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CASE REPORT

Mild Encephalitis / Encephalopathy with Reversible Splenial and Cerebellar Lesions (MERS Type II) in a Patient with Diabetic Ketoacidosis and Hypernatraemia

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ABSTRACT

Mild encephalitis / encephalopathy with a reversible splenial lesion is an uncommon condition that mainly affects young patients and children. Epilepsy and the use of antiepileptic medications are considered to be common associations. Other conditions such as infections, high-altitude cerebral oedema, and metabolic disturbances have also been suggested as causes. Concomitant involvement of the corpus callosum and cerebellum is even rarer. We present a patient with a reversible splenial lesion and concomitant reversible cerebellar lesions within the scope of diabetic ketoacidosis and hypernatraemia.

Key Words: *Encephalitis; Endocrine system; Magnetic resonance imaging*

中文摘要

一名同時患有糖尿病酮症酸中毒和高鈉血症的病人出現伴胼胝體壓部和小腦可逆性病變的輕度腦炎／腦病（MERS II型）

謝家熙、盧承迅

伴胼胝體壓部可逆性病變的輕度腦炎／腦病主要見於年輕患者和兒童。癲癇和服用抗癲癇藥物被認為與此病相關。此外，感染、高原腦水腫和代謝紊亂也可能是病因。此病累及胼胝體和小腦者罕見。本文報告一名同時患有糖尿病酮症酸中毒和高鈉血症的輕度腦炎病人出現伴胼胝體壓部可逆性病變。

INTRODUCTION

Mild encephalitis / encephalopathy with a reversible splenial lesion (MERS) has characteristic clinical and imaging features. The condition has recently been classified into type I and type II. Type I MERS is more common and usually presents as a single lesion in the splenium. Type II MERS is defined when additional lesions of similar signal characteristics are seen in other parts of the brain.

Various associations for MERS have been reported.[1] The underlying pathophysiology remains unclear,

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however. Possible explanations include transient development of intramyelinic oedema and inflammatory infiltrate.[2] Genetic factors might also predispose to this condition as familiar cases of clinical MERS have been reported.[3] We present a case of this rare finding in a patient with diabetic ketoacidosis and hypernatraemia.

CASE REPORT

In January 2014, a 29-year-old man with good past health was admitted for generalised weakness and confusion. He was a social drinker and had no prior history of recreational drug use. Clinically, the patient was confused and dehydrated. Arterial blood gas at admission showed metabolic acidosis (pH 7.26). Blood glucose (41.5 mmol/l; reference range, 3.9-6.1 mmol/l) and sodium level (180 mmol/l; reference range, 136-142 mmol/l) were markedly elevated. He was treated for diabetic ketoacidosis and hypernatraemia in the intensive care unit. His general condition improved and he was transferred to a general medical unit on day 3. Repeated blood tests showed persistent metabolic acidosis (pH 7.28), hyperglycaemia (16.5 mmol/l), and hypernatraemia (157 mmol/l). He subsequently developed progressive cerebellar signs and symptoms on day 6. Urgent computed tomography of the brain was unremarkable.

Lumbar puncture was performed. Biochemistry, microbiology, and serology results were all normal. Magnetic resonance imaging (MRI) of the brain on day 9 showed T2-weighted (T2W) hyperintense lesions with corresponding restricted diffusion over the splenium of the corpus callosum and the left external capsule. The whole cerebellum appeared slightly hypointense on T1W and mildly hyperintense on T2W and fluid-attenuated inversion recovery (FLAIR) images (Figures 1 and 2). Diffuse cerebellar swelling was noted. Restricted diffusion was demonstrated, particularly at the dentate nuclei region.

He was treated empirically for possible central nervous system infection with intravenous ceftriaxone (2 g every 12 hours) and acyclovir (800 mg every 8 hours). His neurological symptoms gradually improved. He was subsequently transferred for convalescence and further rehabilitation. A follow-up MRI scan 1 month later showed mostly resolved splenium and cerebellum abnormalities (Figure 3). His diabetic control was satisfactory with insulin. His fasting blood glucose was 6 mmol/l and sodium level was 140 mmol/l, which were within normal limits. He was otherwise well, with mild residual lower limb weakness. In view of the clinical history and presentation, as well as the laboratory findings, a diagnosis of MERS type II secondary to diabetic ketoacidosis and hypernatraemia was made.

