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CASE REPORT

Isolated palatal weakness without optic neuritis as the presenting manifestation of multiple sclerosis and its diagnostic dilemma with acute disseminated encephalomyelitis in a young boy

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ABSTRACT

We present a case of a young boy who at initial presentation was diagnosed as acute disseminated encephalomyelitis (ADEM) but subsequently on follow-up was diagnosed as multiple sclerosis (MS). Differentiating ADEM from MS in their first presentation can be tricky as the features may not be typical of anyone. The importance lies in the close follow-up of these patients.

Key words: Acute disseminated encephalomyelitis, magnetic resonance imaging, multiple sclerosis, steroids

INTRODUCTION

Acute disseminated encephalomyelitis (ADEM) is a postinfectious acute demyelinating disorder whereas multiple sclerosis (MS) is an immune-mediated disorder with chronic inflammation and variable neuronal injury.[1] ADEM typically has a monophasic course after a febrile episode whereas MS has relapsing nature characterized by optic nerve involvement. The relationship between MS and ADEM is complicated, and difficulties exist in distinguishing them, especially in young children. Although ADEM is more common in children, making an accurate distinction is important as immunomodulatory agents could alter the course of disease.

CASE REPORT

A 12-year-old boy, right-handed presented with a sudden onset change in voice for 5 days before hospitalization. He denied any history of fever, headache, convulsion, or any limb weakness. On examination, there was left-sided palatal weakness without any other neurological signs. Systemic examination was within normal limits. His magnetic resonance imaging (MRI) brain [Figure 1a and b] was suggestive of multiple nodular lesions with peripheral enhancement in cerebral hemispheres, deep periventricular, and juxtacortical white matter, suggestive of ADEM. The patient was evaluated as shown in Table 1 below. The patient was treated with 5 days pulse steroids-intravenous methylprednisolone, followed by tapering course of oral steroids (60 mg oral prednisolone for 8 weeks followed by a taper 5 mg every 2 weeks) and he fully recovered.

However, 5 months after stopping steroids, he presented again with slurring of speech and dribbling of saliva from the right side of the mouth. On examination, he had right-sided upper motor neuron facial palsy with right-sided cerebellar

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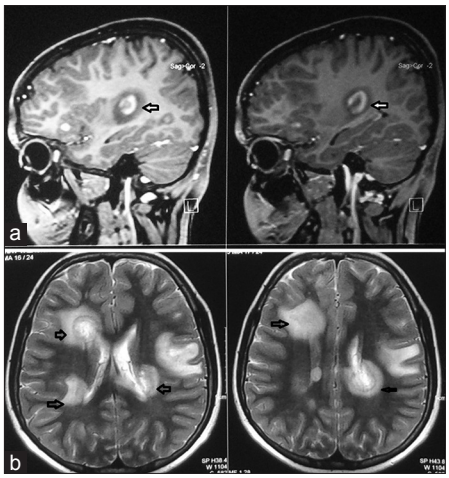

Figure 1: (a and b) T2-weighted magnetic resonance imaging postgadolinium contrast suggestive of multiple nodular lesions with peripheral enhancement in cerebral hemispheres, deep periventricular, and juxtacortical white matter (marked by arrows)

ataxia. Rest of the neurological examination was normal. His repeat MRI was suggestive of near total resolution of old lesions with appearance of new lesions of similar morphology in bilateral frontoparietal periventricular white matter as well as in the right brachium pontis extending to superior cerebellum showing irregular peripheral enhancement [Figure 2a and b].

MRI spine was normal. The patient was retreated with intravenous methylprednisolone and started on azathioprine. Weakness resolved completely with no residual neurodeficit.

Two months later, he presented third time with dragging of the left foot while walking and difficulty in doing work with the left hand. On examination, left-sided pronator drift was present. Power in the left upper and lower limb was Grade 4. He had exaggerated reflexes on the left side with positive Babinski's sign. A repeat MRI was done which showed as compared to previous scan, evidence of disease activity with multiple foci of demyelination in cerebral white matter showing enhancement, and appearance of new lesions [Figure 3a and b].

