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Case Report *Volume 5 Issue 3*

Anterior Vermian Syndrome in SLE - A Case Report and Review of Literature

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Abstract

Neurological and psychiatric manifestations are present in 50-70% of patients with SLE Neurological manifestations involve focal or diffuse CNS involvement and the peripheral nervous system. The prevalence of cerebellar involvement in SLE is 2%. The clinical types of cerebellar involvement include acute pan-cerebellar and focal cerebellar involvement. We report a case of Anterior Vermian Syndrome (AVS) in SLE.

Keywords: Systemic Lupus Erythematosus; Cerebellar Atrophy; Ataxia

Abbreviations

AVS: Anterior Vermian Syndrome; TCDC: Total Cell Differential Count; LFTs: Liver Function Tests; RFTs: Renal Function Tests; PCR: Polymerase Chain Reaction; CSF: Cerebrospinal Fluid.

Introduction

Although neuropsychiatric manifestations are present in 50-70% of patients with systemic lupus erythematosus, cerebellar involvement occurs in less than 2% of cases. Cerebellar atrophy has rarely been reported [1], although cortical atrophy has been detected. The clinical types of cerebellar involvement include mostly pan-cerebellar or focal cerebellar. Here, we report an isolated anterior vermian involvement with cerebellar atrophy.

Case Report

A 25-year-old female patient was diagnosed with SLE in 2008 based on arthritis, fever, and positive ANA, anti-dsDNA, anti-Smith, anti-RNP, anti-Ro, and anti-La antibodies. She had renal involvement, and a renal biopsy revealed membranous nephritis, stage V, with segmental sclerosis and C1q positivity. A cardiac evaluation revealed severe pulmonary artery hypertension. She was treated with hydroxychloroquine, steroids, and later mycophenolate mofetil [2]. In December 2021, she presented with acute onset of unsteadiness of gait, vomiting, and double vision. On examination, the patient had upbeat nystagmus and truncal ataxia but no incoordination of upper and lower limbs or dysarthria. Her higher mental function examination and spino-motor system were within normal limits.

Initial blood investigations revealed anaemia, normal total cell differential count (TC DC), and platelet count. Liver function tests (LFTs) and renal function tests (RFTs) were normal. The cerebrospinal fluid (CSF) analysis was within normal limits. CSF multiple polymerase chain reactions (PCR) did not show any evidence of infection. Anti-NMO and CSF Ig oligoclonal bands were negative. Anti-TPO antibodies were negative. Tests for HIV, HBsAg, and anti-HBc were negative. Chest X-ray, thyroid ultrasound, CT of the chest, thorax, and abdomen, and echocardiogram were all within normal limits. Anti-Yo, anti-Hu, and anti-Ri antibodies were not found. She

was treated with pulse steroids for three days, followed by oral prednisolone over the next four weeks and continued on a maintenance dose. After two months of treatment, her neurological symptoms improved.

