

Endoscopic transsphenoidal surgery and Haddad nasoseptal flap of Rathke's cleft cysts

Cirugía endoscópica transesfenoidal y colgajo nasoseptal de Haddad de quistes de la hendidura de Rathke

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SUMMARY

Introduction: Rathke's cleft cysts (RCC) grow formed from remnants of the embryologic Rathke's pouch and are often found incidentally in the sellar or suprasellar region. These lesions are usually asymptomatic in most people. However, as the cyst progressively enlarges, they may develop symptoms typical of RCC, including headache, vision loss, and endocrine dysfunction. The diagnosis procedure involves an MRI scan of the pituitary gland. The endoscopic transsphenoidal approach has become the most common surgical approach to treat RCC. In the last 15 years, endonasal endoscopy has been widely used in the surgical management of these lesions. **Case report:** A 58-year-old Asian woman was reported mainly with a complaint of headache that had increased over the last two months, occasional nausea and vomiting, and blurred vision experienced for 1 month. A contrast brain MRI showed a suprasellar mass consistent with Rathke's cleft cyst. The patient underwent surgery along with an ophthalmologist

to remove the tumor by endoscopic transsphenoidal approach. **Conclusion:** RCC is a benign lesion that causes symptoms in a minority of patients. Headache, vision loss, and endocrine dysfunction are the most common symptoms. Transsphenoidal surgery is the preferred option for patients with symptoms of RCC.

Keywords: Transsphenoidal, endoscopic, Haddad nasoseptal flap, Rathke's cleft cyst.

RESUMEN

Introducción: Los quistes de la hendidura de Rathke (CCR) crecen formados a partir de remanentes de la bolsa embriológica de Rathke, y frecuentemente se encuentran incidentalmente en la región selar o supraselar. Estas lesiones suelen ser asintomáticas en la mayoría de las personas. Sin embargo, a medida que el quiste crece progresivamente, puede desarrollar síntomas típicos del CCR, como dolor de cabeza, pérdida de la visión y disfunción endocrina. El procedimiento de diagnóstico implica una resonancia magnética de la glándula pituitaria. El abordaje endoscópico transesfenoidal se ha convertido en el abordaje quirúrgico más común para tratar el CCR. En los últimos 15 años, la endoscopia endonasal ha sido ampliamente utilizada en el manejo quirúrgico de estas lesiones. **Presentación de caso:** Se reporta que una mujer asiática de 58 años se quejaba principalmente de dolor de cabeza que había aumentado en los últimos dos meses, náuseas y vómitos ocasionales y visión borrosa desde hacía 1 mes. Una resonancia magnética cerebral con contraste mostró una masa supraselar compatible con un quiste de la hendidura de Rathke. La paciente fue intervenida quirúrgicamente junto

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con un oftalmólogo para extirpar el tumor mediante abordaje endoscópico transesfenoidal. Conclusión: El CCR es una lesión benigna que causa síntomas en una minoría de pacientes. El dolor de cabeza, la pérdida de visión y la disfunción endocrina son los síntomas más comunes. La cirugía transesfenoidal es la opción preferida para pacientes con síntomas de RCC.

Palabras clave: *Transesfenoidal, endoscópico, colgajo nasoseptal de Haddad, quiste de la hendidura de Rathke.*

INTRODUCTION

The pituitary gland is a pea-sized oval structure, suspended from the underside of the brain by the pituitary stalk (known as the infundibulum). It sits within a small depression in the sphenoid bone, known as the sella turcica ("Turkish saddle"). Anatomically, the pituitary gland is a "two-in-one" structure consisting of the anterior pituitary (adenohypophysis) and the posterior pituitary (neurohypophysis) (1,2). In the early phase of pituitary gland development, the adenohypophysis is derived from an outpouching of the roof of the pharynx (stomadeum), called Rathke's pouch. It is composed of glandular epithelium and secretes several hormones. Neurohypophysis consists of nervous tissue. It arises from the embryonic forebrain and is, in essence, an extension of the hypothalamus. The location where the anterior and posterior glands meet is called Rathke's pouch (1). In normal conditions, this pouch closes as the cells develop, but it often leaves a mark in the process, which becomes a fissure. Occasionally, the larger marks can become cysts, commonly called Rathke's Cleft Cysts (RCC) (3).

