A Case of Presumably Rathke’s Cleft Cyst Associated with Postoperative Cerebrospinal Fluid Leakage through Persisting Embryonal Infundibular Recess

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Abstract
Persisting embryonal infundibular recess (PEIR) is a rare anomaly of the third ventricular floor. Only eight cases have been published. In this report, a case of presumably Rathke’s cleft cyst associated with cerebrospinal fluid leakage caused by PEIR is described. An 81-year-old woman underwent endoscopic transsphenoidal surgery for the intra- and supra-sellar cystic lesion. Intraoperatively a hole was confirmed over the sella turcica connecting the sellar cyst and the infundibular recess. Liquorrhea did not occur throughout the procedure. A computed tomography (CT) scan obtained immediately after surgery disclosed accumulation of air in the third and lateral ventricles, in addition to the intra- and supra-sellar region. Air accumulation resolved spontaneously after bed rest for 11 days and she was discharged without neurological deficits. However, she required the second transsphenoidal surgery to repair the sellar floor because of bacterial meningitis caused by liquorrhea on the postoperative day 23. A postoperative 3-tesla magnetic resonance image revealed a deep infundibular recess connecting the sella turcica and the third ventricle, which was considered to be PEIR. To the best our knowledge, this is the first reported case describing the intraoperative findings of PEIR.

Key words: Rathke’s cleft cyst, intraventricular air accumulation, persisting embryonal infundibular recess, cerebrospinal fluid leakage

Introduction
Persisting embryonal infundibular recess (PEIR) is a rare anomaly of the third ventricular floor. Eight cases have been reported,1–8 of which 7 cases were accompanied with hydrocephalus, therefore, increased intraventricular pressure is thought to play an important role. In the four reports,2,5,7,8 PEIR was verified by ventriculography, and in the other three reports1,4,6 by magnetic resonance (MR) imaging. We present a case of presumably Rathke’s cleft cyst associated with postoperative cerebrospinal fluid leakage through PEIR not accompanied with hydrocephalus after endoscopic endonasal transsphenoidal surgery.

Case Report
I. Presentation and examination
An 81-year-old woman presented with temporal quadrantanopsia in the left eye. A 3-tesla MR image showed an 18-mm mass in the sella turcica, which had low signal intensity on T1-weighted image and high signal intensity on T2-weighted image. The optic chiasm was slightly elevated by the cystic mass located between the anterior and posterior pituitary lobes (Fig. 1A–C). No connection was observed between the third ventricle and the sellar cyst. The result of the laboratory tests for adrenocorticotropic hormone, follicle stimulating hormone, luteinizing hormone, growth hormone, insulin-like growth factor, thyroid stimulating hormone, free T3, free T4, and cortisol were within normal limits; however, her serum prolactin level was elevated at 43.85 ng/ml (normal range 5.18–26.53 ng/ml). Considering the size of the lesion, this was presumed to be secondary...
to distortion of the pituitary stalk.

II. Operation

The patient underwent a transsphenoidal surgery with both microscope and endoscope. After incision of the dura, the normal pituitary was divided partially. The cyst wall was punctured. The cyst contained viscous and cloudy liquid suggesting a Rathke’s cleft cyst, and the content was aspirated as much as possible. Unfortunately, the histological examination did not carry out because we could not get enough volume of the cyst wall specimen. A hole was identified over the sella turcica probably connecting the sellar cyst and infundibular recess (Fig. 2). There was a thin membrane covering the hole totally and no laceration of the membrane was detected. Liquorrhea did not occur throughout the procedure. The sella turcica base plasty was made with oxidized cellulose, fibrin glue, and bony nasal septum.

III. Postoperative course

Her symptoms disappeared immediately after surgery. A computed tomography (CT) scan obtained immediately after surgery disclosed accumulation of air in the third and lateral ventricles in addition to the intra- and suprasellar region (Fig. 3). She had been confined to bed rest for 11 days although she did not suffer from liquorrhea. Air accumulation resolved spontaneously on CT scan taken on the postoperative day 12 and she was discharged without neurological deficits. There was no pituitary dysfunction after the surgery.

On the postoperative day 23, she was transferred to our hospital with consciousness disturbance and fever. A CT scan demonstrated intracranial air collection not only in the ventricles but also sylvian fissure, and the interhemispheric fissure. The cerebrospinal fluid examinations revealed bacterial meningitis. Antibiotics were given, and the emergency transsphenoidal surgery was performed. The hole between the third ventricle and the sellar cyst was not confirmed during the surgery. The abdominal fat was packed into the sella turcica and the spinal drainage was placed. The patient was discharged without

Fig. 1 Pre- and postoperative sagittal 3-tesla magnetic resonance (MR) images. T₁-weighted (A), Gd-enhanced (B), and T₂-weighted (C) MR images showing the cystic lesion between the anterior and posterior pituitary lobes. The infundibular recess was elevated by the cyst and there was no connection between the third ventricle and the sellar cyst.

