ISSN: 2578-3335 (Print) 2578-3343 (Online)

Volume 4 | Issue 1 Article 11

2022

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Recommended Citation

Babbar, Shaili; Galati, Jonathan; Corona, Jasmine; and Zahid, Sohail (2022) "Acute Hemiplegia and Ataxia from Lyme Disease," *Cooper Rowan Medical Journal*: Vol. 4: Iss. 1, Article 11.
DOI: 10.31986/issn.2578.3343_vol4iss1.11
Available at: https://rdw.rowan.edu/crjcsm/vol4/iss1/11

Acute Hemiplegia and Ataxia from Lyme Disease

Acute Hemiplegia and Ataxia from Lyme Disease

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ABSTRACT

Lyme disease is a zoonotic illness caused by Borrelia burgdorferi, which present with skin, joint, heart, and central nervous system complications. Central nervous system manifestations of this disease are common, and typically include meningitis, facial nerve palsies, and radiculoneuritis. In this case report, we present a patient who presented with acute right sided hemiplegia and ataxia with negative neuroimaging findings of stroke, inflammation, or mass. Further investigation with cerebrospinal fluid studies and infectious panels revealed the patient had active Lyme disease. The patient improved his motor function, coordination and sensation with ceftriaxone treatment over a few weeks. Acute hemiplegia and ataxia are rare manifestations of Lyme Disease.

Keywords: Lyme Disease, Hemiplegia, Ataxia, Central Nervous System

BACKGROUND:

Lyme disease is a zoonotic illness caused by the spirochete, Borrelia burgdorferi, which is transmitted by ixodid ticks endemic to North America.[1] This disease can manifest as skin, joint, heart, and central nervous system complications. Often, Lyme disease is classified into three stages based on time from tick bite and symptomatology.[1,2] The early localized stage occurs days to weeks after exposure, in which patients commonly have flu-like symptoms and a characteristic “bulls-eye” rash, erythema migrans.[1] The early disseminated stage occurs weeks to months after the initial bite and is marked by multiple erythema migrans lesions, carditis, a facial nerve palsy, or neuroborreliosis.[1] The late disseminated stage, which occurs months to years after the bite, is associated with Lyme arthritis, memory loss, and peripheral neuropathy.[1] We report the case of an older, healthy man who developed acute right-sided hemiplegia and ataxia from neuroborreliosis.

Clinical Findings:

An 80-year-old man with a history of well-controlled hypertension presented to the emergency department with right-sided arm and leg weakness and numbness, right-sided facial droop with loss of sensation, and diplopia for four days. His review of symptoms was otherwise negative. He denied recent illness, travel, or sick contacts. On initial presentation, the patient had a temperature of 97.1âĎL', heart rate of 84 bpm, blood pressure of 125/55 mm Hg, and oxygen saturation of 95% on room air. His skin was warm, dry, without edema, abnormal pigmentation, bleeding, rash or other lesions. He was alert and oriented to self, location, and date. His language exam had intact naming (pen, watch, glasses, and thumb), speech repetition, and comprehension, without aphasia. His recent and remote memory was intact. He had no evidence of visuospatial neglect. His fund of knowledge had appropriate vocabulary and he was able to follow simple axial and appendicular commands. With regards to his cranial nerve exam, his pupils were equal, round, and reactive to light, and his visual fields were intact to finger counting. He had intact extraocular muscles with full range of movement, saccadic pursuit, and end stage nystagmus to the left side that extinguished. His facial sensation was intact to pinprick and symmetric. He had a right sided facial droop with weak eyebrow raise, inability to fully close his right eye, and asymmetric smile. He was hard of hearing on both ears. He had normal palatal elevation, midline uvula, 5/5 strength in his trapezius muscles, and midline tongue.

