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Case Report

Endoscopic Endonasal Surgery for Suprasellar Rathke's Cleft Cyst Mimicking a Dermoid Cyst

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Summary

The differential diagnoses of cystic epithelial masses in the sellar and parasellar regions, when based on clinical findings, imaging, and even histopathological examination, can be challenging. Considerable evidence in the literature supports the existence of a common ectodermal origin of selected sellar and suprasellar cysts, which may account for the overlap of radiological features and pathological transitional states observed among these lesions. Here, we describe a case of suprasellar Rathke's cleft cyst (RCC) mimicking a dermoid cyst, which after successful removal by endoscopic endonasal surgery (EES) by an experienced team of neurosurgeons and otolaryngologists achieved a good clinical outcome. A 30-year-old male was referred with chief complaints of left-sided loss of vision and headache. Magnetic resonance imaging findings indicated possible diagnoses as RCC, craniopharyngioma, or dermoid cyst because the cyst partly contained fluid with a lipid signal. The cyst was resected en-bloc with EES, and the histopathological diagnosis was RCC. RCCs may contain lipid components within the cyst, presumed to be cholesterin crystals due to chronic inflammation. In preoperative imaging of suprasellar cystic lesions, the presence of lipid components in the cyst may not indicate dermoid cyst, and care should be taken to differentiate it from RCC or craniopharyngioma.

Key Words: dermoid cyst, endoscopic endonasal surgery, Rathke's cleft cyst, suprasellar cyst

Introduction

Rathke's cleft cysts (RCCs) arise from the embryonic remnants of the Rathke's pouch and usually manifest as headaches, visual impairment, and endocrine disturbances[1-11)]. It may be difficult to definitively diagnose cystic epithelial masses of the sellar and parasellar regions, based on clinical findings or investigations, including imaging and histopathological examinations. RCCs are often associated with chronic inflammation and squamous epithelialization of the cyst wall, resulting in imaging findings such as wall thickening, contrast enhancement, and heterogeneity of content. Furthermore, Craniopharyngiomas and dermoid cysts are often associated with inflammation and degeneration. However, imaging findings are also atypical in these cases. Therefore, the differential diagnosis of suprasellar cystic lesions modified by inflammation remains dif-

