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 moderated autophony, gait unsteadiness, dizziness and abnormal sound perception in the right ear.

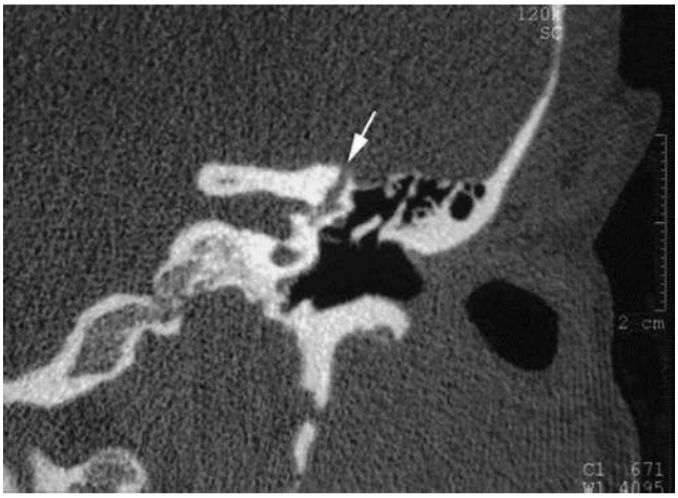
At first, the diagnosis remained elusive as some of the new symptoms were attributed to the neurologic/sensorineural complications of the Lyme disease. However, given the progressive symptoms, he was further evaluated. In 2017, the patient underwent extensive testing including high-resolution CT of the 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 SCCD in this patient derives from a combination of the antecedent skull base defect and the active neuroborreliosis.