His motor exam had normal bulk and tone. The motor exam was notable for Medical Research Council Manual Muscle Testing Scale (MRC) grade 3/5 right shoulder abduction, 3/5 right elbow flexion and extension, 3/5 right finger grip, 2/5 right hip flexion and extension, 3/5 right knee flexion and extension, and 5/5 right foot dorsiflexion and plantarflexion. He had grade 5/5 strength with left shoulder abduction, elbow flexion and extension, left finger grip, left hip flexion and extension, left knee flexion and extension, and left foot dorsiflexion and plantarflexion. The sensory exam was notable for diminished light touch in his right hand, forearm, shin, and thigh. His reflexes were difficult to obtain, but were 1+ in upper extremities bilaterally, 2+ in the left patellar, absent in the right patellar, absent in the Achilles bilaterally, and absent in the adductors bilaterally. On the gait exam, the patient had great difficulty in moving from lying to sitting to standing, and required two person assistance. On ambulation with assistance, he walked with a wide, unsteady gait. He neither exhibited a Romberg sign nor pronator drift. Dysdiadochokinesia and dysmetria were noted on the right side.

Differential Diagnosis:

At the time of initial presentation, the patient's clinical symptoms of acute unilateral weakness and sensation loss in the context of risk factors (advanced age, hypertension) were most concerning for an ischemic or hemorrhagic stroke, which prompted us to pursue immediate CT and MRI imaging.

We then considered infectious and autoimmune etiologies that could explain his presentation. His CSF studies supported a diagnosis of Guillain-Barre Syndrome with Miller Fisher variant,[3] despite denying preceding infectious symptoms or vaccinations. Miller Fisher syndrome, which is marked by an albuminocytologic dissociation on CSF studies, typically presents with ophthalmoplegia, ataxia, and areflexia. It can also be associated with cranial nerve palsies, specifically the facial nerve, and limb motor weakness.[3]

We also considered a broad range of infectious etiologies, including HSV encephalitis, neurosyphilis, cryptococcal meningitis, West Nile Virus encephalitis, and Lyme neuroborreliosis, all of which can have atypical stroke-like presentations. Patients with HSV encephalitis usually have fevers, altered mental status, and seizures. Syphilis is the "great imitator," with a diverse set of clinical signs and symptoms that can mimic most other conditions. Patients with neurosyphilis have varying presentations depending on the stage, but include anything from facial nerve palsies to gummatous diseases to vasculopathy with focal neurologic deficits. Cryptococcal meningitis usually occurs in patients with immunodeficiencies and presents with fever, headache, lethargy, confusion, and light sensitivity. West Nile Virus encephalitis also exhibits acute neurological symptoms, ataxia, and flaccid paralysis, but patients generally also have fever, tremors, headaches, convulsions, and altered mental status. Central nervous system involvement of Lyme disease are common, and typically present as symptomatic meningitis with headaches, fevers, photosensitivity, and neck stiffness.[1] Lyme neuroborreliosis can uncommonly present with cranial nerve deficits, radiculoneuritis, intracranial hypertension, acute mononeuropathy, encephalopathy, and encephalomyelitis.[1] Our patient's symptoms did not exactly match the typical clinical presentations for each of these conditions, so we sent a wide infectious serologic workup, performed a lumbar puncture, and sent the cerebrospinal fluid for additional analyses.

Diagnostic Assessments:

The complete blood count, basic metabolic panel, and hepatic panel showed no abnormalities. A non-contrast head CT (NCHCT) and MRI Brain with and without gadolinium enhancing contrast were first performed due to concern for stroke.

No acute haemorrhage, infarct, or mass was noted on the NCHCT (Figure 1) or the MRI Brain (Figure 2). The MRI showed demonstration of multiple scattered foci of abnormal white matter FLAIR hyperintensities without associated susceptibility artifact, diffusion signal, or associated contrast enhancement (Figures 2). Some of the foci were associated with low T1 signals. Overall the findings were nonspecific, and read as those that may be seen in the setting of chronic microvascular disease with a differential including sequela of prior infection/inflammation. Diffuse parenchymal volume loss was also noted. The MRA Head and Neck were grossly normal with no evidence of vascular stenosis, occlusion,

aneurysm, vascular malformation, abnormal cord signal, or abnormal contrast enhancement. He obtained another MRI with and without gadolinium contrast on his fourth day of hospitalization which showed no changes from his first scan (Figures 3).

