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Case Report

Clinically Mild Encephalopathy with a Reversible Splenial Lesion Accompanying Mumps Virus Infection : a 5-Year-Old Girl Report

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SUMMARY

Aseptic meningitis is known as a mumps complication. However, there are few cases of clinically mild encephalopathy with a reversible splenial lesion (MERS) associated with mumps infection. We report a MERS related to mumps infection in a girl. In the early clinical course, repeating convulsion and consciousness disturbance with hallucination were recognized. Initially, we suspected aseptic meningitis due to mumps, because of her swollen right parotid gland. Cerebrospinal fluid test was performed, but the result was normal. After that, diffusion weighted image of magnetic resonance imaging was added and abnormal signal intensity was recognized in the corpus callosum, so she was diagnosed as MERS. Treatment was performed with steroid pulse therapy and patients was discharged without neurologic sequelae. We need to pay attention to MERS as complication although rare in a mumps infection.

Key Words : clinically mild encephalopathy with a reversible splenial lesion, acute encephalitis, magnetic resonance image

INTRODUCTION

Clinically mild encephalitis/encephalopathy with a reversible splenial lesion (MERS) is a type of acute encephalitic encephalopathy reported recently by Tada et al in 2004[1]. It has long been known about the concern with anti-epileptic drugs[2,3] reported with acute mountain sickness[4], related to this characteristic magnetic resonance imaging (MRI) findings. The clinical feature of MERS is mild compared with other types of acute encephalopathy. Furthermore, characteristic MRI findings of corpus callosum are improved within one week in many cases of MERS[5]. In recent years, Diffusion weighted image (DWI) of MRI has become widespread and reports have increased that MERS is related to various infectious diseases such as ; mycoplasma[6], staphylococcus aureus bacteremia[7], legionella[8], dengue fever[9], rubella[10], rotavirus[11], HHV-6[12] and adeno virus[13] in children[6~12] and adult[13]. Influenza virus type A[14] including 2009 H1N1 influenza[15,16] are the most reported MERS related to childhood infectious diseases, and other various types of viruses are also reported[6~13]. Here in we will report about MERS in a 5-year-girl with mumps virus infection.

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CASE REPORT

The patient is a healthy five year old girl. There were several children with epidemic parotitis in a

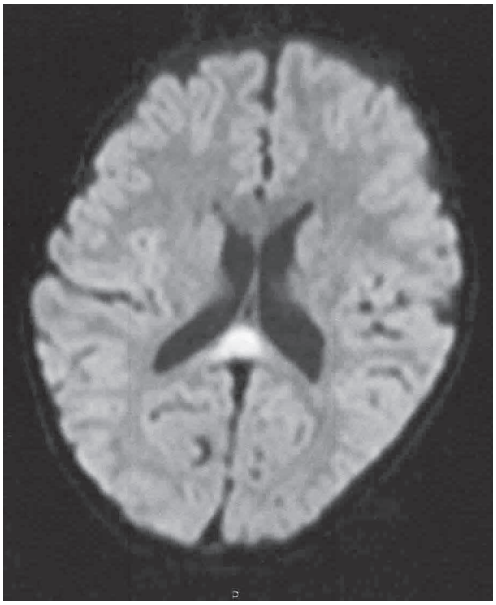

Figure 1
Diffusion weighted image of magnetic resonance imaging (Spin echo ; TE = 3900 msec ; TR = 88 msec, FA = 90) of the patient on admission showed high abnormal intensity with callsal splenial liesion. MRI of MERS is separated into type 1 which shows abnormality in the corpus callosum and type 2 which observes the lesion besides the corpus callosum. Her MRI is diagnosed as MERS type 1.

