A case of steroid-responsive encephalopathy associated with autoimmune thyroiditis (SREAT) in a girl with newly diagnosed Hashimoto thyroiditis

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Background:
SREAT is a rare entity in children, with about 60 cases described to date. It is characterised by acute encephalopathy, elevated anti-thyroid antibodies and exclusion of other causes of encephalopathy like infection, tumour, toxic or metabolic diseases. Typical presentations in children include behaviour changes, psychosis and seizures. The role of anti-thyroid antibodies in the pathogenesis is not clearly understood, and the titre level does not predict severity of disease and relapse. The treatment of choice is intravenous methylprednisolone followed by oral prednisolone.

Case report:
A previously well 11-year-old girl presented to the clinic with short stature and increasing weight gain. She had a diffusely enlarged firm and non-tender goitre. Her free thyroxine was <3.2 pmol/L (8 - 16) and TSH was >450 mIU/L (0.4 - 4). Anti-thyroid peroxidase (TPO) antibodies were elevated at 371 IU/mL (<50). She was treated with thyroxine 50mcg daily. Two weeks later, she presented to the Children's Emergency with behavioural change of 3 days' duration. She was agitated and refused to eat and speak. During hospitalisation, MRI brain and lumbar puncture were normal. An EEG showed generalized cerebral dysfunction. Electrolytes and blood sugar were normal. TSH was 112.71 mIU/ml, free thyroxine 5.7 pmol/L, anti-TPO 145 IU/mL and anti-thyroglobulin 491 IU/mL (<115). Erythrocyte sedimentation rate was elevated at 63 mm/hr (2 - 20). Autoimmune work-up including antinuclear antibody, anti-double stranded DNA antibody, complements and ENA (extractable nuclear antigens) antibodies were normal. Autoimmune encephalopathy panel of the cerebrospinal fluid was negative. In view of her acute encephalopathy, elevated anti-thyroid antibodies and the absence of other causes of encephalopathy, she was diagnosed to have SREAT and treated with 5 days of intravenous methylprednisolone followed by a tapering dose of oral prednisolone. She was also treated empirically with Levetiracetam and 2 days of antibiotics and Acyclovir. On the third day of treatment with methylprednisolone, her behaviour normalised, although she could not recall what the events in the past week. Since recovery, there have been no residual neurological deficits.

Conclusion:
The rarity of SREAT may lead to delayed or under-diagnosis. Clinicians should consider SREAT in children with acute encephalopathy and no other causes are found. Failure to treat may lead to coma, death or permanent neurological deficit.

References
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