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Polyglandular Autoimmune Syndrome Type 2 Presenting With Ventricular Tachycardia

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Abstract **Introduction:** Polyglandular autoimmune syndrome (PGA) describes a condition where dysfunction of two or more endocrine glands occurs in association with circulating organ-specific antibodies directed against the involved glands. **Case Presentation:** A 36 year old female, known type 1 diabetic presented with a 3 week-history of heat intolerance and intermittent palpitations, associated with dyspnea and light-headedness. On examination, a cachexic, lethargic woman with an unrecordable pulse and blood pressure was found. She was found to have sustained monomorphic ventricular tachycardia on cardiac monitor at a rate of 186 beats per minute, confirmed on 12-lead electrocardiogram. The patient was immediately cardioverted, with restoration of sinus rhythm and pulse. Further examination revealed generalized hyperpigmentation with patches of depigmentation, alopecia, a goiter, proptosis and lid lag. Thyroid function test was consistent with thyrotoxicosis and co-syntropin test confirmed adrenal insufficiency. Complete blood count was significant for a macrocytic anemia; with low level of vitamin B12 on testing. A diagnosis of sustained ventricular tachycardia in a patient with PGA type 2 was made and the patient was admitted to the intensive care unit. She was treated with hydrocortisone, fludrocortisone, carbimazole, propanolol and cyanocobalamin. The patient was maintained on amiodarone for one week and she remained in sinus rhythm for the remainder of her hospitalization. **Discussion**: PGA type 2 is characterized by the occurrence of adrenal insufficiency with thyroid autoimmune disease (hypo or hyperthyroidism) and insulin-dependent diabetes mellitus. Electrolyte abnormalities are frequently found in adrenal insufficiency and may predispose to arrhythmias on a background of increased adrenergic effect of thyrotoxicosis. **Conclusion:** The presence of an immunoendocrinopathy warrants the search for other endocrine hypofunction. In situations where a life-threatening arrhythmia is present, urgent identification and treatment of the arrhythmia is top priority.

Keywords: *adrenal insufficiency, autoimmune, polyglandular, thyrotoxicosis, ventricular tachycardia*

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1. Introduction

Polyglandular autoimmune syndrome (PGA) describes a condition where dysfunction of two or more endocrine glands occurs in association with circulating organ-specific antibodies directed against the involved glands [1]. It is divided into types based on the endocrine glands involved and presents with a myriad of clinical features [2]. However, no case of PGA presenting with ventricular tachycardia has been described in the literature.

2. Objective

To report a case of polyglandular autoimmune syndrome (PGA) type 2, presenting with sustained ventricular tachycardia to the emergency unit of a tertiary hospital in Lagos, Nigeria.

3. Case Presentation

A 36 year old female, known type 1 diabetic presented with a 3 week-history of heat intolerance and intermittent palpitations, associated with dyspnea and light-headedness. On examination, a cachexic, fully conscious, but lethargic woman with a body mass index of 15.2kg/m^2 and an unrecordable pulse and blood pressure was found. Cardiac monitor revealed a sustained monomorphic ventricular tachycardia at a rate of 186 beats per minute, confirmed on 12-lead electrocardiogram. The patient was immediately cardioverted, with restoration of sinus rhythm and pulse.

Vital signs obtained after cardioversion were a blood pressure of 92/60mmHg, a pulse of 110 per minute, respiratory rate of 14 cycles per minute and a temperature of 37.0F. Further examination revealed generalized hyperpigmentation with patches of depigmentation consistent with vitiligo, alopecia, a goiter, proptosis and lid lag without ophthalmoplegia or tremors. There was no oral thrush, jugular venous distension, thyroid bruit, or palpable lymph nodes. Examination of the heart revealed normal first and second heart sounds without third or fourth heart sounds. There were no rales on lung exam and no ankle edema.

