Endocrinologic aspects of Langerhans Cell Histiocytosis Complicated with Diabetes Insipidus

許薰惠

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摘要

Abstract

Langerhans cell histiocytosis (LCH) is a rare disorder and may be complicated with hypopituitarism and diabetes insipidus (DI) due to invasion of the hypothalamic-pituitary area. In this study, 10 patients with complete (4) and partial (6) type central Dl were found among 125 LCH patients in our hospital records. The water deprivation test, followed by the pitressin test, was performed to confirm DI. Hypothalamic-pituitary endocrine function tests were carried out on these 10 patients at the initial diagnosis and during follow-up. All patients revealed growth hormone insufficiency in the insulin hypoglycemic tolerance test. Four patients had impairment of cortisol secretion, demonstrated by insulin hypoglycemic stimulating test results. Two patients had poor response in the thyrotropin releasing hormone stimulating test. Two patients had only partial responses in the luteinizing hormone releasing hormone test. Four patients had hyperprolactinemia. All patients underwent surgical treatment followed by chemotherapy and/or radiotherapy. One patient completely recovered from the endocrine disorder, 3 patients required smaller doses of desmopressin, and one patient had normal adrenal, thyroid, and gonadal function. Hypothalamic-pituitary disorders in LCH should not be neglected. Treatment of LCH can partially or completely reverse associated endocrine disorders. Therefore, endocrine studies and hormone replacement should be mandatory for patients with LCH.