Multiple complete ring-shaped enhanced MRI lesions in acute disseminated encephalomyelitis

Kun Eng Lim*, Yuan Yu Hsu, Wen Ching Hsu, Cheng Yi Chan

Department of Radiology, Chang Gung Memorial Hospital, 5 Fu Hsin Road, Kwei Hsian Hsiang, Taoyuan Hsien, Taipei, Taiwan

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Abstract

Acute disseminated encephalomyelitis (ADEM) lesions may or may not enhance with contrast material on contrast T1-weighted magnetic resonance images (T1WI). Enhancing ADEM lesions may show varying patterns of contrast enhancement, which include nodular, diffuse nodular, amorphous, gyral, spotty and incomplete ring-like. Multiple, complete ring-shaped enhanced lesions on contrast T1WI is a rare finding in ADEM. We report such a magnetic resonance imaging (MRI) finding in a 36-year-old female patient with this disease.

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1. Introduction

Acute disseminated encephalomyelitis (ADEM) is an acute inflammatory demyelinating disease of the central nervous system (CNS) that is typically preceded by an infectious illness or a vaccination [1,2]. In some cases of ADEM, no precipitating factor has been traced. It is usually a monophasic illness. Magnetic resonance imaging (MRI) is the most sensitive imaging modality for detection of ADEM lesions. The MRI findings of ADEM are well described [3–7]. Enhancement of ADEM lesions may or may not be seen, depending on stage of the inflammation/demyelination, as well as severity of injury to the blood–brain barrier (BBB). The pattern of enhancement of ADEM is variable, but presenting as multiple complete ring-shaped enhanced lesions is unusual. We here report a case of ADEM presenting with this MRI finding.

2. Case report

A 36-year-old female with a history of chronic pansinusitis for a few years was presented to our hospital with 1 week of mental change. She also suffered from progressive conscious change, unsteady gait, mutism, apathy and personality change for 1 week. She complained about headache, difficulty in concentrating and right hand weakness 1 month ago. The headache was not associated with nausea, vomiting, blurred vision and seizure. She had no history of vaccination, as well as drug abuse recently. She also had no travel history recently too.

On admission, she was afebrile and her neck was supple. Neurological examination revealed mild decreased muscle power of bilateral upper limbs and increased deep tendon reflexes at bilateral lower limbs. No Babinski sign, Kernig sign or cerebellar sign was found.

On admission, the results of all routine blood test, renal function, liver function tests and electrolyte levels were all normal. Antinuclear antibody was positive. Cerebrospinal fluid (CSF) study revealed a normal level of glucose and lactate, normal protein profile and a normal cell count. India ink preparation, acid-fast smear, cryptoccocal antigens, Gram stain and microscopic examinations were negative. The CSF cultures of bacteria were negative. CSF was not sent for oligoclonal studies. The CSF IgG antibodies for herpes simplex-1 and herpes simple-2 viruses were positive.

An EEG showed diffuse cortical dysfunction. Visual-evoked and brainstem auditory-evoked potentials were normal. Median nerve somatosensory-evoked potentials suggested a somatosensory system conduction defect in the peripheral nerve of both arms.

* Corresponding author.

E-mail address: lketh@adm.cgmh.tw (K.E. Lim).
Initial brain computed tomography (CT) showed multiple hypodense lesions in bilateral hemispheres. Brain MRI demonstrated multiple oval-shaped and variable-sized high-intensity lesions in the white matter of bilateral cerebral hemispheres on T2-weighted sequence (Fig. 1). The lesions appeared hypointense on T1-weighted sequence (Fig. 2). The contrast TIWI showed multiple complete ring-shaped enhanced lesions (Fig. 3). Stereotactic brain biopsy was performed in the frontal area 2 weeks after onset. Histopathological examination of biopsy material revealed numerous foamy histocytes, gliosis and perivascular cuffing of lymphocytes (Fig. 4). No microorganism or malignancy was present.

A diagnosis of ADEM was favored after combined typical clinical pictures, CSF analysis, white matter abnormality on brain MRI and pathological examination. After treatment with steroid medication, her neurological condition gradually improved over following days, disturbed conscious state recovered and no other discomfort. The patient remains asymptomatic after 3 months of follow-up. Follow-up brain CT 3 months after the onset of the illness still displayed multiple hypodense lesions but of smaller size at the same sites and no new lesions appeared.

3. Discussion

ADEM is an immunologically mediated inflammatory demyelinating disease of the CNS. Lesions in ADEM have striking similarities to those of experimental allergic encephalomyelitis [8]. ADEM is more common in children than in adult. Although recurrent cases have been reported [6,9], it is usually a monophasic illness. ADEM usually responds to steroid therapy, especially during the acute stage of the disease. Our patient improved rapidly after administration of steroid medication. It usually occurs after vaccination against various viruses or after many viral infections. The disease is characterized pathologically by perivascular inflammation (infiltration of vessel walls and perivascular spaces with lymphocytes, plasma cells and monocytes), edema and demyelination within the CNS. ADEM predominantly involves white matter, but that of deep grey matter involvement is not uncommon, and is now considered part of the radiological spectrum [5,10].
MRI is becoming the imaging modality of choice for detecting the lesions of ADEM. The MRI findings, however, are nonspecific for this disease. The lesions of ADEM are best seen on long TR images. The typical MRI findings are asymmetrical bilateral multiple patchy areas of homogeneous or slightly inhomogeneous increased signal intensity on T2-weighted and proton-density weighted images, and fluid-attenuated inversion recovery sequence [3,4]. On T1-weighted images, iso- to low-signal intensity lesions in the white matter are seen. Bilaterally symmetrical white matter lesions have been reported too. The ADEM-related lesions may be large and confluent occupying almost all of the white matter, but smaller lesions resembling those of multiple sclerosis are reported.

Gadolinium contrast enhancement has been described in ADEM and other demyelinating disease [11,12]. The contrast enhancement seen in the demyelinating processes is caused by breakdown of the BBB, but it may be suppressed by corticosteroids. Enhancement of the lesions after contrast medium injection may or may not be seen, depending on the age of the lesions [3,10,13]. Enhancement of the lesions may also depend on the dosage of contrast agent, magnitude of BBB abnormalities, exact MRI pulse sequence and the time from injection to imaging [4,12]. The pattern of enhancement is nonspecific and may show spotty, nodular, diffuse nodular, amorphous, gyral or incomplete or a complete ring patterns of enhancement [3,6,11]. In our case, multiple complete ring-enhancing lesions in the white matter of the cerebrum were depicted on contrast T1WI. Multiple complete ring-enhanced lesions are very uncommon enhanced MRI pattern in ADEM. The presence of multiple ring-enhanced lesions on contrast T1WI has previously been described in three patients. In two patients, only a few ring-shaped enhancing nearly symmetric lesions were found predominantly in the supratentorial white matter [4]. In the third patient, several irregular ring-enhanced lesions were seen in posterior parietal lobes [11].

The presence of multiple complete ring-enhanced lesions in the cerebral white matter on contrast T1WI is a nonspecific finding for ADEM. When contrast T1WI shows multiple ring-enhancing lesions in the white matter of the CNS, the differentiate diagnosis includes multiple sclerosis, brain abscess, metastatic tumors, tuberculomas, toxoplasmosis and histoplasmosis. The combined clinical data and imaging findings usually can allow differentiation ADEM from these diseases.

In conclusion, we report a rare enhanced pattern of ADEM, multiple complete ring-enhanced lesions on contrast T1WI. Although this MRI finding is not specific, we should keep in mind that this kind of image expression could be ADEM too, in the appropriate clinical setting.

References