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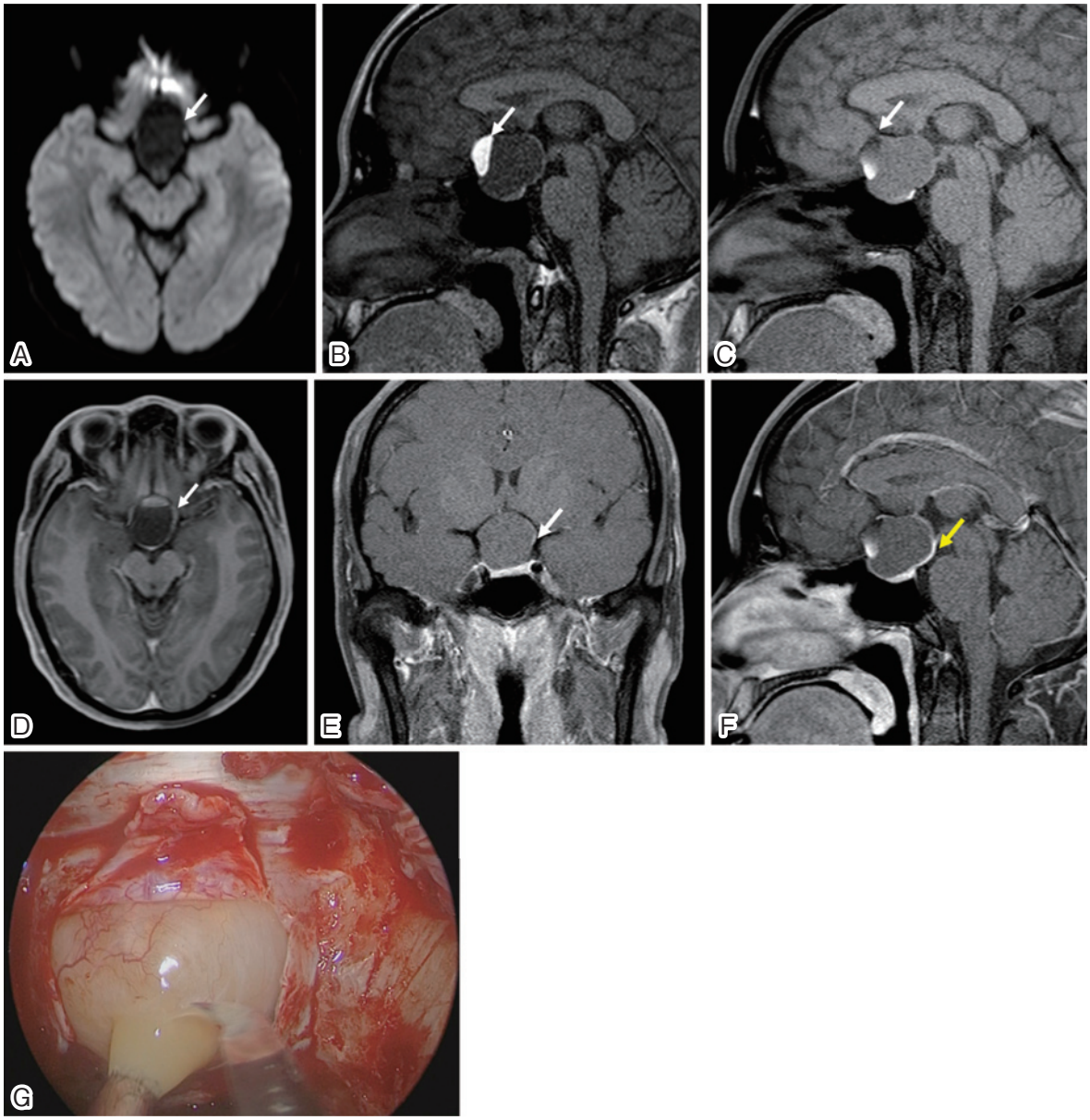

Figure 1 Preoperative magnetic resonance imaging (MRI) and intraoperative endoscopic view

A: Axial view of the diffusion-weighted image reveals a low-signal lesion in the sella and suprasellar area (arrow).

B: Sagittal view of a T1-weighted image shows a cyst with a high-intensity signal in the anterior component of the suprasellar lesion (arrow) and a low-intensity signal in the posterior component.

C: Sagittal view of a T1-weighted image with fat suppression shows a partially fat-suppressed anterior component of cyst (arrow). This suggests that part of the cyst content fluid is lipid.

D, E, F: Axial **(D)**, coronal **(E)**, and sagittal **(F)** views of the T1-gadolinium weighted image with fat suppression showing both the cyst wall with very slight contrast (white arrow) and the pituitary stalk (yellow arrow).

G: On incising the cyst wall under a 30-degree endoscope, the pale, yellowish-white creamy contents were easily aspirated.

ficult. In fact, RCCs with strong squamous epithelialization and ciliated craniopharyngiomas overlap histologically and are difficult to differentiate on imaging[1-8]. This study is the first report of RCCs containing fatty components inside the cysts, mimicking dermoid cysts. Furthermore, we successfully performed a total resection with preservation of endocrine function using endoscopic endonasal surgery (EES) and report on the clinical course of our case.

Case Presentation

A 30-year-old male with no significant medical his-

tory was referred to our hospital with chief complaints of unilateral loss of vision (left) and headache. Magnetic resonance imaging (MRI) demonstrated mainly the suprasellar cystic component of the causative lesion, indicated by the fluid level observed with iso- and high-intensity signals on T1- and T2-weighted imaging, respectively (Fig. 1A-F). Further, the anterior portion of the cyst demonstrated a high-intensity signal on T1-weighted imaging and was suppressed on T1-fat suppression imaging. This supported the presence of fatty droplets within, thereby suggesting a differential diagnosis of RCC or dermoid cyst. The cystic membrane was thinly enhanced on T1-weighted gadolinium scans. On computed tomography (CT), the anterior portion of the cyst also showed a low density, suggestive of fatty components (data not shown). Ophthalmologic examination findings included a marked left visual loss (corrected visual acuity; right/left: 1.2/0.08), left temporal hemianopia, and expansion of a right scotoma. The anterior pituitary function was normal, based on hematological tests. Clinical examination revealed no other neurological deficits. Suprasellar RCC, dermoid cyst, or craniopharyngioma were suspected based on the clinical course and characteristic imaging findings. We elected to perform EES by resecting the suprasellar cyst. Thereby enabling histopathological diagnosis and relieving the patient's symptoms.

