CASE REPORT

Polyglandular Autoimmune Syndrome III with Hypoglycemia and Association with Empty Sella and Hypopituitarism

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A B S T R A C T

A 25-year-old Saudi female with a known case of autoimmune thyroiditis presented to the Emergency Room in stuporous condition. A blood test revealed a blood sugar level of 1.7 mmols/l (30.6 mg/dl). The patient was resuscitated with intravenous glucose. Further evaluations of the patient revealed celiac disease and idiopathic thrombocytopenia with preexisting autoimmune thyroiditis (polyglandular autoimmune syndrome III [PAS III]). The severe hypoglycemia, coupled with 6 years of infertility evaluation, revealed a rare association of empty sella syndrome with hypopituitarism {PAS II}.

Key words: Empty sella syndrome, hypopituitarism, polyglandular autoimmune syndrome III

ملخص البحث:
يعرض الباحثون حالة مريضة تبلغ من العمر 25 سنة، تعاني من التهاب مناعي في الغدة الدرقية أحضرت إلى المستشفى في حالة ذهولية ، اظهر تحليل السكر في الدم انخفاضا قدره (36.3ing/dl) مما استدعى إعطاءها محلول سكر وريديا. أظهر تقييم حالتها وجود حساسية القمح ونقص في الصفائح ، إلى جانب معاناتها من التهاب الغدة الدرقية المناعي. تسمى هذه الحالة بمتلازمة متعددة الغدد المناعي، كما اظهر تقييمها فيما بعد أنها تعاني من العقم ومتلازمة السمع الفارغ وقصور في الغدد النخامية وانخفاض في سكر الدم، ومتلازمة متعددة الغدة المناعـــي (PAS III).

INTRODUCTION

Polyglandular autoimmune syndromes (PASs) are rare disorders characterized by failure of several endocrine and nonendocrine organs associated with immune-mediated destruction. Autoimmune polyglandular syndrome III is a constellation of autoimmune thyroiditis, with another organ-specific autoimmune disease, which cannot be classified as PAS I or PAS II. We are reporting the case of a 25-year-old female with PAS III with a rare association with empty sella turcica and hypopituitarism presenting with severe hypoglycemia. We will discuss the possible association and pathogenesis of empty sella in the setting of PAS III.

CASE REPORT

A 25-year-old Saudi female with a known case of autoimmune thyroiditis (Hashimoto's thyroiditis) presented to the emergency room in stuporous condition. For the past 7 years, she had been treated at a different hospital and prescribed irregular thyroxine replacement to regulate her menstrual cycles. The patient presented

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with fever, sore throat, vomiting, abdominal pain and confusion of 2 days duration.

A blood sugar test by glucocheck revealed a blood sugar level of 1.7 mmols/l (30.6 mg/dl). The patient was resuscitated with intravenous glucose and her cognitive function promptly improved. Previously, she had experienced similar episodes of hypoglycemia following flu-like symptoms. The patient had a history of nausea, occasional vomiting, abdominal pain and weight loss of 20 kg over the past 2 years. There was no history of headache visual disturbance, galactorrhea, polyuria and polydipsia. The patient had a 6-year-old daughter and had been unsuccessful in conceiving since her daughter's birth. Following delivery, she had no history of postpartum hemorrhage but had experienced irregular menstrual cycles. There was no family history of autoimmune disease. Upon physical examination, the pulse was 102/min and the blood pressure was 100/64 mmHg. There was no hypo- or hyperpigmentation of the skin, hyperpigmentation of the buccal mucosa. The rest of physical examination revealed no abnormalities. The radiological imagings were normal; MRI of the pituitary fossa showed a partially empty sella with asymmetry of the residual pituitary gland [Figure 1].

The patient was diagnosed with autoimmune thyroiditis, autoimmune thrombocytopenia, celiac disease (PAS III) and partial empty sella syndrome with hypopituitarism. Corticosteroids were initiated, followed by adjustment by adjustment of the Thyroxine. A gluten-free diet was advised by the dietician. The patient's symptoms improved and blood sugars were maintained within the normal range. Her thrombocytopenia responded quickly with steroid treatment [Figure 2].

