

LETTERS

A case of autoimmune polyglandular syndrome type II presenting with adrenal crisis

Dear Editor,

The autoimmune polyglandular syndromes (APS) form different clusters of autoimmune disorders and are rare endocrinopathies characterized by the coexistence of at least two glandular autoimmune mediated diseases¹. Autoimmune polyglandular syndrome type II is defined by the occurrence of Addison's disease with thyroid autoimmune disease and/or type I diabetes mellitus. Clinically overt syndrome is considered only the tip of the iceberg, since latent forms are much more frequent². Organ-specific autoantibody screening in patients with monoglandular autoimmune endocrinopathies undoubtedly facilitates the identification of those at risk of developing a future APS. Early identification and treatment of another autoimmune endocrine disease may be critical and even life-saving³. Herein, we report a case of adrenal crisis which was precipitated by replacement with levothyroxine without simultaneously adrenal steroid replacement in a patient with APS type II.

A 74-years-old woman came to the hospital due to fatigue, nausea, vomiting and skin hyperpigmentation that had begun two months before her presentation. On clinical examination, the patient had hyperpigmentation throughout the skin, large dark brown areas on the lips and the buccal mucosa and her blood pressure was 90/60 mm Hg. She had been diagnosed with hypothyroidism since five days and had been receiving levothyroxine for two days before admission. Her past medical history as well as her family history was otherwise unremarkable.

Laboratory tests revealed hyponatremia (serum sodium: 130 mEq/L) and hyperkalemia (serum potassium: 5.56 mEq/L). Morning blood cortisol level was <0.2 µg/dL (normal range: 5-25 µg/mL) and ACTH levels were 1248.0 pg/mL (normal range: 9-52 pg/mL). Addison's disease was confirmed and antibodies directed against 21-hydroxylase (OH) were positive, a finding suggestive of autoimmune adrenalitis. TSH levels were 6.62 µU/L (normal range: 0.3-4 U/mL), T3: 204.5 ng/dL (normal range: 60-200 ng/dL), FT4: 1.02 ng/dL (normal range: 0.9-1.7 ng/dL), thyroid peroxidase antibodies (anti-TPO): 314.6 IU/mL (normal range: <34 IU/mL), thyroglobulin (anti-Tg): 613.6 IU/mL (normal range: <115 IU/mL). Autoimmune adrenalitis together with autoimmune thyroid disease was diagnosed, a combination known as autoimmune polyglandular syndrome type II or Schmidt's syndrome⁴. This syndrome has a peak incidence at ages 20-60 years, mostly in the third or fourth decade, and it is common for multiple generations to be affected by one or more component diseases. There is familial clustering and family members of patients are often affected.

Other causes of the adrenal crisis such as abrupt cessation of corticosteroids or an ongoing infection were excluded as she was afebrile with no symptoms, signs or laboratory findings suggestive of infection. It seems likely that the prescription of levothyroxine prior to adrenal steroid hormone replacement in this patient with concurrent Addison's disease precipitated an adrenal crisis. Replacement of levothyroxin increases the cortisol turnover rate in the liver, and this may compromise a failing adrenal gland². There have been rare reports of APS II in children and adults older than 60 years⁵. Our patient did not belong to the preferred-age group and moreover did not mention any family history.

In patients with hypothyroidism and symptoms or signs suggestive of Addison's disease, the possibility of autoimmune polyglandular syndrome type II should be considered. In this case, exclusion of concurrent Addison's disease is necessary before the initiation of levothyroxine replacement as prescription of levothyroxine prior to adrenal steroid hormone replacement may be hazardous.

References

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Conflict of Interest Statement

The authors declare that there is no conflict of interest regarding this manuscript.

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