

Case Report

An Interesting Case of Adrenal Insufficiency with Cardiomyopathy during Index Adrenal Crisis Presentation Followed by Recurrent Pericarditis

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Abstract

Adrenal insufficiency commonly manifests as vague symptoms with or without abnormal lab findings such as hyponatremia, hyperkalemia, hypercalcemia or hypoglycemia. It is very crucial to diagnose adrenal insufficiency in the early stages, as the patient may acutely decompensate due to hypotension, shock, cardiac dysfunction, or respiratory failure. We present a unique case of adrenal insufficiency in a 27-year-old woman who tends to have some degree of cardiac complication with every adrenal crisis episode. Her initial manifestation of adrenal crisis was complex with pericardial effusion and cardiogenic shock from toxic cardiomyopathy due to neurohormonal stress requiring intra-aortic balloon pump and vasopressor/inotropic support. Her cardiac function fully recovered within a few days and returned to baseline prior to discharge. After 1 year she was noted to have a moderate pericardial effusion without cardiac dysfunction during another adrenal crisis episode while being treated for pneumonia. This episode of pericardial effusion resolved within a few days. Since then, she has had multiple episodes of chest pain from pericarditis unrelated to adrenal crisis. This is a rare case of predominant cardiac involvement with every adrenal crisis episode followed by recurrent pericarditis.

ABBREVIATIONS

TTE: Trans-Thoracic Echocardiogram; CTA Chest: Computed Tomography Angiogram of Chest; CT Abdomen: Computed Tomography of Abdomen; ECG: Electrocardiogram; CI: Cardiac Index; SVR: Systemic Vascular Resistance; IABP: Intra-Aortic Balloon Pump; LVEF: Left Ventricular Ejection Fraction; NSAIDs: Nonsteroidal Anti-Inflammatory Drug

INTRODUCTION

Primary adrenal insufficiency/Addison's disease is a rare disorder (prevalence of approximately 120 cases per 1,000,000 in the population). This condition was initially described by Thomas Addison over 150 years ago [1]. This disorder often manifests with vague, nonspecific symptoms, such as fatigue, gastrointestinal symptoms, and weight loss. Physical examination may demonstrate orthostatic hypotension as well as mucosal and skin hyperpigmentation because of elevated melanocyte-stimulating hormone and adrenocorticotropic hormone. Laboratory data may reveal hyponatremia, hyperkalemia, or hypoglycemia. Etiologies for primary adrenal insufficiency include infections,

medications, adrenal hemorrhage, and hereditary disorders. Autoimmune etiology is the leading cause in developed countries. Often autoimmune adrenal disease accompanies other endocrine disorders, like diabetes and thyroid disorders, manifesting as an autoimmune polyglandular syndrome.

Hypovolemic hypotension is the most common cardiovascular complication of Addison's disease. Acute cardiomyopathy and heart failure related to primary adrenal insufficiency have been reported in the literature [2]. Structural myocardial changes have been noted in a small sample of seven patients with newly diagnosed Addison's disease [3]. An autoimmune-mediated phenomenon presenting as dilated cardiomyopathy has also been described in autoimmune polyglandular syndrome [4].

Acute pericardial effusion is predominantly idiopathic in developed countries with no specific identifiable etiology and is presumed to be of viral origin. In 10–20% of the cases, it is associated with identifiable etiologies including autoimmune connective tissue systemic diseases (notably SLE), infections, cancer or trauma [5]. Pericardial fluid accumulation can result in increased pericardial pressure, compression of cardiac

chambers, cardiac tamponade, and subsequent hemodynamic derangements.

Torfoss D et al., in 1997 first described the occurrence of pericarditis with tamponade as an initial manifestation of Addison's disease in a case series of two patients [6]. Tucker et al., postulated that the pathogenesis of pericardial effusion involves not only endocrine failure but also serositis due to derangements in immune complex and immunogenetic status [7].

CASE PRESENTATION

A 27-year-old Caucasian female without any significant medical history except post-traumatic stress disorder had presented to an outlying hospital with complaints of fatigue, recurrent abdominal pain, burning chest pain, dyspnea on exertion, and hypotension. At the outlying facility, she underwent CTA of the chest, which was negative for pulmonary embolus but showed small bilateral pleural effusions and pericardial effusion. CT of the abdomen reported appendicitis and acalculous cholecystitis. She was treated with intravenous fluids and intravenous antibiotics for possible sepsis. However, clinically she started to deteriorate requiring vasopressor support and was intubated for acute respiratory failure. Given her sudden clinical decline and suspicion for ruptured appendicitis, she underwent emergent laparoscopic cholecystectomy and appendectomy. The patient's hemodynamic and respiratory status continued to decline 12 hours post-surgery. She was transferred to our facility for further management. There was no prior history of thyroid problems, autoimmune diseases, recent travel history, or sick contacts. She previously smoked a half pack of cigarettes per day smoked cigarettes and had quit five years before admission. Family history was negative for any autoimmune diseases.