The patient underwent a significant infectious workup, including two lumbar punctures during hospitalization. On the first lumbar puncture, the CSF had a slight pleocytosis (53 WBCs), elevated protein (168.1 mg/dL), elevated glucose (82 mg/dL), elevated albumin (102.7 mg/dL) and no growth of organisms on gram stain (Table 1). The patient's plasma glucose was 171 mg/dL. The following CSF studies were also sent – Lyme, West Nile Virus, Biofire Meningitis/Encephalitis panel, Cryptococcus, EBV, HSV, VZV, Mycoplasma, VDRL, paraneoplastic panel, autoimmune panel, oligoclonal bands, CSF flow, and CSF culture. The following bloodwork was also performed – Lyme antibody, Treponema antibody, West Nile Virus antibody, Cryptococcus antigen, HIV, Quantiferon, Mycoplasma IgG antibody, and Lyme Western Blot. A second lumbar puncture two days later after initiation of IVIG and showed a normal white blood cell count (21 cells/mcL), normal glucose (65 mg/dL), and elevated proteins (178.5 mg/dL) (Table 1).

Therapeutic Interventions:

At first, the patient was started on IVIG for a suspected atypical presentation of Miller Fisher variant GBS. While waiting for the remaining CSF studies, the serum Lyme antibody returned positive from a chemiluminescence immunoassay (Lyme Index IgG 6.91, IgM 6.41, ref < 1.10) (Table 2). He was then started on intravenous ceftriaxone 2 grams every 12 hours for empiric treatment of Lyme neuroborreliosis on his third day of hospitalization. The remaining studies were notable for positive CSF Lyme antibodies from an ELISA polyvalent assay (Lyme optical density 1.198, ref < 0.107), as well as positive IgM and IgG Lyme antibody bands on the Western Blot (Table 2). The rest of the infectious, autoimmune, and paraneoplastic studies in the CSF and blood were negative. The patient was transitioned to ceftriaxone 2 grams every 24 hours to complete a total of 28 days of antibiotic therapy.

Follow-up and Outcomes:

The patient was hospitalized for a total of 15 days and discharged home to complete his ceftriaxone treatment and resume his physical therapy services, as per his request. Upon discharge from the hospital (day 13 of antibiotic therapy), he had no deficits to gross or fine sensation on his right or left side. His motor exam on the right side of his body was notable for 3+/5 right hip flexion, 4/5 right knee flexion and extension, and 4+/5 right elbow flexion and extension, all improved from prior.

He returned for an outpatient visit to both neurology and infectious disease 36 days after he initiated antibiotics (8 days after he completed his antibiotic course). The patient felt that he had been improving since discharge, but still had some weakness with his right leg and was using a rollator to walk around his neighborhood. His motor exam on his outpatient visit was improved from discharge with 5/5 right

shoulder abduction, 5/5 right elbow flexion and extension, 3+/5 right hip flexion, 4+/5 right knee extension, 5/5 right knee extension, and 5/5 right foot dorsiflexion and plantarflexion. Light touch was normal and symmetric in bilateral upper and lower extremities. His coordination was intact with normal finger-nose-finger tests bilaterally and no dysdiadochokinesia. He was able to ambulate with his rollator, but favored his left leg. Because of his significant improvement in the clinical exam, further diagnostic imaging and antibiotic treatment was deferred. The patient continued home physical therapy for the next six months and had gradual improvement in his ambulation.

