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LETTER TO THE EDITOR

Cerebellar ataxia as a first manifestation of systemic lupus erythematosus

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Dear Sir,

Adult-onset cerebellar ataxias can be a diagnostic challenge. Cerebellar ataxia can occur rarely in systemic lupus erythematosus (SLE) but it is very rare for it to occur as the presenting symptom [1]. The following case emphasizes the importance of an early diagnosis of this severe but potentially treatable cause of cerebellar ataxia.

A 52-year-old woman presented with a 2-year history of fatigue and unsteadiness. There was no relevant past medical or family history. Neurological exam revealed direction changing nystagmus, moderate scanning speech dysarthria, severe truncal instability, and bilateral cerebellar incoordination, dysmetria, and intention tremor. She had brisk deep tendon reflexes in both the upper and lower limbs with upgoing plantar responses without muscle weakness or sensory findings. She could take only a few steps with bilateral support and started using a wheelchair a month later.

Routine blood tests, TSH, ACE, HIV, HBV and syphilis were normal/negative. Chest X-ray was normal. Cerebral MRI revealed mild, diffuse cerebellar atrophy with no evidence of focal pathology or specific findings such as hot cross bun sign (Fig. 1).

CSF examination showed a mononuclear cell count 15×10^9/L (≤ 5), protein 0.8 g/L (0.15–0.45), glucose 3.0 mmol/L, oligoclonal bands were positive.

She was treated for presumed autoimmune cerebellitis with methylprednisolone 1000 mg for 5 days followed by prednisolone 1 mg/kg daily tapering over 2 weeks. She was also diagnosed with chronic, minimally active hepatitis C and managed conservatively.

A whole-body 18F-FDG PET–CT revealed no metabolically active lesions. Screening for GAD, antiphospholipid and onconeuronal autoantibodies (Hu, Yo-1, CV-2, PNMA-2, Ri, amphiphysin), ANCA, Borrelia, varicella zoster, and HSV was negative.

She developed arthralgia, fever, weight loss, alopecia and mild lupus dermatitis 3 months after neurologic presentation. Blood tests showed leucopenia (3.6×10^9/L), thrombocytopenia (91×10^9/L), elevated ESR (40 mm/h), positive ANA 1:640 granular type (< 1:160), and anti-dsDNA antibodies level 19.6 IU/mL (< 20). She was diagnosed with SLE.

She received further methylprednisolone 20 mg/day and mycophenolate mofetil 500 mg/day. After a single plasma exchange (PLEX), there was an improvement in sitting balance, tremor and speech, but she still used wheelchair to move around and was severely disabled. She received four more PLEX (each 2–3 months apart), her condition stabilized with minimal further improvement.

We argue that our patient had a cerebellar syndrome as the first presentation of SLE. The diagnosis is supported by mild inflammatory changes in the CSF in the context of a clinical and serological picture consistent with SLE.

An infective encephalitis seems unlikely as the patient had a prolonged time-course of deterioration for infection, had an MRI which lacked meningeal enhancement, and had no evidence of a pathogenic cause on CSF examination.