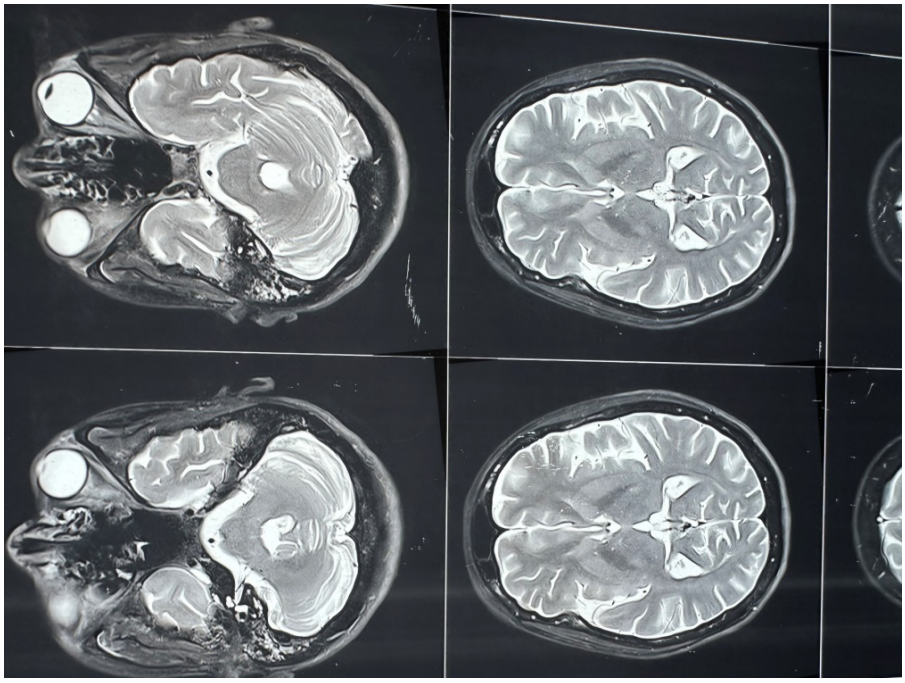

Figure 1: MRI T2 Axial images showing mild cerebellar atrophy with dilated fourth ventricle.

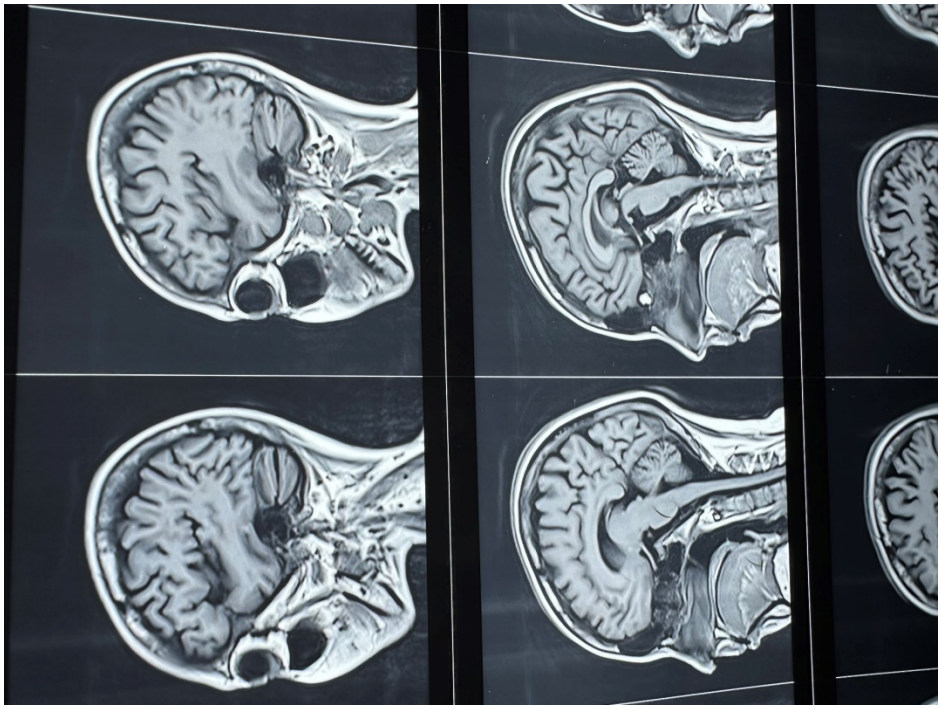

Figure 2: MRI brain T1 saggital image showing cerebllaer and vermian atrophy.

Discussion

The American College of Rheumatology recognizes 19 different NPSLE presentations [3]. NPSLE can be focal or diffuse, and clinical manifestations may range from cognitive dysfunction to acute confusional state, seizures, psychosis, cerebrovascular disorder, movement disorders, peripheral neuropathy, and myelopathies. Isolated cerebellar involvement has not been included in CNS manifestations. Several cases of cerebellar involvement have been reported in the literature, which include pan-cerebellar involvement, unilateral cerebellar involvement, or cerebellar symptoms due to involvement of cerebellar connections in the brainstem [4].

We report a case of isolated anterior vermian syndrome (AVS) in SLE. Our patient presented with acute non-rotatory dizziness and unsteadiness of gait. Neurological examination showed upbeat nystagmus. She could stand unaided but showed side-to-side swaying in natural and tandem gaits and during the Romberg test with feet opposed [5]. There was no dysarthria, tremor, or gaze palsy. The rest of the neurological examination was normal. Most of the cerebellar lesions in SLE reported were pan-cerebellar with acute or sub-acute onset, occurring as an isolated presentation or in association with extra-cerebellar neurological manifestations. A case reported by Appenzeller S, et al. [6] involved only vermian involvement, and the patient's brain MRI showed an isolated lesion in the brainstem.

