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Case Report and Review

Miller-Fisher syndrome associated with unilateral cerebral white matter lesions

Yongfeng Xu [a], Lan Liu [b,*]

[a] Department of Neurology, Second Affiliated Hospital, School of Medicine, Zhejiang University, Hangzhou, PR China
[b] Department of Ophthalmology, First Affiliated Hospital of College of Medicine, Zhejiang University, Hangzhou, PR China

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ABSTRACT

Miller-Fisher syndrome (MFS) is characterized by classical triad of ophthalmoplegia, ataxia and areflexia. The involvement of cerebral white matter in MFS is very rare. We report a typical MFS patient whose brain MRI showed unilateral and extensive involvement in cerebral white matter. We also found mild pleocytosis and raised protein concentration in cerebrospinal fluid. Deficits resolved completely after treatment with intravenous immunoglobulins. Subsequent brain MRI shows cavity formation in involved white matter.

1. Introduction

Miller-Fisher syndrome (MFS), which is characterized by ataxia, ophthalmoplegia and areflexia, is considered to be a variant of Guillain-Barré syndrome (GBS) [1]. It is reported that central nervous system (CNS) could also be involved in MFS [1]. The involvement sites of CNS in MFS are mainly pons, medulla oblangata [1], cerebellar peduncles [2], and occasionally, optic nerves [3]. However, the involvement of cerebral white matter in MFS is very rare. Here, we report a typical MFS patient whose brain MRI showed unilateral and large lesions in cerebral white matter.

2. Case report

A 37-year-old man who had no history of toxic substance exposure or alcohol abuse, no past medical or family history was admitted with double vision and unsteadiness of gait. The patient also had dizziness, left eyelid ptosis and distal numbness on both of the upper limbs. He had antecedent infections 5 days before the onset of the disease. He had no fever on admission and was fully conscious without signs of meningeal irritation. He had asymmetrical partial ptosis and external ophthalmoplegia, and left eye was more severe than the right eye; the light reflex of the pupils was well maintained. Examination of other cranial nerves showed no positive findings. The patient showed no weakness of all the limbs, but all tendon reflexes were absent. No pathological reflexes were found. Finger-to-nose and heel-to-knee tests in both

* *Corresponding author at*: Department of Ophthalmology, First Affiliated Hospital of College of Medicine, Zhejiang University, Hangzhou 310000, PR China. Tel.: +86 0571 87784711; fax: +86 0571 87783122.
E-mail address: flowingblue@126.com (L. Liu).

