Clinical Case Report

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Adult severe encephalitis/encephalopathy with a reversible splenial lesion of the corpus callosum

A case report

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Abstract

Rationale: Clinically mild encephalitis/encephalopathy with a reversible splenial lesion of the corpus callosum (MERS) is a recently identified clinically and radiologically distinct syndrome. Clinical symptoms and lesions on the magnetic resonance imaging (MRI) often disappear in 1 week or a few weeks. However, MERS manifesting as a severe clinical course with significant sequela has not yet been reported.

Patient concerns: A 42-year-old male presented with a 3-day history of headache, fever, and irrational speech. Physical examination showed a body temperature of 39.5°C, dysarthria, dyscalculia, recent memory disturbance, and otherwise normal vital signs. The patient developed status epilepticus and progressive consciousness disturbance. MRI showed abnormal patchy signals in the splenium of the corpus callosum.

Diagnosis: The clinical feature and the characteristic of MRI are mostly consistent with MERS. At the same time, we made a differential diagnosis by testing the NMDARAb, AMPA1Ab, AMPA2Ab, LG1Ab, CASPR2Ab, GABABRAb in CSF and serum.

Interventions: The subject was treated with ganciclovir, antiepileptic, and antipyretic therapy.

Outcomes: The subject was living a virtually normal life with persistent mild memory disturbance. MRI showed that the abnormal signals in the splenium of the corpus callosum had disappeared, but hyperintensity on T2-weighted and FLAIR imaging was noted in the centrum semiovale.

Lessons: MERS is a rare clinicoradiological syndrome, which can manifest as severe symptoms as well. Early diagnosis and treatment should be emphasized, and the diagnostic value of MRI is highlighted. Clinicians should be alert to the potential sequela.

Abbreviations: ADC = apparent diffusion coefficient, DWI = diffusion-weighted imaging, EEG = Electroencephalogram, CSF = cerebrospinal fluid, FLAIR = fluid attenuated inversion recovery, MERS = mild encephalitis/encephalopathy with a reversible splenial lesion of the corpus callosum, MRI = magnetic resonance imaging.

Keywords: case report, corpus callosum, encephalitis, encephalopathy, MRI

1. Introduction

Clinically mild encephalitis/encephalopathy with a reversible splenial lesion (MERS) is a recently identified, clinically, and radiologically distinct syndrome. The definitive pathogenesis of this condition is still unclear, and infection has been considered a common contributor to the incidence of MERS.[1] Clinically, this condition is characterized by neurological deficiencies; the majority of symptoms can resolve completely within 1 month.[2] On magnetic resonance imaging (MRI), the characteristic feature of MERS is a reversible isolated lesion in the splenium of the corpus callosum, which can occasionally spread to the whole corpus callosum and adjacent white matter. These radiological abnormalities usually disappear within 1 week or a few weeks.[3] However, MERS manifesting as a severe clinical course with significant sequela has not yet been reported.

Herein, we report a case with MERS presenting with a severe progressive clinical course.

2. Case report

A 42-year-old, previously healthy male presented to us with a 3-day history of headache, fever, and irrational speech. After administration of compound paracetamol and amantadine hydrochloride, his symptoms were not improved. Physical examination showed a body temperature of 39.5°C, dysarthria, dyscalculia, recent memory disturbance, and otherwise normal vital signs. MRI showed abnormal patchy signals in the splenium of the corpus callosum, which appeared hypointense on T1-weighted imaging and hyperintense on T2-weighted, fluid attenuated inversion recovery (FLAIR), and diffusion-weighted (DWI) imaging (Fig. 1A–E). Three days after admission, he developed epileptic seizure and progressive consciousness disturbance from slight coma to moderate coma. Repeated MRI showed the signal intensity of the splenial lesion declined on FLAIR and DWI, and on an apparent diffusion coefficient (ADC)

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Ethics approval and consent to participate: Though ethical approval was not required for this case report, the patient provided written informed consent for publication and use of all accompanying images.

The authors have no conflicts of interest to disclose.

