Case Report

Spontaneous Rupture of an Intracranial Epidermoid Cyst

Yuki Sakaeyama[1)*] Hiroyuki Masuda[1)] Kosuke Kondo[1)]
Naoyuki Harada[1)] Nobuo Sugo[1)] and Masaaki Nemoto[2)]

[1)]Department of Neurosurgery (Omori), School of Medicine, Faculty of Medicine, Toho University, Tokyo, Japan
[2)]Department of Neurosurgery (Sakura), School of Medicine, Faculty of Medicine, Toho University, Chiba, Japan

ABSTRACT: Background: Ruptured epidermoid cysts can cause aseptic meningitis, but spontaneous rupture is very rare. We report a case of spontaneous rupture of an intracranial epidermal cyst. Case presentation: A 44-year-old man developed occipitalgia and neck pain seven months ago. A brain tumor was detected on computed tomography, but was only followed up. After his headaches worsened, he visited our hospital. The tumor was located in the parasellar region with multiple small lesions in the sylvian fissure. We diagnosed it as a spontaneous ruptured epidermoid cyst and performed tumor resection. The patient did not develop fever or headache after surgery, and the postoperative course was uneventful. Nineteen months after surgery, the tumor has not relapsed. Conclusion: We treated a case of spontaneous rupture of an intracranial epidermal cyst. Since the risk of recurrence increases when the capsule remains, aggressive removal is suggested.

Toho J Med 5 (2): 69–75, 2019

KEYWORDS: epidermoid cysts, spontaneous rupture, aseptic meningitis

Introduction

Epidermoid cysts are congenital non-neoplastic lesions arising from residual embryonic tissues. They are covered by stratified, squamous epithelium with keratinization and are characterized by the lack of skin appendages, unlike dermoid cysts.[1)] Their incidence among brain tumors is 0.9%, the male:female ratio is approximately 7:9, and the average age of development is 50 to 60 years.[2)] Forty to 50% of spontaneous cases occur in the cerebello-pontine angle, 30% occur in the parasellar region, and others occur in the cerebral ventricle or pineal gland.[3)]

Epidermoid cysts are notoriously destructive and cause inflammation, leading to aseptic meningitis after surgery. On the other hand, aseptic meningitis is rarely associated with spontaneous rupture.[4)] We report a case of repeated spontaneous rupture of an intracranial epidermoid cyst with a review of the relevant literature.

Case Presentation

Patient: A 44-year-old man

Chief complaint: headaches

Past history: untreated hyperuricemia and dyslipidemia

Current medical history: Twenty years ago, the patient visited a nearby medical clinic due to frequent headaches. A brain tumor was noted in the vicinity of the pituitary fossa on computed tomography (CT), but was only followed up under a diagnosis of benign tumor. His headaches worsened three years ago; he visited a medical clinic and was prescribed analgesics. Seven months before the

*Corresponding Author: Yuki Sakaeyama, 6-11-1, Omori-Nishi, Ota, Tokyo 143-8541, Japan, tel: 03–3762–4151
e-mail: yuuki.sakaeyama@med.toho-u.ac.jp
DOI: 10.14994/tohojmed.2018-015

Received July 23, 2018: Accepted Dec. 5, 2018
Toho Journal of Medicine 5 (2), June 1, 2019.
ISSN 2189–1990, CODEN: TJMOA2