She arrived at our hospital intubated and on mechanical ventilation and intravenous vasopressors. Admission ECG noted low voltage QRS complexes. Immediate TTE demonstrated LVEF of 10-15% with severe global hypokinesis and moderate sized pericardial effusion without tamponade physiology. TTE performed at the outlying facility 24 hours before transfer was unremarkable with LVEF of 70% and no evidence of pericardial effusion. Labs demonstrated mild hyponatremia with sodium 128 mmol/L, elevated creatinine at 1.22 mg/dL, mild lactic acidosis at 2.2 mmol/L, and mild leukocytosis of 15,110/mcL. Pericardiocentesis was deferred considering that clinical decompensation was secondary to cardiomyopathy rather than pericardial effusion. By then she was on two drug vasopressor support. An emergent right heart catheterization was performed for severe cardiogenic shock that noted CI low at 1.8 liters/hour/meter² and SVR markedly elevated at 1900 dynes. IABP was placed for mechanical support for cardiogenic shock. Intravenous inotropes (milrinone and dobutamine) were initiated in addition to vasopressors (norepinephrine and epinephrine). The patient was also placed on intravenous vancomycin and zosyn for severe sepsis and intravenous high dose steroids for possible adrenal crisis. Random morning serum cortisol was noted to be low at 1.6. microgram/deciliter. She underwent extensive investigations evaluating for etiologies of cardiogenic shock. Serum panels including HIV, hepatitis, ANA, mycoplasma, RF, ferritin, and TSH all resulted unremarkable. Blood and urine cultures were negative. Infectious panels, including respiratory

pathogen array, EBV IgM, and CMV, all resulted unremarkable as well. Her cardiogenic shock was considered to be secondary to toxic cardiomyopathy from neurohormonal stress in the setting of acute illness.

Her CI and SVR improved with IABP, vasopressor, and inotropic support. Her respiratory status and acute renal failure improved with intravenous diuresis. Her hospital course was complicated by post-surgical intraperitoneal hematoma necessitating an abdominal drain and acute blood loss anemia treated with packed red blood cell transfusions.

Limited TTE on day 5 of hospital admission noted marked improvement in LVEF from 10% to 35%, the pericardial effusion regressed to small size and without evidence of tamponade physiology. By day 8, she was weaned off of vasopressors. She continued to be on low dose milrinone and dobutamine in addition to 1:2 IABP support. Repeat TTE noted normalization of left ventricular systolic function with LVEF 60-65%. Subsequently, on day 9, she has weaned off all inotropes, and the IABP was removed. She was extubated on day 11. Later, the abdominal drain was removed, and she underwent extensive physical therapy. After a prolonged 24-day hospital course, she was discharged home on hydrocortisone 10 mg two times a day with concern for possible adrenal insufficiency. Two months later, during an outpatient endocrinology follow up appointment, she was noted to have significantly elevated ACTH and was diagnosed with Addison's disease. Fludrocortisone 0.1 mg once daily was initiated in addition to hydrocortisone 10 mg twice daily.

Twelve months later she had presented to an outlying facility after a syncopal episode and was noted to have pneumonia. Prior to presentation the patient had pleuritic chest pain, fever, and nausea. CT chest demonstrated small pleural effusions and basal infiltrates with pericardial effusion. She was started on IV antibiotics and IV stress dose steroids. Serial TTEs were performed on days 2, 3, and 6 and showed gradual improvement in the size of pericardial effusion from moderate to trivial. Cardiac function was noted to be normal on all of the echocardiograms. She was also started on colchicine in addition to the steroids for pericarditis. She was followed by her cardiologist as an outpatient. She continued colchicine and started on naproxen for pericarditis. Both the medications were discontinued after completion of the course. Repeat TTE 2 months later showed normal ejection fraction without any pericardial effusion.

She was noted to have a few more episodes of pericarditis without hospitalization that resolved with colchicine and NSAIDs.

DISCUSSION

We present a unique case of transient toxic cardiomyopathy and recurrent pericardial effusion in the setting of acute adrenal crisis followed by chronic pericarditis. The patient's index presentation was critical with cardiogenic shock and pericardial effusion both of which had resolved with high dose IV steroid therapy and aggressive supportive therapy with mechanical ventilation, vasopressors, inotropes, and intra-aortic balloon pump.

Impaired hemodynamics with volume depletion and low

cardiac output are common in Addison's crisis. Recovery of cardiovascular hemodynamics can be rapidly achieved with steroid therapy [8]. Understanding the pathophysiological consequences of adrenal insufficiency can provide clues to other mechanisms involved in the course of heart failure and aid in the development of novel therapeutics [9].

Pericardial tamponade, although a rare occurrence, could be the initial manifestation of Addison's disease. In Addison's disease, the existing hypovolemia due to aldosterone insufficiency constitutes an aggravating factor as it significantly reduces the filling pressure of the right ventricle [10]. Early recognition of cardiogenic shock from toxic cardiomyopathy in the setting of an adrenal crisis is of critical importance. Differentiating between septic shock and cardiogenic shock is very important as management could vary drastically. In case of cardiac tamponade, relieving the tamponade with pericardial drainage can be lifesaving [11]. There are a few reported cases of patients with adrenal insufficiency with chronic pericarditis requiring pericardiectomy. Our patient's symptoms have been well controlled so far with medical management.

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