

Addisonian Crisis in a Patient with Schmidt's Syndrome

Introduction

Schmidt's syndrome, also known as polyglandular syndrome type II, is a rare disease with potential life-threatening consequences. It is characterized by multiple immune endocrinopathies, including Addison's disease with thyroid disease and/or type I diabetes [1-3]. We describe a case of Addisonian crisis in a patient with known Addison's disease. Interestingly, she also has type I diabetes mellitus and Graves' disease but was never previously diagnosed with Schmidt's syndrome.

Case

A 67-year old female with Addison's disease, diabetes mellitus type 1, and I-131 induced hypothyroidism secondary to Grave's disease presented with a 1-day history of nausea, vomiting, dizziness, and confusion. She was found on the floor in her home after an unknown duration with blood sugar of 22. She was given 1 ampule of dextrose 50 en route to the hospital. Physical examination was remarkable for bradycardia (49), hypotension (93/30), buccal mucosa hyperpigmentation, and unsteady gait. Her labs were as follows: Na 136 (137-154 mmol/L), K 4.1 (3.6-5.0 mmol/L), BUN 21 (7-20 mg/dL), and sCr 1.35 (0.55-1.05 mg/dL). Other labs included: Serum cortisol <0.16 (6-23 mcg/dL), creatinine kinase 1358 (30-135 U/L), procalcitonin 0.34, TSH 0.25 (0.46-4.7 uIU/mL), free T4 4.21 (1.65-4.07), T3 uptake 36.6 (23.5-40.6%), and thyroglobulin 0.2 (1.5-38.5 ng/mL). Urinalysis was dipstick negative for hematuria with 1-3 RBC casts. Blood cultures remained negative at 72 hours. CT head was unremarkable. CT abdomen/pelvis showed atrophic adrenals. Patient was treated with glucocorticoid (hydrocortisone), mineralocorticoid (fludrocortisone), and fluid support. Levothyroxine dose was decreased due to overmedication. Patient's hemodynamics improved, and her confusion resolved with treatment.

Discussion

Schmidt's syndrome is a compendium of polyendocrine autoimmune diseases (Addison's disease, autoimmune thyroiditis/Graves' disease, type I diabetes) and can be associated with other non-endocrine autoimmune disorders, such as

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myasthenia gravis, Sjogren's syndrome, and rheumatoid arthritis [3]. It is autosomal dominant with variable penetrance and usually affects middle-aged females (female-to-male ratio 3:1) [1]. The diagnosis of this condition can be challenging due to its rarity (1.4 - 4.5 cases/100,000 population), atypical presentation, and variance in clinical symptoms depending on the type and severity of gland involved. Thus, a high index of suspicion is warranted for early diagnosis and appropriate hormonal therapy [1].

References

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