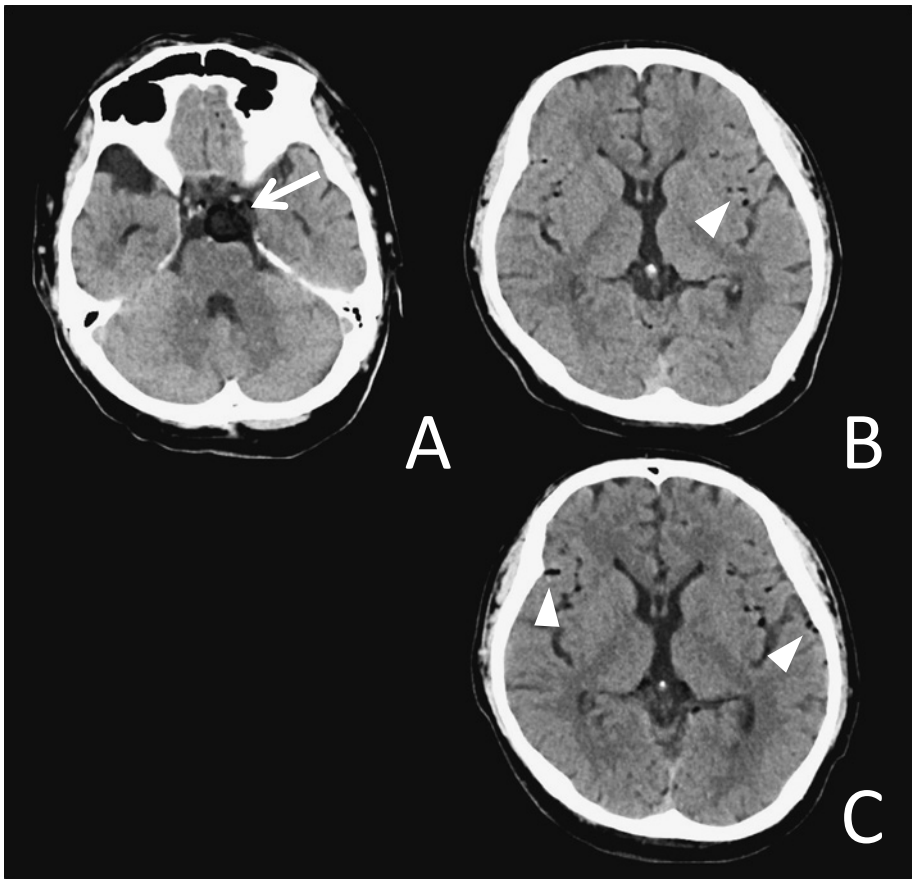

Fig. 1 Preoperative brain CT.
A: Brain CT obtained seven months before the patient's initial visit to our department. A low-density tumor extended from the parasellar region to the prepontine cistern (arrow), and an arachnoid cyst was in the right middle cranial fossa.
B: Seven months before the initial visit, small, low-density lesions were scattered throughout the bilateral sylvian fissures and the subarachnoid space (arrowhead).
C: Four months after the initial visit, the CT revealed that the number of small, low-density lesions had increased (arrowheads).

initial visit to our neurosurgical department, he felt strong pain from the back of the head to the neck, and visited the internal medicine department at our hospital. On CT, a brain tumor was found at the same site and was followed up at the patient's request. As strong headaches developed again one month ago, magnetic resonance imaging (MRI) was performed and the patient was introduced to our department for examination of the brain tumor.

Neurological findings: He was alert with no neurological deficit, except headaches, and no symptoms of meningeal irritation.

Blood test results: C-reactive protein was 0.1 mg/dL, white blood cell count was 6000 /μL, and there were no inflammatory reactions or other abnormal findings.

Neuro-radiological findings: Seven months before his initial visit to our department, the patient underwent a CT scan. It revealed a tumorous lesion of 20 × 15 mm, with low density from the parasellar region to the prepontine cistern, and an arachnoid cyst in the right middle cranial fossa (Fig. 1A). Small, low-density areas were scattered in the bilateral sylvian fissure and the cerebral subarachnoid space (Fig. 1B). On plain CT, four months after the patient's initial visit to our department, the number of small, low-density areas had increased in the bilateral sylvian fissure (Fig. 1C). There was no enhancement effect of the tumorous lesion on contrast-enhanced CT. During the patient's first visit to our department, he underwent an MRI. The tumorous lesions exhibited high signal on T1-weighted images (T1WI), slightly high signal on T2-weighted images (T2WI), low signal on diffusion-weighted imaging (DWI), and high signal based on the apparent diffusion coefficient (ADC). There was no enhancement on

