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RATHKE'S CLEFT CYST ASSOCIATED WITH INTRACRANIAL ANEURYSM: REPORT OF FIVE CASES

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Over the past few decades a considerable numbers of studies have been made on Rathke's cleft cyst. Cerebral aneurysm occurs in approximately 7% of patients with pituitary adenoma, but the factual incidence of Rathke's cleft cyst with cerebral aneurysm remains an open question. To the authors knowledge, only a few reports have been reported in the literature concerning such a combined lesion. From 1980 to July 1997, we have experienced 38 cases of Rathke's cleft cyst. These included 16 males and 22 females with a mean age of 42.5 years (range 11-73 years). They were obtained from the Tokyo Women's Medical College Hospital and its affiliated hospitals to treat Rathke's cleft cyst by surgical procedure and histological verification. We have experienced with five cases (13%) in 38 patients. Our data show that, as compared with pituitary adenoma, incidence of Rathke's cleft cyst associated with cerebral aneurysm was not rare than we have considered previously. Our study documents the necessity and importance of cerebral angiography in patient with tumor of the sellar region prior to the surgical procedure.

Introduction

Rathke's cleft cyst was firstly described by Goldziehen[1] in 1913 and recent reports have contained reviews of various subsets of symptomatic Rathke's cleft cyst. Clinical features were well discussed by Voelker et al[2] in 1991. The real incidence of cerebral aneurysms in Rathke's cleft cyst is as yet unknown. In this manuscript, we are going to report our experience with five cases of Rathke's cleft cyst associated with cerebral aneurysm.

Case Report

Clinical subjects and methods

From 1980 to July 1997, we have obtained 38 cases with Rathke's cleft cyst from Tokyo Women's Medical College Hospital and its affiliated hospitals. There included 16 males (42%) and 22 females (58%) with a mean age of 42.5 years (range 11-73 years). All patients underwent surgery and had their tumors histologically verified. Surgical specimens were examined with hematoxylin and eosion staining. Using the World Health Organization classification, the tumors were classified as Rathke's cleft cyst.

All patients were examined by computed tomography (CT) scanning with or without magnetic resonance (MR) imaging. Cerebral angiography was performed routinely except those who refused angiography or had allergy for positive medium.

Results

We found five case (13%) of 38 patients with Rathke's cleft cyst associated with cerebral aneurysm. As shown in Table, we encountered

Table Characteristics of patients with cerebral aneurysm

Case	Age, Sex	Clinical symptoms	Location of cerebral aneurysm	Outcome
1	73, Male	Headache	Anerior communicating artery	died
2	53, Female	SAH	Anerior communicating artery	good
3	43, Female	SAH	Anerior communicating artery	good
4	66, Female	SAH	Anerior communicating artery	good
5	49, Male	Headache, polyuria, decrease of visual acuity	Bilateral middle cerebral artery	poor

SAH : subarachnoid hemorrhage.

two cases with nonruptured cerebral aneurysms, who had manifested headache. During surgery for aneurysmal clipping three cases of Rathke's cleft cyst were found incidentally. Of these three patients with ruptured cerebral aneurysm, no clinical signs and symptoms had been observed before subarachnoid hemorrhage was presented.

Four patients survived after surgical treatment, but one died with pneumonia.

Illustrative Cases

Case 1

A 73-year-old man visited us for general headache and slowly progressive visual disturbance lasting for a month. He had been under medication for hypertension since more than 20 years ago. On admission, he was 67 kg weight and 159 cm high, with normal dry skin and normal urinary function. Neurological examination revealed infero-lateral quadrant-anopia. Plain craniogram didn't show ballooning, double floor, blistering and abnormal calcification around sellar region. CT showed a round expanding low density mass lesion in- and suprasellar portion, which was not enhanced by the positive medium (Fig. 1).