Previous studies suggested that cerebellar infarction and vasculopathy are possible pathogenetic mechanisms involved in cerebellar ataxia in SLE. In our case, cerebellar ischemia was less likely due to the absence of typical ischemic changes in MRI. The occurrence of cerebellar atrophy in a patient with SLE is exceptional. We found only four cases after searching the PubMed database. In these cases, high-dose corticosteroid therapy, including methylprednisolone pulse therapy, was the main treatment and obtained a relatively good response [7]. In the case reported here, anterior vermian syndrome developed over days to weeks, associated with vermian atrophy and a normal CSF protein level. Cerebellar atrophy was seen in our patient on her initial MRI without focal lesions, reflecting a post-inflammatory sequence. Prompt treatment with intravenous methylprednisolone improved most of her cerebellar symptoms in our case and in cases reported in the literature. It has been suggested that an antibody-mediated reaction could explain her cerebellar symptoms [1]. The possibility of paraneoplastic cerebellar ataxia is less likely due to a normal paraneoplastic antibody panel and negative results for primary malignancy screening. We could not perform anti-neuronal antibody testing due to financial constraints. Some cases subscute cerebellar ataxia was due to antineuronal antibody [8].

A recent study showed newly diagnosed SLE patients showed cerebellar atrophy in 18% of cases, probably due to disease-associated axonal/myelin loss [9]. A recent study of pediatric lupus patients revealed cerebral and cerebellar volume losses in most brain MRIs of pediatric patients with NPSLE manifestations within the first four years of disease presentation. Cerebral and cerebellar volume loss and corpus callosum atrophy occurred in newly diagnosed patients with neurological manifestations before steroid exposure.

Conclusion

Acute Vermian Syndrome (AVS) in SLE is rare. Our case highlights the importance of performing an MRI for SLE patients with cerebellar signs. Although the significance of specific anti-neuronal antibodies in CNS lupus manifestations is still unclear, high-dose corticosteroid therapy is expected to improve neurological symptoms, probably by suppressing the production of pathogenic autoantibodies or cytotoxic processes mediated by cellular and humoral immunity.

References

1. Manto MU, Rondeaux P, Jacquy J (1996) Subacute cerebellar syndrome associated with SLE. Clin Neurol Neurosurg 98(2): 157-160.

2. Johnson RT, Richardson EP (1968) The neurological manifestation of systemic lupus erythematosus. Medicine 47(4): 337-369.

3. Grigor R, Edmonds J, Lewkonia R, Bresnihan B, Hughes GR (1978) Systemic lupus erythematosus: A prospective analysis. Ann Rheum Dis 37(2): 121-128.

4. Macune WJ, Maguire A, Aisen A, Gebarski S (1988) Identification of brain lesions in neuropsychiatric systemic lupus erythematosus by MRI scanning. Arthritis Rheum 31(2): 159-166.

5. Liang MH, Corzillius M, Bae SC, Fortin PR, Gordon C (1999) The American College of Rheumatology nomenclature and case definition for neuropsychiatric lupus syndromes. Arthritis Rheum 42(4): 599-608.

6. Appenzeller S, Cendes F, Costallat LTL (2008) Cerebellar ataxia in SLE. Lupus 17(12): 1122-1126.

7. Chattopadhyay P, Duha D, Philips CA, Saha S (2011) Acute cerebellar ataxia in lupus. Lupus 20(12): 1312-1315.

8. Iwasaki Y, Okamoto A, Shoda H, Takahashi Y, Fujio K, et al. (2012) Subacute cerebellar ataxia developed in a young woman with systemic lupus erythematosus whose cerebrospinal fluid was positive for antineuronal cell antibody. Lupus 21(3): 324-328.

9. Shimomura T, Kuno N, Takenaka T (1993) Purkinje cell antibody in lupus ataxia. Lancet 342(8867): 375-376.