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Acute disseminated encephalomyelitis with medial temporal lesions mimicking acute limbic encephalitis

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Abstract

We describe a 59-year-old Japanese man with acute disseminated encephalomyelitis (ADEM) demonstrating medial temporal lesions on magnetic resonance images (MRI) mimicking acute limbic encephalitis. This patient had antecedent infection and developed urinary retention. Subsequently he showed various neurological manifestations including truncal ataxia, nystagmus and generalized convulsions, suggesting multifocal brain and spinal cord lesions. Brain MRI demonstrated hyperintensity areas restricted to the left hippocampus, amygdala, and inferior temporal gyrus. The cerebrospinal fluid did not show any pleocytosis or elevated protein levels, and had no evidence of herpes virus infection. Serum antineuronal antibodies were not identified. Corticosteroid therapy markedly improved the symptoms. Although these clinical features indicating multifocal central nervous system lesions were compatible with those of ADEM, none of the previously reported patients with ADEM showed lesions confined to the medial temporal lobes mimicking acute limbic encephalitis. Medial temporal lesions are characteristic features of herpetic or non-herpetic acute limbic encephalitis. Findings in our patient suggest that non-herpetic limbic encephalitis patients showing marked response to

corticosteroid therapy represent an atypical form of ADEM.

(171 words)

Key words: ADEM; herpes simplex virus; limbic encephalitis; MRI

Introduction

Acute disseminated encephalomyelitis (ADEM) is a rare inflammatory disorder characterized clinically by antecedent viral infection or vaccination and sudden or acute onset disseminated encephalomyelitis, and pathologically by numerous perivascular foci of demyelination throughout the cerebrum, brainstem, cerebellum and spinal cord [1].

Magnetic resonance images (MRI) findings in ADEM have been reported in several studies; typical ADEM lesions are multifocal white matter lesions in the cerebrum [1,2], and cerebral cortical involvement has also been reported [1,3-6]. In this case report, we describe an ADEM patient with medial temporal cortical lesions mimicking acute limbic encephalitis on MRI.

Case report

A 59-year-old Japanese man developed acute fever for three days. Three days later (day 1), he demonstrated acute urinary retention, and urologic examination showed sensory neurogenic bladder. Cystometry demonstrated detrusor underactivity. On day 4, he complained of memory disturbance with fluctuation. Neurologically (on day 18), he

showed disorientation as to date and place. Lateral gaze nystagmus was disclosed.

Although deep tendon reflexes of the lower extremities were hyperreflexic, the plantar reflexes were flexor. Gait was ataxic and catheterization was required because of severe urinary retention. On physical examinations, no skin lesions, genital ulcers, or oral aphthous ulcers were observed. Uveitis was not detected on ophthalmological examinations. On day 19, consciousness deteriorated and he developed a complex partial seizure followed by secondary generalized convulsions. Since the convulsions persisted, continuous administration of midazolam and mechanical ventilation were required.

Brain MRI on day 19 showed hyperintensity areas in diffusion-weighted, T2 weighted, and fluid attenuated inversion recovery (FLAIR) images in the left temporal lobe involving the hippocampus, amygdala and inferior temporal gyrus (Fig. 1A). The T1 weighted images showed hypointensity with contrast enhancement (Fig. 1B). There were no lesions in the cerebellum, brain stem or spinal cord on the MRI. No apparent abnormalities were disclosed on either hematological or blood chemistry analyses. Serum antinuclear, anti-SS-A, anti-SS-B, and anti-thyroid antibodies were negative. Serum levels of angiotensin converting enzyme were not elevated. There were no serum

anti-neuronal antibodies using immunohistochemistry on sections of rat cerebral cortex.

The cerebrospinal fluid on day 20 did not show any pleocytosis or elevated protein levels.

The oligoclonal bands were negative. Polymerase chain reaction (PCR) measurements of

virus DNA in cerebrospinal fluid were all negative, including herpes simplex virus,

varicella zoster virus, cytomegalovirus, Epstein-Barr virus, human herpesvirus 6, and

human herpesvirus 7.

