Unique Female Case of Hypothalamic Hypopituitarism Associated with MRI Abnormalities in the Pituitary Stalk

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INFILTRATIVE, inflammatory or tumorous involvement of the hypothalamic neurohypophysial system results in central diabetes insipidus (DI) and hypopituitarism. Lymphocytic infundibulo-neurohypophysitis [1] is caused by a possible autoimmune process, in which the posterior pituitary is the principal site and the anterior pituitary functions are spared [1, 2]. Necrotizing infundibulohypophysitis involves a combination of DI and hypopituitarism [3]. In contrast, in lymphocytic adenohypophysitis, the secretion of anterior pituitary hormones is impaired but AVP secretion is reserved [4]. The etiology is clearly related to pregnancy or delivery.

In this paper, we report a unique patient who developed hypothalamic hypopituitarism, hyperprolactinemia and masked DI since her delivery. Endocrinological and MRI findings suggest a plausible diagnosis of unique infundibulohypophysitis.

Case Report

A 39-yr-old woman was referred for amenorrhea and galactorrhea since her first delivery at the age of 32 yr. A diffuse goiter and chronic hepatitis were pointed out at 20 and 39 yrs of age, respectively. One and a half years after the delivery she noticed polydipsia and polyuria, which spontaneously subsided after 6 months. At physical examination, her body height and weight were 152 cm and 41 kg, respectively. The pulse rate was 50/min and the blood pressure was 100/60 mmHg. Mild anemia and soft diffuse goiter were also noted. Visual acuity and the visual field were normal.

Plasma IGF-1 (62 ng/ml), free T4 (0.5 ng/dl) and E2 (<10 pg/ml) were low, and urinary excretion of 17-OHCS (1.6 mg/day) and 17-KS (0.8 mg/day) were below the normal ranges. The basal plasma PRL level (32 ng/ml) was increased. The plasma PRL level was further increased by TRH but not by metoclopramide, and was lowered by dopamine infusion or bromocriptine administration. Plasma TSH showed a delayed increase in response to TRH. Both insulin-induced hypoglycemia and arginine infusion failed to stimulate the secretion of GH (Fig. 1, left panel) and ACTH. In contrast, plasma GH and ACTH levels were normally raised by GH-RH (Fig. 1, right panel) and CRH, respectively. Plasma LH and FSH responses to a single injection of LH-RH were blunted but were normalized after repeated LH-RH administration (400 µg/day for 7 days). Plasma AVP levels did not respond either to hypertonic saline infusion (Fig. 2) or water deprivation.

Head MRI revealed marked thickening of the pituitary stalk with a high intensity signal on T1-weighted scanning. The posterior lobe was not evident (Fig. 3). The intensity of the lesion was not enhanced by gadolinium administration.

After replacement with hydrocortisone (20 mg/day), urinary excretion of 17-OHCS was normalized and the plasma TSH level was decreased. Urine volume was doubled with a decrease in urine osmolality.

Discussion

We report a female patient who developed amen-
orrhea and galactorrhea after her first delivery at the age of 32 yr. Endocrinological findings demonstrated hypothalamic panhypopituitarism and hyperprolactinemia. The plasma PRL level was further increased by TRH. In addition, AVP secretion was impaired and DI was apparent after replacement therapy with glucocorticoid. Head MRI revealed marked thickening of the pituitary stalk with a high intensity signal on T1-weighed scanning. This high intensity signal was not changed after two months, suggesting an inflammatory or a lipid-containing lesion.

Hypothalamo-hypophysial dysfunction is caused by a number of diseases including infiltrative, neoplastic and inflammatory etiologies (Table 1). Among them lymphocytic infundibuloneurohypophysitis involves the pituitary stalk and the posterior lobe due an autoimmune process and T-cell infiltration into the tissues [1]. Necrotizing infundibulohypophysitis involves necrotic changes in the pituitary stalk and the anterior pituitary [3].

**Fig. 1.** GH secretion induced by insulin-induced hypoglycemia or arginine infusion (left panel) and by GHRH (right panel).

**Fig. 2.** Plasma AVP levels during hypertonic saline infusion.

**Fig. 3.** Head MRI (T1-weighed scanning).
The pituitary stalk is enlarged and intensively enhanced on contrast MRI in these cases [1, 3]. The present case was characterized by an enlarged pituitary stalk with a very high signal on T1-weighed MRI images which was not enhanced by gadolinium. Our observations suggest a plausible diagnosis of unique infundibulohypophysitis, although a histological examination is required to confirm our speculation.

### References