Under general anesthesia, a lumbar spinal drain was inserted to prevent postoperative cerebrospinal fluid (CSF) leakage. An experienced team of neurosurgeons and otolaryngologists performed EES by means of a transsphenoidal-transplanum-transtuberculum approach, using both neuro-navigation and visual evoked potential (VEP) monitoring. We performed bilateral posterior ethmoidectomy through the superior meatus, followed by a wide sphenoidotomy, to access and secure the surgical working space, as described previously[12]. We then incised and deviated laterally the inferior halves of bilateral superior turbinates. A mucosal flap was prepared using mucosa on the posterior wall of the sphenoid sinus, and was preserved in the clival recess[13]. The dura mater was exposed after drilling of the bilateral optico-carotid recess, floor of the sella, and planum sphenoidale. After we reversed the dura mater through a U-shaped incision, the cyst wall was identified just behind the dura. On incising the cyst wall under a 30-degree endoscope, the pale, yellowish-white creamy contents were easily aspirated (Fig. 1G), thus draining the cyst in situ. This was presumed to be the cystic content fluid containing fat droplets, suspected preoperatively. We then identified a normal pituitary gland caudal to the cyst, including the contiguous pituitary stalk pressurized by the tumor at the suprasellar cistern. The optic nerve, along with bilateral internal carotid arteries (ICAs), anterior cerebral arteries (ACA), and anterior communicating artery (A-com), were clearly visible through the cyst. The tumor capsule was carefully detached from bilateral ICAs and the superior hypophyseal artery (SHA). Only the SHA branches that fed the cyst wall were coagulated. The capsule was then carefully detached from the surrounding neurovascular structures, after identifying visible boundaries of the cyst limbus. Finally, we carefully dissected through the adhesion tethering the cyst to the upper part of the pituitary stalk, then completely excised the cystic capsule. A red nodule found within the cyst, was also removed from the cyst wall. No changes were observed in the VEP intraoperatively.

Dural reconstruction was performed using the fascia lata as an inlay graft sutured with 6-0 PROLENE® (Ethicon, Tokyo, Japan), and fixed with fibrin glue. The reflected dura mater was repositioned and fixed with suturing. The site was covered with a vascularized sphenoid sinus mucosal flap, which was fixed with fibrin glue, and gently buttressed using Sorbsan® dressing (Alcare, Tokyo, Japan).

The spinal drain was removed on POD 1. On POD 2, the patient was diagnosed with meningitis and administered antibiotic treatment, although fiberoptic otolaryngoscopy showed no signs of CSF rhinorrhea or sinusitis. No other complications, including neurological deficit, anterior pituitary dysfunction, or diabetes insipidus, were encountered postoperatively. MRI (Fig. 2A, B) performed on POD 7 confirmed the complete excision of the cyst. His visual loss and visual field defect resolved on POD 7. The nasal packing was removed at 7 days after the EES, and he was discharged on POD 17 after reconfirming that the meningitis had been resolved, and that there was no late CSF leakage. At the 18-month postoperative outpatient follow-up visit, the patient's vision and endocrine function were

