

CORRESPONDENCE

Lingual myoclonus and neuropsychiatric lupus: a new association?

Dear Editor,

A 56-year-old Chinese woman, recently diagnosed as having systemic lupus erythematosus (SLE) based on malar rash, photosensitivity and a positive assay for antinuclear antibodies, was referred to the rheumatologist because of a sudden onset of depressed level of consciousness 2 days prior to admission. There was no history to suggest infection, head trauma or drug ingestion. Past medical history included hypertension which was well controlled on amlodipine 5 mg daily.

Physical examination revealed a woman who was obtunded with a Glasgow Coma Scale (GCS) score of 8/15 (eye response [E] 1, motor response [M] 5, verbal response [V] 2). Vital signs were normal. She had prominent malar rash and photosensitive rash over the 'V' of the neck. Pupils were equal (3 mm) and reactive to light and fundoscopy was normal. Photophobia and neck stiffness were not demonstrated. Nervous system examination revealed hypertonia in all limbs with brisk tendon reflexes. However, plantar response was down-going bilaterally. Of note, there was no focal neurological deficit. Examination of the other systems were unremarkable.

A diagnosis of active SLE with neuropsychiatric involvement was made. Computed tomography (CT) brain scan demonstrated cerebral atrophy and left basal ganglia infarct. Following this, she underwent a lumbar puncture to exclude central nervous system infection. Cerebrospinal fluid examination was essentially normal.

Laboratory blood investigations showed the following. There was lymphopenia ($0.6 \times 10^9/L$), raised erythrocyte sedimentation rate (ESR) of 80 mm/h, normal C-reactive protein (CRP) of 0.2 mg/dL (normal < 0.5 mg/dL). Liver function test, renal profile, fasting serum cholesterol and thyroid function tests were normal. Antinuclear antibody (ANA) titre was positive (1 : > 2560, homogeneous). Anti-double-stranded DNA (anti-dsDNA (titre 1 : 80) and anti-SSA (Sjögren Syndrome A) antibodies were also positive. Lupus

anticoagulant, anticardiolipin antibody and anti- β 2 glycoprotein I antibody were negative. C3 and C4 levels were normal.

A diagnosis of active lupus with neuropsychiatric involvement was made. Our patient was treated with intravenous methylprednisolone 500 mg daily for 3 days and intravenous cyclophosphamide infusion at 500 mg. She showed gradual improvement in terms of her cognitive function and by day 8 of admission, she was able to open her eyes spontaneously, obey simple commands and verbalize.

On day 24 after admission, she unexpectedly developed constant repetitive protrusion-intrusion movement of the tongue which was associated with drooling of saliva. She was not able to speak or swallow; however, she remained alert. There were no signs of infection and electrolytes were normal. She was promptly referred to the neurologist and a diagnosis of lingual myoclonus was made. Her medications were reviewed and she was clearly not on any drugs that may induce myoclonus. She was treated with a loading dose of intravenous sodium valproate at 1.2 g and subsequently 400 mg three times a day. By the following day, the intensity of lingual myoclonus had decreased considerably; however, she was still unable to speak or swallow. Complete resolution of myoclonus only occurred 2 days later. Further investigations included electroencephalogram (EEG) which was normal. Magnetic resonance (MR) imaging of the brain revealed diffuse cerebral atrophy and an old infarct measuring 10 mm \times 7 mm at the left basal ganglia (Fig. 1). There were no areas of restricted diffusion to suggest an acute infarct. MR angiography of the brain was normal indicating no evidence of vasculitis. In particular, there was no brainstem or cerebellar lesion. Echocardiography and carotid Doppler ultrasound were also normal.