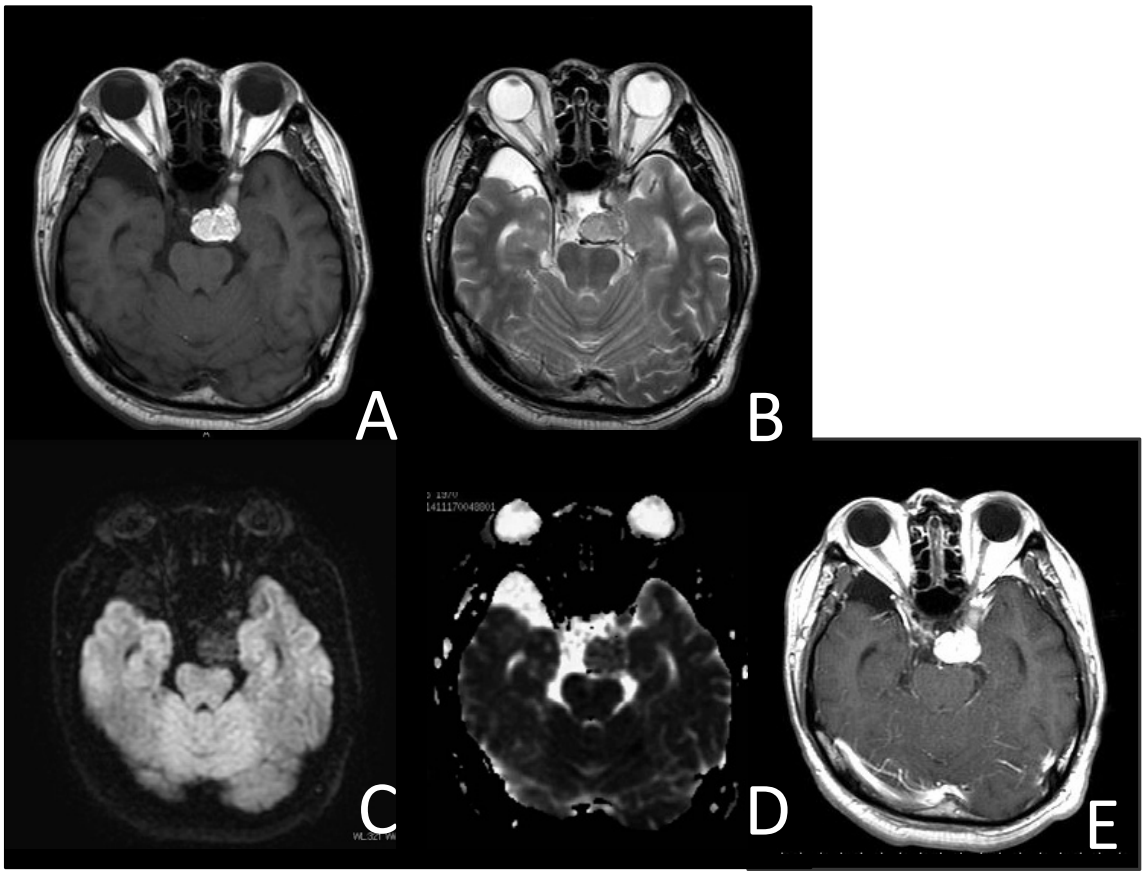

Fig. 2 Brain MRI at the initial visit.
A: On the T1-weighted image, the tumor exhibited high signal.
B: On the T2-weighted image, few high-signal areas were noted.
C: On the DWI, the tumor demonstrated low signal.
D: Based on the ADC, the tumor had high signal.
E: On gadolinium-enhanced MRI, the tumor was not enhanced.

gadolinium (Gd)-enhanced MRI (Fig. 2A-E). Multiple lesions in the subarachnoid space also had high signals on both T1WI and T2WI (Fig. 3A, B), similar to the tumorous lesion.

Surgical findings: We diagnosed the patient with spontaneous rupture of a symptomatic epidermoid cyst. We resected the tumor through the combined transpetrosal approach. The tumor was yellowish, solid, and soft enough to be aspirated easily and removed with a suction tube, with little bleeding from the tumor. It was completely removed while preserving the surrounding arachnoid membrane (Photo 1).

Pathological findings: Only basophilic amorphous material was observed on hematoxylin-eosin staining (Photo 2 A, B). Cytokeratin AE1/AE3 was negative (Photo 2C), and the epithelial membrane antigens were also negative (Photo 2D).

Postoperative course: There was no meningitis or neurological deficit after surgery, and the course was uneventful. Currently, 19 months have passed since surgery. The strong headaches the patient noted before surgery have not reappeared, and there has been no tumor recurrence. In addition, the multiple bilateral lesions scattered in the bilateral sylvian fissure and cerebral subarachnoid space have not changed (Fig. 4A-C).