DISCUSSION:

In this case report, we present a patient with acute hemiplegia and ataxia secondary to suspected Lyme neuroborreliosis. His symptoms greatly improved with intravenous ceftriaxone treatment. The neurologic manifestations of Lyme disease are variable and common, affecting 10-35% of individuals of those infected.[4] Between 25 to 50% of patients with neuroborreliosis recall a previous tick bite, and the time lapse between tick bite and the onset of neurologic symptoms can range from weeks to months.[4] The most common nervous system symptoms include lymphocytic meningitis, facial nerve palsies, and radiculoneuritis, which occur in the early disseminated phase of the virus.[1,2] In later stages of the disease, chronic encephalomyelitis, peripheral neuropathy, and encephalopathy are observed.[1,2]

There have been rare case reports of Lyme disease resembling stroke-like hemiplegia in which patients developed sudden onset facial palsy and one-sided weakness.[5–9] These patients had various presentations in their neuroradiographic imaging. For example in Deloizy et al, the patient had right sided capsulo-thalamic hypodensity on the CT scan and edema of the thalamus and right capsule evident on MRI. However, in Zhang et al, the patient had several small infarcts bilaterally in the parietal lobes, but no recent events as noted by diffusion weighted imaging.[9] Our patient's imaging showed scattered punctate regions of enhancement on T2 FLAIR imaging. It was difficult to ascertain whether these symptoms represented chronic microvascular disease or sequelae of infection in the brain, which prompted us to perform additional testing. Similarly, there have been few case reports of Lyme infection causing acute ataxia.[10–13] Similarly in these cases, there was heterogeneity in the radiographic imaging with multifocal areas of subcortical white matter hyperintensity in some patients[10,11] and no significant abnormalities in others.[12,13] To the best of our knowledge, this is the first case report of an individual who presented with both acute hemiplegia and ataxia.

Once neuroimaging workup is negative, the key to diagnosis in these previous reports and with our patient was an early lumbar puncture and CSF panel. The frequency of lumbar punctures has decreased to around 20% since the adoption of CT scans, which creates risks of missed treatable central nervous system

infections.[14]

In our patient, there was no active lesion observed with brain imaging using CT or MRI that would suggest hemiplegia or ataxia. However, central nervous system infections from B. burgdorferi only rarely cause significant parenchymal inflammation that could be visible on MRI.[15] If visible, said inflammation would resemble focal areas of increased T2 signal and increased contrast enhancement on diffusion weighted imaging.[16] A diagnostic PET imaging study could have potentially shown active inflammation and hypermetabolic areas, but this test was not performed in our patient.[17]

The pathophysiology of our patient's clinical presentation is not fully understood. The Rhesus monkey infected with B. burgdorferi has been an important animal model to study the nervous system manifestations of Lyme disease.[18–20] After inoculation with B. burgdorferi, these Rhesus monkeys developed nerve-sheath fibrosis in the spinal canal, focal demyelination, perivascular lymphocytic infiltration of the spinal cord and peripheral nerves, and necrosis in sensory ganglia.[19,20] These Rhesus monkeys developed a heterogeneous constellation of neurologic symptoms depending on the extent, location, and duration of B. burgdorferi infection,[19,20] which may explain why neurologic symptoms in patients with Lyme neuroborreliosis are so heterogeneous. There is also evidence that B. burgdorferi entrance into the central nervous system leads to a rapid production of cytokines, notably CCL19, CCL21, CXCL12, and CXCL13, triggering a local autoinflammatory process.[15] Other mechanisms have also been proposed, including but not limited to circulating immune complex deposition causing demyelinating disease[21] or neurotoxin release after contact with the virus.[15]

CONCLUSION

Lyme neuroborreliosis can mimic many nervous system disorders.[1] Upon our review of the literature, there have few case reports of acute hemiplegia or acute ataxia caused by Lyme disease.[5–7] While we do not recommend screening for Lyme infection in patients with acute focal neurological deficits,[6] we do recommend early lumbar puncture and CSF studies once neuroimaging is negative, as early administration of antibiotics can prevent further neurologic complications.

Patient Perspective

"I am incredibly grateful to receive the care I received at the hospital. I was first scared about my medical condition. All the doctors and nurses at the hospital were professional, respectful, and caring. I feel much better now. I am amazed that so few people in the world from my disease had the symptoms I had. I am happy to share my story to the public so other doctors and patients can learn from my experience."

