Journal of Advances in Medical Sciences

Case Report

Autoimmune Poly-Endocrinopathies: A Case Report

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Received Date: 14 January 2024

Revised Date: 17 February 2024

Accepted Date: 24 February 2024

Publication Date: 02 March 2024

Abstract

Introduction: Autoimmune poly-endocrinopathies are uncommon disorders characterised by the coexistence of at least two endocrine deficits related to an autoimmune mechanism with sometimes a non-endocrine autoimmune disease. They represent a rare set of inherited disorders characterised by multiple target organ dysfunctions.

Case Presentation: We report the case of a 31-year-old pregnant woman with autoimmune poly-endocrinopathies.

Clinical Discussion: Polyglandular dysfunction". These disorders are usually characterised by the presence of circulating organ-specific antibodies even in the absence of overt clinical disease. Polyendocrinopathy type II is the most common form of polyendocrine dysfunction.

Conclusion: Autoimmune polyglandular syndromes should be kept in mind when dealing with pregnant patients with multi-glandular endocrine dysfunction in order to improve maternal and fetal management.

Keywords

Autoimmune, Polyendocrinopathies, Hashimoto's thyroiditis, Addison's disease.

INTRODUCTION

Autoimmune polyendocrinopathies are uncommon disorders characterised by the coexistence of at least two endocrine deficits related to an autoimmune mechanism with sometimes a non-endocrine autoimmune disease. They represent a rare set of inherited disorders characterised by multiple target organ dysfunctions. They are classified into three types based on their clinical presentations, with types I and II being well-defined entities and type III being poorly defined. The autoimmune nature of these disorders is based on the presence of lymphocytic infiltration of the affected gland, organ-specific autoantibodies in the serum and association with HLA binding genes.

As these individuals have more than one endocrine dysfunction, it is important to look for this syndromic grouping especially during pregnancy in any parturient presenting with a cluster of major endocrinopathies, due to the preponderant maternal-fetal risk, and in the guise of a comprehensive management All our work was reported in accordance with the SCARE criteria and guidelines [8].

CASE PRESENTATION

A 31 year old pregnant woman, I G I P, came at 22 weeks of amenorrhea, for a pregnancy follow-up with a history of T1DM under insulin since the age of 16 years, hashimoto's thyroiditis under ttt for 10 years, followed for an addison's disease under ttt for 5 years, a celiac disease diagnosed since the young age Pregnancy follow-up was unremarkable. The patient underwent an emergency cesarean section for a non-reassuring fetal condition at 35 weeks' gestation, with a large amount of ascites during the surgical exploration. The post-operative course was marked by the appearance of a glycemic imbalance consisting of a profound hypoglycemia at 0.17, and a hyperkalemia at 6. 1, a frank hypoalbuminemia at 18 g/l, on the thoraco-abdomino-pelvic scanner we noted the presence of a polyseritis made up of a bilateral pleural effusion of little abundance, a pericardial effusion of little abundance, an ascites of great abundance which was punctured, the patient was declared discharged at d 10 of the post-partum.

DISCUSSION

Autoimmune poly-endocrinopathies may affect several endocrine glands in the same patient, hence the term 'polyglandular dysfunction'. These disorders are usually characterised by the presence of circulating organ-specific antibodies even in the absence of overt clinical disease. The term "polyglandular autoimmune syndrome" was originally introduced by Neufeld [1], and the classification into three subtypes is based on specific clinical features, with type I autoimmune polyendocrinopathy consisting of a triad of Addison's disease, chronic mucocutaneous candidiasis and hypoparathyroidism. Several disorders may be associated [2]. The onset of symptoms is at an early age, usually in the first decade of life, with males and females equally affected. In type I, there seems to be no correlation with HLA, familial clustering of cases, however, does exist, occurring by an autosomal recessive mode of transmission [3].

Polyendocrinopathy type II is the most common form of polyendocrine dysfunction [4]. The type II classification, or Schmidts syndrome, requires the presence of Addison's disease, autoimmune thyroiditis and/or insulin-dependent diabetes mellitus. The disorder may also be associated with biermer anaemia, gonadal insufficiency, celiac disease [5]. Unlike type I, type II has a later onset with a maximum age of 30 years. There is a strong female predominance, with a dominant mode of inheritance involving the important genes found with the HLA complex on chromosome 6, in particular the HLA-DR genes DR3 and DR4 [6]. Biologically, a number of organ-specific autoantibodies are present in the serum, including antiadrenal, antithyroid and anti-pancreatic antibodies [7].

Treatment of autoimmune poy-endocrinopathies is individualised and based on correction of the major abnormalities. In patients with hypothyroidism and Addison's disease, thyroid supplementation therapy can precipitate a life-threatening Addisonian crisis: it is therefore crucial to assess adrenal function in hypothyroid patients if an autoimmune polyglandular syndrome is suspected before starting replacement therapy. Thyroid function is often improved after the start of glucocorticoid replacement.

Early diagnosis of the type of autoimmune poly-endocrinopathy, especially during pregnancy, is important, not only to initiate adequate replacement therapy for the organs so affected, but also to perform genetic analysis. In families where the syndrome has been documented, relatives should be notified of the early signs and symptoms. In this at-risk population, as well as patients with multiple disorders, screening every 3 to 5 years should take place with blood glucose measurements, thyroid function tests, B12 levels and cortisol levels, such screening can prevent significant morbidity and mortality in these patients and their offspring [8].

CONCLUSION

Autoimmune polyglandular syndromes are rare disorders characterised by the development of multiple endocrine dysfunction of target organs. The syndromes encompass a range of diseases characterised by polyglandular dysfunction. These syndromes should be kept in mind when dealing with pregnant patients with multi-glandular endocrine dysfunction in order to improve maternal and fetal management.

Provenance and Peer Review: Not Commissioned, Externally Peer-Reviewed

Consent: Written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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