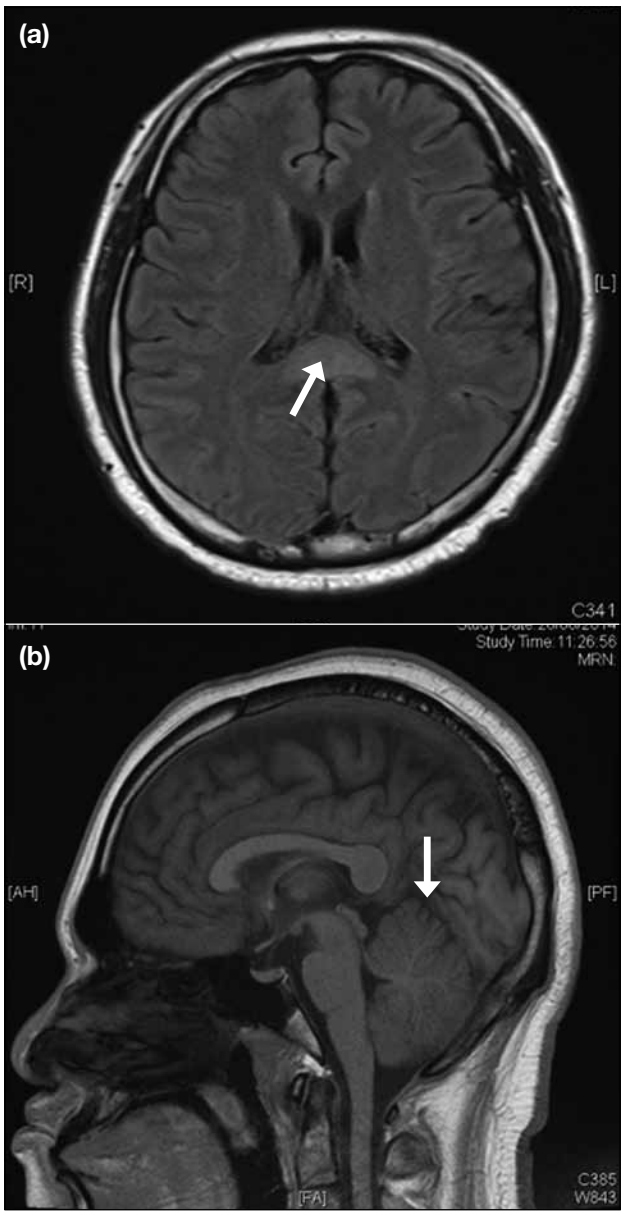

Figure 1. Magnetic resonance images of a patient with mild encephalitis / encephalopathy with a reversible splenial lesion. (a) Fluid-attenuated inversion recovery hyperintense signal at the splenium of corpus callosum (arrow), and (b) T1-weighted sagittal image shows a swollen cerebellum with mild T1 hypointensity (arrow).

DISCUSSION

MERS has only been described recently, but it is gaining more clinical attention with the advancement of imaging technology. Patients can present with non-

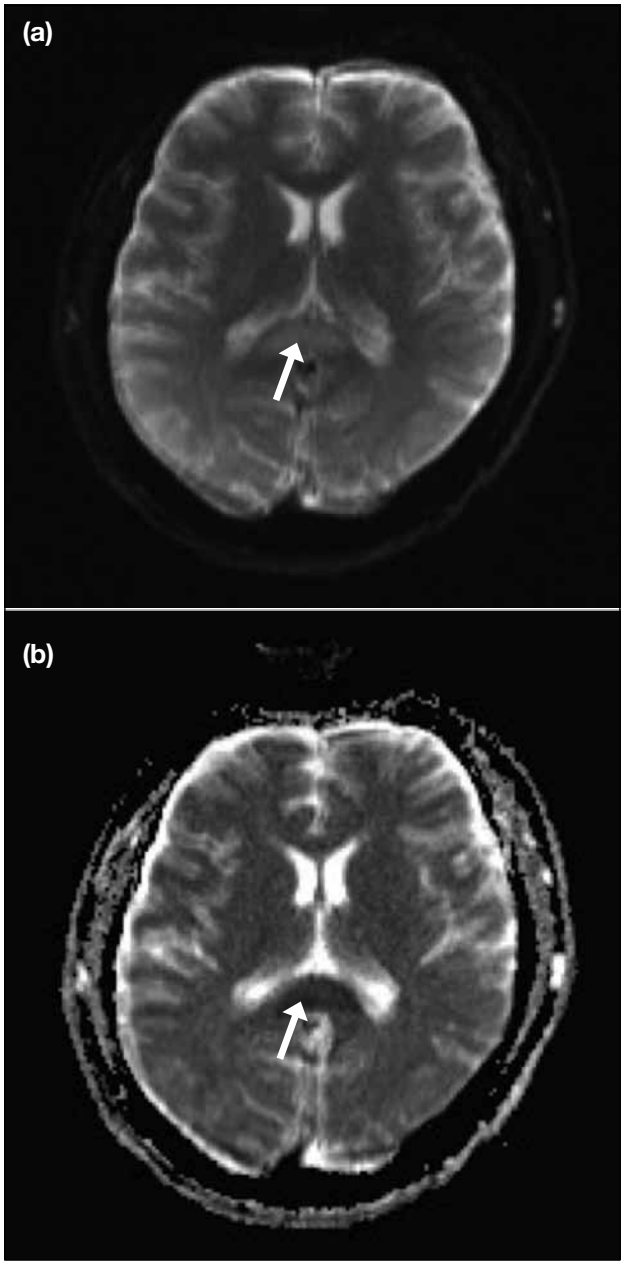

Figure 2. (a) Diffusion-weighted imaging, and (b) apparent diffusion coefficient sequence show restricted diffusion of the corresponding splenial lesion (arrows).