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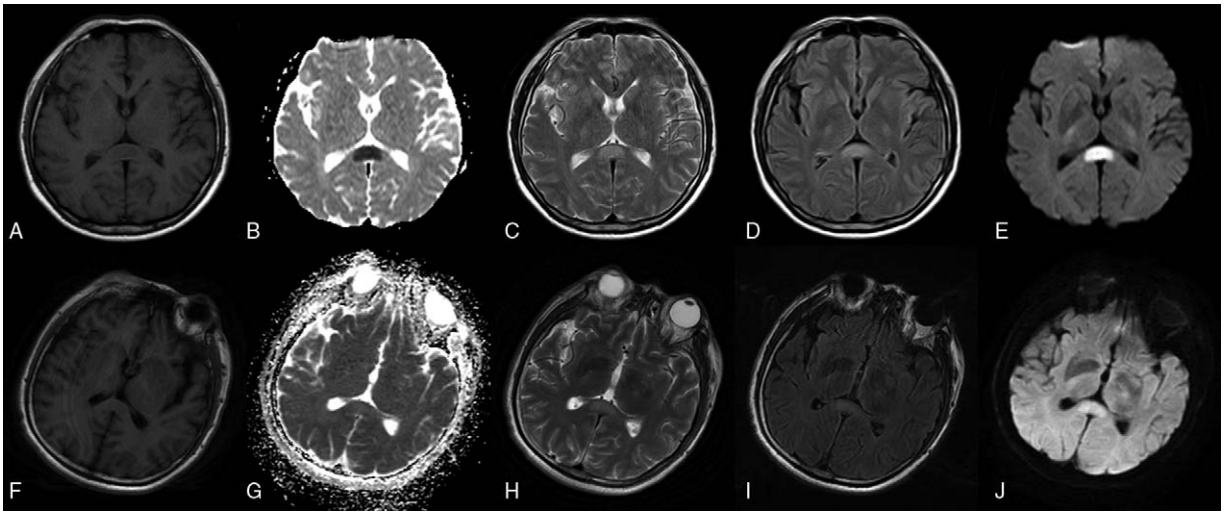

Figure 1. MRI upon hospitalization. MRI on admission showed abnormal patchy signals in the splenium of the corpus callosum, which appear as hypointense on T1-weighted imaging (A) and ADC maps (B) and as hyperintense on T2-weighted imaging (C), FLAIR (D), and DWI (E). Repeated MRI on the fourth day after admission showed the lesion was unremarkable on T1-weighted imaging (F) and ADC maps (G), and the signal intensity of the splenial lesion had declined on T2-weighted imaging (H), FLAIR (I), and DWI (J). ADC = apparent diffusion coefficient, DWI = diffusion-weighted imaging, FLAIR = fluid attenuated inversion recovery, MRI = magnetic resonance imaging.

map, the lesion was unremarkable (Fig. 1F–J). Electroencephalogram (EEG) revealed irregular rhythms with high-voltage waves, and high-amplitude waves (5–6 waves/s) and low-amplitude β waves (20–24 waves/s) were visible. Cerebrospinal fluid (CSF) examination showed a pressure of 180 mmH_2O and negative Pandy test, with the following parameters: leucocyte count, 20×10^6/L; glucose level, 4.36 mmol/L; chlorine, 133.70 mmol/L; and protein, 0.08 g/L. NMDARAb, AMPA1Ab, AMPA2Ab, LG1Ab, CASPR2Ab, GABABRAb were negative in CSF and serum. Bacterial cultures of sputamentum revealed an infection of acinetobacter baumannii. A 16-day regimen of ganciclovir (5 mg/kg/12 h) and a 7-day meropenem (0.5 g/8 h) were scheduled. On the 14th day after admission, his consciousness was restored to a normal level, and the fever and convulsions were completely relieved, while neurological examination showed persistence of recent memory disturbance. After an 8-month follow-up, the patient was living a virtually normal life with continued mild memory disturbance. On MRI the abnormal signals in the splenium of the corpus callosum had disappeared, and hyperintensity on T2-weighted and FLAIR imaging was noted in the centrum semiovale (Fig. 2). A diagnosis of MERS was made.