The patient was treated as pediatric MS and started on a short course of steroid with interferon beta-1a once a week. At 1-year follow-up, he is stable without any further attacks and no neurodeficit.

DISCUSSION

MS is a chronic inflammatory disease of the central nervous system (CNS) characterized by immune-mediated myelin loss with variable degrees of axonal injury. It is characterized by recurrent attacks of neurological dysfunction. ADEM

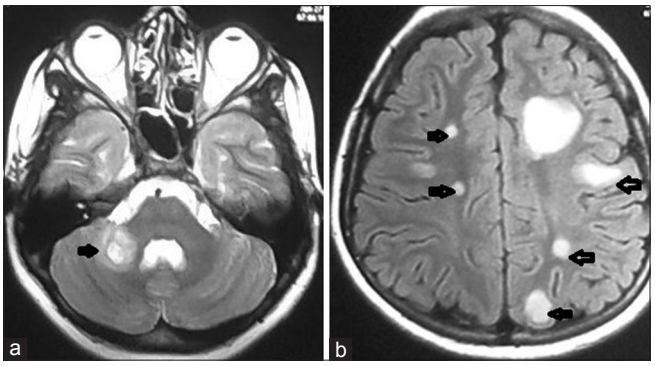

Figure 2: (a and b) Repeat magnetic resonance imaging at second visit showing new lesions of similar morphology in bilateral frontoparietal periventricular white matter, right brachium pontis extending to superior cerebellum showing irregular peripheral enhancement (marked by arrows)

Table 1: Investigations of patient done during hospitalisation

Investigations with normal range in SI units Lab values
CSF protein (15-45 g/l) 0.61
CSF leucocyte count (0-5 lymphocytes) 25 lymphocytes
CSF IgG index (0.28-0.66) 0.72
CSF oligoclonal bands present
HIV/HBsAg/Anti-HCV negative
VDRL negative
ANA negative
VEP Normal
SSEP Normal

is typically known as a monophasic inflammatory demyelinating disorder of the CNS usually following a viral infection. Complete recovery from ADEM is reported at 57%–89%.[1]

Certain clinical, laboratory and radiological features help distinguishing ADEM from MS in first presentation. The mean age of presentation in ADEM is 7 years and in pediatric MS is 14 years. The presence of fever, encephalopathy, and seizures favors ADEM. Optic neuritis is frequently bilateral in ADEM, whereas it is typically unilateral in MS. Cerebrospinal fluid oligoclonal bands and raised IgG index are more often found in MS.

On MRI, the lesions in ADEM often have poorly defined margins, whereas MS lesions have well-defined "plaque-like" margins. There are also differences in the lesion sites. Periaqueductal, corpus callosum, and periventricular white matter lesions are characteristic of MS. By contrast, in ADEM, the lesions tend to be in the deeper white matter with periventricular sparing (only 29%–60% of ADEM patients have periventricular lesions). When the spinal cord is involved in ADEM, the lesion is typically large, swollen, and thoracic. The spinal cord lesions in MS are typically smaller, cervical in location, and more discrete.[2] The presence of periventricular well-defined lesions, relapsing nature, and improvement with interferon favored a diagnosis of MS in our patient.

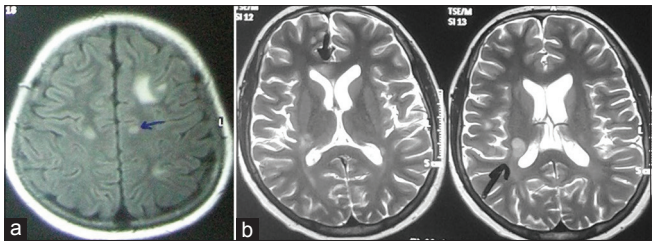

Figure 3: (a and b) Repeat magnetic resonance imaging at third visit showing new foci of demyelination in cerebral white matter (marked by arrows)

The absence of optic nerve involvement in the presence of isolated palatal involvement and irregular large asymptomatic swollen lesions on MRI in initial presentation was highly atypical in our patient.

