—Case Reports—

Mild Encephalitis/Encephalopathy with a Reversible Splenial Lesion in an Adult with Cerebellar Ataxia: A Case Report

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Mild encephalitis/encephalopathy with a reversible splenial lesion (MERS) is a clinicoradiological syndrome characterized by transient mild encephalopathy and magnetic resonance imaging (MRI) findings of a reversible lesion in the splenium of the corpus callosum (SCC). Patients with MERS generally present with central nervous system symptoms such as consciousness disturbance, headache, and seizure; adult-onset MERS with cerebellar ataxia is rare. A 53-year-old man was admitted to our hospital with fever of 1 week's duration, headache, neck stiffness, and gait disturbance. Neurological examination revealed bilateral intention tremor (predominantly affecting the right hand) and gait ataxia. Diffusion-weighted brain MRI showed a focal hyperintense lesion in the SCC. Cerebrospinal fluid analysis revealed elevated levels of mononuclear cells and proteins. Brain imaging with ^{123}I-iofetamine single-photon emission computed tomography showed reduced cerebral blood flow in the left thalamus and right cerebellum. Several diseases, including cerebellar stroke and acute cerebellitis, develop as comorbidities in patients with acute cerebellar ataxia. This case suggests that MERS should be suspected in adults with cerebellar ataxia. (J Nippon Med Sch 2020; 87: 153–156)

Key words: mild encephalitis, encephalopathy, a reversible splenial lesion, cerebellar ataxia

Introduction

Mild encephalitis/encephalopathy with reversible splenial lesion (MERS) is a clinicoradiological syndrome characterized by transient mild encephalopathy and magnetic resonance imaging (MRI) findings of a reversible lesion in the splenium of the corpus callosum (SCC)[1]. Patients with MERS usually present with central nervous system symptoms such as consciousness disturbance, headache, and seizure[1]; adult-onset MERS with cerebellar ataxia is rare. Here, we report a case of adult-onset MERS in a Japanese man with cerebellar ataxia.

Case Report

A 53-year-old man was admitted to our hospital with fever of 1 week's duration, headache, neck stiffness, tremor, gait disturbance, and elevated C-reactive protein level (3.8 mg/dL). His medical and family histories were unremarkable. Neurological examination revealed bilateral upper-extremity numbness, intention tremor, and gait ataxia. The intention tremor was bilateral but predominantly affected the right hand. Brain MRI showed a focal hyperintense lesion in the SCC on diffusion-weighted imaging (DWI) and fluid-attenuated inversion recovery (FLAIR) sequences on the day of admission (**Fig. 1a, b**). The SCC had a low apparent diffusion coefficient (**Fig. 1c**). Moreover, DWI and FLAIR sequences showed no abnormal signal in the cerebellum that would have suggested cerebellar stroke or acute cerebellitis (**Fig. 1d, e**). Brain imaging with ^{123}I-iofetamine single-photon emission computed tomography (SPECT) showed reduced tracer uptake into the left thalamus and right cerebellum (**Fig. 1f**). Analysis of cerebrospinal fluid (CSF) showed increased levels of white blood cells (93 cells/mm^3; 84 mononuclear cells/mm^3 and 9 polynuclear cells/mm^3) and proteins (149 mg/dL) but a normal glucose level (68 mg/dL). These findings indicated viral meningitis, but the pathogen could not be identified.