RCC is often asymptomatic. In some cases, the cyst is discovered incidentally when the patient is undergoing Magnetic Resonance Imaging (MRI) examination (4). Large cysts may cause symptoms as they press on the surrounding tissue structures. An enlarged RCC may cause headaches, visual disturbances, and endocrine dysfunction (3,4). Hyperprolactinaemia and growth hormone deficiency are relatively common endocrinological findings associated with RCC, followed by hypocortisolemia and hypogonadism (5).

MRI remains the preferred modality to diagnose RCC and to differentiate RCC from other cystic sellar lesions. For more valid results in diagnosing RCC, it is necessary to rule out other possibilities, such as arachnoid cysts, pituitary adenoma, and craniopharyngioma (4).

The endoscopic transsphenoidal approach has evolved into the preferred surgical approach to treat RCC. This approach removes the tumor without disturbing the brain or optic nerve (6,7). We report a case of Rathke's cleft cyst successfully treated with a transsphenoidal endoscopic approach, according to the surgical case report guidelines.

CASE REPORT

The Neurosurgery Department consulted a 58-year-old Asian woman who presented a main complaint of headache that increased over the last 2 months, increasingly aggravated, with occasional nausea and vomiting, and blurred vision that has worsened since last month (Figure 1). The headache was slightly relieved when taking analgesics, but only temporarily.



Figure 1. The clinical picture of the patient (with the approval of the patient).

ENDOSCOPIC TRANSPHENOIDAL SURGERY

Physical examination revealed no abnormalities. Examination by an ophthalmologist showed an isochor pupil Ø 2/2 mm, +/- light reflex, VOD 6/60, and VOS 2/60. Anterior rhinoscopy, otoscopy, and pharyngoscopy found no abnormalities. A Contrast-enhanced brain MRI showed a well-defined hyperintense mass with regular margins, size $\pm 2.37 \times 2.19 \times 3.26$ cm

suggestive of suprasellar pressing on the chiasma opticum consistent with Rathke's cleft cyst (Figure 2). Laboratory results showed hypothyroidism with decreased FT4 (0.50 ng/dL). Joint surgery with Neurosurgeons and otorhinolaryngologists collaboration in these skull base approaches was planned for cyst removal.

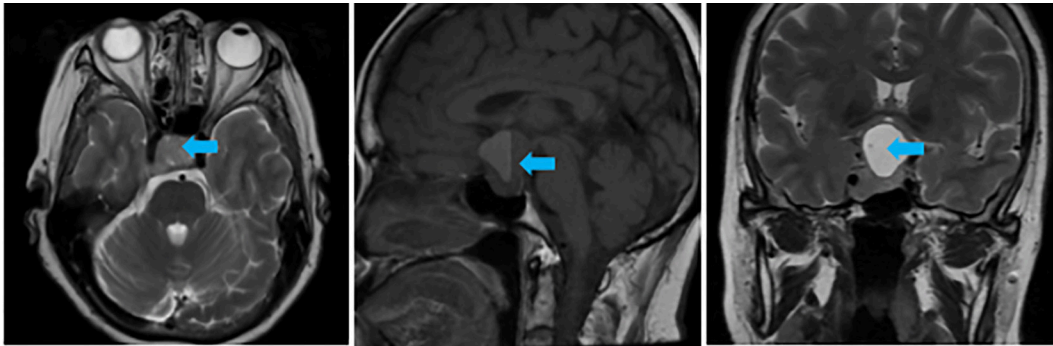


Figure 2. MRI brain with contrast of the patient. Suprasellar mass (blue arrow) pressing the optic chiasma consistent with Rathke cleft cyst. (Images were taken from personal literature).

The surgical procedure was performed under general anesthesia using the Transnasal Endoscopic Surgery technique with Haddad Nasoseptal Flap (HNP) (Figure 3). The sphenoid ostium was identified by cutting the inferior half of the superior concha of the left nasal cavity.

Hadad flap is prepared by making two horizontal incisions on the septum nasi. Free the rostrum from its attachment to the mucosa of the sphenoid sinus, the expulsion of the rostrum, exposing the dura mater and cyst capsule.

