Clinical Case Report

Medicine®

OPEN

Intraparenchymal endodermal cyst with spontaneous intracystic hemorrhage in the temporal lobe of an adult

Xin-Jie Bao, MD[a], Xue-Yuan Li, PhD[a], Qi-Pu Wang, PhD[a], Xin-Yu Ren, PhD[b], Zhi-Yong Liang, PhD[b], Wen-Bin Ma, MD[a], Ren-Zhi Wang, MD[a], Jun-Ji Wei, MD[a,*]

Abstract

Background: Endodermal cysts (EC) are rare but well-known congenial lesions of the central nervous system mainly located in the spinal subdural space. Intracranial ECs are rare and commonly encountered in the posterior cranial fossa as extra-axial lesions; an intraparenchymal location is exceedingly rare. A complete removal is the best surgical strategy and any residue can cause recurrence. It is necessary to exclude EC in patients with intracranial cystic lesions. We present a case of intraparenchymal EC with spontaneous intracystic hemorrhage in the temporal lobe of an adult.

Methods: A 43-year-old man presented with headache and memory deterioration. Brain computed tomography and magnetic resonance imaging showed a slightly enhanced temporal lobe cystic lesion, which was homogenously hyperintense on T1-and T2-weighted images. There was a suspicion of brain abscess at admission. The lesion was totally removed with a left subtemporal craniotomy. Histological examination revealed an EC with intracystic hemorrhage.

Results: The preoperative symptoms were relieved after surgery and 3-month follow-up magnetic resonance imaging found no cystic signs.

Conclusion: This case suggests that EC should be considered in the differential diagnosis of intracranial cystic lesions and a complete removal is the best strategy of choice.

Abbreviations: CNS = central nervous system, CT = computed tomography, EC = endodermal cyst, GFAP = glial fibrillary acidic protein, MRI = magnetic resonance imaging.

Keywords: brain abscess, case report, endodermal cyst, hemorrhage, neurenteric cyst

1. Introduction

Endodermal cysts (ECs) of the central nervous system (CNS) are rare congenital lesions with endodermal origin, also known as neurenteric or enterogenous cysts. CNS ECs mostly occur in the subdural space of the spinal cord[1]; intracranial ECs are very rare and commonly situated in the posterior cranial fossa as extra-axial lesions,[2] whereas an intraparenchymal location is extremely rare. The cyst requires a complete removal and any residue can lead to recurrence. However, due to lack of diagnostic features, ECs, especially those with intracystic hemorrhage, are often misdiagnosed and treated as brain abscess with aspiration. Here, we present 1 case of intraparenchymal EC with spontaneous intracystic hemorrhage in the temporal lobe of a 43-year-old man. We describe the clinical, imaging, and pathological findings and review the literature with the purpose of providing information for clinical treatment.

2. Case report

A 43-year-old male was admitted to hospital for 3-year headache and memory deterioration. He had a 10-year history of otitis media and no history of head trauma. Neurological examination and hematological test were normal. Brain computed tomography (CT) revealed a cystic lesion in the left temporal lobe (Fig. 1A and B). The cyst wall was hyperdense and slightly enhanced, cyst fluid exhibited low density, and some sediment appeared in the cyst. Magnetic resonance imaging (MRI) showed a hypodense cyst wall, hyperdense cyst fluid, and isointense sediment on T1- and T2-weighted images (Fig. 1C and D). Only the cyst wall showed slight enhancement after gadolinium injection (Fig. 1E and F). There was no observable edema around the cyst on both CT and MRI. The patient was diagnosed as having brain abscess based on otitis media history and imaging.

After 3 days of antibiotic treatment, a subtemporal craniotomy was performed for removing the cyst. During surgery, a brain needle could not be inserted into the cyst due to a tough wall and thereby the lesion was excised totally. We found that the

Editor: Yong Liu.

X-JB, X-YL, and Q-PW contributed equally to this article.

The authors have no funding and conflicts of interest to disclose.

[a] Department of Neurosurgery, [b] Department of Pathology, Peking Union Medical College Hospital, Peking Union Medical College, Chinese Academy of Medical Sciences, Beijing, China.

[*] Correspondence: Jun-Ji Wei, Department of Neurosurgery, Peking Union Medical College Hospital, Peking Union Medical College, Chinese Academy of Medical Sciences, Beijing 100730, China (e-mail: weijunji@pumch.cn).

Medicine (2016) 95:46(e4968)

Received: 27 November 2015 / Received in final form: 26 April 2016 / Accepted: 1 September 2016

