Brief Communication

Diabetes insipidus as a presenting manifestation of Rathke's cleft cyst

Manoj Kumar, Deep Dutta, K. S. Shivaprasad, Rajesh Jain, Ankita Sen[1], Dibakar Biswas, Madhumita Mukhopadhyay[1], Sujoy Ghosh, Satinath Mukhopadhyay, Subhankar Chowdhury

Departments of Endocrinology and Metabolism, and [1]Pathology, IPGMER and SSKM Hospital, Kolkata, West Bengal, India

ABSTRACT

Rathke's cleft cysts (RCC) are cystic sellar and suprasellar lesions derived from remnants of Rathke's pouch, lined by cuboidal or columnar epithelium. RCC are usually asymptomatic but can present with headache, visual impairment, panhypopituitarism and hypothalamic dysfunction. Diabetes Insipidus as a presenting symptom of RCC is reported, but rare. We present a case of a 48-year-old male presenting with polyuria and on investigations found to have central diabetes insipidus due to a sellar RCC. Patient underwent transsphenoidal surgery with complete excision with resolution of his symptoms. His polyuria resolved post-surgery without vasopressin replacement, which has never been reported.

Key words: Diabetes insipidus, Rathke cyst, panhypopituitarism

Introduction

The anterior and intermediate lobes of the pituitary gland arise embryologically from Rathke's pouch. Inadequate pouch obliteration results in cysts or cystic remnants at the interface between the anterior and posterior pituitary lobes, found in about 20% of pituitary glands at autopsy.[1] Rathke's cleft cysts (RCC) are usually asymptomatic, but can present with symptoms related to mass effects such as headache, visual impairment, vomitings, features of panhypopituitarism and hypothalamic dysfunction.[1,2]

Case Report

A 68-year-old Indian male patient presented with the complaints of generalized malaise, poor appetite, weight loss, headache and polyuria for last 2 months. He had a chronic ill-looking appearance, a dehydrated tongue and dry skin. Urine output was 5000-5500 mL/day with a history of nocturia and craving for cold fluids. No history of head injury, tubercular meningitis, central nervous system infections or chronic drug intake.

His physical findings on admission were height 156 cm; body weight, 44.5 kg and body mass index, 18.3. Blood pressure was 100/64 mmHg without postural change and pulse rate, 68/min with a regular rhythm. No edema was noted in his legs or feet. He had scant axillary and pubichair. Neurologic examination including visual field was found to be normal.

Laboratory findings were as follows: White blood cell count 6400/mm^3 (neutrophils 51%, lymphocyte 42%, monocyte 5%, eosinophils 2%, basophils 0%); hemoglobin, 13.4 g/dL; hematocrit, 35.5%; red blood cell count, 3.4 × 10^6/mm^3; and platelets, 3.3 × 10^5/mm^3 Serum sodium was 137 mmol/L; potassium, 3.9 mmol/L; and chloride, 103 mmol/L. Blood urea nitrogen was 12 mg/dL; serum creatinine, 0.8 mg/dL; and uric acid, 6.1 mg/dL. Fasting plasma glucose was74 mg/dL; hemoglobin A1c, 4.9% and serum calcium of 9.4 mg/dL.

Anterior pituitary function tests revealed panhypopituitarism (serum cortisol 1.8 μg/dL, Adrenocorticotrophic hormone 15.6 pg/mL, total T3 56 ng/mL, total T4 4.2 μg/dL, thyroid

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Corresponding Author: Dr. Manoj Kumar, Department of Endocrinology and Metabolism, IPGMER and SSKM Hospital, 244 AJC Bose Road, Kolkata - 700 020, West Bengal, India. E-mail: manojkattu@gmail.com

stimulating hormone 1.86 mIU/mL, luteinizing hormone 0.8 mIU/mL, follicle stimulating hormone 1.67 mIU/mL, serum testosterone of 122 ng/dL and serum prolactin of 12 ng/mL. Water deprivation test revealed central diabetes insipidus [Table 1].

Magnetic resonance imaging (MRI) revealed [Figures 1 and 2] a well-defined sellar-suprasellar lesion, which was heterogenous and mildly hyperintense on T1-weighted images and brightly hyperintense on T2-weighted (T2W) images. Internal well-defined hypointense nodule was also seen on T2W images. No obvious post contrast enhancement was seen. Pituitary tissue was compressed and stalk was central. The mass was abutting chiasma, but no obvious mass effect/compression seen. Findings were highly suggestive of RCC.

