Case Report

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Central Diabetes Insipidus Linked to Rathke's Cleft Cyst, Polyuria in a 17-year-old Girl

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A 17-year-old girl presented with polyuria (7 L/day) and polydipsia for one year. Initial urine osmolality was 113 mOsm/kg H_2O. Following 6 h of fluid restriction, serum plasma osmolality reached 300 mOsm/kg H_2O, whereas urine osmolality was 108 mOsm/kg H_2O. Urine osmolality was increased by 427% from 108 to 557 mOsm/kg after vasopressin challenge. The patient was diagnosed with central diabetes insipidus, possibly derived from the atypical occupation of a Rathke's cleft cyst at the pituitary stalk following magnetic resonance imaging with enhancement. She was discharged with desmopressin nasal spray (10 μg); urine output was maintained at 2-3 L/day, and urine osmolality was >300 mOsm/kg. Additional pituitary image studies and evaluation of hypopituitarism should be included in the differential diagnosis of patients with central diabetes insipidus.

Key Words: Central diabetes insipidus, Rathke's cleft cyst, Polyuria

Introduction

Clinical disorders characterized by defective urinary concentrations and a variable degree of polyuria include central diabetes insipidus with arginine vasopressin deficiency, and nephrogenic diabetes insipidus caused by the inability of the kidneys to respond appropriately to arginine vasopressin. According to a study of 79 patients with central diabetes insipidus, the most common cause was idiopathic diabetes insipidus, followed by primary or secondary intracranial tumors or infiltrative diseases such as Langerhans cell histiocytosis, neurosurgery, skull fracture, and autoimmune disease[1,2].

Case Report

A 17-year-old girl (height, 150 cm; weight, 54 kg; body mass index, 24 kg/m^2) presented with polyuria (7 L/day) and polydipsia for one year. She did not have an underlying disease, including diabetes, and denied the use of any medication. Initial urine osmolality was 113 mOsm/kg H_2O, and specific gravity was <1.005. Her vital signs were normal. Laboratory findings were as follows: white blood cells 5,700/mm^3, hemoglobin 13.0 g/dL, and platelets 229 K/mm^3; blood urea nitrogen 8.6 mg/dL and creatinine 0.5 mg/dL; sodium 141 mEq/L, potassium 4.1 mEq/L, and chloride 107 mEq/L; serum osmolality 282 mOsm/kg; and urine sodium 33 mEq/L, potassium 11.1 mEq/L, and chloride 41 mEq/L. Urine analysis revealed no hematuria or proteinuria. Following 6 h of fluid restriction, serum plasma osmolality reached 300 mOsm/kg H_2O, whereas urine osmolality was 108 mOsm/kg H_2O. Fig. 1A shows that urine osmolality was increased by 427% from 108 to 557 mOsm/kg after vasopressin challenge. These remarkable changes in urine osmolality can be used to diagnose cen-

tral diabetes insipidus. Figure (Fig. 2B, C) shows a well-defined hyper signal intensity sellar lesion without an enhanced portion on gadolinium-enhanced fat-suppressed T1-weighted imaging, suggesting Rathke's cleft cyst. Extensive examinations were performed to determine the pathological states of the patient's hypopituitarism (Table 1). Other abnormalities in the release and stimulation of other hormones were not detected. These findings indicate a sole diagnosis of central diabetes insipidus, possibly derived from the atypical occupation of a Rathke's cleft cyst at the pituitary stalk. The patient was discharged with desmopressin nasal spray (10 μg); her urine output was maintained at 2-3 L/day, and urine osmolality was >300 mOsm/kg. She was scheduled for follow-up magnetic resonance imaging (MRI), and pituitary surgery was postponed due to a personal matter.

