An unusual case of autoimmune polyglandular syndrome (APS) II presenting with neurological symptoms and hypoglycemia due to adrenal insufficiency

Case
A 24-year-old, male Caucasian, with alopecia at age 8, and DM1 at age 12, walked into an insulin pump report right hemiparesis and hemipesthesia occurring upon waking up which resolved after eating. After the third episode, he turned off his insulin pump and went to the emergency department. On exam, he was found to have new neurological symptoms. He was started on subcutaneous insulin. A 0.25mg cosyntropin test was performed and blood cortisol was 4 µg/dL at 0, 30 and 60 minutes; baseline ACTH was 454 pg/ml. The test was repeated in the morning: cortisol was 6, 5, and 5 µg/dL at 0, 30 and 60 minutes respectively; and baseline ACTH of 2110 pg/ml. Thyroid stimulating hormone (TSH) and free thyroxine were normal. However, anti-TPO antibody was 115 IU/ml (nl 0 - 34.9 IU/ml). He was started on corticosteroid and mineralocorticoid replacement therapy, and was discharged with multiple subcutaneous insulin injections.

Discussion
Thompson Addison first described a syndrome including weakness, fatigue, anorexia, abdominal pain, orthostatic hypotension, salt craving, and hyperpigmentation. It has been reportedly caused by hereditary disorders, drugs, meningitis, TB adrenalitis, critical illness, and liver disease. In the Western world 80% of cases are caused by the autoimmune adrenal syndrome (Addison’s disease). The syndrome usually ensues within a few years of onset of the underlying disorder, and has a variable presentation, and there are three types: Type I consists of 2 out of 3 of Addison’s disease, hypothyroidism, and autoimmune adrenal insufficiency. Type II consists of adrenal insufficiency and either thyroid disease or other autoimmune disorders other than Addison’s. Type III consists of autoimmune thyroiditis and other autoimmune disorders other than Addison’s. Our patient was diagnosed with APS-1 and had previously been diagnosed with alopecia and DM1. He also had elevated anti-TPO antibodies, indicating Hashimoto’s thyroiditis. Although currently euthyroid, he should be monitored regularly for the development of hypothyroidism. Our patient’s presentation was unusual in that he presented with hemiparesis in the setting of hypoglycemia as a manifestation of hypocortisolism.

References
Kleinberg LO, Haddad LJ, Pitlick SC, et al. Neurologic deficits, such as hemiparesis, can be a presenting manifestation of low levels of glucose. In patients with autoimmune DM1, at stable insulin dose, and no recent lifestyle changes, other etiologies for hypoglycemia should be investigated, including adrenal insufficiency (Addison’s disease).