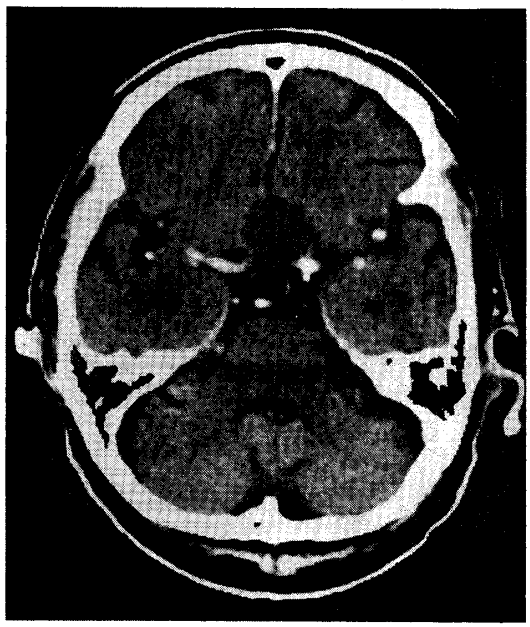

Fig. 1 Case 1: Axial plain CT scan
A round shaped intrasellar mass is noted. The contents of the mass appear low dense.

MR imaging disclosed the lesion as a low intensity to brain on T1 weighted image (Fig. 2) and high intensity on T2 weighted image (Fig. 3). The cyst was rim like enhanced by gadolinium diethylenetriamine pentaacetic acid (Gd-DTPA). Cerebral angiography showed elevation of bilateral A1 portions and anterior communicating artery (Fig. 4). Tumor stain was not observed. The base values of pituitary hormones were all within normal range; thyroid stimulating hormone (TSH) 5.3 MCU/ml, human growth hormone (HGH) 0.8 ng/ml, luteinizing hormone (LH) 2.8 MIU/ml, follicle stimulating hormone (FSH) 7.8 MIU/ml, adrenocorticotrophic hormone (ACTH) 10 pg/ml, prolactine 13.8 ng/ml, and antidiuretic hormone (ADH) 0.6 pg/ml. On stimulating test with thyroid releasing hormone and luteinizing hormone releasing hormone, TSH and LH responded well, FSH less responded, and HGH revealed no response. On glucose tolerance test, cortisol and ACTH responded well, but HGH showed no response.

He underwent surgery by interhemispheric approach. A yellow colored cyst was found in suprasellar region with tough capsular wall.

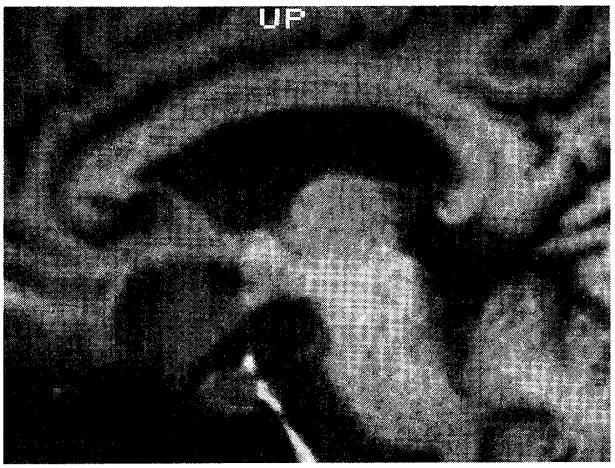

Fig. 2 Case 1: Sagittal T1 weighted MR image shows low signal in mass.

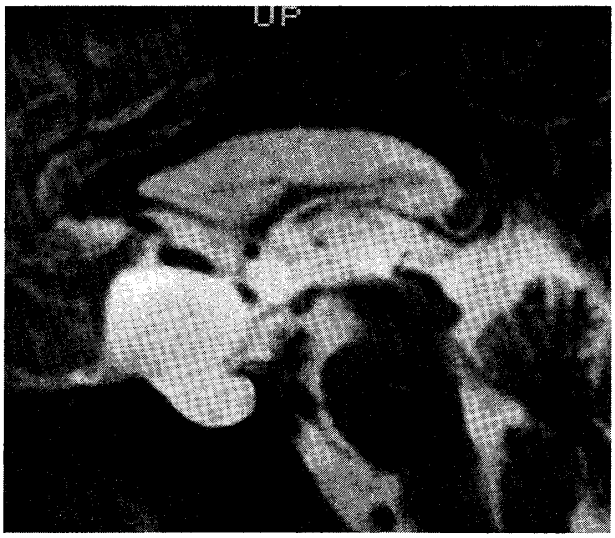

Fig. 3 Case 1: Sagittal T2 weighted MR image shows bright signal in mass.

