

An Unwonted Cause of Acute Cerebellar Ataxia in a Young Lady: A Rare Path to Lupus

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Abstract

Neuropsychiatric manifestations are commonly observed in systemic lupus erythematosus (SLE) patients. In particular, neurological involvement is known to be more common in patients with positive anticardiolipin antibodies and lupus anticoagulants. Nevertheless, cerebellar ataxia has rarely been reported, especially as the first clinical manifestation of neuropsychiatric systemic lupus erythematosus (NPSLE). Cerebral vascular dead tissue or ischemia, vasogenic oedema and counter acting agent intervened cerebral vasculopathy or vasculitic process have been assumed as potential aetiologies of intense cerebellar ataxia connected with SLE. We report the clinical and radiological features of a 38 years old diabetic lady who developed a rapidly progressive cerebellar syndrome as first sign of SLE; no other cause explaining her cerebellar ataxia was found. Imaging of cerebrum in this tolerant uncovered unmistakable cerebellar decay. She was treated with initial pulse intravenous methylprednisolone later oral prednisolone and oral mycophenolate mofetil. There could have been no further movement of her neurological signs after the commencement of treatment. The presence of a cerebellar disorder with obscure etiology with related highlights of conceivable foundational immune system brokenness, ought to be considered in clinical practice for fitting demonstrative workup to give powerful restorative choices.

Introduction

Neuropsychiatric manifestations are present in 50-70% of the patients with systemic lupus erythematosus (SLE) and include a wide variety of central and peripheral neurological manifestations [1]. Cerebellar ataxia is one of the less portrayed neurological appearances of primary lupus erythematosus (SLE). Its prevalence in SLE has been estimated to occur in less than 2% of the cases, even more uncommon as the first clinical manifestation of such autoimmune systemic condition [2-4]. Several possible aetiologies of acute cerebellar syndrome related to SLE have been postulated, including cerebral infarction or ischemia, antibody-mediated dysfunctions and vasculopathy or vasculitis. The greater part of the patients present with intense to subacute beginning cerebellar disorders. Some of them supposedly had vasculitic infarcts in cerebellum on attractive reverberation imaging (MRI). However, the presence of cerebellar atrophy at the outset is even rarer [3]. Volume loss of cerebellum cannot be traced to the known pathogenic mechanisms of lupus. Although auto antibodies are present in such patients, there is no specific pattern attributing to the condition. Anti-Purkinje cell immunizer 5 and anti-neuronal cell antibody6 have been portrayed in the writing without large-scale approval. We report the clinical and radiological features of a patient who showed signs of acute cerebellar syndrome associated to MRI evidence of the right cerebellar hemisphere atrophy. Other possible causes, including infective, metabolic or paraneoplastic causes, drugs, alcohol and demyelinating diseases, are excluded by medical history, physical examination and appropriate investigations. The cerebellar ataxia, which was the main clinical feature in our patient, improved following pulse intravenous steroid administration.

Case Report

A 38 years old diabetic, non-alcoholic pleasant Bangladeshi lady, not known to have any other morbidities or not on any other medications admitted to our hospital with the complaints of 4 days history of vertigo, vomiting and unsteady gait. She had no visual symptoms, headache, convulsion, altered consciousness, loss of consciousness sphincteric disturbances, fever, weakness of any limbs, speech disturbances, hallucination, or other neurological symptoms. No family history of neurological disease was reported. She has been treated for diabetes in the previous 4 years with oral hypoglycemic drug and diabetes was well controlled. There was no history of any pedal swelling, nocturia, hematuria, oliguria, psychosis, menstrual irregularity, oral ulcer, hair loss, joint pain, photo sensitive rash, any abortion, chronic drug usage(including lithium and anti convulsants), exposure to any known toxin or addiction. On admission, the patient's pulse was 80 beats/min, blood pressure 125/70 mmHg, respiratory rate 16/min and SpO2 95% while inhaling room air. Neurological examination showed marked limb and truncal ataxia. She presented kinetic tremor involving right arms and dysdiadikokinesia. Horizontal nystagmus was also present with fast phase towards right. She had an ataxic, wide-based gait with tendency to fall towards right and was unable to perform tandem walk. Deep tendon reflexes were normal and symmetrical. No muscle weakness or sensory disturbance was noted. Mental functions, speech, cranial nerve examination, including ocular movement and fundus examination were normal. Initial blood tests including complete blood count, kidney function test, liver function test, random blood sugar, HbA1c, hepatitis profile, TSH were unremarkable. Urinalysis was normal with no presence of protein, glucose, red blood cells, white cell count or cellular casts. Chest

