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Case Report

Differential diagnosis of MELAS syndrome from viral encephalitis by MRI: A case report

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Abstract

Background and objective: Mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes (MELAS) syndrome and viral encephalitis have little difference and the diagnosis is often uncertain based on clinical manifestations, even biochemical immune indicators couldn't make a definitive diagnosis. However, magnetic resonance imaging (MRI) seems to be able to provide a more definitive diagnosis. Material and method: A sick boy with headache, vomiting and convulsions, and 12 h unconsciousness was admitted to Kunming Children's hospital. After physical examination and history collection, blood and cerebrospinal fluid (CSF) were taken for laboratory tests, including cell count, blood gas analysis. Computer tomography (CT) and magnetic resonance imaging (MRI) of the brain were performed.

Results: MRI was first performed on October 7, 2017. All these finding suggested the diagnosis of viral encephalitis. A repeat MRI was performed on October 23, 2017, which showed resolution of lesions at the bilateral frontal and temporal lobe regions, but emerging new bilateral putaminal lesions, compared with previous MRI examination.

Conclusion: MRI examination could be a useful tool in the diagnosis of MELAS syndrome.

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Keywords: MELAS syndrome; Viral encephalitis; Differential diagnosis; Magnetic resonance imaging

1. Background

Mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes (MELAS) syndrome, is the most common phenotype of mitochondrial disease. It often develops in childhood or adolescence. Muscle histology is an essential component of the diagnostic work-up for mitochondrial cytopathies and is very important in both ruling in the disease as well as ruling out the disease (i.e., alternate diagnoses). A muscle biopsy method must provide tissue for histology,

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electron microscopy, enzymes and DNA and this can be obtained with a suction-modified 5 mm needle. Typical findings on brain imaging include stroke-like findings: Mitochondrial encephalopathy, lactic acidosis and stroke-like episodes (MELAS) is a rare multisystem disorder and is the most common maternally inherited mitochondrial disease. This condition has a special predilection for the nervous system and muscles. In this case report, we discuss a 13-year old male with clinical and radiological features highly suggestive of viral encephalitis syndrome at the beginning. Since this disease is rare and its clinical presentation is complex, it is among the most challenging to diagnose. It is difficult to differentiate between viral encephalitis and MELAS. Magnetic resonance imaging (MRI) and biopsy would provide clue to make accurate diagnosis of MELAS as early as possible.

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2. Ethics statement

The guardians of that participant child provided written informed consent. The study was conducted according to the declaration of Helsinki, implemented by guidelines of the World Health Organization. The experiment was conducted under the supervision of the ethics committee of the Kunming children's hospital.

3. Case presentation

A sick boy was 13-years and 4-months-old, accompanied with seven days of clinical manifestations of headache, vomiting and convulsions, and unconsciousness lasting for 12 h, who was admitted in Kunming Children's hospital on October 3, 2017.

According to his disease history, he had no obvious cause of severe stabbing headache beginning 7 days ago, which concentrated on top of the head and bilateral temporal region. He vomited for seven days, with non-spray-like vomiting, with just stomach contents. He had one episode off ever, up to 38.7 °C, without convulsions and coma.

4. Treatment history

First, he was treated in a local hospital, Fumin District Hospital. Oxygen, penicillin antibiotics, phosphocreatine, mannitol and furosemide were used to treat him. It improved his vomiting, but unconsciousness, paroxysmal irritation, maxillofacial swelling and limb stiffness developed following treatment. Especially, his right hand grab appeared as claw type. Then he was transferred to Intensive Care Unit (ICU) of Kunming Children's Hospital.

5. Physical examination

Under the conditions of oxygen inhalation, his lips were cyanotic, and he was coma-like, without spontaneous blinking, no response to sound and pain stimulation. Facial edema, pupils equal and circular, and slow response to light were observed. He had positive neck resistance and three depression signs. His limb muscle tone was reduced.

6. Laboratory tests

6.1. Blood routine

Leukocyte number concentration was $13.09 \times 10^9/L$; Neutrophil percentage was 92.70%; concentration of high-sensitivity C-reactive protein was 13.44 mg/L. Blood glucose concentration was from 7.53 mmol/L to 10.33 mmol/L.

6.2. Blood gas analysis

Blood gas analysis showed his blood lactate concentration was from 8.4 mg/L to 15.9 mg/L during admission in Kunming Children's hospital.

6.3. Cerebrospinal fluid (CSF) detection

CSF pressure increased to 250 mmH $_2$ O, having tendency of intracranial hypertension syndrome. CSF-white blood cell count reached to 6×10^6 /L of 8×10^6 /L total cell counting and CSF total protein concentration reached to 0.73 g/L. Biochemical analysis illustrate CSF-Glu was 10.25 mmol/L and CSF-chloride was 140.7 mmol/L. At last, the smear microscopy suggested there was no bacteria, acid-fast bacilli, cryptococcus or tuberculosis. Immuno-virological assay results suggested he had not been infected by EB virus, rabies, influenza virus or hand-foot-mouth disease.

6.4. Computer tomography (CT)

Brain CT was employed to exam his condition on October 3, 2017. Multiple low density areas were seen at the right occipital and temporal lobes with the largest $6.3 \text{ cm} \times 5.2 \text{ cm}$ with compression of the right lateral ventricle. Fig. 1. A diagnosis of encephalitis was made with cerebral infarction to be identified.

6.5. Magnetic resonance imaging (MRI)

MRI was performed on October 7, 2017 for the first time. All the constellation of findings suggested the diagnosis of viral encephalitis. The imaging findings included multiple abnormal signals at bilateral cerebellar hemispheres (Fig. 2A), cerebellar herniation (Fig. 2B), abnormal signals at cerebral hemispheres (Fig. 2C), thalami and basal ganglia (Fig. 2D). MRI diffusion weighted sequence showed lesions mainly involving bilateral temporal lobes, occipital lobes, parietal cortex and subcortical white matter. MRI with contrast showed lesions in bilateral occipital lobes, some sections of right temporal lobe.



Fig. 1. The right occipital lobe and temporal lobe show areas of decreased density.

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