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Letter to Editor

Fatal Hashimoto encephalopathy presenting with acute fulminant cerebral edema in a child

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Dear Editor,

Hashimoto encephalopathy (HE) is a rare but controversial entity which is under-diagnosed in children.

A 14-year-old girl presented to us with 1 day history of headache, vomiting, and rapidly evolving encephalopathy. She was afebrile, unconscious and had persistent decorticate posturing. Examination showed sluggishly reacting pupils, papilledema, hyperreflexia, and up going plantars. Child was ventilated and started on antibiotics, acyclovir, and hypertonic saline infusion. Neuroimaging showed diffuse cerebral edema [Figure 1]. Continuous electroencephalogram revealed diffuse slowing without electrographic seizures. Measures for raised intracranial pressure were instituted. Work-up for common infections, metabolic disorders, and autoimmune encephalitis was normal. Lumbar puncture was not done due to elevated intracranial pressure. Serum anti-thyroperoxidase (anti-TPO) antibodies were significantly elevated: >1300 IU/ml (Normal: <5.6 IU/ml).

A diagnosis of HE was made. High dose intravenous methylprednisolone was initiated. In spite of aggressive early medical management, there was neurological deterioration over next 72 h leading to brain death. Neuroimaging repeated after 48 h revealed trans-tentorial herniation and cerebellar tonsillar descent.

Such a fulminant presentation of HE has not been reported before in children. HE presenting as acute encephalopathy in children, has been reported infrequently.[1] Mild cerebral edema has been reported usually following status epilepticus.[2] Death is rare in adult HE, after uncontrolled status epilepticus.[3]

Our patient fulfills all the criteria of HE (raised serum anti-TPO antibodies, encephalopathy, non-specific changes on EEG and MRI with exclusion of known autoimmune disease) except poor responsiveness to steroids. Studies have shown that only around 1/3rd of patients fulfilling other criteria of HE, respond to steroids.[4] This suggests that current diagnostic criteria for HE need revision so that severe cases, like ours, are included. Mattozzi *et al.*[4] suggested that anti-TPO related disorders should include all patients with raised anti-TPO antibodies and neurological involvement, irrespective of steroid responsiveness.

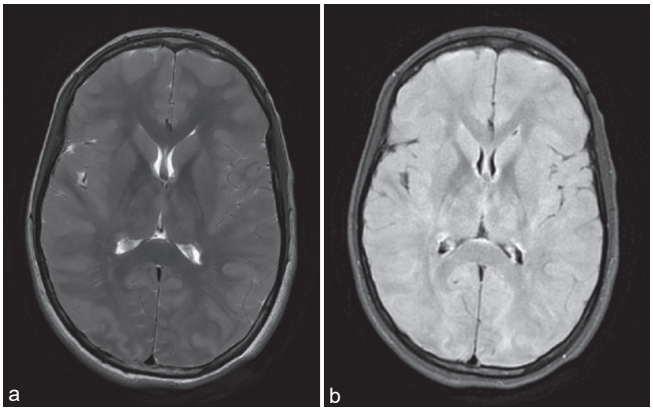

Figure 1: T2 (a) and fluid-attenuated inversion recovery (FLAIR) (b) axial magnetic resonance imaging images of the patient showing diffuse cerebral edema.

Acute fulminant cerebral edema has been described as a phenotype of encephalitis in children.[5,6] These patients had evidence of minimal CNS inflammation, poor response to immunomodulation, and poor outcomes. Infectious etiology could not be identified in most. The clinical and CSF features in these patients raise the possibility of autoimmune process. We propose that a proportion of these patients might have had HE as the underlying etiology.

HE/anti-TPO antibody related autoimmune disorder should be considered as a differential diagnosis in any patient with acute diffuse cerebral edema in the setting of unexplained encephalopathy. The optimal course of treatment in these patients needs evaluation.

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Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Conflicts of interest

There are no conflicts of interest.

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