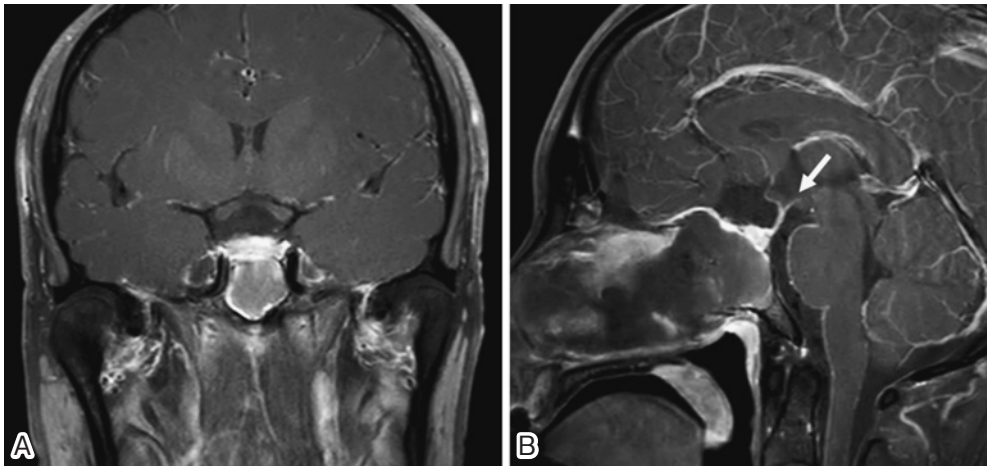

Figure 2 Postoperative magnetic resonance imaging scans
A, B: Coronal **(A)**, and sagittal **(B)** views on T1-weighted gadolinium imaging confirm complete removal of the cyst with pituitary stalk preservation (arrow).

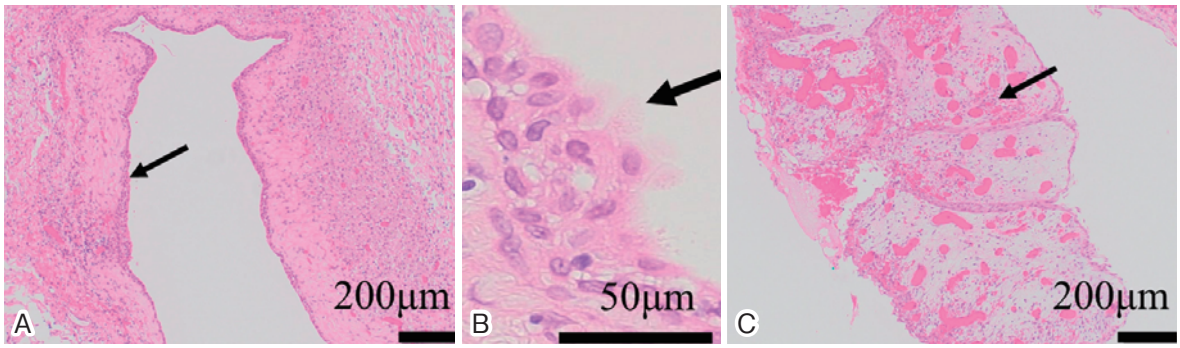

Figure 3 Pathological findings
The pathological diagnosis was Rathke's cleft cyst. **(A)** Histologically, cyst walls covered with monolayer or multilayered squamous (arrow) and cylindrical epithelium were observed (bar 200 μm). **(B)** Some epithelial cells had cilia (arrow) and contained mucus (bar 50 μm). **(C)** Partially noticeable inflammatory cell infiltrates, epithelial shedding, and reactive changes with increased capillary growth are observed (arrow) (bar 200 μm).
(**A**, low-power field; **B**, **C**, high-power field; hematoxylin and eosin)

normal, and MRI showed no recurrence of the cyst.

Histologically, the cyst walls were composed of monolayer or multilayered squamous and cylindrical epithelium, compatible with RCC with squamous metaplasia (Fig. 3). Some epithelial cells were ciliated and contained mucus. Partially noticeable inflammatory cell infiltrates, epithelial shedding, and reactive changes with increased capillary growth were observed.

Discussion

In this case, the preoperative diagnosis was difficult because there were findings suggestive of a dermoid cyst, with fat droplets found within the cyst. To the best of our knowledge, this is the first such case report to document the application of EES for radical resection of an entirely suprasellar RCC, mimicking a dermoid cyst.

