When Vision Blurs and Pressure Falls: The Adrenal Crisis Conundrum[1]

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Introduction

Adrenal insufficiency is a potentially dangerous condition that can be classified as primary, secondary, or tertiary. Secondary adrenal insufficiency (SAI), characterized by decreased adrenocorticotropic hormone (ACTH) levels, is estimated to affect between 150 and 280 people per million. Patients who experience adrenal crisis due to inadequate production of cortisol have a significant mortality risk.

Case:

65-year-old male with a history of chronic atrial fibrillation, hypertension, and right heart failure presented with multiple episodes of diarrhea, amaurosis fugax, dizziness, and lightheadedness for about 1 week. Approximately a month prior to presentation, he was admitted to the ICU for septic shock related to COVID-19. On presentation, he was hypotensive requiring pressors but without evidence of sepsis. He was found to have adrenal crisis in the setting of newly diagnosed adrenal insufficiency. Low cortisol and an inappropriately normal response to ACTH-stimulation test proved adrenal insufficiency, and subsequent workup confirmed secondary adrenal insufficiency. He was treated with hydrocortisone with clinical improvement then started on a prolonged steroid taper. He never needed fludrocortisone. For SAI, MRI did not show pituitary adenoma though it was not a dedicated pituitary study. TTE showed biatrial enlargement and a severely enlarged right ventricle. There was concern for restrictive cardiomyopathy. A PYP scan in 2021 was equivocal. Endomyocardial biopsy resulted with minimal increase in interstitial connective tissue but negative for amyloid and other infiltrative processes.

Discussion:

This case featured an atypical presentation of SAI, particularly in reference to the patient's complaints of vision loss. Typical symptoms include fatigue, muscle weakness, weight loss, nausea, vomiting, and orthostatic hypotension. The differential diagnoses for this case included but was not limited to primary infiltrative disease, autoimmune disease (i.e., vasculitis), hemorrhage, infection, malignancy, and lymphoma. With the laboratory findings of SAI and the patient's complaint of vision loss, a pituitary adenoma was suspected, as the former would be caused by apoplexy and the latter by compression of the adjacent optic chiasm. However, an MRI of the brain did not show a pituitary adenoma, although it was not specific for this purpose. Another suspicion was hypotension by virtue of the adrenal crisis causing ischemia to the optic nerve. Ophthalmology was consulted for this patient and no ischemic findings were found, but rather increased intraocular pressure. Might there be a relation such that the adrenal crisis triggered an increase in intraocular pressure prompting vision loss? That much is not clear but is worth further investigation.

Caring for this patient with an atypical presentation of SAI demonstrates the perplexity of medicine. The probability that this patient's vision loss could have been by virtue of his adrenal crisis, or a pituitary mass is about the same as that that it could have been caused by a separate issue, such as advanced glaucoma as the ophthalmologists reasoned. Regardless, the patient was treated with steroids, the mainstay of adrenal crisis management and he no longer had any visual complaints. This, along with proper education on how to manage adrenal insufficiency, has provided a great mortality benefit.

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[1] Olivier Chabre, Bernard Goichot, Delphine Zenaty, Jérôme Bertherat, "Group 1. Epidemiology of primary and secondary adrenal insufficiency: Prevalence and incidence, acute adrenal insufficiency, long-term morbidity and mortality." Annales d'Endocrinologie, Volume 78, Issue 6, 2017, Pages 490-494. ISSN 0003-4266. https://doi.org/10.1016/j.ando.2017.10.010. (https://www.sciencedirect.com/science/article/pii/S0003426617309198)