Discussion

Epidermoid cysts are classified into three types based on CT findings.[5] Type I, the most frequent, exhibits homogeneous low density on plain CT and no contrast enhancement effect. Since the tumor's main component is cholesterol, it has a low density. Type II exhibits low density in the central part of the tumor and high density in the periphery. The central part has low density due to choles-

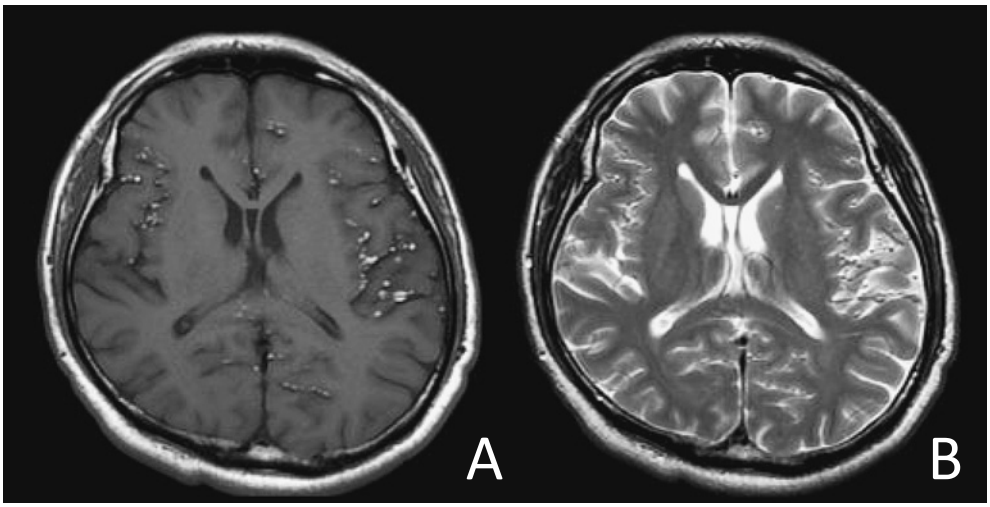

Fig. 3 MRI scan acquired at the same time as that in Fig. 2.
A: T1-weighted image, **B**: T2-weighted image; multiple small high-signal lesions were observed on both T1- and T2-weighted images.

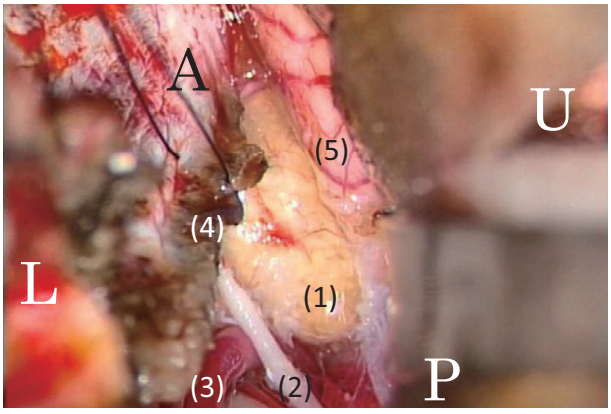

Photo 1 Intraoperative photograph of left combined transpetrosal approach.
The left parasellar tumor was exposed after incising the cerebellar tentorium. The tumor was soft and non-hemorrhagic.
(1) tumor, **(2)** trochlear nerve, **(3)** superior cerebellar artery, **(4)** cerebellar tentorium, **(5)** temporal lobe
A: anterior, **P**: posterior, **U**: upper, **L**: lower

terol, similar to type I, and only the periphery is enhanced due to blood circulation in the capsule or dura mater. Type III exhibits a homogeneous high density in the entire tumor with no contrast enhancement effect. Keratin, calcium, and iron cause the high density. Findings on T1WI and T2WI for epidermoid cysts are nonspecific. In general, they are similar to cerebrospinal fluid; low signal on T1WI and high signal on T2WI are often observed. However, the signal can vary depending on tumor content.[3] The signal on T1WI varies with the protein and albumin concentrations, and the signal on T2WI varies with viscosity. High signal on DWI is important for diagnosing epidermoid cysts. On the other hand, cases with low signal on DWI and high signal on ADC are rarely reported. These cases, where the tumor content has high viscosity, occur more frequently.[6] In this case, the lesion was type I, exhibiting low density on CT, and high signal on both T1WI and T2 WI, so the fat content was probably high. The low signal on DWI also reflects the highly viscous content that was easily aspirated during surgery.

