Hashimoto’s Encephalopathy in an Adolescent Boy

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[Received January 24, 2006; Accepted February 19, 2007]

ABSTRACT

Hashimoto’s encephalopathy is an under-recognized cause of acute encephalopathy both in children and adults. We hereby describe a 12.5 yr old boy with this rare disorder that presented with an acute onset of episodic psychosis with hallucinations along with seizures and had elevated antithyroid antibodies. Symptoms improved with thyroxine replacement and anticonvulsants and EEG normalized 3 mth into follow up. Hashimoto’s encephalopathy should be considered in patients with unexplained encephalopathy and seizures, as prompt recognition and management can lead to an excellent outcome.

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Key words : Pediatric Hashimoto’s encephalopathy; Psychosis; Hypothyroidism; Autoimmune thyroiditis

There are few conditions in clinical neurology as elusive and protean as Hashimoto’s encephalopathy (HE). Although neurological complications of hypothyroidism have been well recognized, HE is a rare disorder in children.1 This is a condition presenting with encephalopathy associated with autoimmune thyroiditis. Adequate information is not available about the frequency of HE in children despite its first description as a distinct clinical condition in adults in 1966.2,3 Since then approximately 90 cases of this disorder has been reported, mostly in adult females.4 The authors hereby report a boy who presented with acute onset psychosis and on investigation was found to have high antimicrosomal antibody titles. In view of the variable clinical presentation, it is underdiagnosed in pediatric patients and a high degree of suspicion is necessary for proper diagnosis.5

CASE REPORT

N, a 12.5 yr old male child presented with history of abnormal behaviour since 2 days preceded by vomiting and low grade fever for 1 day. The child had uncharacteristic behaviour in the form of alternating states of confusion with hyperkinetic – anxious behaviour and decreased affect. There was no history of thyroid disorder or other relevant medical or psychiatric history in the past or in the family or exposure to toxins. On examination the patient had a dull look and rough skin without goiter. Pulse rate was 64/min and blood pressure was normal. Child was cooperative and displayed no insight into the irregularity of his behaviour. At times he was restless noticed to be smiling or talking to self, gesturing in the air. He would claim that he would see ghosts, shout loudly on and off without provocation. All these symptoms would occur episodically, lasting for a few hours and the patient would be amnesic for his abnormal behaviour thereafter. Sleep, appetite and self care were decreased. He used to have episodes of normal behaviour in between for 2-3 hr. Other neurologic examination was within normal limits. While recording
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EEG he had one episode of generalized tonic clonic seizure which lasted for 5 minutes followed by postictal drowsiness for 1 hr. Hemoglobin was 11g/dL with peripheral blood picture showing mild hypocromic, microcytic anemia. Erythrocyte sedimentation rate was 34 mm in 1st hr. Cerebrospinal fluid (CSF) analysis revealed normal cell count, protein and glucose. Bacterial cultures for blood and CSF were negative. Serum and CSF examinations for Herpes and Japanese encephalitis serology were negative. Liver function tests- (Serum bilirubin 1mg/dl, SGOT/PT-14/20 IU, Alkaline phosphate 13 KAU), serum electrolytes and renal function tests (Blood urea 34mg/dl and creatinine 0.6 mg/dl) were normal. Serum calcium and phosphorus were 9.4 mg/dl and 4mg/dl respectively. Random blood sugar level was 104 mg%. Serum ceruloplasmin was 22 mg% and urine for porphobilinogen was negative on 3 occasions. Serum antinuclear antibodies, rheumatoid factor, ds DNA, antícardiolipin antibodies and lupus anticoagulant were negative. Magnetic resonance imaging of the brain was normal. Intercital EEG showed bilateral temporal sharp wave discharges (left more than right) with slow back ground activity while the ictal EEG showed generalized spikes. Thyroid function tests revealed increased serum TSH [80 mIU/mL (N-0.3-5.0 mIU/mL)], decreased freeT4 [0.5 ng/dL (N-0.8-1.8)], and elevated thyroid microsomal antibody titre [1600 IU/mL, (normal <200)]. Technetium pertechnate scan of thyroid showed normal uptake. He was started on thyroxine (10µg/Kg/d) and carbamazepine(15mg/Kg/d) with which he had gradual recovery over the next week in the form of improved interaction and his behaviour completely normalized within 6 wk. There was normalization of serum TSH and free T4 which were 4mIU/ml and 1.2ng/dl respectively, while the microsomal antibodies were still elevated but had decreased (800IU/ml). In the index case as there was significant clinical improvement and normalization of EEG with thyroxine alone, steroids were withheld.