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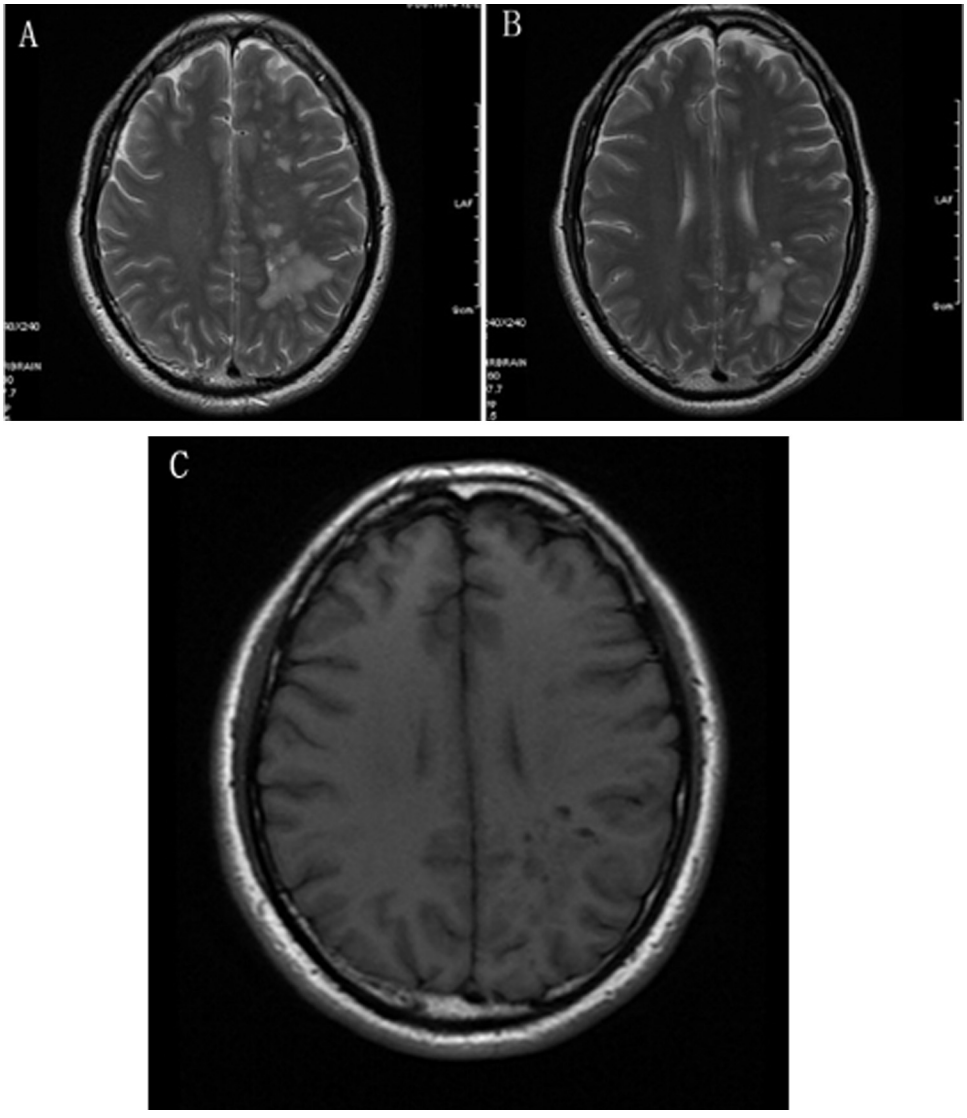

Fig. 1 – T2 weighted MRI on day 6 shows multiple lesions in the juxtacortex, subcortex and deep white matter in the left frontal and occipital lobe (A and B), and T1 weighted MRI shows cavity formation on day 360 (C).

sides were positive, and the patient had cerebellar gait. Pinprick sensation was impaired on the distal part of both of the upper limbs. Cardiovascular, respiratory, gastrointestinal, and ear, nose and throat findings were normal. There were no ticks found on the skin, and no rash.

Biochemical, hematological, liver and renal function investigations were normal. Serological tests for syphilis and HIV were negative. Concentrations of gammaglobulin and complement were normal, auto-antibody screen and rheumatoid factor were negative. Chest radiography and ECG showed no abnormality. Lumber puncture showed that pressure was 200 mmH_2O, and CSF contained 10 lymphocytes/mm^3 and 0.26 g/l protein (on day 7). On day 6, brain MRI showed multiple lesions in the juxtacortex, subcortex and deep white matter in the left frontal and occipital lobe (Fig. 1A and B). The signals were hypointensive in T1-weighted images and isointensive in DWI images (not shown). The lesions were not enhanced after contrast administration (not shown). The cerebellum and brainstem were not involved. Brain CT angiography and Orbital CT scan were normal (not shown). Electromyography of limb muscles was normal. Neurophysiological examination showed tibial H reflexes were absent. However, the motor and sensory nerve conduction velocity in ulnar, median, tibial, peroneal and sural nerves was normal, and distal motor latencies were normal. Intravenous immunoglobulin (IVIg) (0.4 g/kg per day for 5 days) was started on day 5. Tendon reflexes re-appeared and pinprick sensations of upper limbs were normal on day 18, and gradual improvement in ataxia and ocular movement ensued in the following 2 weeks until the time when recovery was complete (day 60). On day 18, lumber puncture showed that the pressure was 195 mmH_2O, the CSF contained 0.58 g/l protein and 14 lymphocytes/mm^3. Brain MRI of day 180 and day 360 were similar to that of day 6, but cavity formation in the involved matter could be found in brain MRI of day 360 (Fig. 1C). No recurrence was seen in one-year follow up.