SUMMARY

Multiple sclerosis is considered the most frequent demyelinating disorder of the Central Nervous System (CNS) among young adults, yet is very rare before 10 years old. Acute disseminated encephalomyelitis is a monophasic, polysymptomatic disorder that involves the CNS white matter with demyelinating lesions, which usually occurs after systemic viral infections. These two demyelinating diseases can present initially as an acute focal neurological syndrome and they can be difficult to distinguish. We describe a case of a nine-year-old girl that presented initially with dysphonia, gait ataxia, eyelid myokymia and brainstem disturbances. This was her second episode; the first episode was at the age of four years old. She recovered without neurological sequelae. The brain magnetic resonance imaging (MRI) demonstrated multiple demyelinating lesions in the white matter, cortical regions of the frontal lobe, periventricular distribution, internal capsule, corpus callosum and cerebellum. The purpose of the presentation of this case was to highlight the similarities between these two entities, since the clinical picture and neuroimaging are difficult to distinguish, mainly in relation to the first episode.

Keywords

multiple Sclerosis, multiphasic acute disseminated encephalomyelitis, childhood

Multiphasic acute disseminated encephalomyelitis and differential with early onset multiple sclerosis

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Multiple Sclerosis (MS) is considered the greatest demyelinating disorder in young adults, yet rare before 10 years. The overall incidence of acquired demyelinating syndromes in children and adolescents ranges from 0.6 to 1.66 per 100 000 children per year (1,2). Acute disseminated encephalomyelitis (ADEM) is a single-phase, polysymptomatic disorder involving central nervous system blanking, leading to demyelinating lesions secondary to systemic viral infections, often reaching the age of 5 years of age (3). For confirmation diagnosis, there is no specific biological marker test or confirmatory test, the MRI being considered the elected exam. Analysis of the cerebrospinal fluid may be useful, showing pleocytosis lymphocytic cells without oligoclonal bands and elevation of albumin. These pathologies may present with a focal neurological syndrome whose differential diagnosis is difficult to distinguish.

We describe a 9-year-old girl with a family health history, that eight days before admission she had gastroenteritis, and on admission presented difficulty walking, dysphonia and dysphagia. Neuro-psychomotor development was normal until that time. At the age of 4, she presented a similar condition accompanied by altered consciousness and coma that was interpreted as viral meningoencephalitis, evolving without sequelae.

Physical examination revealed eyelid myokymia on the right, ataxia, dysphonia, left upper limb monoparesis, left central facial paralysis and involvement of the X and XII cranial nerves.

Current brain MRI revealed multiple demyelinating lesions in the white matter in the frontal and periventricular regions involving the internal capsule, corpus callosum and cerebellum (Figure 1). Cerebrospinal fluid found a slight increase in immunoglobulins (12.7%) and absence of oligoclonal bands. Our patient met the criteria for multiphasic acute disseminated encephalomyelitis (MDEM): i) Two clinical events meeting criteria for acute disseminated encephalomyelitis, separated in time by greater than 3 months, and ii) No evidence for clinically-silent new lesion formation on MRI between acute disseminated encephalomyelitis episodes (4). The patient was medicated with intravenous pulsotherapy of methylprednisolone and acyclovir, obtaining a good
recovery in three weeks.

The International Paediatric Multiple Sclerosis Study Group defines ADEM as i) a first polifocal, clinical CNS event with presumed inflammatory demyelinating cause; ii) encephalopathy not explained by fever, systemic illness, or postictal symptoms; iii) no new clinical and MRI findings emerging 3 months or more after the onset; iv) brain MRI is abnormal during the acute (3 mo) phase with diffuse, poorly demarcated, large (> 1-2cm) lesions predominantly involving the cerebral white matter (5).

The distinction between ADEM, MDEM or MS has been previously explored with no satisfactory consensus. Historically, ADEM was defined as the initial presentation of disseminated encephalomyelitis and MDEM as the occurrence of new symptoms in the setting of a history of ADEM.

The hallmark of this new category was the occurrence of two clinicoradiographic episodes of disseminated encephalomyelitis separated by at least three months. The clinical findings were defined as being new or a re-emergence of prior symptoms. If the patient sustained three or more episodes, it was classified as having a chronic inflammatory demyelinating disorder (5).

Our patient had an interval of five years between the first and the second clinical episode, the time between them can vary from three months to 33 years as reported by Numa et al. (6).

The presence of antibodies directed against anti-myelin oligodendrocyte protein (MOG) occurs in monophasic demyelinating disorders, particularly, in younger children and patients with ADEM. However, up to 1/3 of these children with MOG-abs will relapse within 2 years. Recent cohorts have suggested that a significant percentage of patients with recurrent optic neuritis, multiphasic demyelinating encephalomyelitis, ADEM associated with optic neuritis and neuromyelitis optica spectrum disorders have MOG-abs (7,8).

A diagnosis of MS can be confirmed by the presence of recurrent clinical demyelinating events and/or MRI evidence for new lesions involving different regions of the CNS. Implementation of the 2010 revised McDonald criteria may allow for diagnosis to be made at the time of the first demyelinating syndrome if imaging demonstrates silent lesions in two of the four regions typical for MS, at least one of which enhances with gadolinium. When criteria are not met at the time of the first event, new clinical attacks and/or serial imaging demonstrating accrual of lesions are needed to confirm the diagnosis of MS (9).

The purpose of the presentation of this case was to highlight the similarities between these two entities, since the clinical picture and neuroimaging are difficult to distinguish, mainly in relation to the first episode. Considering that 25% of cases with ADEM evolve to MS, evolutionary studies of neuroimaging are recommended.

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References


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