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Panhypopituitarism Due to Hemochromatosis

Hemokromatozise Bağlı Panhipopituitarizm

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Abstract

Hemochromatosis is an iron storage disease. Panhypopituitarism is a clinical condition in which the anterior pituitary hormones are deficient. Herein, we report a rare case of panhypopituitarism due to hemochromatosis. *Turk Jem 2013; 17: 125-6*

Key words: Hemochromatosis, hypopituitarism, liver cirrhosis

Özet

Hemokromatozis bir demir depo hastalığıdır. Panhipopituitarizm ise ön pituiter bez hormonlarının yetersiz olduğu klinik durumdur. Bu yazıda hemokromatozise bağlı panhipopituitarizmli nadir bir olgu bildirilmiştir. *Turk Jem 2013; 17: 125-6*

Anahtar kelimeler: Hemokromatozis, hipopituitarizm, karaciğer sirozu

Introduction

Hemochromatosis is an iron storage disease characterized by iron deposition in parenchymal cells due to increased intestinal iron absorption. Iron overload leads to tissue damage and dysfunction particularly in the liver, pancreas, heart, joints, and pituitary gland. Panhypopituitarism is a clinical condition in which the anterior pituitary hormones are deficient. Herein, we report a rare case of hemochromatosis.

Case Report

A sixty-two-year-old woman was admitted to our hospital with the complaints of weakness, body swelling and confusion. On admission, her general condition was poor; regarding her state of consciousness, she had a tendency to sleep, and she had an apathetic face and bronze-colored skin. Past medical history included hepatitis C, esophageal varices and a liver biopsy performed 1.5 years ago which revealed the presence of micronodular cirrhosis with accumulation of hemosiderin. Laboratory values on admission are shown on Table 1. Abdominal ultrasonography showed chronic parenchymal liver disease, splenomegaly (15.5 cm) and cholelitiasis. She was hospitalized in the intensive care unit with the diagnosis of chronic parenchymal liver disease and hepatic encephalopathy. We performed pituitary hormone tests for the possibility of pituitary involvement due to hemochromatosis. The values were: LH: 1.12 [14.2-52.3] IU/ml, FSH: 2.77 [19.3-100.6] IU/ml, GH: <0.05 ng/ml, prolactin: 0.657 [3-23] ng/ml, cortisol: 3.54 [5-23] µg/dl. An ACTH stimulation test was done for pituitary insufficiency (Synacthen 1 mg IM). At 0th minute, the cortisol level was 3.54 µg/dl, 30th minute - 20 µg/dl and at 1st hour, it was 10 µg/dl. Therefore, primary adrenal insufficiency was excluded. Contrast-enhanced computed tomography of the pituitary was normal. Thus, the patient was diagnosed as having panhypopituitarism. Methylprednisolone 40 mg IV initially and levothyroxine on the fifth day were administered. She recovered with this therapy and discharged with prednisolone 7.5 mg/day and levothyroxine 0.1 mg/day.

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Table 1. Laboratory values

	Patient's values	Normal values		Patient's values	Normal values
AST (U/L)	39	[13-40]	AFP (IU/ml)	5.4	[0.5-5.5]
ALT (U/L)	28	[7-45]	Hb (g/dl)	10.5	[12-18]
GGT (IU/L)	21	[3-96]	Fe (mcg/dl)	127	[50-175]
ALP (IU/L)	85	[32-213]	Ferritin (ng/ml)	581	[10-250]
T. BIL (mg/dl)	5.3	[0.3-1.2]	IBC (mcg/dl)		
D. BIL (mg/dl)			fT3 (pg/ml)	120	[250-450]
PT (sec)	4.3	[0-0.2]	fT4 (pg/ml)		
Alb (g/dl)			TSH (IU/ml)	0.39	[1.8-5.2]
	36.7	[11-16]		< 1	[0.8-2.7]
	2.5	[3.5-5.5]		0.79	[0.4-4.2]

AST: aspartate aminotransferase, ALT: alanine aminotransferase, GGT: gamma-glutamyl transferase, ALP: alkaline phospatase, T.BIL: total bilirubin, D.BIL: Direct bilirubin, PT: Prothrombin time, Alb: Albumine, AFP: Alpha-fetoprotein, Hb: haemoglobin, Fe: serum iron level, IBC: iron binding capacity, fT3: free triiodothyronine, fT4: free tetraiodothyronine, TSH: tiroid stimulating hormone

Discussion

Hereditary hemochromatosis is an autosomal recessive disorder caused by mutations in the HFE gene on chromosome 6 (1). The outcome of this genetic mutation is excessive absorption of dietary iron leading to parenchymal iron overload and subsequent tissue damage. It typically presents in the 4th and 5th decades, and while phenotypic expression is variable; iron overload in the liver and skin predominate. Hemochromatosis causes hypopituitarism by deposition of iron in the anterior pituitary. After diabetes, hypogonadotrophic hypogonadism is the most common endocrinopathy in hemochromatosis. Recently, an Irish study reported a prevalence of hypogonadism of only 6.4% in its male hemochromatosis patients (2). Transferrin saturation (calculated from the ratio of serum iron concentration to total iron-binding capacity expressed as a percentage) is regarded as the best screening test for hereditary hemochromatosis (3). Liver biopsy is not required for diagnosis in all patients but is obviously useful in assessing disease progression (3,4).

In conclusion, hemochromatosis may rarely cause panhypopituitarism by accumulation of hemosiderin. The clinician should consider pituitary involvement if the state of the consciousness diminishes in patients with cirrhosis due to hemochromatosis. Hepatic encephalopathy might be confused with panhypopituitarism. Additionally, panhypopituitarism may lead to a change in consciousness in the presence of secondary hypothyroidism. Hyponatremia, which may accompany hypothyroidism, may also cause confusion and lethargy. Imaging is usually normal and patients are often wrongly labeled as having idiopathic hypopituitarism. The differential diagnosis should be made; otherwise the treatment might be delayed. Therefore, in consistent with Lewis et al., we recommend that iron studies are performed in all patients who present with hypopituitarism and normal pituitary imaging (5).

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