Central Diabetes Insipidus Caused by Nonspecific Chronic Inflammation of the Hypothalamus: Case Report

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A case of central diabetes insipidus (DI) caused by nonspecific chronic inflammation of the hypothalamus was reported. As the central DI was associated with acute posterior multifocal placoid pigment epitheliopathy with an immunogenic predisposition, and human leukocyte antigen class I antigen A2 and class II antigen DR4 were found, it might be a case of autoimmune reaction.

KEY WORDS: Acute posterior multifocal placoid pigment epitheliopathy; Diabetes insipidus; Human leukocyte antigen; Hypothalamus; Nonspecific chronic inflammation

Several conditions affecting the hypothalamus may lead to central diabetes insipidus (DI). Hoshimaru et al. [4] made the first report of central DI resulting from nonspecific chronic inflammation localized in the neurohypophysial system. We report a case of central DI resulting from nonspecific chronic inflammation of the hypothalamus.

Case Report

A 54-year-old fisherman presented with a 5-year history of polydipsia and polyuria. A diagnosis of idiopathic DI was made, and he was placed on a course of 1-diamino-8-D-arginine vasopressin (DDAVP) in another hospital. Five years later, he became aware of a decrease in left visual acuity. He was referred to our hospital, and a diagnosis of acute posterior multifocal placoid pigment epitheliopathy (APMPPE) of the left eyeball was made. A computed tomography scan showed a small isodense hypothalamic lesion with marked enhancement after intravenous administration of contrast agent (Figure 1). T1-weighted and T2-weighted magnetic resonance images revealed an isointense mass in the hypothalamus, and this lesion was more strongly enhanced in the periphery than in the center after intravenous administration of Gd-DTPA (Figure 2).Treatment with prednisolone was started for the APMPPE. His visual acuity gradually improved, but the treatment had no effect on the hypothalamic lesion. We considered the lesion might be a craniopharyngioma or a glial tumor, and a biopsy was performed using the interhemispheric approach. A whitish-yellow soft mass was identified in the floor of the anterior third ventricle and was biopsied. The specimens showed nonspecific chronic lymphocytic nongranulomatous inflammation (Figure 3). The postoperative course was uneventful, but a daily dose of DDAVP was slightly decreased. He was discharged 1 month after the operation, but he presented with noninsulin-dependent diabetes mellitus despite the release of prednisolone. Therefore, treatment with gliclazide 20 mg daily was begun.

Preoperative endocrinologic studies confirmed slight hyperprolactinemia (47.0 ng/mL) with a minimal response to a thyrotropin-releasing hormone stimulation test, and hypogonadism with a blunted response to administration of luteinizing hormone-releasing hormone. The level of angiotensin converting enzyme (ACE) was normal. A tuberculin test was negative. A chest roentgenogram showed no hilar lymphadenopathy nor abnormal calcification of pulmonary apex. Cerebrospinal fluid examination was not preoperatively performed.

Human leukocyte antigen (HLA) analysis was done, and the HLA class I antigens A2, A26(10), BW59, BW61(40), CW1, and CW3, and the HLA class II antigens DR4 and DRW6 were found. Further immu-
Figure 1. A plain computed tomography scan demonstrating a small isodense hypothalamic lesion (A). The lesion was strongly enhanced (B).

Figure 2. A magnetic resonance (MR) T₁-weighted image showing an isointense mass lesion in the hypothalamus (A). This lesion was more strongly enhanced in the periphery than in the center after intravenous administration of Gd-DTPA (B). An MR T₂-weighted image showing this lesion to be isointense (C).
nologic studies were performed, and antinuclear and antipituitary antibodies were negative.

Discussion
Diabetes insipidus (DI) is divided into two major divisions: primary DI and secondary DI [7]. Primary DI includes familial and idiopathic DI. Secondary DI is the result of damage to the hypothalamic-neurohypophysial system from trauma, neoplasms, vascular disease, infectious disease, or systemic disease including sarcoidosis [7,11]. Hoshimaru et al [4] made the first report of central DI resulting from nonspecific chronic inflammation localized in the neurohypophyseal system, and three of their seven patients were histologically identified. Herein, we reported a patient with central DI of the same etiology—nonspecific chronic inflammation, and the lesion was precisely localized in the hypothalamus. However, the site of the lesion in Hoshimaru’s patients was either the pituitary stalk or the posterior lobe of the pituitary gland. Although we were unable to determine the cause of this nonspecific chronic inflammation in the hypothalamus, it might be a case of autoimmune reaction to the neurohypophyseal system similar to that Scherbaum and Bottazzo [9] reported in the case of autoantibodies to vasopressin-secreting cells of the human hypothalamus in 11 of 30 patients with idiopathic DI.

Tuberculosis and sarcoidosis were excluded from the differential diagnosis as the result of the negative tuberculin test, normal level of ACE, and normal chest roentgenogram. The pathologic finding was not only incompatible with tuberculosis and sarcoidosis but also with histiocytosis X. Lymphocytic hypophysitis and giant-cell granulomatous hypophysitis are well known as inflammatory diseases of the pituitary gland [1,2,8,10]. Several authors have analyzed HLA typing in patients with lymphocytic hypophysitis and supported an autoimmune basis for the disease [2,10]. In our case, the presence of HLA class I antigen A2 and HLA class II antigen DR4, which have been associated with a variety of autoimmune diseases [3,5] is worth noting. On the other hand, it has been suggested that patients with APMPPE may have an immunogenetic predisposition for acquiring this disease because of an increased prevalence of HLA-B7 and HLA-DR2 antigens in these patients [12]. Therefore, we think that the nonspecific chronic inflammation of the hypothalamus in our case was an immunologic response to the neurohypophyseal system.

References


