

Neuroborreliosis Manifesting with Multiple Cranial Neuropathies

Case Report

Joseph S^{1,2} and Taimur Malik M^{1,2*}

¹Department of Neuroscience, Geisinger Neuroscience Institute, United States

²Department of Neurology, Geisinger Commonwealth School of Medicine, United States

***Corresponding author:** Taimur Malik M, Assistant Professor of Neurology, Geisinger Neuroscience Institute 100 N. Academy Ave. Danville, PA 17822, Tel: 570-271-6590; Fax: 570-271-5874; Email: mmalik@geisinger.edu

Copyright: © 2021 Joseph S, et al. This is an open access article distributed under the Creative Commons Attribution License, which permits unrestricted use, distribution, and reproduction in any medium, provided the original work is properly cited.

Article Information: Submission: 29/12/2020; Accepted: 23/03/2021; Published: 26/03/2021

Abstract

It is imperative to rule out neuroborreliosis in a patient presenting with multiple cranial neuropathies in a Lyme endemic area to prevent long term complications associated with the disease. Interpretation of serological tests is critical. With a recent history of tick bite and positive IgM antibodies, some patients do not have the IgG antibodies against Lyme within 30 days of the exposure. Doxycycline effectively treats Lyme disease but must be given for 28 days.

Keywords: Neuroborreliosis; Multiple cranial neuropathies; Doxycycline

Introduction

Neuroborreliosis is a common manifestation of Lyme disease, usually presents with facial nerve weakness, but other cranial nerves can also be affected. It is essential to rule out other causes of multiple cranial neuropathies before starting the treatment to prevent further complications. About 5-10% of the patient would present with cranial neuropathies [1].

Case Report

A 48-year-old female with no significant past medical history presented to an outside hospital with a one-week history of progressive bilateral facial weakness, increased lacrimation, and difficulty chewing. As per the patient, she was in her usual state of health when she noticed a tick bite about two weeks back. She initially started having a dull bilateral occipital headache after the tick bite, which later resolved after a few days. She denies having any recent fevers, weight loss, sensory or motor weakness in her extremities, or altered awareness. At presentation, her vital signs were stable. General physical examination was unremarkable except for difficulty

in sensorineural hearing in both ears. Neurological examination revealed dis-conjugate gaze, bilateral lower motor neuron facial nerve weakness, and sensorineural hearing loss. Her basic laboratory investigations including complete blood, comprehensive metabolic profile, chest x ray, urine analysis did not show any abnormality, while the Lyme testing showed a positive IgG/IgM antibody value of 7.28 with 1 IgG and 3/3 IgM positive antibodies. The significant IgM bands were 23,39,41. She later underwent an MRI (magnetic resonance imaging) of the brain with and without contrast, which showed enhancement of bilateral facial nerves (canalicular segments), bilateral trigeminal nerves (cisternal segments), and left cochlear nerve, as shown in Figure 1B, 1E, 1F. A spinal tap showed normal CSF (cerebral spinal fluid) protein and glucose with 1 white cell, while the bacterial, viral, and fungal cultures were negative for any infectious etiology. Patient was diagnosed with neuroborreliosis secondary to her positive tick bite history, serological testing, and imaging findings. She was later started on Doxycycline 100 mg twice a day for 28 days, with close follow-up scheduled in the neurology outpatient setting. The patient did not have the follow-up MRI imaging yet.

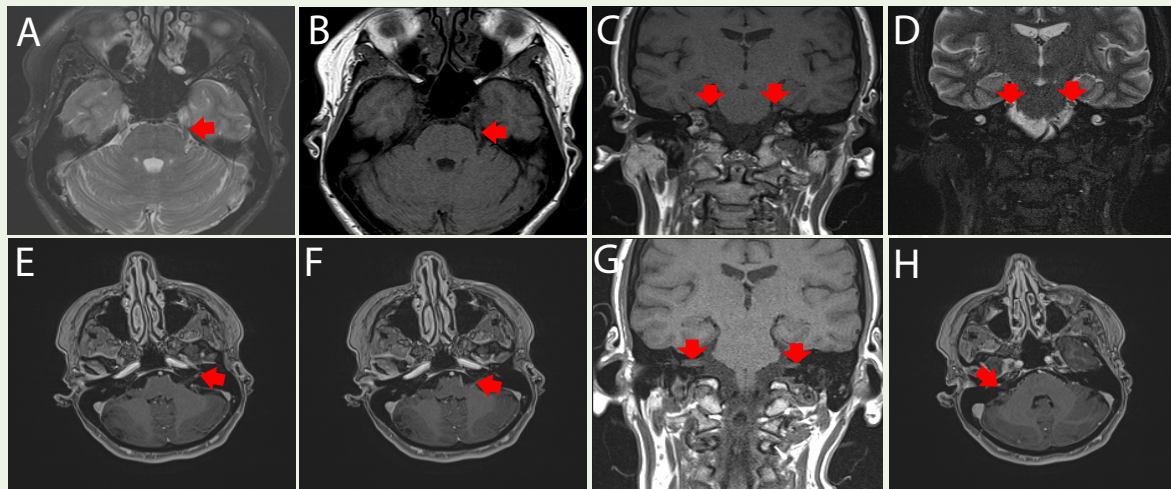


Figure 1: (A&B) Axial T2 fat sat thin sequence of MRI brain with and without contrast showing bilateral trigeminal nerve cisternal segment enhancement with associated T2 hyperintensity as shown with the red arrows. (B) Axial T1 sequence of MRI brain with and without contrast showing bilateral trigeminal nerve cisternal segment as shown with the red arrows. (C) coronal T1 sequences of MRI brain with and without contrast showing bilateral trigeminal nerve cisternal segment as shown with the red arrows (E&F) Axial T2 fat sat thin sequences of MRI brain with and without contrast showing bilateral facial nerve canalicular segment enhancement as shown with the red arrow. (G) Coronal T1 sequence of MRI brain with and without contrast showing bilateral facial nerve canalicular segment enhancement as shown with the red arrows. (H) Axial T2 fat sat thin sequence showing cochlear nerve enhancement as shown with the red arrow.

