

Case Report

Pituitary apoplexy associated with Rathke's cleft cyst : A report of two cases

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Although pituitary apoplexy associated with Rathke's cleft cyst is rare, we have experienced two cases. A 28-year-old man had sudden onset of headache, nausea, and vertigo in March 2004. Brain magnetic resonance imaging (MRI) revealed a pituitary fossa mass with suspected intratumoral bleeding. The mass was surgically removed using a trans-sphenoidal approach. The pathological findings indicated Rathke's cleft cyst. The other case was a 32-year-old woman who developed headache in September 2004. Brain MRI showed intratumoral bleeding at a pituitary lesion mass. The trans-sphenoidal approach was again used to remove the tumor and hematoma. The microscopic findings of Rathke's cleft cyst were almost identical to those of the first case. Symptoms resolved postoperatively in both patients. Although pituitary apoplexy is usually caused by pituitary adenoma, Rathke's cleft cyst should be considered in the differential diagnosis.

Key words : pituitary apoplexy, Rathke's cleft cyst

Introduction

With the relatively recent development of magnetic resonance imaging (MRI), Rathke's cleft cyst has become easier to diagnose. Rathke's cleft cyst is macroscopically identifiable in about 20% of autopsy cases, but clinical symptoms do not occur if the cyst is smaller or shows no growth in mass size⁵). Pituitary apoplexy associated with Rathke's cleft cyst is rare, with only 7 cases reported in the literature^{2,3,4,7,9}). We report 2 cases in which bleeding appeared as a likely cause of pituitary apoplexy associated with Rathke's cleft cyst.

Case 1

A 28-year-old man experienced severe headache, nausea, and vertigo in March 2004, and visited a clinic. Brain MRI was undertaken because of continuing severe headache, and showed a pituitary mass with suspected intratumoral bleeding. He was admitted to our hospital for further examination and treatment. Neurological examinations identified no abnormality other than headache. Hormone levels appeared normal from laboratory tests, and no clinical sign of hypopituitarism was noted. Brain MRI revealed a pituitary mass of mixed intensity on T1- and T2-weighted imaging. The cyst wall was well depicted on gadolinium-enhanced imaging (Fig.

