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AN INTERESTING CASE OF ADDISONS DISEASE

Endocrinology

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ABSTRACT

Addison's disease is a disorder of chronic adrenal insufficiency [1]. Tuberculosis is said to have accounted for one of the most common causes due to bilateral adrenal gland destruction; autoimmune disease accounts for nearly 70-90% currently with the remaining being caused infectious diseases, metastatic cancer, lymphoma, adrenal haemorrhage or infarction, or drugs.

KEYWORDS

Addison's disease, Polyglandular autoimmune syndrome type 2.

INTRODUCTION

We are reviewing a case of a young female a known hypothyroid since the age of 12 years on medication who presented to the OPD with complaints of dysphagia, nausea, vomiting and fatigue. On examination she was found to have pallor and uniform homogenous hyperpigmented skin. Further investigations revealed a low serum cortisol level and a serum ACTH level more than 2000pg/ml with Anti-TPO antibodies more than 1000 IU/mL. Imaging revealed enlarged thyroid glands with multiple nodules. This was found to be a case of Polyglandular autoimmune syndrome type 2. Patient was started on corticosteroid therapy and is under regular follow up.

CASE REPORT

A 23-year-old female, known case of hypothyroidism on irregular treatment presented to the OPD with complaints of dysphagia more to solids than liquids, nausea, vomiting, fatigue for 1 week. History of chronic weight loss was present. On examination she was found to be severely anaemic suggesting that the dysphagia may have been due to a post cricoid web. Her skin appeared homogenously hyperpigmented for which she gave a history of hyperpigmentation having started 1 year ago and was left unevaluated as patient was unaware of its clinical implications. Her blood pressure was 90/60, with a pulse rate of 88/min and a O2 saturation maintaining at 94% at room air. Systemic examination was unremarkable. All base line investigations were sent. Hb was 10.5 with peripheral smear showing microcytic hypochromic anaemia. UGI scopy revealed granular mucosa in the body and antrum possibly being H.Pylori induced and no evidence of any other abnormality. Ultrasound neck revealed enlarged thyroid gland with multiple nodules. Further investigation related to the work up on the causes of hyperpigmentation with Addison's disease in mind showed low 8 AM cortisol levels (0.054 ug/dL). Serum ACTH levels were more than 2000pg/ml. Serum Anti-TPO antibodies were more than 1000IU/L. The above paraments suggests an autoimmune aetiology and an accompanying autoimmune related hypothyroidism[10]. Parathyroid hormone levels were normal. A diagnosis of Autoimmune polyglandular syndrome type 2 was made; hypoparathyroidism which is a characteristic finding in APS type 1 is not found in APS type 2 [9].RBS was 99mg/dL which was sent to rule out Type 1 diabetes mellitus seen in 11 % cases of APD-2.Patient was started on corticosteroids ,thyroid supplementation, fluids and measures to correct the underlying anaemia. Patient was symptomatically better on day 7 of admission and was discharged with steroid and thyroxine supplementation. Patient is under regular follow up for monitoring improvement and dose adjustment.

Prior 1 year

On presentation

DISCUSSION

Patients affected with APS-2 have a course characterized by at least two of the following three endocrinopathies: type 1 diabetes, autoimmune thyroid disease, and Addison's disease [2]. Women are affected more with APS-2. other manifestations related to autoimmune conditions can also develop, which includes celiac disease, alopecia, vitiligo, primary ovarian insufficiency, and pernicious anaemia [8]. The onset of APS-2 classically occurs in young adulthood, much ahead than the onset of APS-1. Currently, there is no gold standard investigation to diagnose APS-2 in patients but testing for autoantibodies may be supportive in assessing disease risk, since the related autoantibodies are often detectable years before disease onset [3]. Examples are antibodies to thyroid peroxidase in autoimmune thyroid disease, to glutamic acid decarboxylase in type 1 diabetes, and to 21-hydroxylase in autoimmune Addison's disease [6].

Treatment and follow-up of autoimmune polyendocrine syndrome: In general, management of autoimmune polyendocrine syndromes consist of hormone-replacement therapy as required and treatment of complications. Patients must have a minimum of two follow-up visits every year. It is mandatory to check all siblings of patients even if the siblings are adults and apparently well. Screening for 21-hydroxylase is beneficial in assessing the risk of the development of adrenal insufficiency [5]. Treatment of APS-2 should focus on replacement of deficient hormones in accordance with recent guidelines for treating the key components of APS-2. The clinician should be vigilant that a patient with APS-2 is at increased risk for the development of other organ-specific autoimmune disease.

CONCLUSION

This case report is to highlight the importance of early work up and management of young female patients, presenting with typical symptoms, hyperpigmented skin and immature sexual development which should prompt a diagnostic work up for autoimmune polyglandular endocrinopathies. Such patients must be on regular follow up in order to prevent them from going into an adrenal crisis [4]; patients may also be screened for other autoimmune induced endocrinopathies most importantly to pick up ones that are of new onset [7].

CONFLICT OF INTEREST

Conflict of interest declared as none.

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