Case Report

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Rare Clinico-radiological Syndrome Mimicking Transient Ischemic Attack - A Case Report

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Abstract

Mild encephalitis / encephalopathy with reversible splenial syndrome (MERS) is a rare clinico-radiological entity characterisedcharacterized by mild to moderate neurological symptoms following a prodrome. Numerous infectious causes have been documented, most common being viruses and few bacteria (E. coli, Salmonella, Streptococcus & Legionella). MRI typically shows an isolated lesion in the splenium of corpus callosum, rarely in other white matter regions. Here, we discuss a 13 years old boy presenting to us like transient ischemic attack but turning out to be MERS on neuroimaging. The boy improved symptomatically in 72 hrs with spontaneous resolution within four weeks.

Introduction

Mild encephalopathy / encephalitis with reversible splenial lesion (MERS) is characterized by neuropsychiatric manifestations. MRI findings of the reversible lesions in the splenium of corpus callosum, and excellent clinical outcomes. It was initially described by Tada et al in 2004.[1] Exact incidence is not known but cases reported mostly in children than adults. Its prevalence in a radiological series is 3%.[2]

Case Report

Thirteen years old developmentally normal boy, with no significant past history, presented with a prodrome of fever and myalgia for two days followed by two episodes of unresponsiveness and altered consciousness spanning over six to eight hours. During the episode, child was unresponsive with vacant stare but not associated with tonic - clonic movements. Child complained of bifrontal headache after the period of unresponsiveness. Parents also reported subtle changes in behaviour like lethargy, slow response to oral commands and excessive sleepiness.

On examination, he was conscious, oriented and afebrile. CNS examination revealed a decrease in the power of distal muscle groups (wrists and ankles) with power of 3 / 5 along with weak finger grip. He also had high stepping gait while walking. There was no alteration in tone. His deep tendon jerks were normal. His higher mental functions, cranial nerves, cerebellum, sensory examination were all within normal limits. We did fundoscopy which was normal. There was no bowel, bladder involvement. There were no signs of meningeal irritation.

Initial investigations (Complete blood picture, urea, creatinine, liver function test, electrolytes, lipid profile, peripheral smear, INR) were unremarkable. MRI Brain contrast with spectroscopy revealed a moderately defined T1 hypointense, T2 / FLAIR intermediate signal intensity lesion involving the splenium of corpus callosum, along the midline. MR spectroscopy showed NAA peak with Cho / NAA ratio of 0.4; no lipid,

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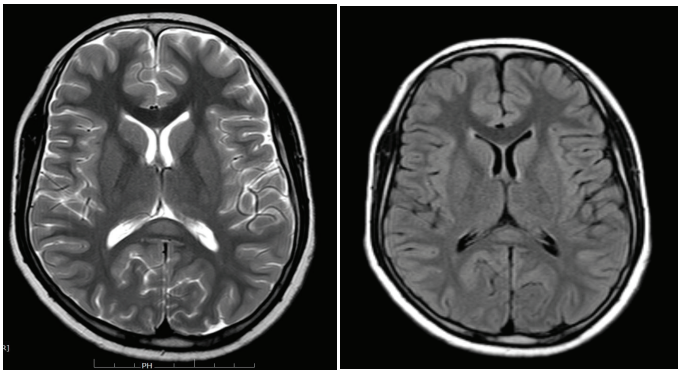
lactate peak. Rest of the brain parenchyma and angiography was normal. The findings were suggestive of a transient splenial lesion.



a b

Figure 1: a) T2 weighted axial MRI shows an ill defined, hyperintense lesion in the splenium of corpus callosum b) The lesion appears partly suppressed on FLAIR sequence.

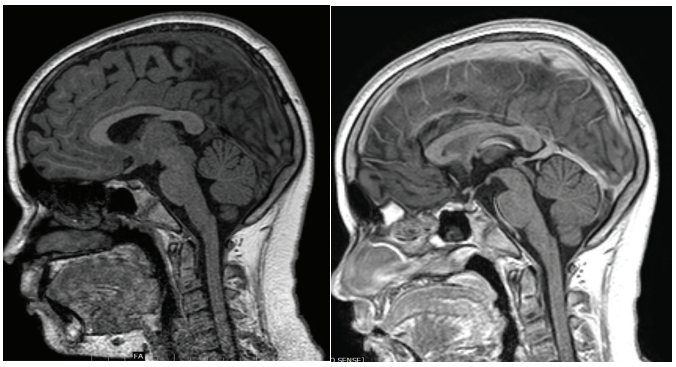


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Figure 2: a) T1 weighted axial MRI shows an ill defined, hypointense lesion in the splenium of corpus callosum b) The lesion shows non enhancement on post contrast image

In view of the clinical features and CNS involvement, we did CSF analysis, which was reported to be normal. Rapid assays for viral, bacterial agents were not done due to unavailability of the facility at our centre. There were no further episodes and patient started improving symptomatically with supportive management over 72 hours. Follow-up CNS examination after four weeks was normal.

Discussion

Mild encephalitis / encephalopathy with reversible splenial lesions or MERS can present with a wide range of signs and symptoms. It is usually preceded by a prodrome of fever, vomiting, diarrhoea and cough, one to three days before the onset of symptoms.[3,4] The most common neurological symptom is delirious behaviour, followed by disturbance of consciousness, and seizures. The associated infectious agents include influenza, Escherechia coli, Streptococcus pneumonia, Legionella, Mycoplasmapneumoniae, mumps, rotavirus, measles, varicella and CMV.[5-9] Sometimes, it may be seen in children who have been rapidly withdrawn from AED or who have received toxic levels of AED.[1] There has been one case report with Kawasaki disease being the cause of MERS.[10]

MERS is classified into type 1 and type 2 based on MRI. Type 1 is with only corpus callosal changes and type 2 involving other areas of white matter along with corpus callosum.[8] The exact pathophysiological mechanism of MERS is not yet known. Two main mechanisms were proposed – intramyelinic oedema and inflammatory infiltrates. However, both these mechanisms fail to explain why splenium of corpus callosum is the site of involvement.[1,7] Repeat MRI typically at four weeks shows complete resolution of the lesion. This resolution of the lesion is the hallmark of this phenomenon.

This child showed typical prodromal symptoms with altered consciousness and distal muscle weakness. MRI brain contrast showed Focal T2 / FLAIR hyperintense lesion with restricted diffusion along the midline in splenium of corpus callosum and the child recovered with symptomatic management. Hence, our case fulfils the diagnostic criteria proposed by Hoshino et al.[3] This child fits into Type 1 MERS. The patient did not have any recurrence of symptoms on follow-up four weeks later. Repeat MRI was hence not done due to financial constraint. Most cases of children with MERS improve with minimal or no intervention at all. In severe cases, injection methyl prednisolone, intravenous immunoglobulin and anti-epileptic drugs have been given along with empirical antibiotics and antivirals as treatment of cause.[4]

Conclusions

Although uncommon, any child presenting with delirium, MERS should also be considered as a possible etiology. A delirious child with MRI brain showing an isolated lesion in the splenium of corpus callosum, along with a prodromal infection and typical neurological features clinches the diagnosis of MERS. Outcome is excellent with majority of patients having a full recovery within four weeks with no major intervention. Hence, early recognition of this rare entity can avoid unnecessary investigations / treatment and provides reassurance about good outcome.

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