REFERENCES

1. **Burgdorfer W**. Lyme borreliosis: ten years after discovery of the etiologic agent, Borrelia burgdorferi. *Infection*. 1991 Jul-Aug;19(4):257-62. Review. PubMed PMID: 1917043.
2. **Ballard HS, Bottino G, Bottino J**. The association of thrombocytopaenia and Lyme disease. *Postgrad Med J*. 1994 Apr;70(822):285-7. PubMed PMID: 8183774; PubMed Central PMCID: PMC2397887.
3. **Logigian EL, Kaplan RF, Steere AC**. Chronic neurologic manifestations of Lyme disease. *N Engl J Med*. 1990 Nov 22;323(21):1438-44. PubMed PMID: 2172819.
4. **Feder HM Jr, Johnson BJ, O'Connell S, Shapiro ED, Steere AC, Wormser GP; Ad Hoc International Lyme Disease Group, Agger WA, Artsob H, Auwaerter P, Dumler JS, Bakken JS, Bockenstedt LK, Green J, Dattwyler RJ, Munoz J, Nadelman RB, Schwartz I, Draper T, McSweegan E, Halperin JJ, Klempner MS, Krause PJ, Mead P, Morshed M, Porwancher R, Radolf JD, Smith RP Jr, Sood S, Weinstein A, Wong SJ, Zemel L**. A critical appraisal of "chronic Lyme disease". *N Engl J Med*. 2007 Oct 4;357(14):1422-30. Review. Erratum in: *N Engl J Med*. 2008 Mar 6;358(10):1084. Agger, WA [added]; Artsob, H [added]; Auwaerter, P [added]; Dumler, JS [added]; Bakken, JS [added]; Bockenstedt, LK [added]; Green, J [added]; Dattwyler, RJ [added]; Munoz, J [added]; Nadelman, RB [added]; Schwartz, I [added]; Draper, T [added]; McSweegan,. PubMed PMID: 17914043.
5. **Minor LB, Solomon D, Zinreich JS, Zee DS**. Sound- and/or pressure-induced vertigo due to bone dehiscence of the superior semicircular canal. *Arch Otolaryngol Head Neck Surg*. 1998 Mar;124(3):249-58. PubMed PMID: 9525507.
6. **Ward BK, Carey JP, Minor LB**. Superior Canal Dehiscence Syndrome: Lessons from the First 20 Years. *Front Neurol*. 2017 Apr 28;8:177. doi: 10.3389/fneur.2017.00177. eCollection 2017. Review. PubMed PMID: 28503164; PubMed Central PMCID: PMC5408023.
7. **Mau C, Kamal N, Badeti S, Reddy R, Ying YM, Jyung RW, Liu JK**. Superior semicircular canal dehiscence: Diagnosis and management. *J Clin Neurosci*. 2018 Feb;48:58-65. doi: 10.1016/j.jocn.2017.11.019. Epub 2017 Dec 7. Review. PubMed PMID: 29224712.
8. **Benamira LZ, Alzahrani M, Saliba I**. Superior canal dehiscence: can we predict the diagnosis? *Otol Neurotol*. 2014 Feb;35(2):338-43. doi: 10.1097/MAO.0000000000000230. PubMed PMID: 24448294.
9. **Heidenreich KD, Kileny PR, Ahmed S, El-Kashlan HK, Melendez TL, Basura GJ, Lesperance MM**. Superior Canal Dehiscence Syndrome Affecting 3 Families. *JAMA Otolaryngol Head Neck Surg*. 2017 Jul 1;143(7):656-662. doi: 10.1001/jamaoto.2016.4743. PubMed PMID: 28384775; PubMed Central PMCID: PMC5824203.
10. **Niesten ME, Lookabaugh S, Curtin H, Merchant SN, McKenna MJ, Grolman W, Lee DJ**. Familial superior canal dehiscence syndrome. *JAMA Otolaryngol Head Neck Surg*. 2014 Apr;140(4):363-8. doi: 10.1001/jamaoto.2013.6718. PubMed PMID: 24526223.
11. **Chilvers G, McKay-Davies I**. Recent advances in superior semicircular canal dehiscence syndrome. *J Laryngol Otol*. 2015 Mar;129(3):217-25. doi: 10.1017/S0022215115000183. Epub 2015 Feb 6. Review. PubMed PMID: 25655361.
12. **Nelson RF, Hansen KR, Gantz BJ, Hansen MR**. Calvarium thinning in patients with spontaneous cerebrospinal fluid leak. *Otol Neurotol*. 2015 Mar;36(3):481-5. doi: 10.1097/MAO.0000000000000552. PubMed PMID: 25122599.
13. **Schutt CA, Neubauer P, Samy RN, Pensak ML, Kuhn JJ, Herschovitch M, Kveton JF**. The correlation between obesity, obstructive sleep apnea, and superior semicircular canal dehiscence: a new explanation for an increasingly common problem. *Otol Neurotol*. 2015 Mar;36(3):551-4. doi: 10.1097/MAO.0000000000000555. PubMed PMID: 25118577.
14. **Bi WL, Brewster R, Poe D, Vernick D, Lee DJ, Eduardo Corrales C, Dunn IF**. Superior semicircular canal dehiscence syndrome. *J Neurosurg*. 2017 Dec;127(6):1268-1276. doi: 10.3171/2016.9.JNS16503. Epub 2017 Jan 13. Review. PubMed PMID: 28084916.
15. **Merchant SN, Rosowski JJ**. Conductive hearing loss caused by third-window lesions of the inner ear. *Otol Neurotol*. 2008 Apr;29(3):282-9. doi: 10.1097/mao.0b013e318161ab24. Review. PubMed PMID: 18223508; PubMed Central PMCID: PMC2577191.
16. **Cho YW, Shim BS, Kim JW, Kim TS, Ahn JH, Chung JW, Lee KS, Yoon TH, Park HJ**. Prevalence of radiologic superior canal dehiscence in normal ears and ears with chronic otitis media. *Laryngoscope*. 2014 Mar;124(3):746-50. doi: 10.1002/lary.24281. Epub 2013 Jul 8. PubMed PMID: 23794324.

17. **Manara R, Lionello M, de Filippis C, Citton V, Staffieri A, Marioni G**. Superior semicircular canal dehiscence: a possible pathway for intracranial spread of infection. *Am J Otolaryngol*. 2012 Mar-Apr;33(2):263-5. doi: 10.1016/j.amjoto.2011.05.006. Epub 2011 Jul 23. Erratum in: *Am J Otolaryngol*. 2012 May;33(3):377. Dosage error in article text. PubMed PMID: 21784554.