nursery school where she is enrolled. She had no history of mumps vaccination nor mumps infection. She had a fever over 38 degrees since morning. The right parotid gland was painful and swollen. Moreover, she suddenly laughed out loud several times. On the same evening, she developed the first febrile seizure with bilateral symmetry for 5 minutes during sleep. After about an hour, she had a second tonic-convulsion of bilateral symmetry, after which she was transported to our hospital. While being transported in an ambulance, she developed myoclonic seizures for a few minutes. When she arrived at the hospital, her level of consciousness almost improved (Japan Coma Scale ; JCS 1) and conversation with doctors and parents was possible. After observing for several hours at a outpatient department, she was able to walk and go back home. But she was brought back and hospitalized after having a 3-minute tonic-clonic convulsion in her car on her way home. While being hospitalized, she suddenly pointed to the wall and said "There is a fish", after which she could not answer her name. We estimated her consciousness level was JCS 3. Based on convulsions with clusters and delirium state, we tentatively diagnosed it as aseptic meningitis due to mumps. In the neurological examination, meningitis signs were not noted. To investigate the cause of this symptom, we performed blood examination but found no abnormality other than high amylase level of 872 IU/L and the sodium value was 133 mEq/L. The IgM value of mumps virus by the enzyme immunoassay (EIA) method was positive as 1.84 (reference value less than 0.08). The cerebrospinal fluid (CSF) examination showed a cell count of 2/3, glucose level of 43 mg/dl, and other values were all normal. The cerebrospinal fluid pressure was also normal. Virus isolation from CSF by polymerase chain reaction (PCR) method was performed, and the result was negative. Radiological examination with the DWI of brain MRI detected an abnormal intensity in the corpus callosum (Figure 1). Thus, she was diagnosed with MERS due to mumps. She was then given steroid mini-pulse therapy with 10 mg/kg of methyl-prednisolone (mPSL) for 3 days. Starting with mPSL pulse therapy, her consciousness improved completely the following day. After 3 days of mPSL therapy, she was discharged without convulsion recurrence. Electroencephalogram showed normal background activity on 3rd day from the admission. The abnormality of the corpus callosum was improved when examined by MRI on the 11th day.

DISCUSSION

In mumps infection, aseptic meningitis, hearing loss, orchitis, and other complications are known. Among them, the most frequent complication of childhood mumps infection is aseptic meningitis. The increase in the cell count of CSF is recognized in 65% or more of children with mumps. However, clinically diagnosed as sterile meningitis is in 1 to 3% of childhood mumps infection. In addition 2.5/1000 cases (0.0025%) of childhood mumps may complicate to meningoencephalitis that causes neurological sequelae. Importantly, the mortality of meningoencephalitis associated with childhood mumps is 0.5 to 2.3%[17]. Our patients also had a parotid gland swelling, disturbance of consciousness and repeated seizures. CSF examination was performed, but the number of cell count did not rise.

Hoshino *et al*. reported clinical statistical study on MERS in Japan during 2007–2010, from 210 pediatric hospitals. In this article, MERS is second common type of acute encephalopathy in Japan following acute encephalopathy with biphasic seizures and late reduced diffusion (AESD), and in 153 cases of MERS, the viral pathogens were influenza in 53 cases (34.4 %), rotavirus in 18 (11.7%), mumps virus in 6 (3.9 %), and HHV-6 in only 3 (2.0%). Notably, there were 5 cases (3.3%) following bacterial infections[18]. Mumps was the third prominent cause of MERS in this article.

For mumps-related MERS, there is a report that has developed after mumps vaccine[19,20]. Our patient was not vaccinated with mumps vaccine. It is important to enlighten not only aseptic meningitis caused by mumps but also complications of MERS. The incidence of aseptic meningitis as a side effect of mumps vaccine is 0.05% . Nagai *et al*. reported the onset of meningitis by the vaccine is low compared to the natural infection of mumps[21]. However, the incidence of MERS associated with mumps vaccine is unknown.

Finally, we discuss the relation between clinical symptoms in this case and MERS. In children with acute encephalopathy, there are many cases that recognize febrile convulsion and consciousness disorder as the initial symptoms. However many of the MERS do not develop with convulsions in clinical feature. The initial symptoms of common to MERS are consciousness disorders[12]. So, early diagnosis is difficult without MRI findings. Although the detailed mechanism of MERS has not been elucidated, abnormal signals of MERS on MRI are often recognized not only in the cerebral cortex causing convulsions but also in corpus callosum and deep white matter causing consciousness disorders. On the other hand, convulsions are often recognized in childhood meningitis. Moreover, meningeal signs are slight difficult to be observed in childhood meningitis. Viral meningitis is also well known as a complication of mumps infection in childhood. For these reasons, we initially considered aseptic meningitis associated with mumps. After that, finally an MRI was performed to confirm the diagnosis of MERS. In conclusion, we need to pay attention to MERS as a complication although rare in a mumps infection.

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