Common epidermoid cysts are covered by keratinized stratified squamous epithelium and keratin is layered to fill the cystic space.[3] In this case, the surgically removed tumors were pathologically examined, but only basophilic amorphous material was noted. The reason for this may be that the capsule was damaged by the tumor rupture, and the only remaining content was predominantly fat. In previous reports, the pathology of the capsule could not be clarified, but the cause for this remains unknown.[7] Since both the CT and MRI revealed fat, it is necessary to distinguish such lesions from lipomas. Intracranial lipomas are more common in the fissure around the corpus callosum and in the suprasellar region. In addition, lipomas strongly adhere to the surrounding brain tissues, involving nerves and blood vessels in the tumor,[8] which was different from the intraoperative findings in our case. Furthermore, to the best of our knowledge, there are no reports of spontaneous intracranial lipoma rupture. Thus, we diagnosed this case as an epidermoid cyst rupture.

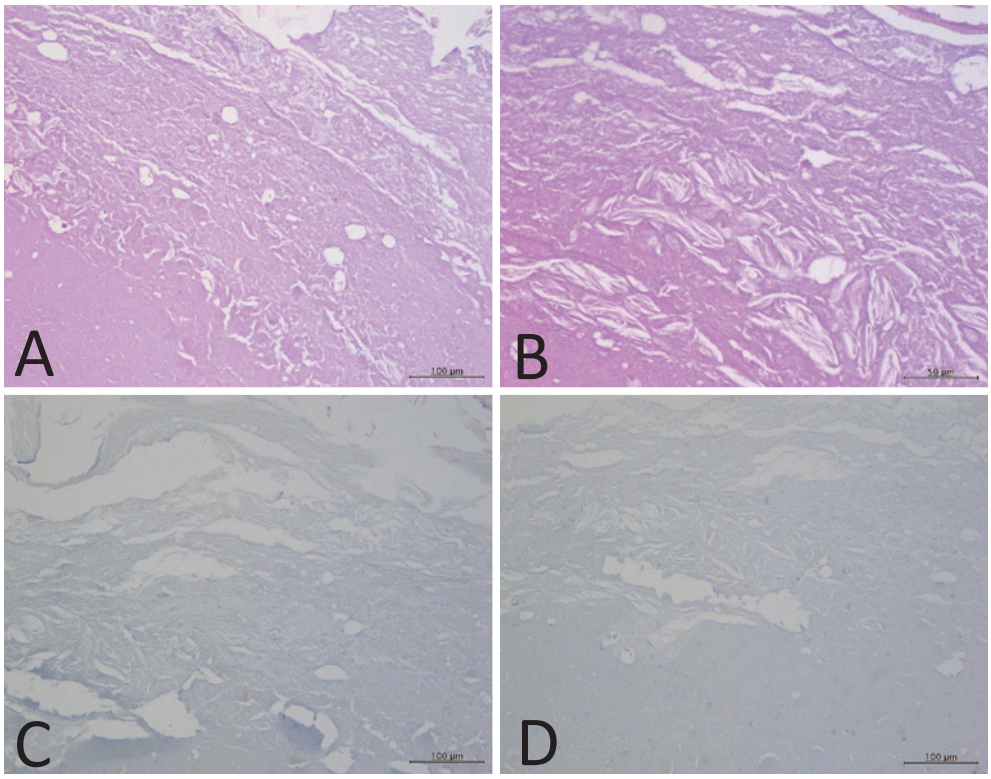

Photo 2 Histological and immunohistological findings.
A: By hematoxylin-eosin staining (×20), only basophilic amorphous material was observed.
B: Hematoxylin-eosin staining (×40).
C: Staining for cytokeratin AE1/AE3 (×20) was negative.
D: Staining for epithelial membrane antigen (×20) was also negative. No significant pathological findings were noted.