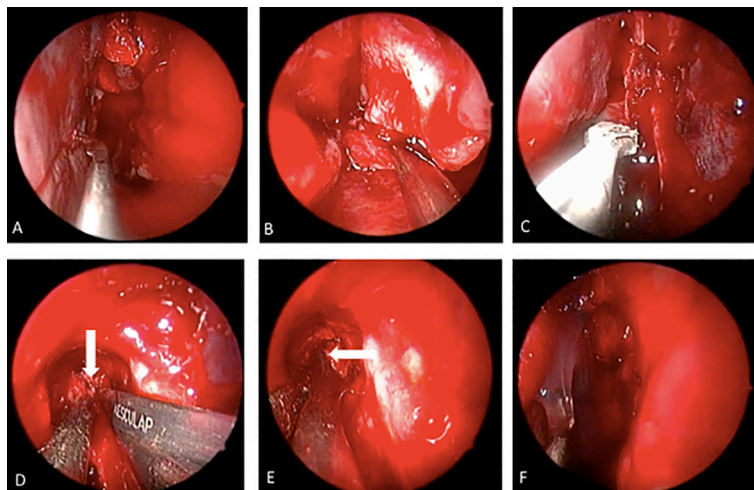


Figure 3. Transsphenoidal endoscopic surgery procedure. (A) Making two horizontal incisions on the septum for the Hadad flap. (B) Elevation of the septal mosaics for the nasoseptal flap. (C) Sculpting the rostrum. (D) opening the pituitary gland layer for visualization of the cyst (white arrow) (E) curettage and debulking the cyst (F) applying fibrin glue to attach the flap. (Images were taken from personal literature).

The patient was admitted to the Intensive Care Unit (ICU) postoperatively. General condition was good. The patient was conscious, no active bleeding from the nasal cavity. The patient received board-spectrum antibiotics and analgesics. The anterior tamponade was removed 3 days later, and the neurosurgeon discharged the patient after being treated for 5 days after surgery.

The patient was controlled for the first time in the outpatient department with complaints of reduced headache and slight pain in the surgical site. The patient submitted the histology results, which showed ciliated columnar epithelium in the characteristics of Rathke's cleft cyst (Figure 4).

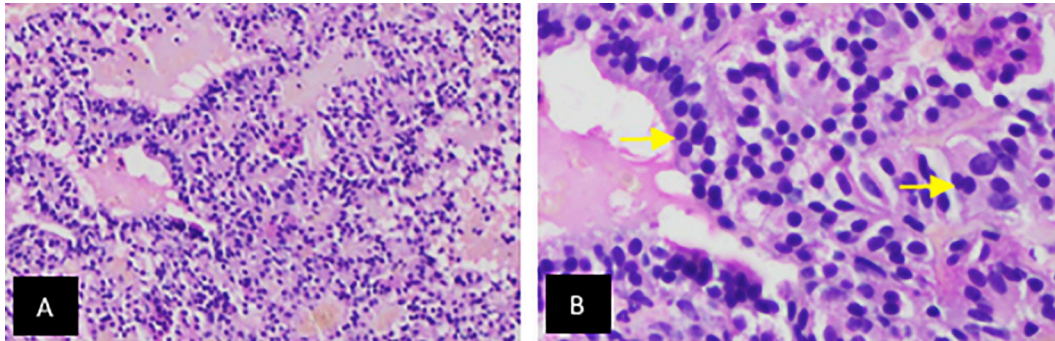


Figure 4. Histology results of the operated tumor. Hematoxylin and eosin staining at (A) 20x and (B) 100x show a ciliated columnar epithelium (yellow arrow). (Images were taken from personal literature).

DISCUSSION

Rathke's cleft cysts (RCCs) are relatively common, benign, nonneoplastic, and intra- and suprasellar lesions originating from the remnant of Rathke's pouch lesions composed typically of a thin cyst wall enclosing a mucous, gelatinous, or caseous liquid core (6). Most RCCs are small and asymptomatic, with an incidence of up to 22 % on routine autopsies (7). A few may grow large enough to cause the tumor mass pressure on the surrounding tissue structures and cause significant pituitary dysfunction, including temporary and permanent diabetes insipidus (DI), visual field deficit, and headache, particularly frontal episodic headache. Cysts are sometimes found for different reasons when patients perform MRIs (4).