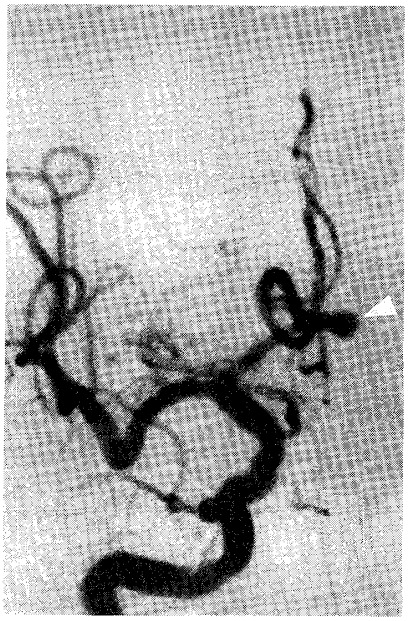

Fig. 4 Case 1: Right carotid angiogram antero-posterior projection subtraction print
Lateral displacement of supraclinoid segments and elevation of anterior cerebral artery. There is an aneurysm projecting medially, at the origin of the anterior communicating artery (arrow head).

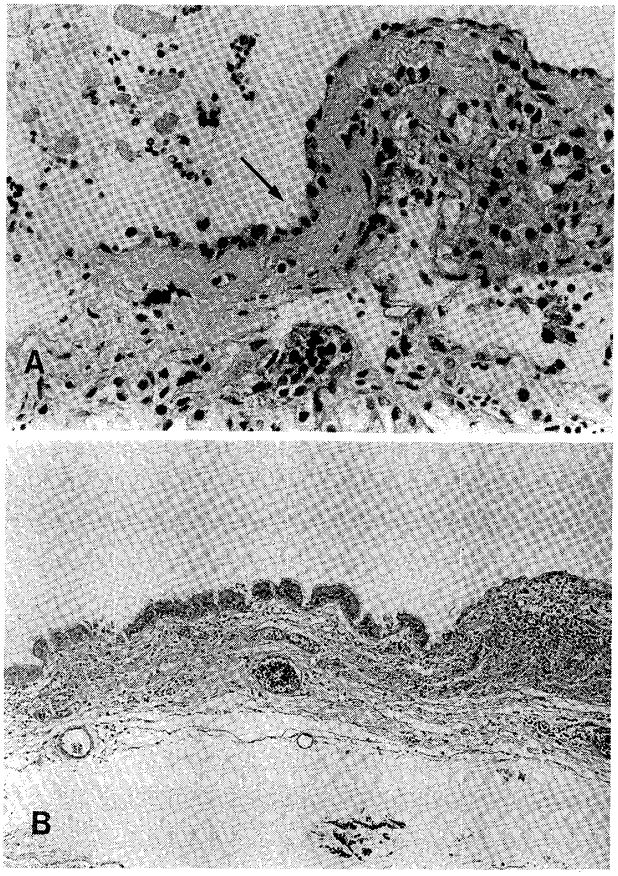

Fig. 5 Case 1
A: Pathological findings confirm Rathke's cleft cyst consisting of a single layer of ciliated cuboidal or columnar epithelium.
B: The wall is composed of one layer of cuboidal epithelial cells.

The mucous fluid was aspirated without leaking the content out. The capsular wall was easily separated from the surrounding cortex after collapsing the cyst. Cyst contents was golden yellow material. Cerebral aneusysm was exposed behind the cyst with tight adhesion to dorsal wall of the cyst. Pituitary stalk was not identified. After clipping of aneurysmal neck,

supradiaphragmal part of the cyst was removed except the small segment adhered to the aneurysmal dome. The residual lesion under the diaphragm was evacuated.

After surgery, pan-hypopituitarysm was observed: TSH 1.4 MCU/ml, HGH 1.1 ng/nl, LH 0.4 MIU/ml, ADH 0.3 pg/ml. Diabetes insidious and hyponatremia with slight consciousness disturbance were controlled by water balance and with vasopressin. However, hemiplesia due to residual subdural effusion exacerbated his clinical course, which made him bedridden for more than half year. He was finally lost by pneumonia. Pathological examination: The wall was composed of the layer of ciliated columnar epithelium. In some parts of the wall, lining epithelium have been lost (Fig. 5A • B).