RCCs arise from benign remnants of the Rathke pouch, typically in the sella, and may sometimes have suprasellar extensions[1,2]. Sanno N, et al. reported that 5.3% of cysts were enlarged, 15.9% were shrunken, and 76.5% were unchanged in the study of 139 RCCs with a mean follow-up period of 26.9 months. In general, surgical treatment is indicated in symptomatic cases, especially in those with obvious visual dysfunction[3]. Entirely suprasellar RCCs are challenging to resect[1] because of their proximity to the optic chiasma and to the pituitary infundibulum. RCCs with a suprasellar

component, as compared to their sellar counterparts, commonly cause visual dysfunction, are more difficult to extract completely (38%-55%), recur more frequently (16%-29%), and are associated with higher postoperative endocrine morbidity. Preoperative symptoms, including visual dysfunction and headache, are less likely to improve with surgery[1,2]. These factors must be considered during the treatment of an RCC with a suprasellar component. In our patient, we considered a dermoid cyst[2-7] and craniopharyngioma[2,6,8,10-12,15] in addition to RCC as a part of the preoperative differential diagnosis, and therefore attempted complete cyst resection to prevent recurrence.

We note that EES performed through the transsphenoidal-transplanum-transtuberculum approach[8-12] by an experienced team of neurosurgeons and otolaryngologists contributed to its success. The effectiveness of the procedure was supported by correction of the patient's visual impairment and by the lack of postoperative complications, including transient or permanent diabetes insipidus, postoperative pituitary dysfunction, or neurological deficits. No recurrence was detected in a limited evaluation at the short-term follow-up. In any event, careful monitoring with long-term follow-up is necessary.

Results of animal studies and reports of transitional cystic epithelial masses or lesions that crossover from typical to more aggressive pathological subtypes have collectively provided a solid foundation for the theory that selected sellar and suprasellar cystic lesions may have a common ectodermal origin[5]. Histological features indicative of transitional pathological states beyond that of a simple and benign RCC include squamous metaplasia, stratified squamous epithelium, and ciliated or mucinous goblet cells in squamous-papillary craniopharyngiomas[5]. Several studies have identified key clinical imaging, and histopathological features that can be used to differentiate these lesions[5].

In this patient, we diagnosed a rare RCC with contents suggestive of fat droplets on imaging, characteristic of dermoid cysts. The fatty droplet contents of the cyst were suppressed on preoperative T1-fat suppression imaging and were confirmed during intraoperative cyst aspiration. Cholesterin crystals, which are composed of phospholipids in cell membranes, associated with chronic inflammation, have also been reported to be found in cysts in RCCs and craniopharyngiomas, and are particularly likely to be found in large cysts in RCCs[16-20]. Fujio et al.[19] reported that xanthogranulomas characterized by cholesterin crystal and hemosiderin deposition are the last stage of chronic inflammation in RCCs or craniopharyngiomas. No cholesterin crystals were found in the pathological investigations of our case. However, in the intraoperative findings of our case, the yellowish-white creamy intracystic solution reflected cholesterin crystals and was thought to be the cause of the fat-suppressed T1 high-signal lesion on preoperative MRI. Furthermore, the histopathological features of the extracted RCC support the aforementioned theory of a possible common ectodermal origin of selected sellar and suprasellar cystic lesions[5]. Further analysis in other cases is needed to confirm this hypothesis.

In conclusion, RCCs may also contain fat droplets inside the cysts, which are presumably cholesterin crystals due to chronic inflammation. In preoperative imaging of suprasellar cystic lesions, the presence of lipid components in the cyst may not be indicative of dermoid cyst, and care should be taken to differentiate it from RCC or craniopharyngioma.

Our experience shows that EES is effective for achieving successful, complete suprasellar cystectomy in cases wherein a definitive diagnosis may not have been possible during the preoperative work-up and may improve the clinical outcomes in affected patients with suprasellar RCCs that mimic dermoid cysts. We hope that this case report will help clinicians diagnose and treat similar lesions and that a comprehensive study of the high recurrence rate of suprasellar RCCs will lead to the establishment of evidence-based treatments that will lead to good, long-term outcomes.

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Author contribution

YM and HA were involved in study design. All authors critically revised the article, commented on drafts of the manuscript, and approved the final report.

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