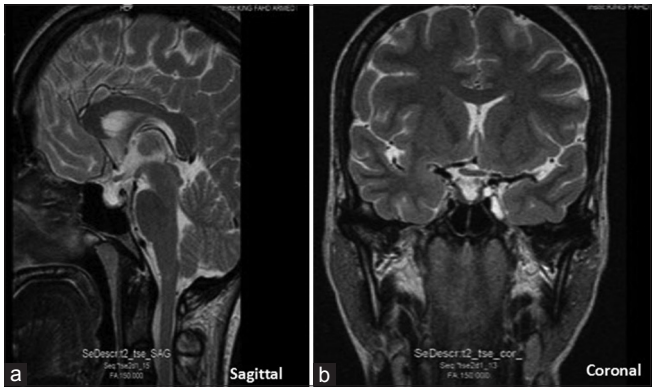

Figure 1: (a) Coronal section of pituitary magnetic resonance imaging showing empty sella with no obvious pituitary gland tissue. (b) High signal T2 sagittal section of same pituitary showing empty sella.

DISCUSSION

PAS is defined as multiple endocrine gland insufficiencies accompanied by autoimmune disease of endocrine and nonendocrine systems. Schmidt described a case of non-tuberculous adrenal gland dysfunction with thyroiditis in 1926. Neufeld defined PAS I, PAS II and PAS III and its subtypes in 1980.[1] PAS III is a combination of autoimmune thyroiditis with other organ-specific autoimmune disease. In this case, a 25-year-old Saudi female had suffered from autoimmune thyroiditis with poorly controlled thyroid function for 7 years. Patient had a history of celiac disease. The laboratory findings revealed iron deficiency anemia, low total protein, low albumin and positive anti-transglutaminase immunoglobulin A and immunoglobulin G. Confirmation with jejunal biopsy could not be done since the patient refused to have an endoscopy.

She has asymptomatic chronic mild thrombocytopenia. On exclusion of other causes of chronic thrombocytopenia, such as drugs, autoimmune diseases, viral hepatitis and idiopathic thrombocytopenia a diagnosis of autoimmune thrombocytopenia was made. Thrombocytopenia secondary to celiac disease is rarely described and was unlikely as it responded to steroids. With the combination of autoimmune thyroiditis, celiac disease and idiopathic thrombocytopenia, we diagnosed her with PAS III.[2] Even though autoimmune thrombocytopenia is an uncommon association with PAS, its association with PAS II and PAS III has been reported in several cases in the literature.[3-7] The association of celiac disease with PAS is also reported in few case reports.[8-10] It is quite interesting to note that the present case with PAS III is associated with empty sella and hypopituitarism, manifested by severe secondary adrenal insufficiency presenting with

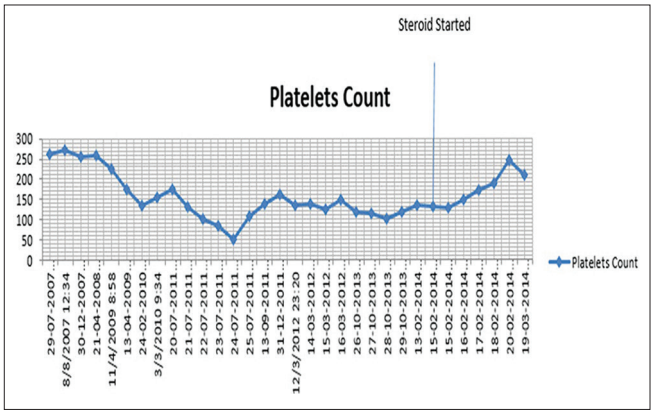

Figure 2: The graph showing chronic thrombocytopenia and brisk response to corticosteroids.

hypoglycemia, hyperprolactinemia and hypogonadism. Very few cases of PAS associated with empty sella have been reported in the literature.[10-13] The pathogenesis of empty sella and hypopituitarism is debatable in this case with PAS III. The primary empty sella which is not an uncommon condition, may be considered as a coincidental finding with PAS III. Although some studies claim deficiencies of one or more pituitary hormones the evidence for such deficiencies is not convincing in primary empty sella.[14] This patient had poorly controlled autoimmune thyroiditis for several years. The resultant thyrotrophic hyperplasia might have damaged the anterior pituitary gland leading to hypopituitarism and subsequently empty sella. This scenario is extremely rare and only four cases have been reported in the literature.[15] The other speculation could be the empty sella with hypopituitarism might have been the late consequence of autoimmune hypophysitis, which is a known association of PAS.[16] There are several case reports where the empty sella is described as the final outcome of lymphocytic hypophysitis.[12,17,18] This interesting case demonstrates PAS III with a rare association of empty sella syndrome and hypopituitarism.

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Conflicts of interest

There are no conflicts of interest.

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