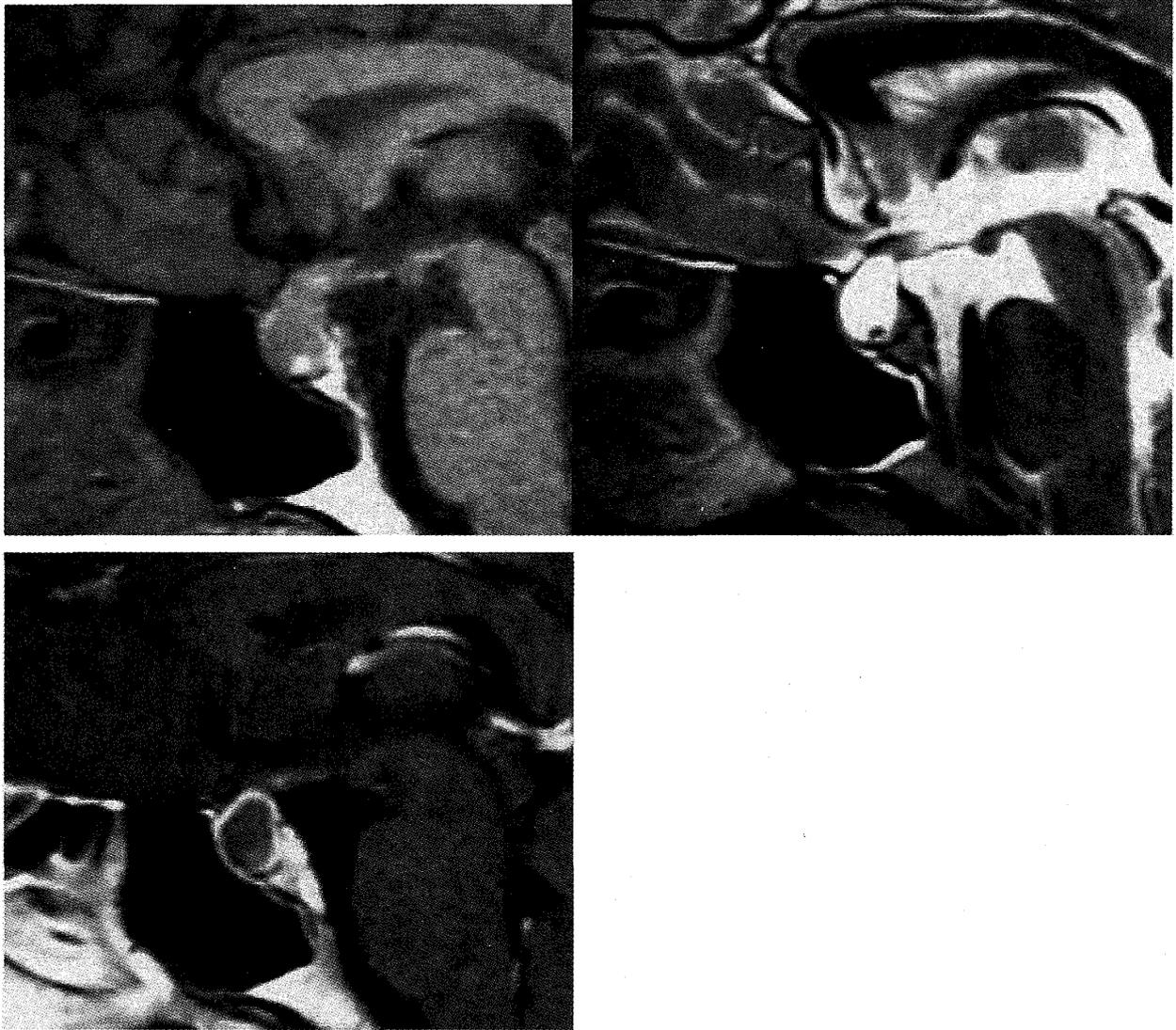


Fig. 1 T1-weighted MR image demonstrating an intrasellar iso-intensity cystic mass with high intensity spot. T2-weighted MR image showing high-intensity lesion with low-intensity spot. T1-weighted image with Gd-DTPA administration demonstrating ring like enhancing the cyst wall.

1). Our first impression was pituitary apoplexy associated with pituitary adenoma, with differential diagnoses of craniopharyngioma, Rathke's cleft cyst, or other congenital tumors. The tumor was removed using a trans-sphenoidal approach. Pathological findings indicated Rathke's cleft cyst with hematoma. Necrosis and squamoid epithelium revealed squamoid change of Rathke's cleft cyst (Fig. 3A). The patient's headache resolved postoperatively.

Case 2

A 32-year-old woman experienced sudden onset of severe headache in September 2004. Although she tried over-the-counter medications for headache, they proved ineffective. The patient then visited a local clinic, where left temporal upper quadrant anopsia was noted. Brain MRI identified a pituitary mass, and she was admitted to our hospital. Neurological findings were normal and visual disturbance improved after admission, but she complained of continu-