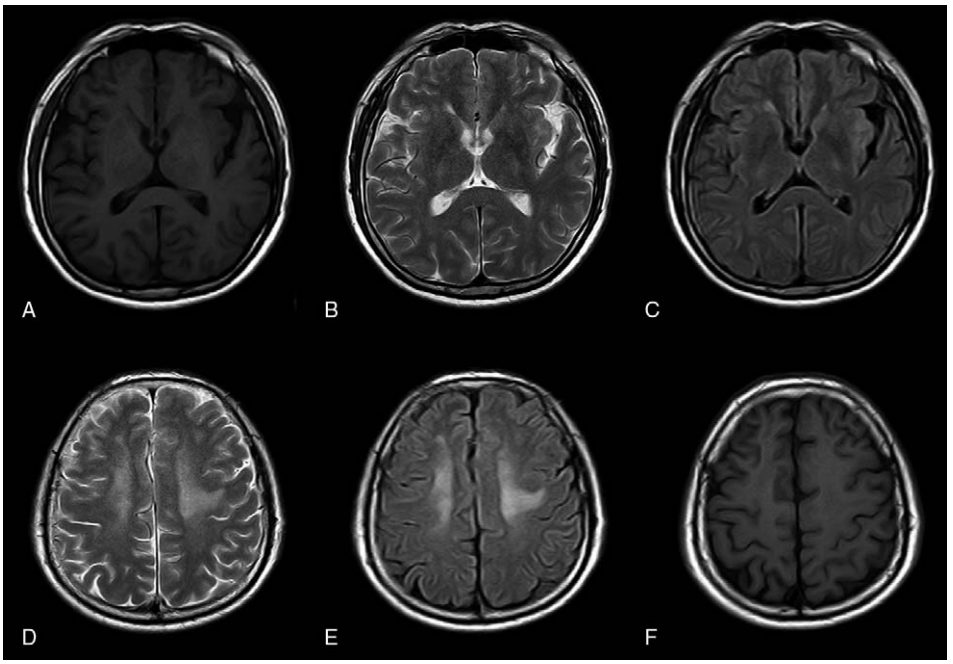

Figure 2. MRI at the 8-month follow-up. MRI showed that the previous abnormal signals in the splenium of the corpus callosum had disappeared on T1-weighted (A), T2-weighted (B), and FLAIR (C) imaging. Additionally, hyperintensity on T2-weighted (D) and FLAIR (E) imaging was noted in the centrum semiovale, which was unremarkable on T1-weighted imaging (F).

3. Discussion

MERS was originally reported and nominated as a clinically and radiologically distinct syndrome.[4] Takanashi et al studied the clinicoradiological characteristics of MERS in a 54-case cohort. They found the common clinical symptoms were delirium, confusion, and epileptic seizures, all of which would disappear within 1 week.[5] In the current case, the patient presented with much more severe symptoms including persistent fever, progressive consciousness disturbance, and epilepsy from status epilepticus to generalized tonic clonic seizure. Moreover, these symptoms last for 2 weeks, and the memory disturbance remained after an 8-month follow-up. Thus, the clinical symptom spectrum of MERS still requires further research.

The characteristic MRI features of MERS are symmetrical oval hypo- to isointensity on T1-weighted imaging in the splenium of the corpus callosum, which appear as hyperintensity on T2-weighted, FLAIR, and diffusion-weighted sequences and hypointensity on ADC maps; there is usually no enhancement.[4] According to the MRI patterns, MERS is classified into 2 forms: type I, MERS limited to the splenium of the corpus callosum; and type II, MERS involving the splenium of the corpus callosum as well as subcortical or deep white matter. The latter form is usually associated with a longer clinical course and neurological sequela. In the current case, typical MERS intensities were observed in the splenium of the corpus callosum and had disappeared by the 8-month follow-up; nevertheless, delayed symmetrical lesions in the bilateral centrum semiovale were noted. These characteristics were consistent with type II MERS.

The definitive etiology and pathogenetic mechanism are still unclear. According to the literature, the potential causes include viral or bacterial infection, epilepsy, antiepileptic drug withdrawal, metabolic disturbance, and drug-related toxicity. The present case showed premonitory symptoms including fever and headache, and antibody for autoimmune encephalitis was negative, which was consistent with viral infection. Some researchers found that the interleukin-6 level in the CSF was elevated, which suggested an immunological response might play an important role in the pathophysiology of MERS.[5]

There is still no consensus regarding the treatment of MERS. In the literature, some evidence shows that MERS responds well to a combined antiviral and steroid pulse therapy.[2] In the current case, after administration of ganciclovir, the abnormal signals in the splenium of the corpus callosum disappeared. However, white matter lesions were noted and the memory sequela remained, and thus, much longer observation was still needed.

4. Conclusion

MERS is a rare clinical and radiological syndrome, and its clinical symptoms can be severe. Clinicians should be alert to the potential neurological sequela. Early diagnosis and treatment should be emphasized, and the diagnostic value of MRI is highlighted by the present case.

Author contributions

Conceptualization: Gang Yao.
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Project administration: Gang Yao, Xi-jing Mao, Bo-chi Zhu, Ting-min Yu.
Supervision: Ting-min Yu.
Writing – original draft: Xi-jing Mao.
Writing – review & editing: Gang Yao.

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