A diagnosis of acute limbic encephalitis was made. We started acyclovir 1,500

mg/day intravenously on day 19. Acyclovir was administered for 14 days. On day 20, we

added intramuscular injection of dexamethasone 16 mg/day. The doses of dexamethasone

were reduced to 8 mg/day on the 5th day after initiation of dexamethasone, and

dexamethasone was administered for 8 days in total. On day 25, seizures disappeared, and

consciousness became clear. Brain MRI revealed no obvious improvement of the

hyperintense lesions in the left medial temporal lobe (Fig. 1C), however, in the

T1-weighted images apparent contrast enhancement was disappeared (Fig. 1D). On day

35, urinary retention disappeared without medication. The patient presented with mild

memory disturbance and retrograde amnesia. Brain MRI in a year after the onset

demonstrated both improvement of the hyperintense lesions and mild atrophy in the left medial temporal lobe with no additional lesions. He had no episode of relapse for 3 years after clinical remission.

Discussion

Although MRI findings were confined to the limbic systems on the left side, there was no evidence of herpetic viral infections, supporting the initial diagnosis of non-herpetic acute limbic encephalitis [7]. While the presence of various neurological manifestations, including neurogenic bladder, nystagmus, truncal ataxia and epilepsy, suggested multifocal lesions of the brain and spinal cord; taken together with antecedent infection, the clinical features in this patient, showing marked response to steroid therapy, were compatible with those of ADEM [1,6]. Although autoimmune inflammatory disorders, metabolic encephalopathy, Hashimoto's encephalopathy and brain tumors could show medial temporal lesions and ADEM-like features [8-10], he had no evidence of having such diseases. He had no serum antineuronal antibodies indicating no evidence of paraneoplastic limbic encephalitis. The medial temporal lesion mimicking acute limbic

encephalitis, absence of pleocytosis in the cerebrospinal fluid, and absence of cerebellar and spinal cord lesions detected on MRI are quite atypical for ADEM. However, urinary retention was common in ADEM [11]. Concerning the limbic lesions, only one case of acute hemorrhagic leukoencephalitis, a more severe form of ADEM, has been reported to have features mimicking herpetic encephalitis [12]. Although multifocal white matter lesions are found in ADEM patients, several ADEM patients have been reported with cerebral cortical lesions [1,3-6], suggesting that medial temporal lobes could be affected in patients with ADEM.

Concerning normal CSF findings and the absence of demyelinating lesions detected on MRI, several ADEM patients that lacked pleocytosis or MRI findings have been reported [4,13]. The diagnosis of ADEM requires the following: acute onset, symptoms and signs of disseminated multifocal central nervous system involvement and an antecedent infection or vaccination [1,6,13]; MRI findings are frequently normal even in patients with severe clinical manifestations [13]. Taken together with these findings, although the possibilities of another cause of acute limbic encephalitis are not fully excluded, our patient indicates the existence of an atypical form of ADEM with lesions

confined to the limbic system on MRI.

In conclusion, we described a patient with an atypical form of ADEM characterized by medial temporal lesions mimicking acute limbic encephalitis.

(891 words)

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Figure legend

Fig. 1: Brain magnetic resonance images (MRI) on day 19 (A,B). Fluid attenuated inversion recovery (FLAIR) axial image (A) showed hyperintensity areas in the left medial part of the temporal lobe. T1-weighted coronal image with gadolinium enhancement (B) showed the enhanced left inferior temporal gyrus and edema in the left amygdala. Brain MRI on day 25 (C,D). FLAIR image (C) revealed no apparent improvement of the hyperintense lesions in the left medial temporal lobe, however, T1-weighted image with gadolinium enhancement (D) revealed no apparent contrast enhancement. FLAIR image in a year after the onset demonstrated both improvement of the hyperintense lesions and mild atrophy in the left medial temporal lobe (E).