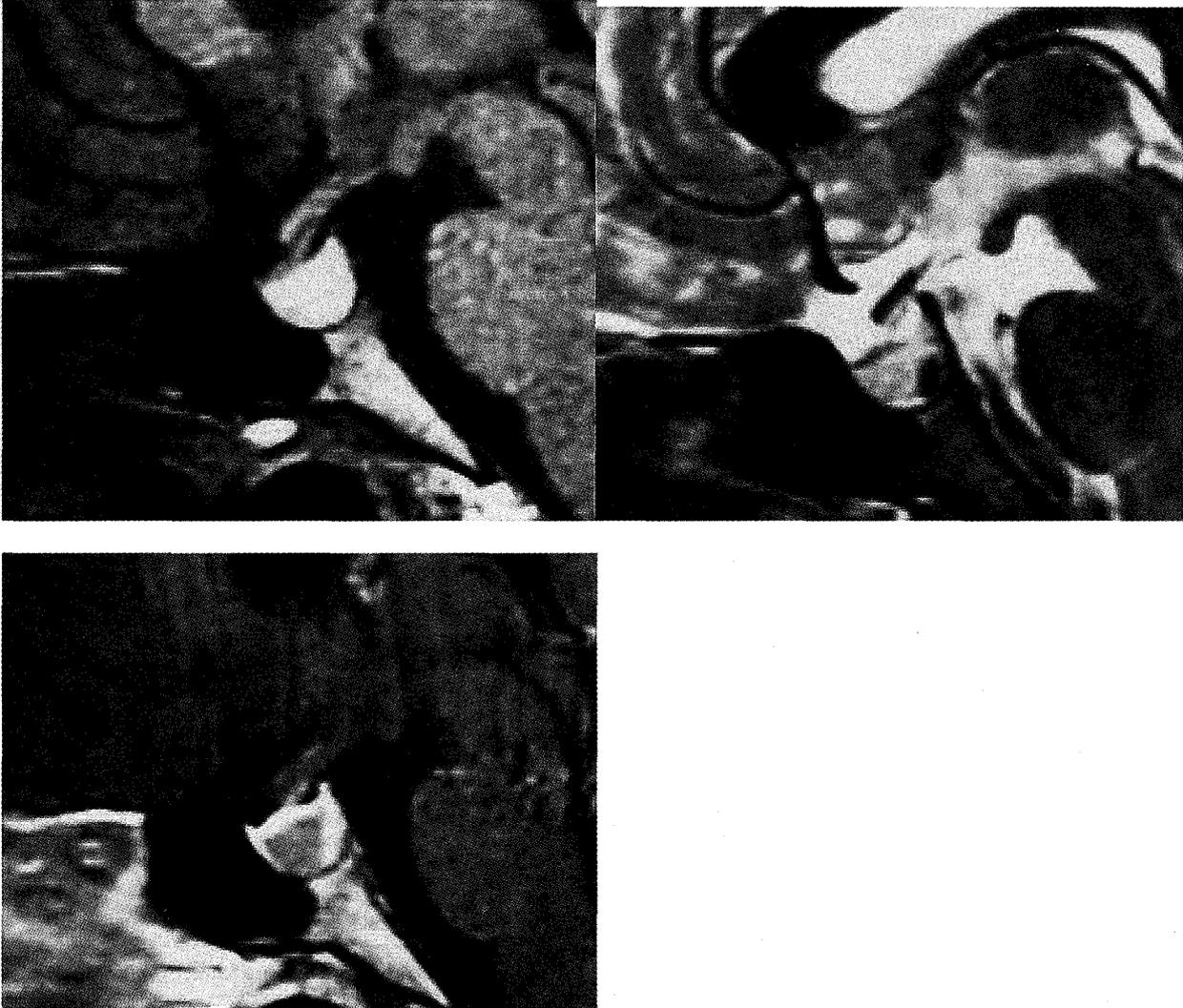


Fig. 2 T1-weighted MR image demonstrating an intrasellar high-intensed cystic mass with high spot. T2-weighted MR image showing almost same findinds with case 1. Gd-DTPA administration demonstrating the cyst wall.

ous headache. Hormone levels were normal and showed no hypopituitarism. Repeated brain MRI showed decreased pituitary mass size with decreasing compression of the optic chiasm (Fig. 2). Although surgery did not seem indicated because of the absence of clinical symptoms other than headache, it was performed to determine the cause of bleeding and the decrease of mass effects. The pathological diagnosis was Rathke's cleft cyst, with findings of some glandular structure and a few epithelia, suggestive of ciliated figures (Fig.3B). The postoperative course was uneventful and her headache disappeared.

Discussion

Rathke's cleft cyst is considered to arise from a remnant of the stomodeum⁸⁾. The cyst is macroscopically noted in about 20% of autopsies, but rarely causes clinical symptoms⁵⁾ except in a few cases.

Histologically, the wall of Rathke's cleft cyst consists of a single or pseudostratified epith-

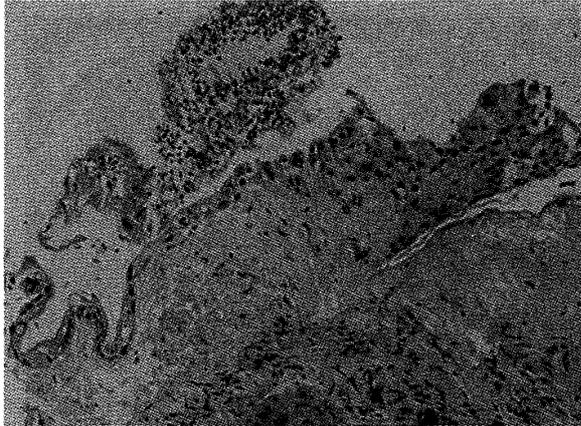


Fig. 3A A vacuum with hematoma presenting. Necrosis and squamoid epithelium suggesting the squamoid change of Rathke's cleft cyst.