radiograph did not show any abnormalities. Cranial magnetic resonance imaging (MRI) with gadolinium ruled out acute ischemic infarct, parenchymal hemorrhage, mass lesion and demyelinating process. More specifically, the cranial MRI failed to reveal any lesion particularly in the posterior fossa that could support the findings of acute cerebellar ataxia. But right cerebellar hemisphere seemed atrophied (Figure 1,2). In a perfect world, a positron outflow tomography or a useful MRI should be possible to show areas of brokenness.

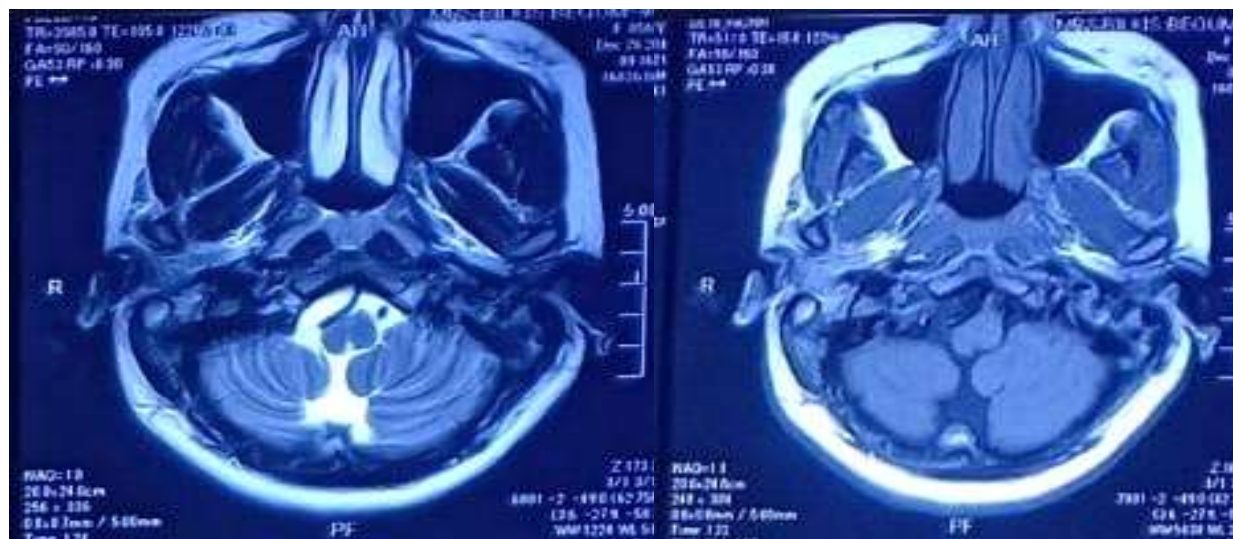


Figure 1,2: MER of brain revealed right sided cerebellar atrophy. There was no evidence of ischemic change.

She was started treatment symptomatically with intravenous prochlorperazine, oral flunarizine and betahistine but she failed to improve in next 2 days. There after CSF study was done which revealed normal opening pressure, cell counts, biochemistry, gram stain, culture and ADA (adenosine deaminase). In the absence of an infectious or vascular cause, the work-up for an autoimmune disease was pursued. Anti-nuclear antibody by immunofluorescence in HEp-2 cells was strongly positive with fine speckled pattern. Anti-dsDNA antibody was also positive 124 U/ml (positive >75 u/ml). ENA profile, c-ANCA, p-ANCA, anti phospholipid antibody were negative. There was low C3 value of 65 mg/dL and low C4 value of 6 mg/dL (normal C3 is 80–178 mg/dl, for C4 is 12–42 mg/dl) The patient thus fulfilled four of the 11 Systemic Lupus International Collaborating Clinics Classification (SLICC) criteria.