Discussion

Lyme disease is a tick-borne illness caused by *Borrelia burgdorferi*. About 15% of the patients infected with Lyme disease will have neurological manifestations, including 7th cranial nerve palsy, painful radiculopathy, and meningitis [2]. Careful history about the tick exposure and appearance of erythema migrans (bull's-eye rash) are important clues for the correct diagnosis. The accurate interpretation of the serological tests is important to ensure that proper treatment is instituted to prevent long-term complications to prevent mortality and morbidity.

Although there is a broad differential diagnosis for multiple cranial neuropathies which include vascular causes like brain stem infarcts, cavernous sinus thrombosis, intracranial aneurysms, vasculitis, traumatic causes including skull-based fractures, intracranial dissection, autoimmune conditions like multiple sclerosis, sarcoidosis, acute inflammatory demyelinating polyneuropathy including Guillain-Barré syndrome (GBS) and Miller Fisher variant, infectious etiologies including Human Immunodeficiency Virus (HIV), varicella-zoster, neurosyphilis, chronic fungal meningitis, neoplastic causes like brain stem gliomas, leptomeningeal carcinomatosis, metastatic tumors, metabolic causes like thyroid ophthalmopathy, thiamine deficiency and Melkersson-Rosenthal syndrome [3]. In our patient, we excluded these differentials with the help of an MRI of the brain as shown in Figure 1. CBC, urine analysis and CSF analysis were negative for any infectious etiology but positive for Lyme serology with a spinal tap showing a protein of 45 and normal glucose, normal metabolic profile with normal liver and kidney function tests.

As per the previous guidelines from the Center of Disease Control (CDC), a two-tier testing is recommended which includes to have an initial enzyme-linked immunosorbent assay (ELISA) testing for the

specific antigens, and if the sample is positive or borderline positive it undergoes Western blot testing to confirm the diagnosis. If 5/10 specific IgG bands are present in patients with symptoms of more than 30 days or 2/3 IgM bands present with symptoms of less than 30 days, the Western blot is considered positive [4].

A new modified two-tier testing algorithm has also been approved by the CDC, which is as accurate as the standard two-tier testing utilizing only the enzyme immunoassay (EIA). If the initial test is positive, another EIA is performed to confirm with a different set of antigens [5,6]. Prompt treatment with Doxycycline 200 mg daily for 2-4 weeks or in a patient who is allergic to Doxycycline, Amoxicillin 1.5 gm daily is considered effective, while Azithromycin or Clarithromycin can be used if an alternative agent is needed. Ceftriaxone 2 gm daily is also viewed as efficacious [7,8].

It is important to have a broad differential in a patient with multiple cranial neuropathies, but a concise history, optimal serological markers, and imaging need to be done to prevent long-term complications.

References

- Halperin JJ (2010) Nervous system Lyme disease. *J R Coll Physicians Edinb* 40: 248-255.
- Feder HM Jr, Johnson BJ, O'Connell S, Shapiro ED, Steere AC, et al. (2007) A critical appraisal of "chronic Lyme disease". *N Engl J Med* 357: 1422-1430.
- Zeng W, Geng S, Niu X, Yuan J (2010) Complete Melkersson-Rosenthal syndrome with multiple cranial nerve palsies. *Clin Exp Dermatol* 35: 272-274.
- Lantos PM, Rumbaugh J, Bockenstedt LK, Falck-Ytter YT, Aguero-Rosenfeld ME et al. (2021) Clinical Practice Guidelines by the Infectious Diseases Society of America, American Academy of Neurology, and American College of Rheumatology: 2020 Guidelines for the Prevention, Diagnosis, and Treatment of Lyme Disease. *Neurology* 96: 262-273.

5. Branda JA, Linskey K, Kim YA, Steere AC, Ferraro MJ (2011) Two-tiered antibody testing for Lyme disease with use of 2 enzyme immunoassays, a whole-cell sonicate enzyme immunoassay followed by a VisE C6 peptide enzyme immunoassay. *Clin Infect Dis* 53: 541-547.
6. Mead P, Petersen J, Hinckley A (2019) Updated CDC recommendation for serologic diagnosis of Lyme disease. *MMWR Morb Mortal Wkly Rep* 68: 703.
7. Shapiro E (2014) Clinical practice. Lyme disease. *N Engl J Med* 370: 1724-1731.
8. Pfister HW, Preac-Mursic V, Wilske B, Schielke E, Sörgel F, et al. (1991) Randomized comparison of ceftriaxone and cefotaxime in Lyme neuroborreliosis. *J Infect Dis* 163: 311-318.