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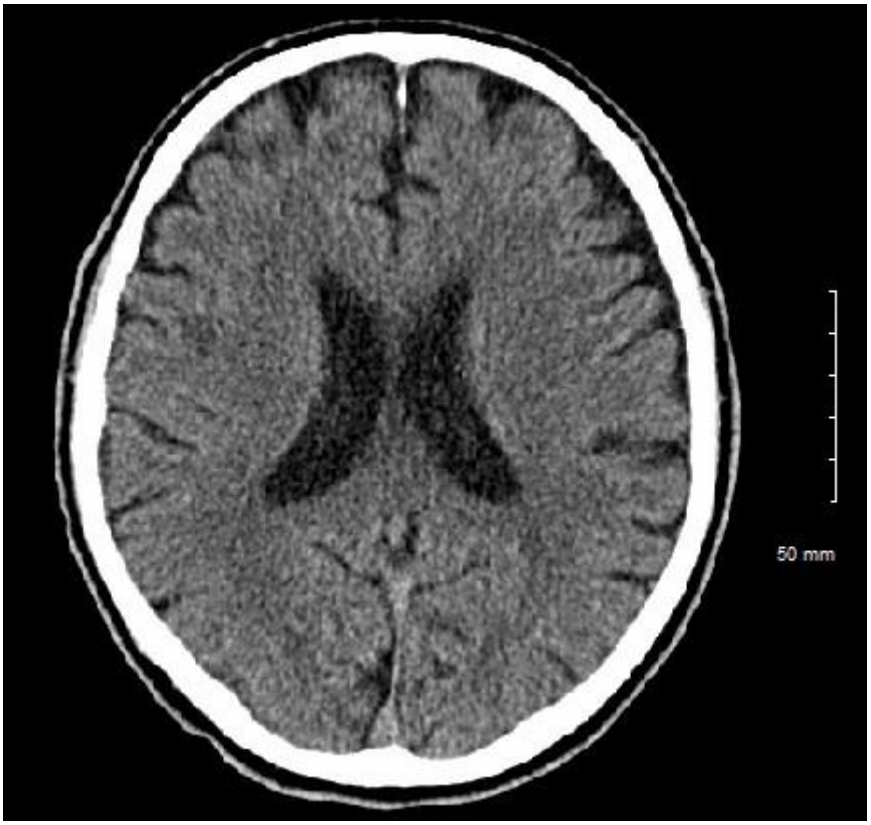

Figure 1 Noncontrast Head CT showing no acute haemorrhage, infarct, or mass.