A final diagnosis of lupus cerebellitis with right sided cerebellar atrophy was made. Treatment was initiated with injection methylprednisolone (MP) 1 g i.v. 5 days followed by oral prednisolone 1mg/kg body weight along with 2 gram daily of oral mycophenolate mofetil. The patient answered expeditiously with progressive enhancement of all cerebellar elements inside the resulting 10 days. After 3 months of follow-up, her neurological features improved. As a conclusion, her cerebellar decay can be credited to SLE.

Discussion

Intense cerebellitis is a fiery problem most regularly found in kids. It is rare in adults. The pathophysiology is not completely understood. The etiology is regularly obscure however might be optional to drugs, paraneoplastic or parainfectious causes, or a particular microbe (e.g., EBV, HSV, or *Mycoplasma pneumoniae*), headache (88%), nausea and vomiting (88%), and fever (71%). Altered consciousness is seen in only 29% of patients. Treatment depends on the hidden reason and frequently comprises of antimicrobial and antiviral treatments and steroids.

Intense cerebellar brokenness in SLE can be because of SLE movement itself as a piece of neuropsychiatric SLE (NPSLE), or can be because of intense dispersed encephalomyelitis, diffusely penetrating glioma or lymphoma, drug-related fiery cycles or vasculitis.⁷ Clinical course and MRI picture of these cases are characteristic, and were absent in our present case.

The estimated prevalence of neuropsychiatric signs in SLE is between 12% and 95%.⁸ According to the 1999 American College of Rheumatology consensus statement for NPSLE, 19 neuropsychiatric syndromes have been defined. Further arrangement included diffuse mental or neuropsychological signs and central neurological disorders. Localized central nervous system (CNS) involvement is represented by focal NPSLE [9]. However, cerebellar symptoms or ataxia, such as those seen in our patient, has not been described previously as manifestations in the NPSLE spectrum.

The CNS is viewed as immunologically interesting because of the presence of firmly controlled and exceptionally prohibitive blood-mind and blood-CSF hindrance. Strangely, the two mind districts specially designated via autoimmunity are the limbic framework and the cerebellum. Numerous immune-mediated diseases affect the cerebellum and the mechanisms for each differ. These may include paraneoplastic cerebellar degeneration, Miller Fisher syndrome, postinfectious cerebellitis and cerebellar ataxia associated with connective tissue diseases [9]. Interestingly, cerebellar involvement in SLE occurs only in less than 2% of reported cases [6,10]. Possible causes of cerebellar ataxia related to SLE are cerebral ischaemia, vasogenic oedema and antibody-mediated dysfunction [10]. Because specific markers of cerebellar involvement in SLE have not been validated, a response to therapy can be accepted as a surrogate marker of immunopathogenesis [11]. Ahmed et al. investigated fifteen instances of patients with SLE giving cerebellar side effects. They found that everything except one case happened in females with 80% of patients being somewhere in the range of 15 and 34 years old. Cerebellar contribution was available at the hour of SLE conclusion in five patients, and, except for one quiet who was determined to have SLE 12 years after cerebellar brokenness was noted, beginning was 1 month to 14 years after SLE finding.

The examination of decision for cerebellar ataxia is cerebrum MRI.⁷ Functional examinations like positron outflow tomography (PET), practical MRI, or single photon discharge mechanized tomography (SPECT) can exhibit sketchy areas of brokenness in mind regions unaffected on regular MRI, which recommends an uncoupling of metabolic cycle free of obstacle to blood stream. The instrument of these metabolic disturbances is obscure at this point. For our situation, customary MRI bombed additionally to uncover any sore that could make sense of the intense cerebellar ataxia. Because of serious monetary

imperatives and strategic issues we were unable to carry out useful mind imaging in our patient. Prompt treatment with i.v. methylprednisolone reverses the cerebellar dysfunction in most cases, as was evident in our case. Delay in initiation of therapy is associated with poorer prognosis [12].