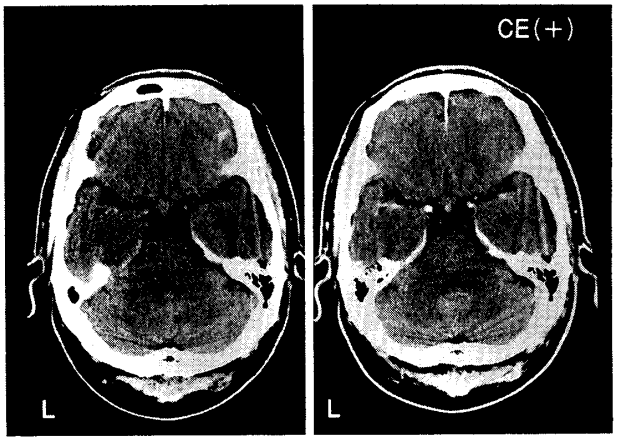

Fig. 6 Case 5: Preoperative study
CT reveals low density round mass in suprasellar region.

Case 5

A 49-year-old man was admitted to our hospital with headache, polyuria and decrease of visual acuity. The remainder of the neurological functions were found to be normal. Stimulating test and base values of pituitary hormones were all with-in normal range. CT revealed low density round mass in suprasellar region (Fig. 6).

Cerebral angiography documented that he had nonruptured middle cerebral aneurysms bilaterally (Fig. 7). This patient was operated to clip right middle cerebral artery aneurysm firstly. On exploration, after neck clipping, we found a yellowish round cyst in suprasellar region and performed partial resection of the cyst wall. He developed left hemiparesis after surgery.

Pathological examination: The cyst wall lined by single layer with ciliated columnar epithelium. The tumor was composed of groups of epithelial cells arranged around blood vessels.

Discussion

Rathke's cleft cysts are found at the rate

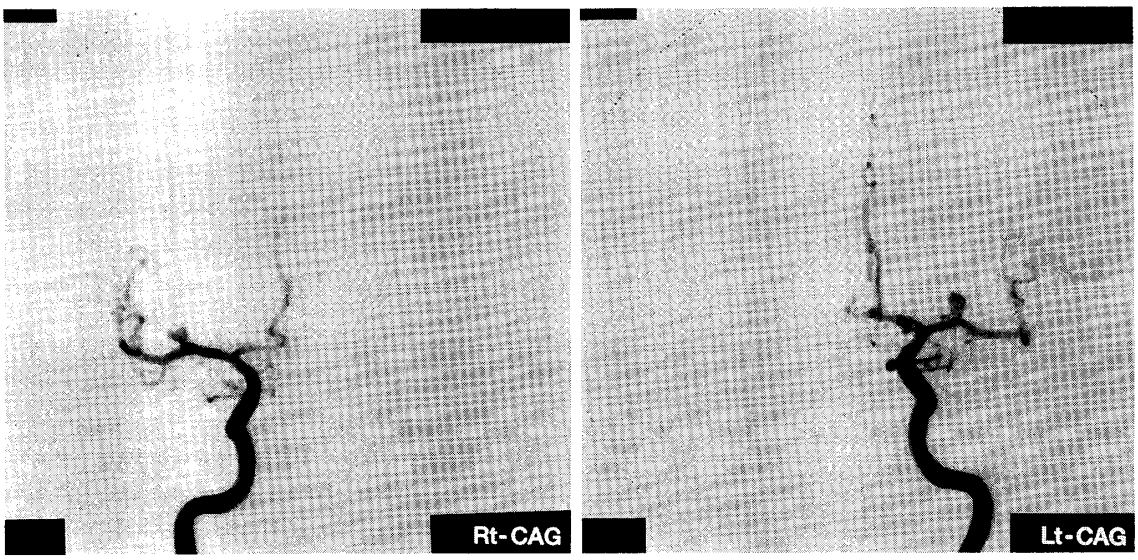

Fig. 7 Case 5: Cerebral angiography shows bilateral nonruptured middle cerebral aneurysms.

from 13% to 33%[3)~8)] in the autopsy studies, and the numbers of the patients with incidentally observed Rathke's cleft cyst may gradually increase since the advent of new neuroradiological imaging[9)~14)]. There are four possible combinations of cerebral aneurysms and Rathke's cleft cysts, namely, ① symptomatic Rathke's cleft cyst with ruptured cerebral aneurysm; ② asymptomatic Rathke's cleft cyst with nonruptured cerebral aneurysm; ③ symptomatic Rathke's cleft cyst with nonruptured cerebral aneurysm; ④ asymptomatic Rathke's cleft cyst with ruptured cerebral aneurysm.