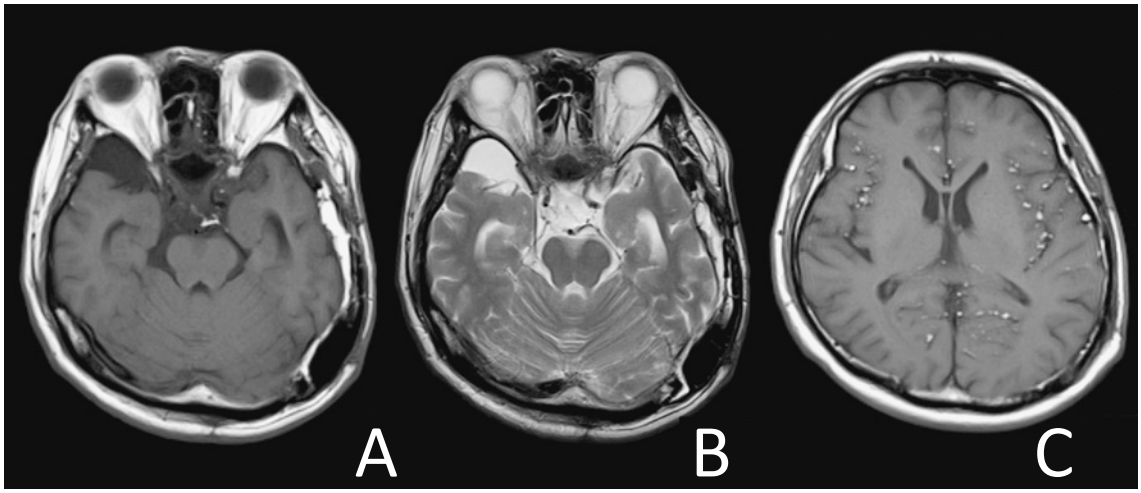

Fig. 4 MRI 19 months after surgery.
No tumor recurrence was observed (**A**: T1WI, **B**: T2WI). **C**: The multiple small lesions had not changed.

Thus far, 24 cases of spontaneously ruptured intracranial epidermoid cysts, including our case, have been reported (Table). The patients' average age was 38.3 years, which is slightly younger than that of all patients with epidermoid cysts, and there was no characteristic site of occurrence for ruptured cysts. Among the initial symptoms, headaches developed in 70%, and 54% were symptoms suggesting meningitis, such as fever, headache, and vomit-

Table Summary of previous reports of spontaneously ruptured epidermoid cysts.

No.	Author	Age/ Sex	Location	Initial symptom	Operation
1	Velamati R, 2013	3/F	CPA	fever, headache, vomiting	total removal
2	Schwartz JF, 1978	4/F	prepontine	fever, headache, vomiting	total removal
3	Aristegui FJ, 1998	7/M	anterior of medulla	fever, headache, vomiting	subtotal removal
4	Guan Z, 2015	13/F	CPA	headache, photophobia, vomiting	total removal
5	Laster DW, 1977	28/F	suprasellar	headache	removal
6	Crossley GH, 1990	32/M	posterior fossa	fever, nystagmus, ataxia	ND
7	Pampliega-Pérez A, 2003	36/M	CPA	fever, headache, ataxia	non operation
8	Carvalho GA, 2000	36/F	CPA	trigeminal neuralgia	removal
9	Laster DW, 1977	38/M	parasellar	headache	removal
10	Kitayama J, 1996	40/F	pineal region	headache, vomiting	non operation
11	Maeda Y, 1990	40/F	quadrigeminal cistern	headache, vomiting	subtotal removal
12	Wei Y, 2015	41/M	suprasellar	headache, vomiting	subtotal removal
13	Miyagi Y, 2000	44/M	anterior cranial fossa	headache, convulsion	subtotal removal
14	Achard JM, 1990	44/M	3rd ventricle	fever, headache, vomiting	stereotaxic aspirations
15	Loumiotis I, 2012	46/M	prepontine	dizziness, headache, atxia	VP shunt
16	Itoh T, 1988	59/M	multiple	convulsion, LOC	removal
17	Shaw, A S, 2005	59/M	parasellar	headache, cough	removal
18	Hao S, 2010	61/F	CPA	headache, vomiting	subtotal removal
19	Hsieh CH, 1996	63/F	middle & posterior cranial fossa	ataxia	removal
20	Nakai T, 2015	67/M	CPA	headache, nausea, disorientation	subtotal removal
21	Barat JL, 1990	ND	cavernous sinus	ND	subtotal removal
22	Bollar A, 1989	ND	parasellar	ND	ND
23	Gormley WB, 1994	ND	ND	ND	ND
24	Present case	44/M	prepontine	headache	total removal

F: female, M: male, CPA: cerebello-pontine angle, ND: not determined, LOC: loss of consciousness, VP: ventriculo-peritoneal

ing. The epidermoid cysts' fragile structures cause the cells to separate easily, leading to self-destruction because of growth inhibition in a limited space.[9] This is a possible mechanism for their spontaneous rupture.