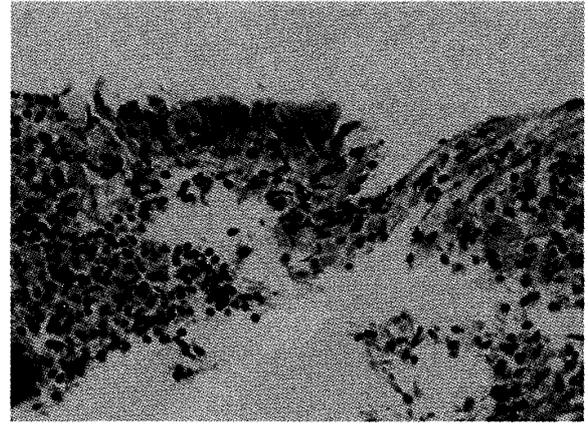


Fig. 3B Some glandular structures presenting, and a few epithelia showing of ciliated figures.

Table. 1

authors	years	age	sex	intraoperative findings
Onesti	1990	25	F	intratumoral hemorrhage and necrosis
Kleinchmidt-DeMasters	1995	32	M	semisolid necrotic cystic contents
Kleinchmidt-DeMasters	1995	51	F	semisolid necrotic cystic contents with hematoma
Kleinchmidt-DeMasters	1995	28	F	inspissated cyst contents
Kurisaka	1998	46	F	bloody contents
Nishioka	1999	46	F	blood clot and mucous materials
Fukushima	2001	67	F	old clot and granulation tissue
present case 1	2005	28	M	old clot
present case 2	2005	32	F	debris and granulation tissue

elium with an underlying layer of connective tissue¹⁰).

The epithelium may contain ciliated cells, goblet cells or squamous cells¹⁰).

Usual clinical symptoms of Rathke's cleft cyst are headache, pituitary dysfunction, and visual disturbance, generally as a result of local mass effects¹). Some rare symptoms such as aseptic meningitis caused by rupture of cyst contents into the subarachnoid space or abscess formation within the cyst have been reported¹⁰).

On MR imaging, the cyst is round and located in the sella or extended suprasellar region. Contrast signals on T1- or T2-weighted imaging range from high to low, depending on factors such as cystic contents or protein^{1,6,8,10}). Pituitary apoplexy associated with Rathke's cleft cyst is very rare, and has been reported in only 9 cases, including the 2 cases discussed here (Table 1)^{2,3,4,7,9}).

In regard to the mechanism of pituitary apoplexy associated with Rathke's cleft cyst,

Nishioka et al. found an old hematoma and granular tissue in the cyst, and discussed the possibility that the apoplexy is caused by the rupture of vessels in the granular tissue⁷⁾. Fukushima reported a case of pituitary apoplexy in which the pituitary stalk was compressed by increasing hematoma in the cyst, causing small vessels to rupture²⁾.

In case 1, histological study showed that cystic space was present and included hematoma and necrosis. Microscopy in case 2 revealed a glandular structure and ciliated epithelium. These findings indicate that the mechanism of pituitary apoplexy, with rupture of vessels in the granular tissue or small vessels of the cyst, is the same as that reported by Fukushima²⁾ and Nishioka⁷⁾.

In regard to operative indications, if a cyst bleeding in a pituitary lesion was found and was associated with headache and hemianopsia, surgical intervention would be carried out. However, patients with absent or decreasing symptoms, as in case 2, would be followed up conservatively with imaging.

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ラトケ嚢胞に伴って発症した下垂体卒中の2症例

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要 約

ラトケ嚢胞に伴って発症する下垂体卒中は極めて稀である。今回我々は、ラトケ嚢胞に伴った下垂体卒中の2症例を経験した。症例は次の通りである。症例1：28歳男性で突然の頭痛と嘔気、めまいにて発症。MRIでトルコ鞍内から鞍上部に混合信号域を認め、出血を伴った嚢胞性病変が疑われた。経蝶形骨洞的腫瘍摘出術施行し、術後症状は消失した。症例2：32歳女性。鎮痛剤無効の頭痛のためMRI施行し、同様にトルコ鞍内に出血が疑われたため、経蝶形骨洞

的腫瘍摘出術施行した。術後症状は消失し、病理所見はいずれも出血を伴ったラトケ嚢胞であった。

下垂体卒中は主に下垂体腺腫に伴って生じることが多く、ラトケ嚢胞に伴った下垂体卒中の報告は、これまで渉猟しえた文献では、本2症例を含め9症例のみであった。ラトケ嚢胞からの出血の機序や治療方法など、我々の経験も含め若干の文献学的考察を加え報告する。