Acute Adrenal Insufficiency Due to Symptomatic Rathke’s Cleft Cyst

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A 65-year-old Japanese man who suffered from secondary hypopituitarism due to Rathke’s cleft cyst is reported. Although computed tomography failed to detect any pituitary abnormality, magnetic resonance imaging demonstrated the presence of a cystic intrasellar mass, initially suggesting craniopharyngioma or abscess. Operative findings revealed Rathke’s cleft cysts within the pituitary fossa which resulted in secondary hypopituitarism. Among cases of secondary hypopituitarism with abnormal findings in the pituitary, symptomatic Rathke’s cleft cysts should be included in the differential diagnosis of adrenal insufficiency.

Key words: hypopituitarism, MR imaging, Rathke, adrenal crisis

Introduction

Rathke’s cleft cysts are usually incidental autopsy findings and are asymptomatic (1–3). Although symptomatic Rathke’s cleft cysts are extremely rare, recent advances in high-resolution computed tomography (CT) and magnetic resonance imaging (MRI) provide the means by which to detect them preoperatively (1–8). Rathke’s cleft cysts may become symptomatic when the size increases and compresses the surrounding tissues: the hypothalamus, hypophysis, optic nerve and chiasma, and third ventricle. The most common presenting symptoms are, therefore, impairment of visual acuity and/or visual field defects, hypopituitarism, and headache (9, 10). We are reporting a case of Rathke’s cleft cyst who had acute adrenal insufficiency and in whom abnormal pituitary findings were detected by MRI preoperatively. Initially craniopharyngioma or pituitary abscess was suggested, however, the post-operative specimens revealed the histological characteristics of Rathke’s cleft cyst. Symptomatic Rathke’s cleft cysts may be more common than suspected. Therefore this possibility must be considered when acute adrenal insufficiency is encountered.

Case Report

A 65-year-old Japanese man was referred to us for evaluation of loss of consciousness on June 13, 1990. He had no familial history of this symptom. On June 7, he had a sudden onset of headache with nausea and vomiting. The following day, high grade fever of 40°C and psychiatric abnormality appeared. On physical examination, his systolic blood pressure was 70 mmHg. His skin was dry and pale with depigmentation of nipples. Body hair was sparse and pubic hair formation was of the female type. He had loss of temporal recession. There was no gynecomastia or galactorrhea. No other neurological focal deficits were observed.

Laboratory investigation revealed decreased serum sodium (118.7 mEq/l [normal 135–147]) and serum chloride (91.1 mEq/l [92–108]). Plasma adrenocorticotropic hormone (ACTH) was less than 10 pg/ml [<50]. Serum growth hormone (GH) was 0.4 ng/ml [0.3–5.0] and serum thyroid-stimulating hormone (TSH) was less than 0.1 mU/ml [0.3–3.2]. Serum prolactin, luteinizing hormone (LH) and follicle-stimulating hormone (FSH) levels were 0.5 ng/ml [1.5–9.7], 0.8 mIU/ml [5–20] and 3.4 mIU/ml [15–30], respectively. Plasma antidiuretic hormone (ADH) was normal (0.8 pg/ml). Serum T3 and T4 levels were 51.0 ng/dl and 5.4 μg/dl, respectively.
Serum cortisol was significantly decreased [0.5 μg/dl (5–15)] and serum testosterone was markedly decreased [53.3 ng/dl (250–1,100)]. Intravenous injection of insulin-induced hypoglycemic test revealed a low response of ACTH with undetectable cortisol levels. Corticotropin-releasing hormone (CRH) loading test demonstrated no response to ACTH. Testing with thyrotropin-releasing hormone (TRH) and LH-releasing hormone (LHRH) showed low responses of TSH, LH and FSH, consistent with deficiencies of anterior pituitary hormones. Rapid ACTH test revealed low response of cortisol (from 0.5 to 5.0 μg/dl). Electrocardiogram (ECG) disclosed low voltage in limb leads and negative T waves in precordial leads.

A plain X-ray of the skull showed a normal sella turcica, and a CT scan including enhancement revealed no detectable abnormalities in intra- or supra-sellar areas.

By MRI, it could be discerned that the intrasellar cystic mass did not elevate the optic chiasma (Fig. 1). The pituitary mass was markedly hyperintense on the T1-weighted image and slightly hyperintense on T2-weighted image. This cystic intrasellar mass was diagnosed as craniopharyngioma or abscess preoperatively, and hydrocortisone therapy (20 mg/day) was begun.

During transsphenoidal surgery, a greenish yellow mucoid mass was found within the intrasellar space and this was quite easily suctioned. The intrasellar space was repeatedly soaked with pure alcohol to prevent recurrence. Histological examination revealed that almost all the surgically removed materials could be stained with HE, PAS and mucicarmine. The capsule was composed of a single layer of ciliated columnar epithelial cells intermingled with goblet cells without atypical cells, which was compatible with a Rathke’s cleft cyst (Fig. 2).

The patient recovered from the operation uneventfully. With hydrocortisone replacement, he has been completely relieved of his symptoms.

**Discussion**

The secondary adrenocortical insufficiency observed in the present case developed as an adrenal crisis. ACTH deficiency occurs in association with multiple pituitary tropic hormone deficiencies. Unlike primary adrenal insufficiency, the patient had no pigmentation and the serum ACTH levels were undetectable, indicating pituitary dysfunction. Over time, with hypopituitarism, the patient gradually worsened, and a sudden attack of adrenal insufficiency was finally encountered. Based on this concealed hypopituitarism, not only sudden changes in the pituitary lesion like cyst abscess (11, 12) or bleeding (13), but also general infection (acute febrile reaction) could occur to compromise this chronic state and trigger acute adrenal insufficiency. The operative and histological findings revealed that the secondary adrenal insufficiency was due to a symptomatic Rathke’s cleft cyst. The majority of sellar or parasellar epithelial cysts are remnants or derivatives of Rathke’s cleft (14). These cysts are usually very small and rarely cause symptoms. However, once they become large enough to compress or destroy the surrounding tissues, the gradual appearance of hypopituitarism is predicted. Before the availability of CT and MRI, the accurate diagnosis of symptomatic or asymptomatic Rathke’s cleft cysts was quite difficult. The MRI characteristics of Rathke’s cleft cysts have been carefully analyzed by Asari et al (4). According to their data MRI findings are influenced by the cystic content of Rathke’s cleft cysts—the so-called cerebrospinal fluid-like mucoid or the presence of blood. The present case demonstrated the presence of greenish yellow mucoid materials in the cyst, which was consistent with the MRI pattern of hyperintensity on T1-weighted and slight hyperintensity on T2-weighted images. Heteroge-
Symptomatic Rathke's cleft cyst

Homogeneous patterns within the hyperintensity in the MRI suggest that the cystic content is often mucoid. However, it is unlikely that MRI alone can reliably distinguish Rathke's cleft cysts from other cystic lesions such as craniopharyngioma, degenerative cystic pituitary adenoma or intrasellar arachnoid cyst. CT could not detect the pituitary lesion in the present case although typical cases of Rathke's cleft cysts usually appear as homogeneous, intrasellar hypodense masses, with or without capsular enhancement (1, 5–8). Despite these limitations, findings on CT and MRI may facilitate an accurate preoperative diagnosis. Rathke's cleft cysts should be, therefore, considered in the differential diagnosis of a patient presenting with acute adrenal insufficiency.

References