The patient continued to receive another two doses of intravenous cyclophosphamide at 3-weekly intervals. As there was no recurrence of lingual myoclonus and the

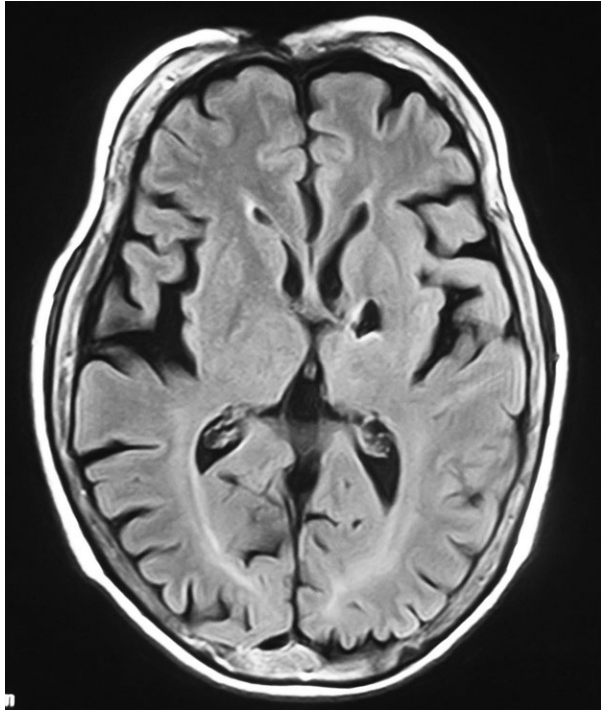


Figure 1 Brain magnetic imaging showing diffuse cerebral atrophy and an old infarct measuring 10 mm × 7 mm at the left basal ganglia.

patient showed good recovery, a decision was made to discontinue further cyclophosphamide therapy. Sodium valproate was also ceased after 6 months. Subsequent treatment included azathioprine as maintenance therapy, addition of hydroxychloroquine and gradual tapering of prednisolone. She recovered with no residual neurological deficit and to date, her lupus had remained quiescent without any flares.

DISCUSSION

According to the American College of Rheumatology nomenclature and case definitions for neuropsychiatric lupus syndromes, motor disorders form one of the 19 neuropsychiatric syndromes observed in lupus.¹

We describe a case of SLE with neuropsychiatric involvement that presented with a rare movement disorder, that is, lingual myoclonus. To the best of our knowledge, the occurrence of lingual myoclonus as a manifestation of neuropsychiatric lupus has not been previously reported.

A literature search revealed that cases of lingual myoclonus were indeed very infrequent and lingual

myoclonus remains a rare entity which is poorly understood. Most of the cases reported were associated with an underlying abnormality. Kulisevsky *et al.*,² reported a case that was associated with Arnold-Chiari malformation while Lee *et al.*,³ and Postert *et al.*,⁴ reported one case each associated with craniovertebral junction abnormalities and brainstem ischemia, respectively. Cases of isolated lingual myoclonus were also described by Gobernado *et al.*,⁵ Troupin *et al.*⁶ and Bettoni *et al.*⁷ The case by Gobernado *et al.*,⁵ was similar to ours, whereby the EEG was normal, MR imaging of the brain did not reveal any brainstem or cerebellar lesion and the patient demonstrated good response to sodium valproate. The basal ganglia infarct in our patient demonstrated in both CT scan and MR imaging of the brain was old and most likely related to hypertension and age. Furthermore, basal ganglia lesions would not result in lingual myoclonus.

The etiology and pathogenesis of lingual myoclonus in our patient remained unclear. Of note it occurred 3 weeks after pulsed corticosteroid was given. We believe that complete disease activity may not be arrested instantaneously even with pulsed corticosteroid and it was still possible for new disease manifestations to occur during the early phase of treatment. We could only speculate that this clinical manifestation was a consequence of either vascular thrombosis or vasculitis, albeit there was no obvious lesion noted in the brainstem or cerebellum. Electrolyte imbalance, drugs and infection that could induce this condition have been excluded.

Myoclonus in general is indeed one of the commonest forms of movement disorders; however, lingual myoclonus is rare. The site of pathology in lingual myoclonus arises from the brainstem. Medications that are frequently used to control myoclonus include sodium valproate, clonazepam and levetiracetam.⁸

In conclusion, this case suggests that lingual myoclonus is possibly a manifestation of neuropsychiatric lupus, even though no pathologic lesion was demonstrated on MR imaging/MR angiography of the brain.

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