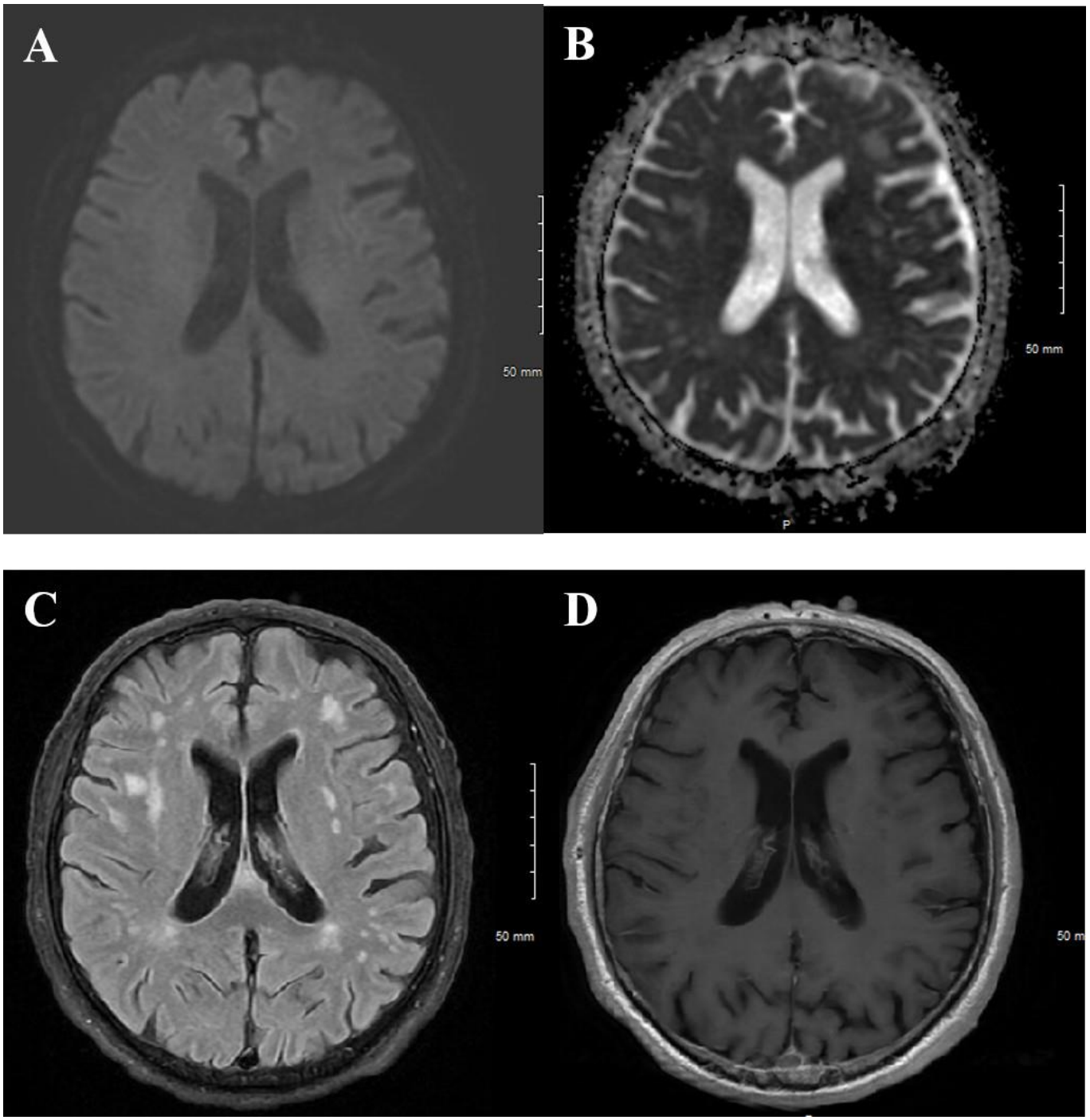

Figure 2 Axial DWI (A) with no signal rich areas and Axial ADC (B) with nocorresponding low signal intensity areas to indicate stroke. T2 FLAIR (C) withscattered punctate regions of enhancement. T1 with contrast (D) with no activeareas of enhancement.