DISCUSSION

HE is a syndrome of cerebral symptoms seen in patients with serologic evidence of autoimmune thyroid disease. Antibody titres do not correlate with the clinical presentation. The prevalence of HE in children is unknown although the estimated prevalence in adults is 2.1 /100000. Thyroid status varies greatly and patients may be either euthyroid, hypothyroid and rarely even hyperthyroid.

The clinical features of HE are heterogenous, and a high degree of suspicion is necessary for diagnosis. Two types of presentation are observed in adults – a) a vasculitic type, with repetitive stroke like episodes, such as hemiparesis, aphasia, ataxia, and only mild cognitive impairment and b) a diffuse progressive type, with insidious onset of dementia, seizures, psychotic episodes, or altered consciousness, a significant overlap may occur. Myoclonus, tremors, and seizures may occur in both types. The present case had the second type of presentation. Behavioral presentation with psychosis has been rarely described in pediatric cases. Myxedematous psychosis was unlikely as a cause of symptomatology as patient had acute onset of positive symptomatology of psychosis and seizures with mild hypothyroidism and high antimicrosomal antibody titre. This feature has also been stressed by earlier authors. Seizures are seen in 66% of patients, especially in children. Generalized tonic clonic followed by complex partial seizures, are the most common types of seizures seen. Focal motor seizures and recurrent status epilepticus have also been described.

The most common EEG abnormality described is the diffuse slowing of the background rhythm as was seen in the index case who also had independent bitemporal-onset ictal discharges, right more than left. The temporal lobe may be particularly affected in some patients with HE. The MRI studies are negative in almost 50% of cases, but mild cerebral atrophy, infarction, focal mesiotemporal, basal ganglia and white matter abnormalities have been detected. Cerebral isotope studies or brain scans manifest abnormalities consisting of global, focal, or symmetric multifocal areas of decreased perfusion.

The diagnostic criteria of these cases has been stated in a recent editorial by Doherty. The clinically probable cases can be classified based on findings of a conducive clinical history; positive antibody titres, transient sharp or triphasic activity and slowing on an EEG with or without focal T2 abnormalities on MRI. The gold standard for this diagnosis will be by brain biopsy proven vasculopathic lesion including demyelination with or without inflammatory markers. The index case qualifies all the above features except the biopsy which was not undertaken in view of the normal MRI and rapid clinical response to therapy.

Several pathogenic mechanisms have been suggested to explain the encephalopathy like cerebral edema, autoimmune cerebral vasculitis, existence of an unidentified common antigen between the brain and the thyroid gland or a central excess of thyroid releasing hormone.

Although recognized treatment studies are absent, most reports have shown that glucocorticoids are effective. Out of 12 patients in a series of HE who were treated with thyroxine alone 8 were found to have improved similar to our case. Improvement of the EEG and neuropsychologic testing are considered as tools for monitoring the response and the length of therapy. The prognosis of HE when treated is generally good however some patients may experience relapses. Adolescents with
this condition may experience residual cognitive deficits even when early recognition and prompt treatment are instituted which was not seen in the index case.11

This case underscores the importance of thyroid screening in children who present with acute neuropsychiatric manifestations. It is a potentially treatable encephalopathy and hence pediatricians who may be unacquainted to this entity must become aware so that prompt therapy may be instituted for a good outcome.

REFERENCES