No tumor capsule was observed in this case, suggesting its disappearance due to prolonged tumor inflammation rather than local capsule rupture. Moreover, the tumor content was repeatedly scattered due to the capsule's absence.

Surgical removal is the first choice of treatment for symptomatic epidermoid cysts. Total removal, including the capsule, reduces the recurrence rate; however, it is often difficult. Frequently, only 50 to 80% of the epidermoid cyst can be removed completely[10] because the rupture-induced inflammation may cause the capsule to bind to the surrounding structures, making it impossible to separate.[11] Of the 24 reported cases of spontaneous rupture, 18 were removed. Of these, 2 cases underwent malignant transformation, leading to the patient's death after surgery, although malignant transformation of epidermoid cysts is extremely rare.[12, 13] In this case, at least three ruptures were suspected to have occurred over 20 years. Therefore, since such cysts are symptomatic and intractable, aggressive surgical treatment is necessary.

Conclusion

We treated a case of an intracranial epidermoid cyst that ruptured spontaneously. Since the risk of recurrence increases when the capsule remains, aggressive removal is suggested.

Conflicts of interest: None declared.

References

1) Nishizawa S. Parasellar epidermoid and dermoid cysts -MRI findings and surgical strategy-. Progress in Neuro-Oncology. 2015; 22: 1-8.
2) Report of Brain Tumor Registry of Japan (2005-2008). Neurol Med Chir (Tokyo). 2017; 57: 9-102.
3) Nagasawa D, Yew A, Safaee M, Fong B, Gopen Q, Parsa AT, et al. Clinical characteristics and diagnostic imaging of epidermoid

tumors. J Clin Neurosci. 2011; 18: 1158-62.
4) Dobre MC, Smoker WR, Moritani T, Kirby P. Spontaneously ruptured intraspinal epidermoid cyst causing chemical meningitis. J Clin Neurosci. 2012; 19: 587-9.
5) Ito Y, Kaku M, Kodama T, Matsukado Y. Intracranial high density epidermoid --classification of CT scan findings and chemical analysis of the cystic content. No Shinkei Geka. 1980; 8: 645-8.
6) Bohara M, Yonezawa H, Hanaya R, Takeshita S, Sumida M, Arita K. Posterior fossa epidermoid cysts presenting with unusual radiological appearances -two case reports-. Neurol Med Chir (Tokyo). 2011; 51: 85-8.
7) Maeda Y, Fujita T, Mabuchi E, Kanoh M, Tsujimura T. Epidermoid of the quadrigeminal cistern -case report-. Neurol Med Chir (Tokyo). 1990; 30: 59-62.
8) Nakao S, Yamamoto T, Fukumitsu T, Ban S, Motozaki T, Sato S, et al. Cerebellopontine angle lipoma. Case Report. Neurol Med Chir (Tokyo). 1988; 28: 1113-8.
9) Kitayama J, Toyoda K, Fujii K, Ibayashi S, Sugimori H, Sadoshima S, et al. Recurrent aseptic meningitis caused by rupture of a pineal cyst. No To Shinkei. 1996; 48: 1147-50.
10) Conley FK. Epidermoid and dermoid tumors: clinical features and surgical management. In: Wilkins RH, Rengachary SS, editors. Neurosurgery. New York: McGraw-Hill; 1985. p. 668-73.
11) Ulrich J. Intracranial epidermoids. A study on their distribution and spread. J Neurosurg. 1964; 21: 1051-8.
12) Nishio S, Takeshita I, Morioka T, Fukui M. Primary intracranial squamous cell carcinomas: report of two cases. Neurosurgery. 1995; 37: 329-32.
13) Murase S, Yamakawa H, Ohkuma A, Sumi Y, Kajiwara M, Takami T, et al. Primary intracranial squamous cell carcinoma -case report-. Neurol Med Chir (Tokyo). 1999; 39: 49-54.