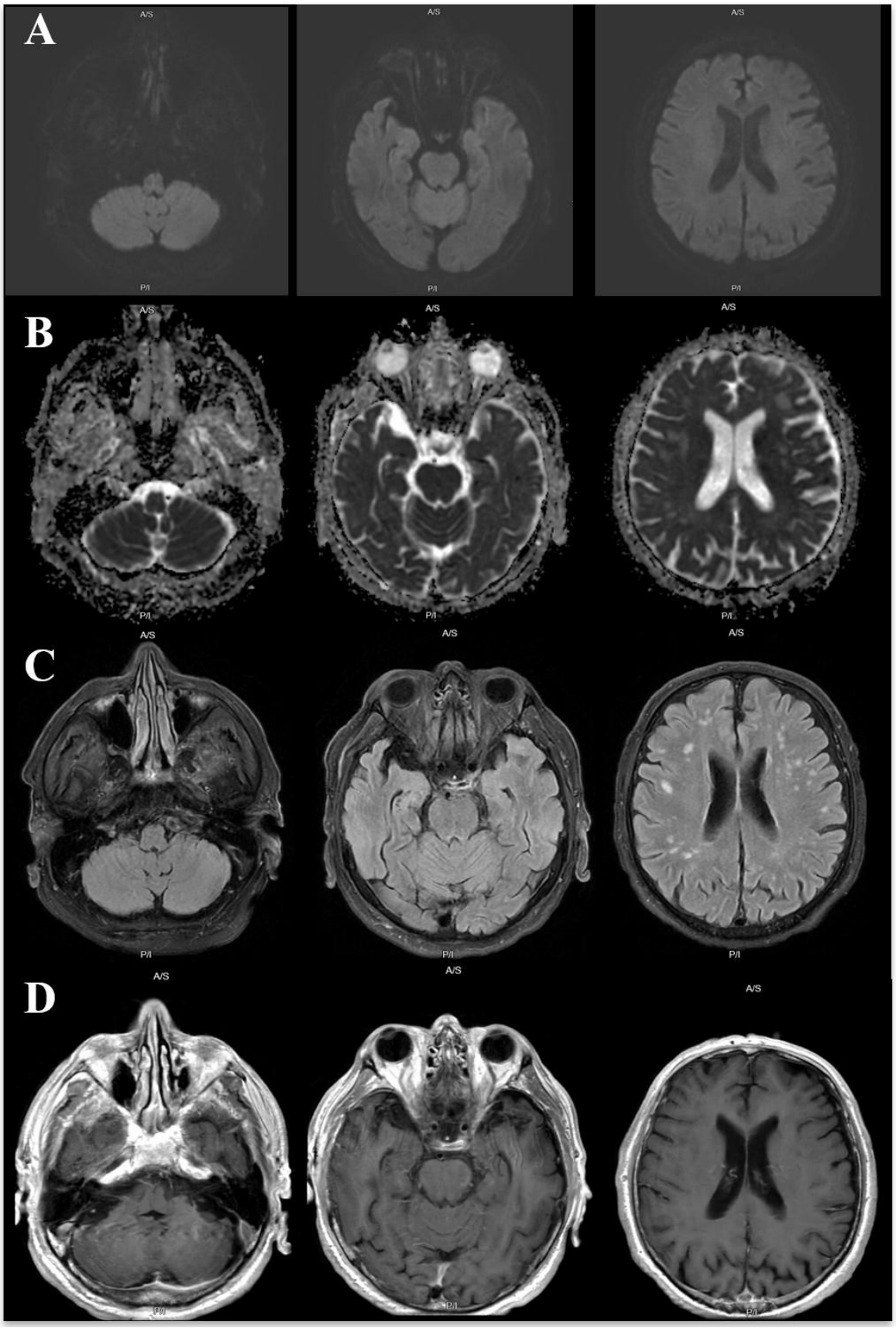

Figure 3 Repeat MRI imaging on fourth day of hospitalization showing axial DWI (A) with nosignal rich areas, axial ADC (B) with no low signal intensity areas to indicatestroke, T2 FLAIR (C) with redemonstration of scattered punctate regions ofenhancement and T1 with contrast (D) with no active areas of enhancement.

Table 1 CSF Serologies

Tube	Initial Lumbar Puncture	After 2 Days IVIG
Color	Colorless	Colorless
Appearance	Clear	Clear
WBC Count	53 cells/mcL	21 cells/mcL
RBC Count	4 cells/mcL	10 cells/mcL
Polys, CSF	2%	4%
Lymphs	82%	83%
Monocytes	16%	14%
Glucose	82 mg/dL	65 mg/dL
Protein	168.1 mg/dL	178.5 mg/dL
Albumin	102.7 mg/dL	

Table 2 Lyme Studies

Lyme CSF Antibody	Reactive Optical Density 1.198 (ref <0.107)
Lyme VISE IgG/IgM AB	IgM: 6.41 (ref < 1.10) IgG: 6.91 (ref < 1.10)
Lyme Western Blot IgM	Positive
Lyme Western Blot IgG	Positive