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# TYPE II POLYGLANDULAR AUTOIMMUNE SYNDROME PRESENTED WITH DIABETIC KETOACIDOSIS; A RARE CASE

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## ABSTRACT
Type II polyglandular autoimmune syndrome is characterized by two or more of the endocrinopathies. Commonly involved endocrinopathies are primary adrenal insufficiency, Graves' disease or autoimmune hypothyroidism, type 1 diabetes mellitus and primary hypogonadism. We reported a 22 years old female with autoimmune hypothyroidism, primary adrenal insufficiency, vitiligo, alopecia and insulin dependent diabetes mellitus who was diagnosed as a case of PGA type II syndrome. It is important to highlight, if a patient has an autoimmune disease that should be screen for other autoimmune conditions. This patient was improved on thyroxine, insulin and steroid replacement therapy.

**KEYWORDS:** Type II polyglandular autoimmune syndrome, Autoimmune hypothyroidism, Insulin dependent diabetes mellitus, Vitiligo, Alopecia.

## INTRODUCTION
Polyglandular autoimmune syndrome type II is characterized by two or more of the endocrinopathies. It is also known as Schmidt's syndrome.[1] PGA 2 should be considered if immune dysfunction affects two or more endocrine glands and other nonendocrine disorders. These are primary adrenal insufficiency (Addison's disease), type 1 diabetes mellitus, Grave's disease, autoimmune thyroiditis and other autoimmune disorders like myasthenia gravis, primary hypogonadism, vitiligo, alopecia and serositis and pernicious anaemia.[2]

## CASE REPORT
A 22 Years old bit obese unmarried female was presented in our emergency department with complaints of malaise, generalised weakness and increased thirst for 2 weeks followed by low grade fever, nausea and vomiting, abdominal discomfort and feeling faint for 2 days. On enquiry her relatives told that she was taking thyroxine 100µg for hypothyroidism for 4 months. On the day of admission, patient was dehydrated and looking anxious. She had no pallor and icterus. Her temperature was 100$^{o}$F and pulse rate was 110/ min hypovolumic. Her systolic B.P-80/64 mmHg and respiratory rate was-22/min. On physical examination vitiligo was present on lips and left lower limb. Alopecia was also present. Her abdomen was soft without any organomegaly. Rest of the systemic examinations did not reveal any abnormalitiy. Her random blood glucose was >500 mg/dL. Arterial blood gas analysis revealed pH 7.10, $pCO_2$ 38 mmHg, $pO_2$ 94 mmHg, $HCO_3$ -13.6). Urine dipstick was positive for ketones (+++) and glucose (+++). Patient was diagnosed as a case of diabetic ketoacidosis (DKA). Standard protocol of DKA was followed. She was managed with normal saline, regular insulin drip and broad spectrum antibiotics. Her BP was not picked up even after five hours of appropriate fluid resuscitation while normalize of blood glucose. Patient was re-evaluated and a short synacthen test with a baseline ACTH was carried out. Hydrocortisone 100mg was administered to the patient immediately. BP was improved after Hydrocortisone therapy. It was suggestive of primary adrenal insufficiency. Her significant Arterial Blood Gas analysis (ABG) reports are summarised in table 1. Her completed blood count showed haemoglobin (Hb)-13.5 g/dl, total leucocyte count- 12600/mm$^3$, diffential leukocyte count: neutrophils 84%, lymphocytes 14% and platelet count 2,00000/mm3. Smear examination for malaria parasite was negative. Her renal function tests, liver function tests, serum electrolytes, fasting lipid profiles, serum calcium and serum phosphate were within normal limits. Thyroid profile showed thyroid stimulating hormone (TSH) 16.0 uIU/mL (normal range 0.4–5.0), antithyroid peroxidase 2200 IU/mL (normal range 60), free thyroxine 0.9 ng/dL(normal range 0.7–2.0 mg/dL). Chest X-ray, Abdominal Sonography, 2D-Echocardiography and ECG were normal. Enzyme-linked immunosorbent assay for human immunodeficiency virus, Australia antigen for Hepatitis B and antibody against Hepatitis C virus were

negative. On the basis of history, examination and laboratory findings patient was diagnosed as a case of polyglandular autoimmune type II syndromes having autoimmune hypothyroidism, primary adrenal insufficiency, vitiligo, alopecia and insulin dependent diabetes mellitus. Patient was improving day by day and got discharged on 13th day of admission.

**Table 1.**

| ABG parameters | Significant ABG during KGMU hospitalisation | | | | |
|---|---|---|---|---|---|
| | Normal range | 1st day | 2nd day | 3rd day | 4th day |
| pH | 7.35-7.45 | 7.1 | 7.2 | 7.32 | 7.38 |
| Pco2(mmHg) | 35-45 | 38 | 39 | 44 | 42 |
| Po2(mmHg) | 80-100 | 94 | 92 | 96 | 96 |
| HCO3mEq/L | 22-26 | 13.6 | 18 | 21.8 | 22.0 |

## DISCUSSION

Type II polyglandular autoimmune syndrome is a female predominant rare endocrine disorder with a frequency of 1.4–2.0/100 000. It is characterised by autoimmune adrenal failure, autoimmune thyroid disease and/or type 1 diabetes. Other autoimmune conditions associated with it are pernicious anaemia, vitiligo, hypergonadotrophic hypogonadism, chronic autoimmune hepatitis, coeliac disease, autoimmune diabetes insipidus and rarely lymphocytic hypophysitis, stiff-person syndrome and myasthenia gravis.[3] Vitiligo and alopecia are present in our case though less common in PGA 2 than PGA1. Vitiligo is caused by antibodies against the melanocyte.[1] A variety of autoantibodies are seen in PGA type II which are 21 hydroxylase autoantibodies (21OHAb) directed against adrenal cortex, thyroperoxidase (TPOAb), thyroglobulin (TGAb) and TSH receptor autoantibodies (TRAb) directed against thyroid; insulin (IAA), and glutamic acid decarboxylase autoantibodies (GADAb) directed against the endocrine pancreas.[4] The primary adrenal insufficiency is because of an autoimmune process that destroys the adrenal cortex. When there is evidence of both humoral and cell-mediated immune mechanisms directed at the adrenal cortex, it is often associated with autoimmune destruction of other endocrine glands referred as polyglandular autoimmune syndromes. However, first-degree relatives of patients with autoimmune primary adrenal insufficiency express these antibodies and have an increased risk of developing adrenal insufficiency of up to 10%.[2] Our case was found to have autoimmune hypothyroidism, primary adrenal insufficiency, vitiligo, alopecia and insulin dependent diabetes mellitus who was finally diagnosed as a case of Type II polyglandular autoimmune syndrome. We are reporting this case because the patient recovered on thyroxine, insulin and steroid replacement therapy.

## CONCLUSION

It is important to highlight in this case that if a patient has an autoimmune disease or patient fails to respond with appropriate treatment of uncontrolled T1DM or its acute complications, patient should be reassessed for other possible endocrine abnormalities. Early diagnosis and management with prompt supportive care can reduce the morbidity and mortality of these patients by preventing complications.

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