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Original article

Clinically mild encephalitis/encephalopathy with a reversible splenial lesion of corpus callosum in Chinese children

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Abstract

Objective: To investigate the characteristics and etiology of mild encephalitis/encephalopathy with a reversible splenial lesion (MERS) in Chinese children.

Methods: We collected ten pediatric MERS patients from local hospital and enrolled another nineteen patients by reviewing the available literatures. The information of enrolled patients about clinical features, laboratory data, treatment strategies and prognoses were collected for further analysis.

Results: A total of 29 children, the median age of twenty-nine patients was (4.09 ± 3.64) years old. The male-to-female ratio was 1.42:1.0. The major cause of MERS was viral infection. 18 patients had consciousness disturbance which was the most prominent syndrome. 18 patients had transient seizures and only one needed anticonvulsant treatment for long. 9 patients were observed serum sodium levels <135 mEq/L. The cells and protein of cerebral spinal fluid (CSF) were increased in 3 patients. In all patients, brain MRI evaluation revealed typical lesion in splenium of the corpus callosum (SCC). 5 patients had additional lesions involving the periventricular white matter or bilateral centrum semiovale diagnosed. 3 patients were treated with antivirus treatment because of virus infection. 7 patients received corticosteroid. 2 patients received intravenous IVIG. As a result, all patients had fully recovered without neurological residual.

Conclusions: The result of present study suggests that Chinese children with MERS might have favorable prognosis, although there is still no guideline for treatment.

Keywords: Mild encephalitis/encephalopathy with a reversible splenial lesion (MERS); Splenium of the corpus callosum (SCC); Children; China

1. Introduction

Mild encephalopathy with a reversible splenial lesion (MERS) is a clinico-radiological syndrome [1]. The typical neurologic symptoms of MERS include delirious behavior, decreasing conscious level and seizures, always followed by prodromal illness such as fever, cough, vomiting or diarrhea and so on. Generally, the main magnetic resonance imaging (MRI) finding during MERS acute episode reveals a lesion in the SCC, sometimes also extending to other areas of the corpus callosum or adjacent parenchymal white matter [2]. Most reports agree that the prognosis of this acute encephalopathy is favorable. Patients without any major

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complications may recover completely within a few days, and lesions in MRI can completely or almost completely resolve within days to weeks as well. Therefore, understanding the characteristics of the disease is helpful to accurate diagnosis and avoidance of unnecessary treatment.

MERS has been largely described in case reports or case series world widely, especially in Japan. However, to our knowledge, reports with regard to Chinese pediatric patients suffered from MERS are rare so far. In this study, ten Chinese children who presented with MERS from local hospital, together with nineteen MERS cases from reported literature were collected. Data with regard to clinical features, including clinical courses, laboratory, MRI findings and electroencephalograph (EEG) findings, therapy and prognosis, was extracted and analyzed to elucidate the possible characteristics and pathogenesis of Chinese pediatric MERS.

2. Materials and methods

According to the diagnostic criteria of Takanashi [3], pediatric MERS patients were identified by reviewing the inpatient database of local hospital from August 2008 to March 2016 after receiving full approval from local hospital ethics committee. Furthermore, the other cases of MERS were enrolled by reviewing previous studies about Chinese pediatric patients from August 2004 to March 2016. Specifically, utilizing keywords 'mild encephalitis/encephalopathy with a reversible splenial lesion' or 'reversible splenial lesion' and 'child', we conducted a full retrospective review though browsing journal search engines both in English and Chinese, including PubMed, EMBASE, OVID, CNKI, Cqvip, Wangfang Data and Science China. Then, the information of enrolled patients about clinical features, laboratory data, treatment strategies and prognoses were collected for further analysis.

3. Results

3.1. Enrolled patients and search results

According to the diagnostic criteria, 10 pediatric MERS patients were identified by reviewing the inpatient database of local hospital. 12 Refs. [4–15] were initially found from the electronic databases according to search strategy, but only a total of 6 reports containing 19 patients were included after excluding the reports which were lack of integrated clinical data [4–9]. As a result, there were 10 patients from local hospital along with 19 patients from literatures retrieval were enrolled for subsequent analysis totally.

3.2. Clinical manifestation

The median age of 29 patients was (4.09 ± 3.64) years old (rang 3M–14Y). The male-to-female ratio was 1.42:1.0. Non specific prodromal symptoms were observed prior to encephalopathy in all patients, including fever ($n = 22$), vomiting ($n = 14$), diarrhea ($n = 9$), abdominal pain ($n = 3$) and cough ($n = 4$). All patients were presented with symptoms of acute encephalopathy. These symptoms mainly included seizure ($n = 18$), and changes of conscious level which were manifested as confusion ($n = 3$), irritability ($n = 4$), and drowsiness ($n = 11$). Among the 18 patients with seizures, 11 patients presented with general tonic–clonic and lasted less than 5 min, 17 (94.4%) of them occurred once and didn't need any anti-convulsant management. Besides, the rest of neurological symptoms occurred containing headache ($n = 4$), dysphagia and dyslalia ($n = 1$), paroxysmal numbness of limb ($n = 1$), and acute urinary retention ($n = 1$) (Table S-1).

3.3. Auxiliary examination

Lumbar punctures were performed in all 29 patients for diagnostic purposes. Except for 3 cases, whose protein levels and cell counts in the cerebral spinal fluid (CSF) were elevated: The initial white cell count and protein in CSF of patient 1 was 198×10^9/L and 178 mmol/L, while the results of the other 2 patients were 480×10^9/L (WBC) and 1420 mmol/L (protein), 348×10^9/L (WBC) and 780 mmol/L (protein). The rest of 26 patients' protein levels and cell counts were normal in CSF. The glucose levels of CSF were normal in all patients (Table S-2). The bacterial cultures on CSF were absent in all patients. White blood cell counts in peripheral blood in 4 cases were high, and the levels of C-reactive protein (CRP) of 5 cases were high, 3 cases were normal, while the rest of cases had not supply enough data in detail (Table S-2). The levels of serum AST, LDH, CK-MB in all patients from local hospital were higher than normal. The median value for AST was 54.5 U/L (range: 42–109), 276 U/L (range: 180–496) for LDH and 49 U/L ((range: 30–113) for CK-MB in these patients, respectively (Table S-2). 9 patients suffered from hyponatremia (Na < 135 mmol/L), the most serious case among them from local hospital (Na < 120 mmol/L) lasted two weeks despite supplying sodium per-day by intravenous infusion (Table S-2). All the patients received serum and fecal pathogenic examination using rapid antigen-detection assay or PCR, the results included 5/16 (31.2%) rotavirus virus (HRV), 4/16 (25%) mycoplasma pneumonia (MP), 2/16 (12.5%) herpes simplex virus (HSV), 2/16 (12.5%) coxsackie virus, 2/16 (12.5%) adenovirus, 2/16 (12.5%) echoviruses, 1/16 (6.3%) Influenza A virus (INFA),

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