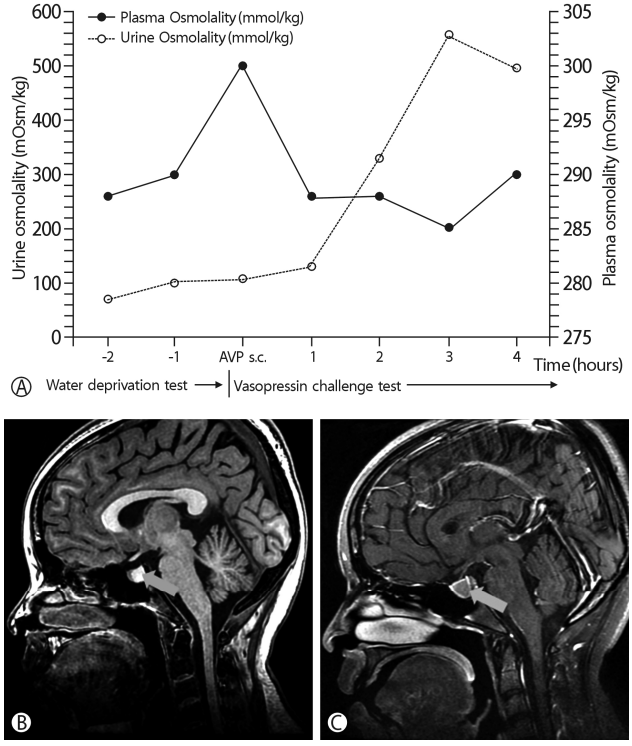

Fig. 1. (**A**) Water deprivation and arginine vasopressin challenge test; (**B**, **C**) Sagittal T1-weighted magnetic resonance (MR) image shows well-defined hypersignal intensity sellar lesion (**B**), without enhancing portion on gadolinium-enhanced fat-suppressed T1-weighted image (**C**).

Discussion

Rathke's cleft cysts are the most common incidentally discovered benign sellar cysts derived from remnants of Rathke's pouch, the same structure from which craniopharyngiomas arise. In MRI, they typically appear as well-demarcated cystic lesions with homogeneous intensity signals and are sometimes combined with thin cyst wall enhancement[3]. Among patients with symptomatic Rathke's cleft cysts, 20% showed panhypopituitarism on admission. The serum prolactin levels did not exceed 200 ng/mL, and hypopituitarism was not correlated with cyst size[4].

We present a case of central diabetes insipidus accompanied by Rathke's cleft cyst. Additional pituitary image studies and evaluation of hypopituitarism should be included in the differential diagnosis of patients with central diabetes insipidus.

Table 1. The endocrinological data of various challenge tests for pituitary hormones

	Basal	30 min	60 min	90 min	120 min	Normal response
Glucose (mg/dL)	38	174	98	89	84	<40 mg/dL in insulin intolerance test
GH (ng/mL)	6.6	8.2	15.5	18.4	10.8	GH peak >5 ng/mL in insulin intolerance test
ACTH (pg/mL)	55	610	359	138	79	20-100 pg/mL in peak level with CRH test
Cortisol (μg/dL)	7.7	15.5	16.5	14.9	14.0	>20 μg/dL or >Δ7 μg/dL with CRH test
TSH (IU/L)	5.7	44.4	46.8	33.1	19.1	Peak TSH ≥2.5-fold with TRH test
Prolactin (ng/mL)	12	140	200	220	139	>2-fold of basal with TRH test
LH (IU/L)	12	67	121	136	153	>Δ10 IU/L (males: >4 × basal; females: >3 × basal) with GnRH test
FSH (IU/L)	7.1	11.5	18.4	23.2	30.9	>Δ2 IU/L with GnRH test

GH, growth hormone; ACTH, adrenocorticotropic hormone; CRH, corticotropic-releasing hormone; TRH, thyrotropin-releasing hormone; TSH, thyroid-stimulating hormone; LH, luteinizing hormone; FSH, follicle-stimulating hormone; GnRH, gonadotropin-releasing horm

Consent

Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

Conflict of Interest: The authors declare that they have no competing interests.

Acknowledgments

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References

1. Kimmel DW, O'Neill BP: Systemic cancer presenting as diabetes insipidus. Clinical and radiographic features of 11 patients with a review of metastatic-induced diabetes insipidus. Cancer 52(12):2355-2358, 1983
2. Maghnie M, Cosi G, Genovese E, et al.: Central diabetes insipidus in children and young adults. N Engl J Med 343(14):998-1007, 2000
3. Bonneville, F. Cattin, F. Marsot-Dupuch, K.Dormont, D. Bonneville, J. F. Chiras, J: T1 signal hyperintensity in the sellar region: spectrum of findings. Radiographics 26(1): 93-113, 2006
4. Zhong W, You C, Jiang S et al. Symptomatic Rathke cleft cyst. J Clin Neurosci 19(4):501-508, 2012