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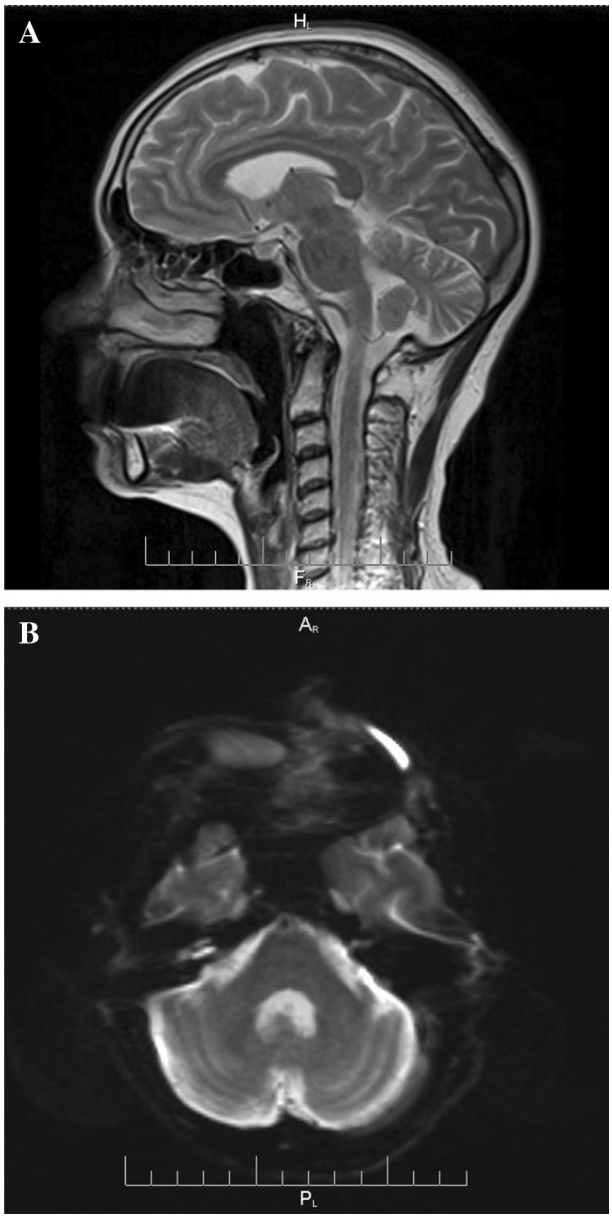

Fig. 1 MRI showed mild cerebellar atrophy with no evidence of focal pathology. **a** T2W imaging, **b** diffusion-weighted imaging

Another important differential diagnosis is paraneoplastic cerebellar degeneration but our patient underwent negative screening for malignancy and paraneoplastic antibodies.

Cerebellar degeneration due to SLE is rare. Ahmed et al. found only 15 cases in the literature [1]. The majority of patients were women at the age 15–34, and had bilateral cerebellar ataxia which was diagnosed after SLE. There were only few reports of cerebellar ataxia developed before SLE was diagnosed [1, 2]. SLE can also affect the brainstem, and our patient had upgoing plantars and brisk reflexes which may correspond to a degree of brainstem involvement [3]. A number of mechanisms could be responsible for cerebellar involvement in SLE, including vascular (premature atherosclerosis, microvascular thrombosis) and inflammatory (cytokine- and antibody mediated). The latter is favored in our patient due to the lack of chronic ischemic changes on MRI of the brain. The diffuse cerebellar ataxia seen in SLE may be mediated by anti-dsDNA antibodies which can cross-react with NMDA receptors but a clear mechanism has not been elucidated [1, 4, 5].

Progressive cerebellar ataxias may occur due to genetic or neurodegenerative conditions. The lack of family history or autonomic disturbance, rapid progression over 2 years, inflammatory CSF profile and modest, but definite, response to PLEX favor an immune-mediated cause over spinocerebellar degeneration or multiple system atrophy.

There are several reports of full recovery of cerebellar ataxia in SLE treated with immunosuppressive agents such as prednisolone 30–40 mg/day or 1 mg/kg, or pulse—methylprednisolone followed by oral prednisolone +/− azathioprine or cyclophosphamide [1, 5]. However, the success of treatment depended on a number factors, such as time of the development of ataxia in relation to manifestation of SLE. In the majority of reported cases, ataxia developed after SLE, or simultaneously, which means that the most patients were already treated with immunosuppressive agents prior to the development of ataxia. In our case, ataxia developed as a first sign of SLE, but treatment was given relatively late, when irreversible damage to the cerebellum appears to have already occurred. Earlier or more aggressive immunotherapy, ideally administered in combination with antiviral therapy for hepatitis C in the case of our patient, may have produced a better neurological outcome.

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Compliance with ethical standards

Conflict of interest None declared.

Ethical approval This article does not contain any studies with human participants or animals performed by any of the authors.

Patient consent for publication Next of kin consent obtained.

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