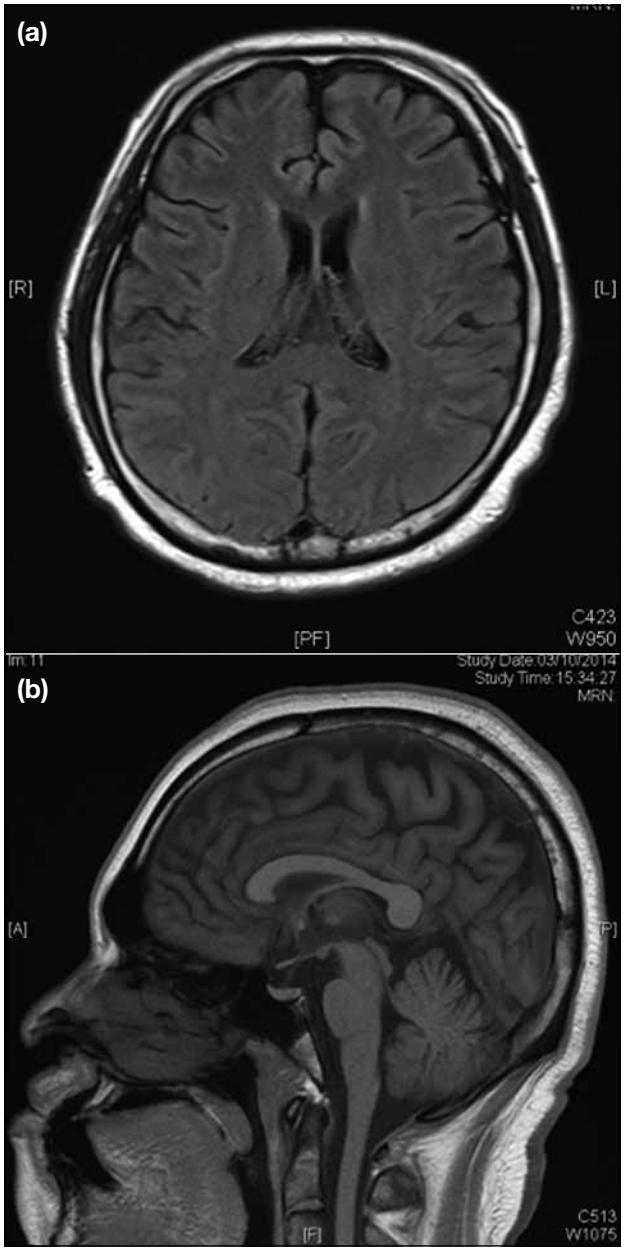

Figure 3. Follow-up magnetic resonance images show (a) complete resolution of the splenial lesion, and (b) resolved cerebellar swelling and normal signal intensity.

specific prodromal symptoms that mimic viral infection. Encephalopathy symptoms such as speech difficulties, drowsiness, decreased consciousness, delirium, seizures, irritability, agitation, and disorientation evolves with disease progression.[4] Delirium is considered to be the most common neurological symptom according to a multicentre study in Japan.[5]

The condition is classified into type I and type II depending on whether there is additional extra corpus callosum white matter involvement. The splenial lesion tends to linger in follow-up MRI, however. It is believed that type II MERS has a better prognosis with complete resolution usually within 2 weeks. Type I MERS is more commonly associated with clinical encephalitis / encephalopathy.

MERS lesions are hyperintense on MRI in T2W and

FLAIR sequences. On MRI, the splenial lesion can be further divided into two types according to its appearance. An oval-shaped well-circumscribed focal lesion is usually located in the middle of the splenium, whereas a wider, more infiltrative, lesion involves the entire splenium ('boomerang sign'). Diffusion-weighted imaging is more sensitive for the detection of early lesions. Single-photon emission computed tomography is another sensitive investigation for identifying a hypoperfused region that may remain normal on MRI, and is especially useful in screening for MERS type 2. MR spectroscopy usually demonstrates normal n-acetylaspartate-to-creatine and choline-to-creatine ratios, suggesting that there is no neuroaxonal damage or demyelination.

Differential diagnoses include posterior reversible encephalopathy syndrome, multiple sclerosis, lymphoma, and pontine myelinolysis. Acute disseminated encephalomyelitis (ADEM) is an important consideration in patients with encephalitis / encephalopathy. ADEM, however, usually shows multiple bilateral and asymmetrical hyperintense lesions in the subcortical white matter on T2W sequence. Resolution of lesions in ADEM usually takes several months, contrary to the rapid disappearance of lesions in MERS.

Treatment is based on the underlying causes, for example, correction of metabolite disturbance or titration of an anti-epileptic regimen. MERS has a favourable prognosis and hence conservative approach is recommended. A limited follow-up MRI to demonstrate resolution should reassure patients and clinicians.

CONCLUSIONS

MERS is a clinico-radiological diagnosis, characterised by mild encephalitis or encephalopathy associated with a reversible lesion at the splenium of corpus callosum. Because of its benign nature, it is an important differential diagnosis from other sinister diseases affecting the corpus callosum such as tumour or demyelination.

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