After admission, he was treated prophylactically with

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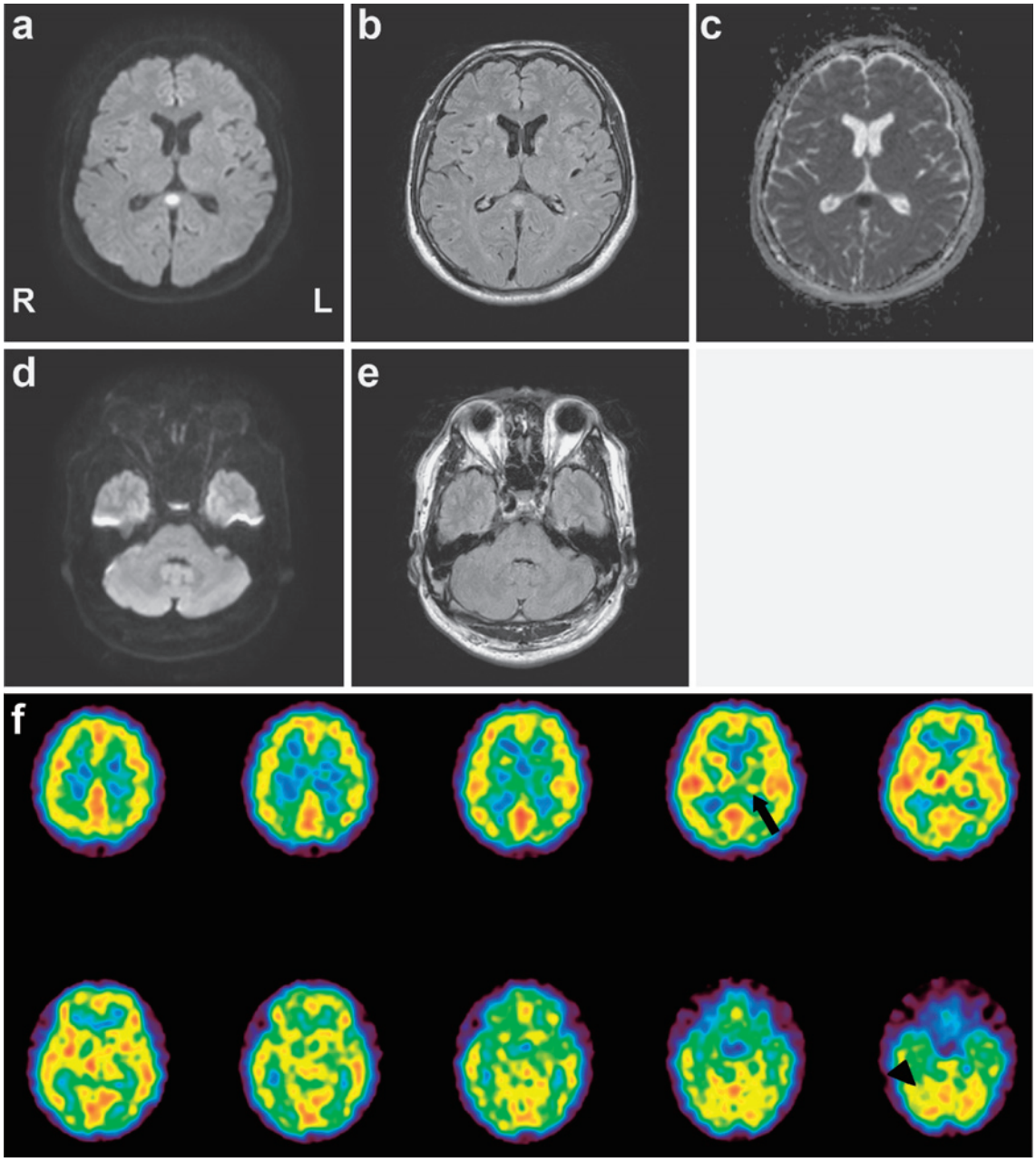

Fig. 1 Representative brain MRI and ^{123}I-IMP SPECT images

a. DWI image showing hyperintense lesions in the splenium. **b**. FLAIR image showing hyperintense lesions in the same area. **c**. Image of the SCC showing a low ADC value. **d**. DWI image showing no abnormal signals in the cerebellum. **e**. FLAIR image showing no abnormal signals in the cerebellum. **f**. ^{123}I-IMP SPECT brain image. The arrow indicates reduced tracer uptake into the left thalamus. The arrowhead indicates reduced tracer uptake into the right cerebellum.

Abbreviations: ^{123}I-IMP SPECT, ^{123}I-iofetamine single-photon emission computed tomography; ADC, apparent diffusion coefficient; DWI, diffusion-weighted imaging; FLAIR, fluid-attenuated inversion recovery; MRI, magnetic resonance imaging; SCC, splenium of corpus callosum

intravenous acyclovir (10 mg/kg for 8 h/d over 7 days) and intravenous methylprednisolone pulse therapy (1,000 mg/d for 3 days), because of the risk of herpes simplex encephalitis. A follow-up brain MRI scan at 14 days after the initial examination showed that the abnormal signal intensity in the SCC had disappeared (**Fig. 2a-c**). Follow-up CSF analysis on the same day revealed decreased levels of white blood cells (15 mononuclear cells/mm^3) and proteins (41 mg/dL). A polymerase chain reaction-based test for herpes simplex virus DNA in the CSF yielded negative results. Cerebellar ataxia significantly improved within 14 days. On the basis of the