Fig. 2 Intraoperative endoscopic view. A small hole (*) existed above the sella turcica. A hole was identified over the sella turcica probably connecting the sellar cyst and infundibular recess. There was a thin membrane (arrowheads) covering the hole totally and no laceration of the membrane was detected. However, the membrane covering the hole might be partially lacerated intraoperatively at the first operation.

Fig. 3 Postoperative computed tomography scan obtained immediately after surgery disclosed air accumulation. Note that air accumulated only in the sellar and suprasellar region, the third ventricle and lateral ventricle.
any neurological deficits 1 month after the second surgery.

On the 14th month after the second operation, a 3-tesla T₁-weighted and T₂-weighted MR image demonstrated empty sella and a deep infundibular recess which connected the third ventricle floor and sella turcica (Fig. 4A, B; arrowheads).

**Discussion**

An infundibulum of the hypothalamus, also called as the pituitary stalk, is composed of two parts: the proximal hollow part and the distal solid part. The cavity of the proximal part of the neurohypophysis originates from the infundibular sac, which develops during the third embryonal week. The cavity of the proximal part of the infundibulum is an extension of the third ventricle and is called as the infundibular recess. The distal part of the infundibulum is obliterated and is referred to as the infundibular stem. By the time the fetus reaches 45 mm in length, the distal part of the infundibular sac is obliterated by cellular proliferation and ultimately differentiates to form the obliterated part of the infundibulum and the posterior pituitary lobe. In some mammals, such as the cat or the bear, the entire embryonal infundibular recess persist extending through the tubular pituitary stalk to the posterior lobe of the pituitary. In contrast, in adult humans and apes the infundibular recess is usually truncated.

The mechanism for developing PEIR is controversial. The authors of previous publications proposed that the increased intraventricular pressure developed PEIR, because of the presence of hydrocephalus in the majority of reported patients with PEIR. On the other hand, Steno et al experienced a case of PEIR without hydrocephalus and proposed the etiology of PEIR is a developmental anomaly caused by lack of obliteration of the distal part of the primary diencephalic evagination. In the present case, PEIR was not accompanied by hydrocephalus, which supports the latter hypothesis. The relationship between PEIR and Rathke’s cleft cyst remains unclear and there is no reported case of PEIR associated with Rathke’s cleft cyst. However, Rathke’s cleft cyst may play an important role to express the symptoms in this case. The distal part of PEIR had been covered by Rathke’s cleft cyst before the first surgery and the Rathke’s cleft cyst shrank during the surgery, which lead to opening of the distal part of PEIR. The hole endoscopically detected at the top of cyst is considered to be PEIR. The membrane covering the hole might be partially lacerated intraoperatively at the first operation. The cerebrospinal fluid did not flow out possibly due to low cerebrospinal pressure. As a ground for thinking low cerebrospinal pressure, the membrane covering the hole was recessed. On the other hand, the entrance may have a check valve system, that is one-way traffic from the sellar cyst to the third ventricle. The air could accumulate in the third ventricle through the hole; however, the cerebrospinal fluid intercepted by the check valve system could not flow out to the sella turcica. The postoperative 3-tesla MR imaging revealed a deep infundibular recess which seemed to connect sella turcica and the third ventricle. If there does not exist PEIR, the accumulation of the air only in the third and lateral ventricles and the intra- and supra-sellar region cannot be explained. As to the treatment of the patient, we made the patient be on strict bed rest for 11 days and confirmed the spontaneous resolution of the air accumulation; however, the patient suffered from cerebrospinal fluid leakage followed by bacterial meningitis. Consequently surgical intervention was required to repair the sellar floor. This means that direct surgical intervention is indispensable to avoid postoperative cerebrospinal fluid leakage whenever we encounter intraventricular air accumulation after the transsphenoidal surgery.

We reported a case of presumably Rathke’s cleft cyst associated with intraventricular air accumulation caused by PEIR following transsphenoidal surgery. Based on our experience, the authors caution that cerebrospinal fluid leakage may occur whenever intraventricular air accumulation is detected on postoperative CT scan. Urgent surgical intervention for repairment of the sellar floor is required to avoid meningitis. This is the first case report demonstrating the endoscopic view of PEIR, and a 3-tesla MR image of PEIR.

**Conflicts of Interest Disclosure**

There is no conflicts of interest in this article.
References


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