Diabetes Insipidus as Initial Presentation of Rathke's Cleft Cyst

Cheng-Xian Yang, Ming Feng, Kan Deng, Xiao-Hai Liu, Xin-Jie Bao, Ren-Zhi Wang

Department of Neurosurgery, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing 100730, China

To the Editor: A 56-year-old Chinese man was referred to Peking Union Medical College Hospital because of polydipsia (9000 ml/24 h) and polyuria (7000 ml/24 h) for over 20 days accompanied with intermittent moderate headache in frontal and temporal areas of both sides for about 2 weeks. The patient underwent a brain magnetic resonance imaging (MRI) with contrast in a local healthy facility, revealing that there was an irregular pituitary lesion measuring 1.1 cm × 1.4 cm × 2.2 cm. Apparently, the diagnosis of central diabetes insipidus (DI) was made, but the pituitary lesion remained to be elucidated. The patient received desmopressin acetate tablets (1.2 mg, q.n.) to ameliorate polydipsia (5000 ml/24 h) and polyuria (4000 ml/24 h). In our hospital, another brain MRI with enhancement was performed demonstrating a round cystic lesion in sellar region measuring 1.99 cm × 0.84 cm × 1.08 cm. The MRI signal feature was hypo- to iso-intense on T1-weighted images (T1WI) with rim enhancement and iso- to hyper-intense on T2-weighted images (T2WI) [Figure 1]. Moreover, hyperintense signal of pituitary posterior lobe on T1WI was not detected with consistence to the typical MRI signal of central DI. Endocrine evaluations demonstrated no pituitary hormone disorders. The ophthalmic test showed no visual disturbance. The past history of this patient was unremarkable. Taken together, the admission diagnoses of pituitary abscess and Rathke's cleft cyst (RCC) were carefully considered and discriminated before operation. The patient underwent an endoscopic endonasal transsphenoidal surgery. During the procedure, yellowish-white mucous content was seen and drained out. Moreover, white cystic wall was piecemeal resected. Cerebrospinal fluid leakage occurred unexpectedly, and sellar repair and reconstruction were subsequently conducted. In the postoperative period, intracranial infection happened but was controlled by parenteral antibiotics therapy. Desmopressin acetate therapy was then tapered to a maintenance dosage of 0.05 mg for every 8 h. The histopathological findings proved the sellar lesion to be RCC.

RCC commonly remains an asymptomatic sellar entity. Patients with rare symptomatic RCC usually present with headache, visual disturbance, and partial hypopituitarism.[1] However, DI as the initial presentation of RCC is extremely rare. DI usually occurs following pituitary damage, such as trauma, abscess, and hypophysitis. In our case, the lack of hyperintense signal in pituitary posterior lobe on T1WI is observed indicative of the neurohypophyseal dysfunction, verifying central DI firmly. Previous researches demonstrate that pituitary MRI features of RCC are variable according to intracystic content. Rim enhancement with contrast on T1WI is generally considered diagnostic of pituitary abscess but also can be detected in RCC, especially in infected RCC.[2,3] It is difficult to distinguish pituitary abscess and infected RCC according to MRI features alone. Pituitary abscess is rare and usually causes severe DI, hypopituitarism, and headache,[4] whereas the hormone levels were normal in our patient. Thus, infected RCC is a preferred diagnosis which explains both aberrant MRI changes and normal endocrine results. In the procedure, no obvious pus is detected, but surgical

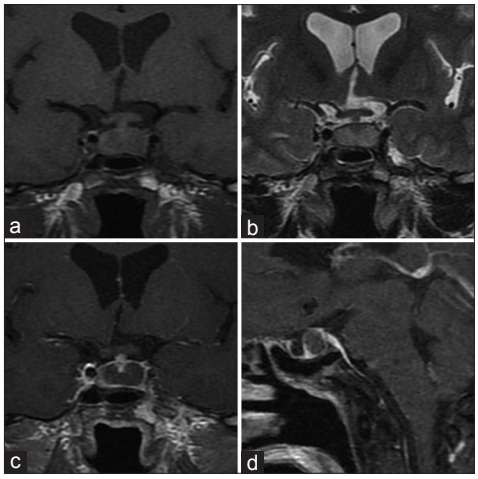

Figure 1: T1-coronal MRI (a). T2-coronal MRI (b). T1-coronal (c) and sagittal (d) MRI with contrast showing rim enhancement of the lesion. MRI: Magnetic resonance imaging.

Address for correspondence: Dr. Xin-Jie Bao, Department of Neurosurgery, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing 100730, China
E-Mail: xinjieabao@163.com

Access this article online

Quick Response Code:

Website: www.cmj.org

DOI: 10.4103/0366-6999.201612

Received: 26-12-2016 **Edited by:** Yuan-Yuan Ji
How to cite this article: Yang CX, Feng M, Deng K, Liu XH, Bao XJ, Wang RZ. Diabetes Insipidus as Initial Presentation of Rathke's Cleft Cyst. Chin Med J 2017;130:751-2.

findings of the content are described as yellowish-white mucous materials. The limitation of our case is that we did not perform bacterial and cytological examinations of the fluid content though results of bacterial culture in infected RCC are mostly negative according to related case reports. Taken together, infected RCC is the most likely diagnosis resulting in DI in our case.

The etiology of infected RCC remains unclear. Patients with infected RCC commonly have no remarkable history of serious infection or immunosuppressive drug intake. The incidence of DI in infected RCC is significantly lower than that of pituitary abscess and hypophysitis. Visual dysfunction and headache are the two most common symptoms in infected RCC. In terms of endocrine results of infected RCC, hyperprolactinemia and partial hypopituitarism stay the most seen endocrine dysfunctions. Pituitary MRI studies demonstrate that infected RCC is usually isointense on T1WI with peripheral enhancement and hyperintense on T2WI. In conclusion, infected RCC is rare and requires comprehensive differential diagnoses.

When infection in sellar region is highly suspected, a transsphenoidal surgery is preferably recommended because of limited infection exposure and minimal invasiveness, but the risk of cerebrospinal fluid leakage should be noticed.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

References

1. Larkin S, Karavitaki N, Ansorge O. Rathke's cleft cyst. Handb Clin Neurol2014;124:255-69. doi: 10.1016/B978-0-444-59602-4.00017-4.
2. Naama O, Gazzaz M, Boulahroud O, Elmoustarchid B. Infection of a Rathke cleft cyst: A rare cause of pituitary abscess. Surg Infect (Larchmt) 2014;15:358-60. doi: 10.1089/sur.2013.069.
3. Famini P, Maya MM, Melmed S. Pituitary magnetic resonance imaging for sellar and parasellar masses: Ten-year experience in 2598 patients. J Clin Endocrinol Metab 2011;96:1633-41. doi: 10.1210/jc.2011-0168.
4. Liu F, Li G, Yao Y, Yang Y, Ma W, Li Y, *et al.* Diagnosis and management of pituitary abscess: Experiences from 33 cases. Clin Endocrinol (Oxf) 2011;74:79-88. doi: 10.1111/j.1365-2265.2010.03890.x.