Case report

Multiphasic disseminating encephalomyelitis (MDEM)

Bimsara Senanayake[1]

Sri Lanka Journal of Neurology, 2013, **2**, 17-18

Index words: Acute disseminating encephalomyelitis (ADEM), Multiphasic disseminating encephalomyelitis (MDEM), Post chicken pox CNS demyelination

Introduction

Acute disseminated encephalomyelitis (ADEM) is an immune mediated disease of the brain which usually follows a viral infection or vaccination[1]. However it could occur following other infections or spontaneously. Although it affects all age groups, most reported cases are in children and adolescents, with an average age around 5 to 8 years[2]. A full recovery is seen in 50 to 75% of cases; while up to 70 to 90% recover with some minor residual disability[3]. The average time to recover is one to six months. It produces multiple inflammatory lesions in the brain and spinal cord, particularly in the white matter. Usually these are found in the subcortical and central white matter and cortical gray-white junction of both cerebral hemispheres, cerebellum, brainstem, and spinal cord[4]. When the patient suffers more than one disseminated demyelinating episode, it is called recurrent disseminated encephalomyelitis (RDEM) or multiphasic disseminated encephalomyelitis (MDEM)[5,6]. Such a case of MDEM following chicken pox infection in a 20-year-old Sri Lankan patient is reported here. Relapsing remitting multiple sclerosis (RRMS) is the closest differential diagnosis. MR imaging findings and the clinical history makes RRMS less likely in this case.

Case history

A 20-year-old female developed lower limb weakness with a mid thoracic sensory level i.e. features compatible with transverse myelitis in February 2012. She suffered from chicken pox a week prior to this initial neurological deficit. She was unable to walk and was bed bound with no sphincter control. She was treated with intravenous steroid pulses to which she responded. However her cognitive skills were poor with occasional confusion, but seizures were not reported. The MRIs showed large bilateral subcortical and central white matter lesions which were not enhancing (Figure 1). Periventricular lesions were not a prominent feature. She was readmitted in April 2012 with weakness of lower limbs for which she again received treatment with IV methyl prednisolone. She gradually improved from this event and was discharged. In September 2012 she again developed right upper limb weakness followed by right lower limb weakness – a 'stroke like' event. She was re-admitted and imaged again. The right 'hemiplegia' again improved with IV methyl prednisolone therapy. Repeat MRI did not show any new lesions or GAD enhancement. Previous brain lesions were seen along with the cervical cord lesions and cord swelling. Visual evoked responses were bilaterally normal. Cerebro spinal fluid analysis too did not reveal any abnormality.

Figure 1. The MRI of the brain showing large bilateral subcortical and central white matter lesions which were not enhancing.

Discussion

Relapsing remitting multiple sclerosis (RRMS) and recurrent or multiphasic disseminated encephalomyelitis (MDEM) are considered different diseases[5]. However ADEM could be confused with the first episode of MS and RRMS needs to be considered in cases of MDEM. However they differ in 2 important ways[5].

a) MS occurs in people who are genetically vulnerable, a feature noticeably lacking in ADEM. ADEM is usually post infectious or post vaccination

b) Even more importantly the sharp edges of the typical MS lesion have been recognized by neuropathologists as pathognomonic for MS; not described in ADEM

[1] *Institute of Neurology, National Hospital of Sri Lanka.*

In a study comparing children with ADEM/MDEM complex with those with MS some features **were, more commonly seen with ADEM/MDEM group**[6],

1. A pre-demyelinating infectious disease
2. Encephalopathy
3. Polysymptomatic presentation
4. Seizures – favoring a diagnosis of ADEM
5. Blood leucocytosis

MRI showed that subcortical white matter lesions were almost universal in both groups, though peri ventricular lesions were more common in multiple sclerosis. Follow-up MRI revealed complete or partial lesion resolution in 90% and no new lesions in the ADEM/ MDEM group. It has been demonstrated that relapses may occur immediately after ADEM. If these relapses are thought to represent part of the same acute monophasic immune process, the term 'multiphasic disseminated encephalomyelitis' (MDEM) is used.

MRI imaging is the most useful tool so far to differentiate RRMS from MDEM in the absence of any specific biomarker. In ADEM/MDEM, FLAIR sequence and T_2-weighted images show abnormalities than in T_1-weighted images. The lesions were predominantly in the white matter. Absolute and relative sparing of the peri ventricular white matter occurred in 56 and 78% of images, respectively. Involvement of the deep and subcortical white matter was nearly universal. The supra tentorial white matter lesions were universally asymmetrical. In MS T_2- images demonstrated the lesions best. A large majority of patients have periventricular lesions. The lesions were also prevalent throughout the subcortical white matter (92% of images). No patients had any cortical grey matter involvement. The thalami and basal ganglia were involved infrequently (25 and 8%, respectively). Fifty-eight per cent of the images had at least one lesion over 1 cm in size.

Post infectious acute disseminating encephalomyelitis with recurrent symptoms and CNS demyelination should be differentiated from RRMS. ADEM/MDEM complex generally has a good long term prognosis.

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