range of symptoms seen in this disease.⁷ The clinical findings range from autophony, tinnitus (could be pulsatile), low frequency hearing loss, phonophobia, aural fullness to vestibular symptoms of torsional eye movement, disequilibrium and vertigo.⁷ The diagnosis of SSCD is difficult and challenging. The cornerstone of diagnosis requires an abnormal vestibular evoked myogenic potential (VEMP) test and a characteristic (Figure 1) high-resolution CT of the temporal bones.⁸

The etiology of SSCD appears to be multifactorial. Both acquired as well as congenital forms have been described. The presence of this disease in young children and family members, suggests a genetic correlation as relatives share similar skull base morphologies.⁹⁻¹¹ In addition, literature search reveals

loose associations between SSCD and elevated intracranial pressure, obstructive sleep apnea (resulting in dehiscence), trauma, inflammation, Paget's disease and infections.¹²⁻¹⁵ The management of SSCD is multidisciplinary and the mainstay of therapy is surgical reinforcement of the SSC roof, reinforcement of the round/oval window or plugging of the osseous defect.¹⁴

Many articles correlate SSCD with various infectious processes involving ears such as chronic otitis media or the brain such as brain abscesses.^{16,17} Our patient had no prior history of ontological diseases, family history of SSCD, head trauma or prior CNS infection. The only known infectious process in this patient is the neuroborreliosis.

Our patient is the first case of concurrent SSCD and late Lyme disease/neuroborreliosis. This raises the possibility of an association between neuroborreliosis and SSCD. This association remains speculative, as these two entities may be unrelated synchronous events in the same patient. We, however, hypothesize that the symptomatic SSCD in this patient derives from a combination of the antecedent skull base defect and the active neuroborreliosis.

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