Thyroid function test revealed a free triiodothyronine (fT3) of 7.5pmol/L (reference range 3.8-6.0), free thyroxine (fT4) of 30pmol/L (reference range 7.2-16.4) and thyroid stimulating hormone (TSH) of 0.01Miu/L (reference range 0.37-3.50), consistent with thyrotoxicosis. Basal cortisol was extremely low (3.5nmol/L), with 30- and 60-minute samples following high dose co-syntropin administration of 92.9nmol/L and 105.5nmol/L respectively. Due to unavailability at our laboratory, plasma adrenocorticotropic hormone (ACTH) was not tested. Anti-thyroglobulin and anti-thyroid peroxidase antibodies were positive. Serum chemistry revealed hyponatremia (120mmol/L), potassium of 5.0mmol/L, mild hyperchloremic metabolic acidosis and fasting blood glucose of 136mg/dL. Complete blood count was significant for a macrocytic anemia; with low level of vitamin B12 on testing. Electrocardiogram (EKG) revealed sinus tachycardia and a prolonged QT interval. Her echocardiogram revealed normal heart chamber size and wall thickness, with normal systolic and diastolic function and without significant valvular abnormalities or pulmonary hypertension.

A diagnosis of sustained ventricular tachycardia in a patient with PGA type 2 was made and the patient was admitted to the intensive care unit. Her electrolyte abnormalities were corrected and she was transfused with 2 units of packed red blood cells. Hydrocortisone was started intravenously initially and then continued orally. In addition, fludrocortisone together with divided daily doses of carbimazole and propanolol, together with cyanocobalamin were instituted. The patient was maintained on amiodarone for one week and she remained in sinus rhythm for the remainder of her hospitalization. She was discharged thereafter and is currently being followed up at the endocrinology, hematology and cardiology clinics, without recurrence of ventricular arrhythmias.

4. Discussion

Polyglandular autoimmune syndrome is an immunoendocrinopathy which comprises of dysfunction in multiple endocrine glands. PGA type 2 is characterized by the occurrence of adrenal insufficiency with thyroid autoimmune disease (hypo or hyperthyroidism) and insulin dependent diabetes mellitus. PGA type 1 on the other hand is characterized by hypoparathyroidism, mucocutaneous candidiasis and adrenal insufficiency [1]. Electrolyte abnormalities are frequently found in adrenal insufficiency type 2 and may predispose to arrhythmias on a background of co-existent thyroid dysfunction in PGA type 2, which increases susceptibility to arrhythmias posed by the increased adrenergic effect of thyrotoxicosis. However, arrhythmias due to thyroid dysfunction tend to be supraventricular (including paroxysmal atrial tachycardia, atrial fibrillation, and atrial flutter). No case of PGA type 2 presenting with ventricular tachycardia was found in our search of the literature.

This patient presented with unstable ventricular tachycardia and co-existent adrenal crisis, manifesting with shock and electrolyte abnormalities (hyponatremia, borderline hyperkalemia, mild hyperchloremic metabolic acidosis). Even after cardioversion with restoration of sinus rhythm, she had relative hypotension. A normal response to the high-dose co-syntropin (ACTH) stimulation test is a rise in serum cortisol concentration after either 30 or 60 minutes to a peak of ≥18 to 20 mcg/dL (500 to 550 nmol/L) [3,4]. A normal response to the high-dose (250mcg intravenously) co-syntropin stimulation test excludes primary adrenal insufficiency [5] and most patients with secondary adrenal insufficiency. This patient's adrenal insufficiency was confirmed by an extremely low basal cortisol and very minimal response to co-syntropin. The etiology of the adrenal insufficiency was presumed to be autoimmune, given the presence of thyroid autoantibodies and the presence of vitiligo and pernicious anemia which are other autoimmune diseases. Although plasma ACTH could not be measured due to unavailability of the assay in our laboratory, the generalized hyperpigmentation found in this patient suggests primary adrenal insufficiency.

The etiology of the ventricular tachycardia seen in this patient with a structurally normal heart on echocardiography remains unclear, but it may have been due to electrolyte abnormalities. Our patient had Prolonged QT interval on EKG which returned to normal following correction of hyponatremia and institution of fludrocortisone to correct her mineralocorticoid deficiency. Prolonged QT is a marker of abnormal myocardial repolarization [6] which may cause a characteristic life-threatening arrhythmia called torsades de pointes, a form of polymorphic ventricular tachycardia [7,8], which was not seen in our patient. However, hyponatremia has not been previously described as a cause of prolonged QT interval.

5. Conclusion

The presence of an immunoendocrinopathy warrants the search for other endocrine hypofunction. Early recognition and replacement therapy can be lifesaving. In situations where a life-threatening arrhythmia is present, urgent identification and appropriate treatment of the arrhythmia is top priority.

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