Subcortical and periventricular white matter lesions indicative of small infarcts or edema are commonly observed in the MRI scans of adults with SLE [13]. Quantitative volumetric measures of cerebral and corpus callosum atrophy in adult patients progress over a relatively short period of time and correlate with disease duration, history of NPSLE, aPL positivity and cognitive impairment.¹⁴ Cumulative corticosteroid dosing has been associated only with gray matter atrophy, in particular hippocampal and cerebral atrophy¹ [14-16]. In a recent study of newly diagnosed adult patients with SLE, cerebral atrophy was found in 18% of patients, probably as a result of disease-associated axonal/myelin loss, and was unrelated to higher corticosteroid dosing [17]. Although cerebellar changes are associated with neurocognitive deficits in other inflammatory and ischemic disorders, there are minimal data on cerebellar involvement in patients with SLE [2,18]. In a very recent study of pediatric lupus patients, researchers have found cerebral and cerebellar volume losses in most of the brain MRI of pediatric patients with NPSLE manifestations within the first 4 years of disease presentation [18]. Cerebral and cerebellar volume loss and corpus callosal atrophy occurred in newly diagnosed patients with neurologic manifestations prior to steroid exposure [18]. This may reflect sequelae of active NPSLE and not just physiologic corticosteroid effects. Less prominent volume loss in patients with previous nephritis may be related to more aggressive control of systemic inflammation or vasculopathy by previous immunosuppressive agents [18].

As far as anyone is concerned, the assessment of pooled data viewing patients with NPSLE introducing as cerebellar ataxia has not been finished in the writing. However, multiple case reports on the subject have been cited since 1988 [12]. The patients described are female and between 14 and 41 years old. These individuals were previously diagnosed with SLE, eventually developing cerebellar ataxia as a neuropsychiatric manifestation of the disease [6,12,19]. Greater part had imaging proof, uncovering a cerebellar infarct, singular sores in the intersection between the pons and the medulla, or cerebellar decay.

Most reports of cerebellar ataxia in SLE showed reaction to immunosuppression with high-portion corticosteroid. Immunomodulators such as azathioprine, cyclophosphamide or hydroxychloroquine were also beneficial. Subsequently, greater part of patients had great results with progress of neurological side effects after treatment.

The patient's neurological signs suggested a disorder affecting right hemispheres of the cerebellum. The rhythm of the sickness movement could be made sense of by a demyelinating problem, a neoplasm or a vasculitic condition. In this persistent, the chance of a first assault of demyelinating messes like different sclerosis was thought of, yet there were no interesting elements on mind imaging. Last analysis of the patient as SLE was affirmed with the assistance of immune system markers. Reports of cerebellar ataxia in lupus demonstrated response to immunosuppression with high-dose intravenous corticosteroid.²⁰ In rare instances, a biopsy of cerebellum revealed dense microglial infiltration and perivascular

neutrophilic invasion [21]. As unambiguous markers of cerebellar friendship in SLE have not been approved, a reaction to treatment can be acknowledged as a proxy marker of immunopathogenesis. Nonetheless, treatment with mycophenolate mofetil in such a circumstance has not been accounted for. So to finish up, we report a fascinating. So to close, we report an intriguing instance of cerebellar decay with regards to SLE with clinical reaction to a joined routine with mycophenolate mofetil and oral steroid.

Conclusion

Lupus cerebellitis is an exceedingly rare disorder that needs to be considered in any patient presenting with ataxia that persists despite treatment of other potential underlying causes. The appearance of an acute or subacute cerebellar syndrome with no obvious cause, especially together with emerging associated autoimmune systemic features during the clinical course, should suggest for possible SLE. Such diagnostic hypothesis, albeit not frequent, is crucial due to implications for clinical practice in order to provide a prompt therapy and to improve clinical outcome.

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