

# Delayed CSF Leak with Encephalocele After Rathke's Cleft Cyst Fenestration

Zachary Christian, MD<sup>1</sup>; Kevin L Li, MD<sup>1</sup>; Tran Locke, MD<sup>1</sup>; Meha Fox, MD<sup>1</sup>; William Yao, MD<sup>2</sup>; Ali Jalali, MD,PhD<sup>3</sup>; K. Kelly Gallagher, MD<sup>1,3</sup>

<sup>1</sup>Baylor College of Medicine- Otolaryngology Head & Neck Surgery, <sup>2</sup>University of Texas Houston- Otolaryngology Head & Neck Surgery, <sup>3</sup>Baylor College of Medicine Neurosurgery

# Baylor College of Medicine

# Abstract

- A case of a delayed cerebrospinal fluid (CSF) leak with an encephalocele after Rathke's cleft cyst fenestration has not been reported in the literature.
- Here we present a patient who had a CSF leak with an encephalocele 16 months after a Rathke's cleft cyst fenestration
- Awareness of this delayed postoperative complication informs for close post-operative surveillance

# Introduction

### Images



- Endoscopic cyst fenestration and drainage is the most common surgical treatment for Rathke's cleft cysts [1]
- Rates of postoperative CSF leak range from 0-21% [2-21]
- Most cases present within 3 months of surgery
- One case of delayed CSF leak (209 months) has been reported [2]
- No prior cases of delayed CSF leaks with an encephalocele.
- Here we present a patient with a CSF leak associated with an encephalocele 16 months after Rathke's cleft cyst fenestration.

# **Case Description**

A 28-year-old female initially presented in 2016 with primary amenorrhea. She was noted to have hypothyroidism and mullerian agenesis, but further workup revealed a 2.4 cm cystic sellar mass on MR imaging (Figure 1). She was otherwise asymptomatic, so she opted for observation with serial imaging. Surveillance imaging 4 years later revealed that the mass grew to 4.9 cm in size, causing mass effect on optic chiasm (Figure 2). She underwent an endoscopic endonasal approach to fenestrate and drain the cyst in 2022. Biopsy of the cyst wall was consistent with Rathke's cleft cyst. Because there was no intraoperative CSF leak, the dural defect to the sphenoid sinus was left open to reduce the risk of re-accumulation. She had no evidence of CSF leak at 6 weeks postoperatively, but she was lost to follow up thereafter. 16 months later, she presented with persistent clear rhinorrhea and headache. MR images revealed herniation of basal brain structures through the sellar defect at the prior surgical site (Figure 3). She underwent expanded endoscopic endonasal approach to reduce the encephalocele. The defect was repaired with an abdominal fat graft and a nasal septal flap. At her first postoperative visit 3 weeks later, the nasal cavity was debrided. No CSF leak was noted. She continued to do well at her 3-month postoperative visit.

Figure 1: Sagittal MRI demonstrating 2.6 cm sellar/suprasellar cystic mass in 2016



Figure 2: Sagittal MRI demonstrating a Rathke's Cleft Cyst that grew to be 4.9 cm in size in 2022



#### **Table 1.** Review of Postoperative CSF Leak Rates and Time to CSF Leak Postoperatively by Author

Author	Postop CSF leak rate	Time to CSF leak
Alsavaf[3]	(0/45) 0%	Not applicable
Ozawa[6]	(0/14) 0%	Not applicable
Arko[4]	(0/30) 0%	Not applicable
Mendelson [10]	(0/15) 0%	Not applicable
Madhok[12]	(0/35) 0%	Not applicable
Kuan[15]	(0/7) 0%	Not applicable
Barkhoudarian[16]	(0/36) 0%	Not applicable
Yamada[14]	(1/27) 3.7%	Not reported
Jiang [21]	(1/13) 7.7%	Not reported
Nakase[7]	(2/35) 5.7%	Not reported
Millesi[17]	(2/39) 5%	Not reported
Park[8]	(2/73) 2.7%	Not reported
Frank[13]	2/22 (9.1%)	Not reported
Seo[5]	(2/32) 6.3%	Not reported
Solari[9]	(2/29) 6.9%	POD5
Xie[11]	3/23 (13.0%)	Not reported
Marcus[19]	13/61 (21.3%)	POD 2-29

Figure 3: Sagittal MRI demonstrating CSF leak with basal brain herniation through the sellar defect

### Discussion

- Cyst marsupialization has a higher risk of cyst re-accumulation, but lower risk of CSF leak when compared to total cyst resection [1]
- No correlation between rates of intraoperative and postoperative CSF leaks [1]
- Patients can present with meningitis[2] or other complications in a delayed fashion
- Important to have ongoing follow-up after surgery

## Conclusions

- A postoperative CSF leak is an uncommon complication of endoscopic transsphenoidal surgery for Rathke's cleft cysts.
- Clinicians must be aware of the risk of developing an encephalocele through the unrepaired dural defect in order to better counsel patients preoperatively and advocate for close postoperative surveillance.
- Abdominal fat grafts with a nasoseptal flap are reasonable repair options in these cases.

