A Young Man In Shock: Expect The Unexpected
B Vahid, A Esmaili

Citation

Abstract
We report a case of primary adrenal insufficiency that presented with shock. The patient was a young man with no skin hyperpigmentation. Primary adrenal insufficiency should be considered in patients with shock regardless of presence of skin hyperpigmentation. Adrenal insufficiency was suspected after findings of hyponatremia, hyperkalemia, and hypoglycemia on blood chemistry. Corticosteroid and mineralocorticoid replacement was started and the patient recovered fully.

CASE REPORT
A 26 year-old man presented to the emergency department with fever, nausea, vomiting, and diarrhea for 2 days. Ten days before presentation, he was treated with oral prednisone for swelling of lower and upper lips. Swelling of lips was presumed to be due to allergic reaction to an unidentified allergen. At presentation, the patient looked ill and in distress. Physical examination revealed temperature of 100.8°F, blood pressure of 65/40 mmHg, heart rate of 120 beats/min, and respiratory rate of 26 breaths/min. Small non-tender lymph nodes were palpable in posterior cervical region and bilateral inguinal areas. Jugular veins were not distended. Chest and cardiac exam were unremarkable.

Abdominal exam revealed a soft and non-tender abdomen. Spleen and liver were not palpable. Skin pigmentation was not present. Despite 5 liters of normal saline infusion over 2 hours, hypotension persisted. An echocardiogram revealed normal left ventricular function and no evidence of pericardial effusion. Chest radiograph was normal. Blood chemistry showed sodium of 125 mmol/L, potassium of 6.9 mmol/L, chloride of 92 mmol/L, bicarbonate of 18 mmol/L, glucose of 61 mg/dl, and creatinine 2.7 mg/dl. White blood cell was 15.3X10^3 /L and hemoglobin was 15.9 g/dl. Based on presentation and blood chemistry, adrenal crisis was suspected. Baseline plasma cortisol level was 1.7µg/dl. Sixty minutes after the administration of 25 units (0.25 mg) of cosyntropin intravenously, plasma cortisol level was 2.4µg/dl. Hydrocortisone 100mg intravenously every 6 hours was started that resulted in normalization of electrolytes, creatinine, and blood pressure. Two sets of Blood cultures and a urine culture were negative. Human immunodeficiency virus antibody was negative. Computed tomography (CT) scan of chest was normal. CT scan of abdomen was normal except for bilaterally atrophic adrenal glands (Figure1).

Figure 1
Figure 1: Abdominal CT scan showing bilateral atrophic adrenal glands (circles).

Serum Adrenocorticotropic Hormone (ACTH) level was 439 pg/ml (normal less than 70 pg/ml), serum aldosterone level was not detectable, and serum direct renin level was elevated (608 µU/ml). These findings are compatible with primary adrenal insufficiency. Anti-adrenal antibody was positive with elevated titers (1:20). Adrenal crisis secondary to autoimmune adrenalitis was diagnosed. Patient was discharged home on oral prednisone and fludrocortisone acetate.

DISCUSSION
Destruction of adrenal gland tissue results in primary adrenal insufficiency. Secondary adrenal insufficiency is due to malfunction of hypothalamic-pituitary region. Autoimmune adrenalitis accounts for 80-90% of cases of primary adrenal insufficiency in developed countries (1). Causes of adrenal insufficiency are summarized in table1(1,2,3,4).
Primary adrenal insufficiency has a prevalence of 93-140 per million and an incidence of 4.7-6.2 per million in white populations. Patients with acute adrenal insufficiency typically present with hypotension, abdominal pain, vomiting, and fever. Skin hyperpigmentation is seen in patients with primary adrenal insufficiency. Although hyponatremia can be seen in both primary and secondary adrenal insufficiency, hyperkalemia and raised creatinine are seen only in primary adrenal insufficiency (1). In this case, we believe the patient had secondary adrenal insufficiency and primary adrenal insufficiency due to recent prednisone therapy. This may explain why the patient had no skin hyperpigmentation. Unexpected adrenal crisis has been described in young patients that presented with shock. Physicians should have high index of suspicion for diagnosis of adrenal crisis. Absence of skin hyperpigmentation is not enough to exclude the diagnosis.

CORRESPONDENCE TO
Bobbak Vahid, MD Thomas Jefferson University 1015 Chestnut Street, Suite M-100 Philadelphia, PA 19107 Tel: 215 955-6591 Fax: 215 955-0830 Bobbak.vahid@mail.tju.edu

References

### Table 1: Causes of adrenal insufficiency

| Primary adrenal insufficiency | Autoimmune adrenalitis  
| | • Isolated autoimmune adrenalitis  
| | • Autoimmune adrenalitis as part of APS  
| Infectious adrenalitis |  
| | • AIDS  
| | • CMV infection  
| | • Cryptococcosis  
| | • Histoplasmosis  
| | • Coccioidiomycosis  
| Bilateral adrenal hemorrhage |  
| | • Meningococcal sepsis  
| | • Antiphospholipid syndrome  
| Adrenal infiltration |  
| | • Lymphoma  
| | • Sarcoidosis  
| | • Metastatic disease  
| | • Amyloidosis  
| | • Hemochromatosis  
| Secondary adrenal insufficiency | Corticosteroid treatment  
| | Autoimmune lymphocytic hypophysitis  
| | Tumor of hypothalamic-pituitary region  

Figure 2
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Author Information

Bobbak Vahid, M.D.
Department of Pulmonary and Critical Care Medicine, Thomas Jefferson University

Ali Esmaili, M.D.
Department of Medicine, Thomas Jefferson University