It is not clear that factual incidence of the combination Rathke's cleft cysts and cerebral aneurysm. Cerebral aneurysm occurs in approximately 7% of patients with pituitary adenoma[15)16)]. In our search of 57 cases of Rathke's cleft cyst detected[17)~25)] in the literature, there were only two cases associating with intracranial aneurysms[24)25)].

Trokoudes et al[24)] reported a patient, who had suffered hyperprolactinemia for more than 10 years with an internal carotid aneurysm. They treated pituitary adenoma and Rathke's cleft cyst through transsphenoidal approach and associated aneurysm was left as it was. Yamamoto et al[25)] reported another case. Rathke's cleft cyst was found during direct surgery for anterior communicating aneurysm and partially removed. We described our experience in case of above mentioned combination ② and ③. In our first case, a small berry type nonruptured intracranial aneurysm was found incidentally with preoperative cerebral angiography.

In our case 2, 3 and 4, an asymptomatic Rathke's cleft cyst was found incidentally during surgery for adjacent ruptured intracranial aneurysm and the size of the lesion was less than 7 mm in diameter.

Rathke's cleft cyst with intracranial aneurysm incidentally discovered in our case 2, 3 and 4 might have less influences in blood flow to aneurysm. In our case 1, physical strain might have affected on aneurysmal dome through continuous compression.

On these our experience, Rathke's cleft cyst with intracranial aneurysm is not so rare as we have considered previously. We have come to the conclusion that cerebral angiography should be performed not only in patients with pituitary adenoma but also in patients with in- and para-sellar cyst. And in such patients, knowledge of the coexistence of cerebral aneurysm should be taken into account in surgical planning.

The indications for surgery on nonruptured asymptomatic cerebral aneurysms are still unclear. The cases herein show that asymptomatic aneurysms, especially anterior communicating aneurysm, should be clipped and Rathke's cleft cyst exposed in the operation. Once asymptomatic Rathke's cleft cyst was exposed during surgery on ruptured cerebral aneurysm, the cyst should be dismissed under allowable circumstances.

Recent advance of MR angiography and CT angiography could help us to confirm the coexistence of cerebral aneurysm noninvasively.

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脳動脈瘤を伴ったラトケ嚢胞5症例の経験

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ラトケ嚢胞は，画像診断技術の発達に伴い近年報告例が増加している腫瘍である．しかしラトケ嚢胞と脳動脈瘤の合併の報告例は少なく，我々の渉猟し得た範囲では過去2例の文献報告をみるのみである．1980年より1997年7月までに我々の施設および関連病院においてラトケ嚢胞と診断されたものは38症例である．これらのうち動脈瘤を合併した症例を5例経験したので報告する．2例は下垂体部腫瘍症例に術前検査として脳血管撮影を施行したところ動脈瘤が偶然に発見された症例である．3例はくも膜下出血で発症し，そのクリッピング手術の際に，嚢胞性腫瘍が鞍上部に偶然発見されたものである．全症例について切除標本に対し病理学的検査を施行し，ラトケ嚢胞と診断した．

ラトケ嚢胞に対する脳動脈瘤の合併頻度は不明であるが，我々はラトケ嚢胞38症例中5例（13%）と高率に動脈瘤の合併を経験した．下垂体腺腫においては約7%に脳動脈瘤の合併を認めるとの報告がなされているが，ラトケ嚢胞についても脳動脈瘤の合併は稀なものではないと考えられた．我々の経験した症例数は，いまだ十分ではなく今後の症例の積重ねが必要と考えられるが，下垂体部の腫瘍を認めた際には脳動脈瘤の存在を考慮し，脳血管撮影を施行すべきと考えられた．また，近年のMRAや3次元CTの進歩により，より非侵襲的に動脈瘤の合併を検索することが可能となってきていると考えられる．

ラトケ嚢胞は良性腫瘍であるが，視神経障害や下垂体機能低下といった症候を伴うものは積極的な外科治療の必要性を有するものと考えられる．しかし，正常下垂体と腫瘍の位置関係や合併する動脈瘤の位置などにつき，十分に術前検討を加えることが必要と考える．くも膜下出血で発症し偶然ラトケ嚢胞が発見された場合には，術者の疲労度や時間的余裕を考慮して，可能ならば嚢胞切除を行うことが望ましいと考える．