Transsphenoidal surgery with complete excision of the mass was carried out and histopathological examination revealed cyst wall with columnar lining suggestive of RCC. Patient was started on replacement doses of levothyroxine, hydrocortisone tablets and testosterone injections. His symptoms improved and he was doing well in follow-up. Surprisingly his polyuria improved in postsurgical period without vasopressin replacement.

Table 1: Results of water deprivation test

	Urine osmolality (mosm/kg)	Serum osmolality (mosm/kg)	Urine output
Baseline	234	298	120
1 h	269	305	80
2 h	325	310	50
1 h post-injection vasopressin	573	298	10
2 h post-injection vasopressin	564	297	15

Discussion

RCC arises from the remnants of Rathke's pouch, which is an invagination of the stomodeumand is formed by the fourth gestational week.[3] Rathke's pouch is normally obliterated by the proliferation of the anterior and posteriorlobe of the pituitary gland, which forms a thin residual cleft in the gland. This cleft persists as a cyst lined with columnar or cuboidal epithelium of ectodermal origin. Failure of obliteration with the proliferation of the lining cells with accumulation of secretions may result in cyst formation between anterior and middle lobes. RCC is usually asymptomatic and found in about 20% of normal pituitary glands in autopsy. RCC is an intrasellar cyst containing mucoid material. The most common presenting symptoms are due to mass effect such as headache, visual disturbance. Panhypopituitarism and hypothalamic dysfunction may however, also occur.

The mean age of presentation is 38 years with the highest frequency in the fifth decade and marked female preponderance. Incidence of hypopituitarism varies from 12% to 100% in different studies.[4-6] The most frequent cases involve hyperprolactinemia, followed by gonadotropindeficiency, pan-hypopituitarism, hypothyroidism and hypocortisolism. Our case showed panhypopituitarism without hyperprolactinemia. Diabetes insipidus is thought to be a result of stalk impairment.

It is important to differentiate RCC from other neoplastic lesions, such as craniopharyngioma and pituitary adenoma. Pre-operative diagnosis in most reported cases were pituitary adenoma.

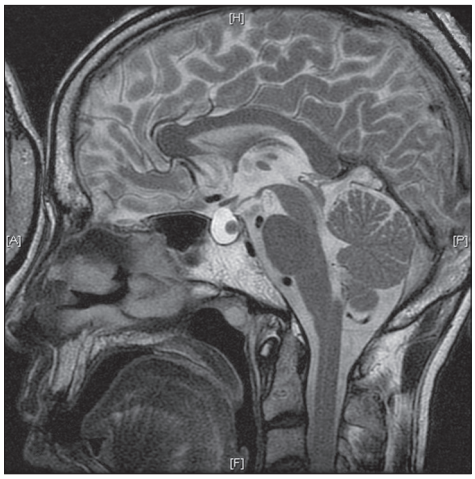

Figure 1: Magnetic resonance imaging pictures well-defined sellar-suprasellar heterogenous lesion and mildly hyperintense on T1-weighted (T1W) images and brightly hyperintense on T2-weighted (T2W) images suggestive of Rathke's cyst

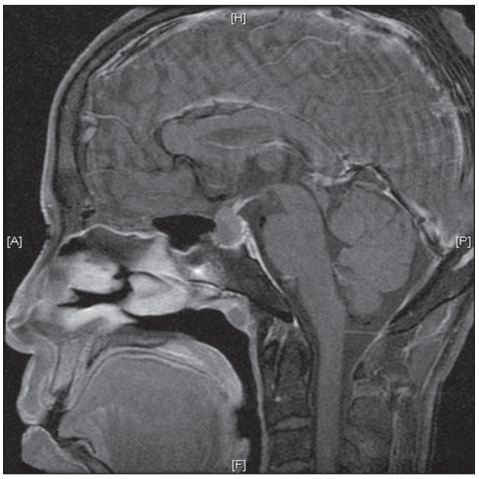

Figure 2: Magnetic resonance imaging pictures well-defined sellar-suprasellar heterogenous lesion and mildly hyperintense on T1-weighted (T1W) images and brightly hyperintense on T2-weighted (T2W) images suggestive of Rathke's cyst

Transsphenoidal surgery is preferred because there is a danger of damage to the hypothalamus and optic apparatus. Surgical treatment is generally recommended even when patient has mild symptoms or signs, increased prolactinor a cyst size of more than 10 mm. In general, the prognosis after a partial removal of the cyst wall or simple aspiration of the cyst seems to be good even though the cyst recurs.

Patients with small-sized RCC or even asymptomatic, should be followed-up regularly with an MRI. Systematic endocrinological examination is recommended once a year.

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