Adrenal

ADRENAL CASE REPORTS

Pituitary Macroadenoma Masked by Iatrogenic Adrenal Insufficiency: A Diagnostic Blind Spot

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Background: Exogenous steroid use is the most common cause of central adrenal insufficiency. Depending on the duration and strength used, it may take months to years for the hypothalamic-pituitary-adrenal (HPA) axis to recover after the steroid is stopped. We report a case of iatrogenic hypoadrenalism with persistent suppression of the HPA axis for 13 years, discovered later to be due to a second pathology.

Case: A 48 year old lady presented in 2005 with weight gain of 20 kg over 1 year and florid Cushingoid features. 9AM cortisol was undetectable (<12 nmol/L). History taking revealed use of oral dexamethasone at various dosages over the past 9 years for her knee pains. A diagnosis of iatrogenic adrenal insufficiency was made. She was started on hydrocortisone replacement, and was advised to stop the over-the-counter steroids. By 2011 her short Synacthen test (SST) showed much improved functioning of the HPA axis (cortisol 182 (0 min) -> 329 (30 min) -> 408 nmol/L (60 min) [N peak>500 nmol/L]), and she was back to her usual body weight. However, subsequent monitoring revealed declining trend of 9AM cortisol from 135 nmol/L (2013) -> 99 nmol/L (2014) -> 57 nmol/L (2015) -> 64 nmol/L (2016) -> 18 nmol/L (2017) [N 166–507 nmol/L]. Hydrocortisone compliance and abstinence from exogenous steroids was confirmed with the patient. The ongoing hypofunction of the HPA axis was continually attributed by multiple physicians to her history of prolonged use of dexamethasone. In 2018, at the age of 60, the lady presented with new onset headaches, blurred vision, and bitemporal hemianopia for 3 months. MRI showed a 1.8x1.8x3.5 cm (WxAPxH) pituitary mass with suprasellar extension compressing the optic chiasm. Blood tests revealed panhypopituitarism: SST cortisol 17 -> 59 -> 49 nmol/L, ACTH 2.7 pmol/L [N <10.1 pmol/L]; fT4 9.5 pmol/L [N 12–22 pmol/L], TSH 0.97 mIU/L [N 0.27–4.2 mIU/L]; LH <0.1 IU/L, FSH 0.52 IU/L (menopause at age of 48); IGF-1 27 μg/L [N 41–279 μg/L]; prolactin 17 mIU/L [N 102–496 mIU/L]. After partial excision of the mass her vision improved, but remained dependent on hydrocortisone and thyroxine supplements. The lesion was pathologically proven to be a pituitary macroadenoma.

Discussion: This case presents the uncommon course of a patient who had almost recovered from iatrogenic hypoadrenalism, only to lapse back into worsened central adrenal insufficiency, as part of panhypopituitarism related to an undiagnosed pituitary mass. In retrospect, the unusually protracted state of hypocortisolemia and the atypical waxing and waning HPA axis should have alerted one to consider alternative etiologies at work. As LH-FSH and GH deficiencies commonly develop before ACTH and TSH deficiencies in most pituitary macroadenomas, a lower threshold for testing the other anterior pituitary hormones followed by imaging of the pituitary could have picked up the tumor earlier in this patient.

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Polyglandular Autoimmune Syndrome Type II in a Patient Presenting With Hypotension After Acute Illness

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Introduction: Polyglandular Autoimmune Syndrome Type II (PAS-2) is a rare disorder characterized by two or more endocrine diseases (primary adrenal insufficiency, autoimmune thyroid disease, type 1 diabetes). Of these, the most common is autoimmune thyropathy, followed by Type 1 Diabetes (1). These endocrinopathies rarely have concurrent onset. This case reports a 22-year-old male, recently recovered from severe pneumonia, who presented to the Emergency Department in acute adrenal crisis and was diagnosed with PAS-2.

Case: A 22-year-old male with past medical history of celiac disease presented with abdominal discomfort, nausea, vomiting, fatigue and dizziness for 1–2 weeks. Review of systems included a 20-pound weight loss over several months. Recent history included a hospitalization at another facility three weeks prior for pneumonia and septic shock requiring admission to the ICU and vasopressor treatment. He was not discharged on any medications. He was afebrile with heart rate of 105/min, blood pressure 78/48 mm Hg and BMI of 17.2. Physical exam revealed dry mucous membranes and mild diffuse abdominal tenderness. Skin was warm and dry without hyperpigmentation. Laboratory values included sodium 123 (135 - 146 mmol/L), potassium 6.8 (3.3 - 4.8 mmol/L), glucose 47 (70 - 100 mg/dL), TSH 37.67 (0.3 - 5.00 uIU/mL), and FT4 0.7 (0.7 - 1.7 ng/dL). He was started on fluids and intravenous hydrocortisone. Cortisol and ACTH levels drawn prior to the initiation of steroids resulted at 0.6 (6–20 mcg/dl) and 977 (9 - 46 pg/mL) respectively. Additional labs included: aldosterone < 1 ng/dL, 21 hydroxylase antibody positive, TPO antibody > 1000 (0 - 100 Units) and GAD-65 antibody > 47 IU/mL (<5 IU/mL). Levothyroxine was initiated after hydrocortisone. Blood glucose was elevated during hospitalization, peaking at 227 mg/dL. He was discharged on prednisone, fludrocortisone and levothyroxine. At 2 week follow up, he reported overall improvement in health and was pleased with a weight gain of 12 lbs. Blood glucose remained mildly elevated (123 - 143 mg/dL).

Conclusion: The patient had pneumonia and septic shock septic at an outside hospital three weeks prior to presentation. There was no record of steroid administration or suspicion of adrenal insufficiency. We postulate that his severe illness contributed to significant depletion of his adrenal reserve, and therefore he presented to our facility a short time later in overt adrenal crisis. Adrenal crisis is unusual to be the first presentation of PAS-2. It is important to have a high index of suspicion for adrenal insufficiency and PAS-2 in patients presenting with severe illness or hypotension who have known autoimmune disorders.

Reference: 1. Kahaly, G.J., Frommer, L. Polyglandular autoimmune syndromes. J Endocrinol Invest. 2018; 41: 91–98.