# Contact

### Zachary Christian Baylor College of Medicine One Baylor Plaza, NA 102, Houston TX 77030 Zachary.Christian@bcm.edu 214-960-6621

### References

1.	Madapoosi, A., et al., Rathke's cleft cyst marsupialization and repair with a free mucosal graft - Video case report and literature review. Am J Otolaryngol, 2022. 43(5): p. 103519.	
2.	Lee, J.J., et al., Delayed Cerebrospinal Fluid Leakage After Treatment of Skull Base Tumors: Case Series of 9 Patients. World Neurosurg, 2019. 132: p. e591-e598.	
3.	Alsavaf, M.B., et al., Endoscopic endonasal marsupialization of Rathke cleft cysts: clinical outcomes and risk factors analysis of visual impairment, pituitary dysfunction, and CSF leak. Pituitary, 2023. 26(6): p. 696-707.	
4.	Arko, L.t., et al., Endonasal Endoscopic Fenestration of Rathke's Cleft Cysts: Whether to Leave the Fenestration Open or Closed? J Neurol Surg B Skull Base, 2021. 82(Suppl 3): p. e101-e104.	
5.	Seo, S.H., et al., Surgical Management and Long-Term Results of Rathke's Cleft Cyst. J Korean Neurosurg Soc, 2023. 66(1): p. 82-89.	
6.	Ozawa, H., et al., Risk factor for cerebrospinal fluid leak after endoscopic endonasal skull base surgery: a single-center experience. Acta Otolaryngol, 2021. 141(6): p. 621-625.	
7.	Nakase, K., et al., Long-term outcomes and potential predictive recurrence factors after endonasal endoscopic surgical treatment of symptomatic Rathke's cleft cysts. Neurosurg Rev, 2024. 47(1): p. 85.	
8.	Park, J.K., E.J. Lee, and S.H. Kim, Optimal surgical approaches for Rathke cleft cyst with consideration of endocrine function. Neurosurgery, 2012. 70(2 Suppl Operative): p. 250-6; discussion 256-7.	
9.	Solari, D., et al., Endoscopic Endonasal Approach in the Management of Rathke's Cleft Cysts. PLoS One, 2015. 10(10): p. e0139609.	
10.	Mendelson, Z.S., et al., Endoscopic transsphenoidal surgery of Rathke's cleft cyst. J Clin Neurosci, 2015. 22(1): p. 149-54.	
11.	Xie, T., et al., Endoscopic endonasal resection of symptomatic Rathke cleft cysts. J Clin Neurosci, 2011. 18(6): p. 760-2.	
12.	Madhok, R., et al., Endoscopic endonasal resection of Rathke cleft cysts: clinical outcomes and surgical nuances. J Neurosurg, 2010. 112(6): p. 1333-9.	
13.	Frank, G., et al., Transsphenoidal endoscopic approach in the treatment of Rathke's cleft cyst. Neurosurgery, 2005. 56(1): p. 124-8; discussion 129.	
14.	Yamada, H., et al., Long-Term Outcomes of Endoscopic Cyst Fenestration for Rathke Cleft Cyst. World Neurosurg, 2022. 161: p. e282-e288.	
15.	Kuan, E.C., et al., Preventing Restenosis of Marsupialized Rathke Cleft Cysts Using a Nasoseptal Flap Lining. Laryngoscope, 2019. 129(10): p. 2258-2261.	
16.	Barkhoudarian, G., et al., Rathke's cleft cysts: a 6-year experience of surgery vs. observation with comparative volumetric analysis. Pituitary, 2019. 22(4): p. 362-371.	
17.	Millesi, M., et al., Endoscopic Treatment of Rathke's Cleft Cysts: The Case for Simple Fenestration. Brain Sci, 2022. 12(11).	
18.	Lee, J.A., et al., Endonasal Endoscopic Surgery for Pediatric Sellar and Suprasellar Lesions: A Systematic Review and Meta-analysis. Otolaryngol Head Neck Surg, 2020. 163(2): p. 284-292.	
19.	Marcus, H.J., et al., Rathke's cleft cysts following transsphenoidal surgery: long-term outcomes and development of an optimal follow-up strategy. Acta Neurochir (Wien), 2020. 162(4): p. 853-861.	
20.	Fraser, S., et al., Risk factors associated with postoperative cerebrospinal fluid leak after endoscopic endonasal skull base surgery. J Neurosurg, 2018. 128(4): p. 1066-1071.	
21.	Jiang Z, Yu M, Jiang Y, Peng Y. Endoscopic endonasal resection of symptomatic Rathke cleft cysts: clinical outcomes and prognosis. Neurosurg Rev. 2019 Sep;42(3):699-704. doi: 10.1007/s10143-018-01058-0. Epub 2018 D	
	10. PMID: 30535966 © Poster Template by Genigraphics® 1.800.790.4001 v	