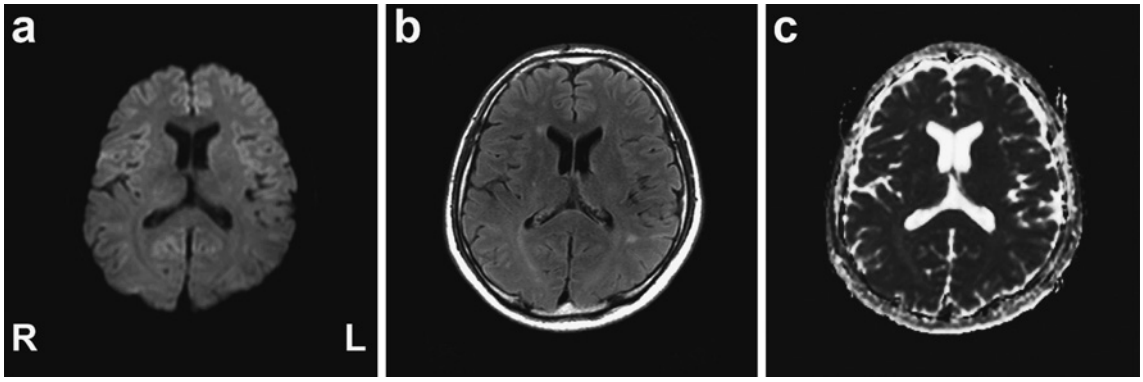

Fig. 2 Representative follow-up brain MRI images
a. DWI image showing no abnormal signals in the splenium. **b**. FLAIR image showing no abnormal signals in the same area. **c**. Image of the SCC showing no abnormal ADC values. Abbreviations as in Fig. 1.

characteristic transient splenial lesion and reversible clinical symptoms, MERS with concomitant viral meningitis was diagnosed.

Discussion

Adult-onset MERS is rare, and cerebellar ataxia is clearly present in only 10.3% of adults with MERS[1]. Here, we describe a case of adult-onset MERS with cerebellar ataxia. The patient also had clinical findings indicative of viral meningitis. Earlier studies reported that MERS can be triggered by pathological causes such as viruses (e.g., influenza virus, mumps virus, and rotavirus), bacteria (e.g., *Salmonella enteritis* and *Escherichia coli* O157), drugs (e.g., antiepileptic drugs), and electrolyte imbalances (e.g., hyponatremia)[1,2]. Legionella, mumps virus, and rotavirus have been reported as pathogens associated with MERS with cerebellar ataxia[1,3,4]; however, the cause of MERS frequently remains unidentified. No infectious pathogen or blood biochemical abnormality was detected in our patient.

The typical clinical symptoms of MERS include reversible disturbances of consciousness, fever, headache, and seizure that completely resolve within 1 month[5]. Our patient had bilateral intention tremor, gait ataxia, and a solitary SCC lesion visible on MRI scans. The exact function of the SCC has not been identified; however, congenital and acquired lesions of the SCC can cause confusion, dysarthria, and ataxia[6].

Few reports have described SPECT findings for patients with MERS[3,7]. Although one previously reported patient had reduced cerebellar hypoperfusion in SPECT images, the present study is the first to report SPECT evidence of hypoperfusion unilaterally, in the thalamus, and contralaterally, in the cerebellum[3]. Thalamic nuclei, especially the ventralis posterior lateralis pars oralis, have disynaptic fibers that connect the contralateral cerebellar dentate nucleus and primary motor cortex through the dentatothalamocortical pathway[8,9]. Reduced tracer uptake in the right cerebellum, including the dentate nucleus, indicated crossed cerebellar diaschisis (CCD) due to hypoactivity of the contralateral thalamus. Similarly, a previous report described a patient with hemiataxia whose SPECT images revealed contralateral thalamic hemorrhage and ipsilateral CCD[10]. In our patient, the fact that bilateral intention tremor predominantly affected the right hand might have been attributable to left thalamic hypoactivity.

Several conditions present as comorbidities of acute cerebellar ataxia, including infections (e.g., acute cerebellitis), poisoning (e.g., alcoholic cerebellar degeneration), immune-mediated disorders (e.g., anti-glutamic acid decarboxylase ataxia and Miller-Fisher syndrome), structural and vascular conditions (e.g., tumors and cerebellar stroke), and metabolic disorders (e.g., Wernicke encephalopathy and biotinidase deficiencies)[11]. The present case suggests that MERS should be considered a potential comorbidity in adults with cerebellar ataxia. Future studies should use neuroimaging techniques such as SPECT and positron emission tomography to clarify functional associations between the SCC, thalamus, and cerebellum and determine the mechanism underlying cerebellar ataxia accompanied by MERS.

Conflict of Interest: None declared.

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