In cases of more than one clinical episode, distinguishing between recurrent or multiphasic ADEM from MS has been a matter of controversy. Whether their pathogenesis is the same or they are a part of the same spectrum is still unclear. It is accepted that if the second episode occurs more than 6 months later, recurrent disseminated encephalomyelitis/multiphasic disseminated encephalomyelitis is less likely.[3] In the series of Schwarz *et al*.,[4] 35% of the forty patients with an initial diagnosis of ADEM developed clinically definite MS over a mean observation period of 38 months. In a retrospective study of 21 patients of Cohen *et al*.,[5] 8 developed recurrences, of which 2 were definite MS.[4] As per International Pediatric MS Study Group criteria[6] [Table 2], the first episode had characteristics of ADEM. However, on follow-up, appearance of new lesions separated in time and space favored a diagnosis of pediatric MS [Table 3]. The absence of polysymptomatic episodes without encephalopathy ruled out multiphasic ADEM [Table 4]. Our patient initially presented with monophasic ADEM-like picture; however, his course of illness eventually led to a diagnosis of pediatric MS and was started on short course of steroid and weekly interferon beta-1a.

Although steroids significantly improve acute episodes of demyelination, recurrent demyelinating disorders, such as MS and neuromyelitis optica (NMO) required long-term immunomodulation. azathioprine currently is the most favored long-term immunomodulator used in NMO. Interferon beta and glatiramer acetate are currently recommended for MS. However, azathioprine may be a suitable alternative in a resource-limited setting.[7] As per the experience of few studies and cases,[8] azathioprine was deemed suitable in our patient.

Table 2: IPMSSG criteria for diagnosis of ADEM

ADEM (Acute Disseminated Encephalo-Myelitis)
First clinical event with presumed inflammatory or demyelinating cause, with acute or subacute onset that affects multifocal areas of CNS.
The clinical presentation must be polysymptomatic and must include encephalopathy
Event should be followed by improvement, either clinically or MRI or both but there may be residual deficit
No history of a clinical episode with features of a prior demyelinating event
No other etiologies can explain the event
New or fluctuating symptoms/signs/MRI findings occurring within 3 months of the inciting ADEM event are considered part of acute event

Table 3: IPMSSG criteria for diagnosis of Pediatric MS

Pediatric MS (Any of the Following)
Two or more nonencephalopathic (e.g., not ADEM like) clinical CNS events with presumed inflammatory cause, separated by more than 30 days and involving more than one area of the CNS
One nonencephalopathic episode typical of MS associated with MRI findings consistent with 2010 Revised McDonald criteria for dissemination in space (DIS) and in which a follow-up MRI shows at least one new enhancing or nonenhancing lesion consistent with dissemination in time (DIT) MS criteria
One ADEM attack followed by a nonencephalopathic clinical event, 3 or more months after symptom onset, that is associated with new MRI lesions that fulfill 2010 Revised McDonald DIS criteria
First, single, acute event that does not meet ADEM criteria and whose MRI findings are consistent with the 2010 Revised McDonald criteria for DIS and DIT (applies only to children≥12 years old)

Table 4: IPMSSG criteria for diagnosis of Multiphasic ADEM

Multiphasic ADEM
ADEM followed by a new clinical event also meeting criteria for ADEM, but involving new anatomic areas of the CNS as confirmed by history, neurologic examination, and neuroimaging
The subsequent event must occur at least 3 months after the onset of the initial event
Presentation at subsequent event must be polysymptomatic including encephalopathy, with neurologic symptoms or signs that may differ or remained same as the initial event
The second ADEM event can involve either new or a re-emergence of prior neurologic symptoms, signs and MRI findings

CONCLUSION

Distinguishing between ADEM and MS is very complicating and challenging. In cases where atypical features of either are present, it should be labeled as clinically isolated syndrome and should be closely monitored for the course of illness. Differentiating the two has prognostic and treatment implications as the use of immunomodulators can reduce exacerbations,[3] MRI activity and, to some extent, the progression of disability in MS. Close follow-up is the key.

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Conflicts of interest

There are no conflicts of interest.

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