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Case Report

Pituitary Stalk Interruption Syndrome: A Case Report

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Abstract

Panhypopituitarism occurs as a result of the insufficiency of all hormones produced in the anterior pituitary gland. Pituitary stalk interruption syndrome (PSIS) is a rare congenital syndrome leading to hypopituitarism. The anterior pituitary hormones should be evaluated in patients with the signs and symptoms of hormone insufficiency and magnetic resonance imaging of the pituitary should be performed for the etiology. Although PSIS is a rare cause of hypopituitarism, it must be treated with the replacement of the insufficient hormones. In this case report, we evaluated a PSIS case presented with panhypopituitarism.

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Introduction

Panhypopituitarism is the name given to the clinical situation that occurs as a result of the insufficiency of all hormones produced in the anterior pituitary gland. Pituitary stalk interruption syndrome (PSIS) is a rare congenital syndrome that is usually presented with the hypoplastic pituitary gland, undemonstrated pituitary stalk, and ectopic neurohypophysis. PSIS is an intrinsic pituitary cause of panhypopituitarism. Available data suggests that complex genetic patterns and environmental factors work together for PSIS.[1,2] Although the disease onset varies regarding age, it can typically be seen in pediatric ages as combined anterior pituitary hormone deficiencies and can progress to panhypopituitarism even in adulthood. Therefore, lifelong follow-up of patients is essential for sufficient management. In this case, we aimed to report a PSIS case presented with panhypopituitarism.

Case Report

A 22-year-old female patient, who was followed up with a diagnosis of panhypopituitarism, was admitted to our endocrinology outpatient clinic with complaints of anorexia, weakness, nausea, and vomiting. It was discovered that she had an involuntary weight loss of approximately 5% in the last 2 months and did not comply with levothyroxine sodium and fludrocortisone treatments. Also, she was amenorrheic. During the physical examination, her blood pressure was

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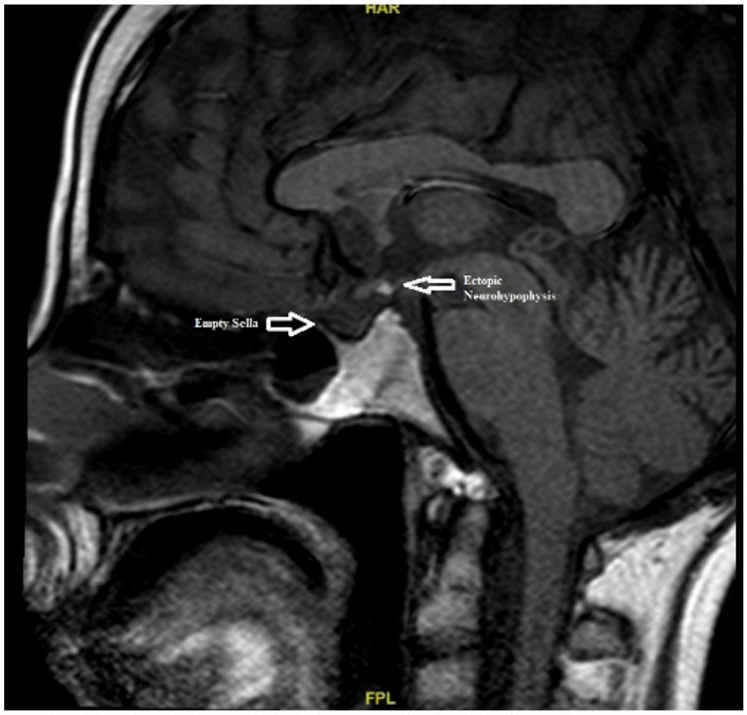

Figure 1. Ectopic neurohypophysis and empty Sella view in Sella MRI.

100/60 mmHg, pulse rate was 130 beats/min, rhythmic and body temperature was measured to be 36.4 °C. Cardiovascular assessments were normal. No pathological finding was found in other physical examinations. In the laboratory investigation during admission, her serum creatinine was 0.75 mg/dL, sodium 136 mmol/L, potassium 3.8 mmol/L, AST 21 U/L, and ALT 9 U/L. As a result of the clinical and laboratory evaluation, the patient was suspected to have adrenal insufficiency and administered hydrocortisone and hospitalized in the endocrinology clinic. Her follow-up and treatment were initiated. In the examinations performed during hospitalization, her anterior pituitary hormone levels were lower than normal; ACTH: 5 ng/L, TSH: 0.07 mU/L, free T3: 1.73 ng/L, free T4: 1.10 ng/dL, FSH: 0.24 IU/L, LH: <0.09 IU/L, GH <0.05 ug/L, somatomedin C (IGF-1) <15, cortisol 0.8 µg/dL, estradiol 10 ng/L, and progesterone <0.1 µg/L. Sella magnetic resonance imaging (MRI) showed an image compatible with ectopic neurohypophysis, empty Sella syndrome, and PSIS (Figure 1). Oral hydrocortisone and oral contraceptive replacement treatments were initiated in appropriate doses for the patient, and other hormonal treatments were arranged accordingly. After observation of clinical improvements, the patient was discharged and asked to attend regular outpatient follow-ups.

Discussion

As in our case, hypopituitarism may present with various clinical findings. The anterior pituitary panel should definitely be examined in patients who have signs of hormone insufficiency. After the diagnosis is made, magnetic resonance imaging of the pituitary gland should be performed for the etiology. Although PSIS is a rare cause of hypopituitarism, its treatment should include the replacement of the insufficient hormones like thyroxine, glucocorticoids, sex steroids and growth hormone when necessary. The prognosis of the disease is good under regular follow-ups and treatments.

Conflict of Interests

Authors declare that there are none.

Acknowledgment

This study has been presented in 17th Uludag Internal Medicine National Winter Congress, 6th Bursa Family Medicine Association National Congress, 11th Uludag Internal Medicine Nursing Congress, 5–7 March 2021, Bursa, Turkey.

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