Symptoms of RCC usually occur in the 4th or 5th decade of life, with a slightly higher prevalence in women (3). Blurred vision is caused when the cyst grows into the brain cavity, pressing on

the optic nerve (3,4). Increased pressure on the normal gland also can cause hypopituitarism (5). Symptoms of hypopituitarism include loss of appetite, weight loss, fatigue, decreased energy, decreased mental function, and dizziness (10). This 58-year-old Asian woman presented a main complaint of headache that increased over the last 2 months, with occasional nausea and vomiting, and blurred vision.

All patients with cystic sellar lesions should undergo a multidisciplinary preoperative evaluation consisting of appropriate ophthalmological, endocrinological, and neuroimaging studies (3,5). Laboratory examinations include serum levels of prolactin, thyroid function, adrenocorticotrophic hormone (ACTH), and insulin-like growth factor-1 (IGF-1) (5).

MRI remains the primary modality in diagnosing RCC and differentiating RCC from other cystic sellar lesions. On MRI images, RCC often appears as round or ovoid lesions located in the sellar region. The intensity of the cystic contents on contrast-enhanced MRI images

shows high variability on T1 and T2 sequences and correlates with the nature of the cystic contents (4). A Contrast-enhanced brain MRI of the patient showed a well-defined hyperintense mass with regular margins, size $\pm 2.37 \times 2.19 \times 3.26$ cm suggestive of suprasellar pressing on the chiasma opticum consistent with Rathke's cleft cyst (Figure 2).

The gold standard for confirming the diagnosis of RCC is histopathological analysis. Hematoxylin and eosin analysis usually shows a simple columnar or cuboidal epithelium, often with ciliated or mucinous goblet cells (11). Pseudostratified columnar cells are also frequently observed. In this case, it was found to be ciliated columnar epithelium, as seen in Figure 4.

These lesions are usually asymptomatic in the majority of people and do not require surgical management and can be monitored using periodic imaging evaluation (3). However, if progressive symptoms occur, such as persistent headaches, visual disturbances, and underlying laboratory evidence of endocrine dysfunction, surgery is the main treatment option (6,7). In this patient, there were symptoms of persistent headache and visual disturbances that had worsened over the past 2 months. Therefore, surgery was the best option for the patient.

There are two surgical approaches: transphenoidal endonasal surgery and transcranial surgery (craniotomy) (4). Transsphenoidal endonasal surgery is preferred for treating the pituitary gland (6,7). The advantages of endonasal surgery over craniotomy surgery include a minimally invasive approach, anatomically more direct targeting, no facial scarring, less brain and neurovascular structures trauma, faster devascularisation of the blood supply to the tumor, and good visualization of relevant anatomical structures (6). We, therefore, decided to perform endoscopic transsphenoidal surgery on this patient.

Nie et al. (13) compared the clinical outcomes of transcranial and endoscopic endonasal surgery (EES) in skull base surgery. Of the total 273 patients studied from this retrospective study, The EES group had a greater gross total resection (GTR) rate (89.8 % EES vs. 77.3 % TCS) and a lower rate of hypopituitarism (53.4 % EES vs. 68.1 % TCS). More patients in the EES group

with preoperative visual deficits experienced improvement after surgery (74.5 % EES vs. 56.3 % TCS). However, Salama (12) believes that a transcranial approach is still needed in some cases of large pituitary adenomas and significant expansion laterally, anteriorly, or superiorly, as well as in patients with recurrence after previously undergoing a transsphenoidal approach (14).

Closing skull base defects with an endoscopic approach using a nasoseptal flap (also known as Hadad-Bassagasteguy flap) is an option. The Hadad-Bassagasteguy flap is a vascularised pedicle flap of the mucoperiosteum and mucoperichondrium of the septum nasi blooded by the nasoseptal artery, a terminal branch of the internal maxillary artery (15).

CONCLUSIONS

Rathke's cleft cysts are benign that cause symptoms in a small proportion of patients. Headache, vision loss, and endocrine dysfunction are the most common complaints. Surgery with a transsphenoidal endoscopic approach is the first choice in RCC cases. It offers satisfactory results that correlate well with symptomatic improvement, improved pituitary function, and minimal complications.

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