http://dx.doi.org/10.1097/MD.0000000000004968

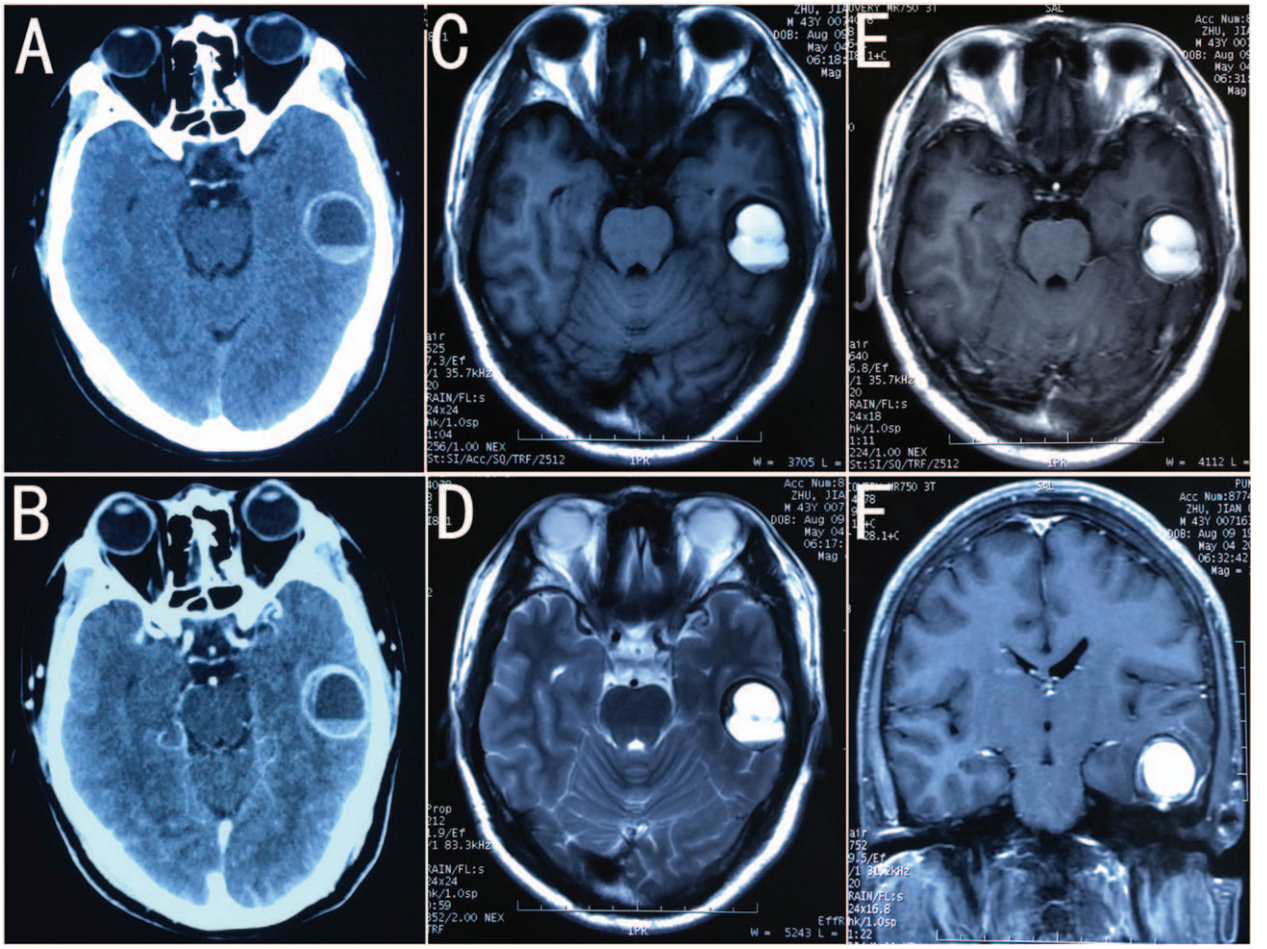

Figure 1. Brain CT and MRI at admission. Nonenhanced (A) and enhanced (B) CT; T1-weighted (C) and T2-weighted (D) MRI; axial (E) and coronal (F) enhanced T1-MRI. CT = computed tomography, MRI = magnetic resonance imaging.

cyst was of intraparenchyma and had a sharp demarcation with adjacent brain tissue. The cyst wall was grayish and solid enough to resist a brain needle (Fig. 2). The cyst contained about 10 mL dark brown mucoid fluids. After contents were collected, the cyst was fixed in 10% formalin, dehydrated, and embedded in paraffin for histological test. Histological examination of the wall showed a thin fibrous membrane lining with columnar epithelial cells and plenty of macrophages, myofibroblasts, inflammatory cells, and cholesterol crystal (Fig. 3A–C). Immunohistochemical staining showed that epithelium was positive for cytokeratin AE1/AE3 (Fig. 3B) and carcinoembryonic antigen CD68 (Fig. 3C) but negative for glial fibrillary acidic protein (GFAP), S-100, and vimentin (not shown), ruling out the epithelium of ectodermal origin. Sporadic staining of CD34 and smooth muscle actin was observed in cyst wall, supporting the cyst of endodermal origin (Fig. 3D and E). Moreover, some epithelial cells stained positive for Ki-67 (Fig. 3F). Cytology of cyst fluid revealed a mixture of white blood cells and red blood cells, indicating intracystic hemorrhage. Sterile culture of cyst fluids showed no bacterial growth. A final diagnosis was EC based on histological findings.

After surgery, the patient suffered a transient sensory aphasia and occasional headache. Three-month follow-up showed a completely restored headache and memory deterioration, and showed no cystic signs on brain MRI (Fig. 4). Three-year follow-up found no evidence of symptom recurrence.

3. Discussion

Intracranial ECs were first reported in 1962 by Small[3] and about 80 to 100 cases have been reported to date.[2] They are usually encountered as extra-axial lesions and rarely associated with hemorrhage. We described a case of intraparenchymal EC with intracystic hemorrhage in the temporal lobe, which is the first report of this kind to our knowledge.

Pathogenesis of CNS EC is thought to associate with persistence of primitive endodermal cells in neurenteric canal in early embryogenesis.[4] This theory explained the location of ECs in the spinal subdural space and posterior fossa but not those with supratentorial location. Other theories include anomalous migration of endodermal cells along neurenteric canal cephalad and to lateral positions,[5] which deciphered the decreasing occurrence of ECs from spine to head and lateral supertentorial position. Our case could be explained with a combination of 2 theories, which displayed a fascinating entity of uncertain embryopathogenesis.[5]

The primary complaint of patients with intracranial ECs is headache. Other symptoms include gait dysfunction, ataxia, and motor, sensory, and memory disturbances.[2,6] These signs may result from mass effect, inflammation, or irritation of adjacent brain tissues.[2] Supratentorial ECs are reported to have a larger size and later onset of symptoms than those in the posterior fossa.[2] The most frequent presentation was seizure, followed by

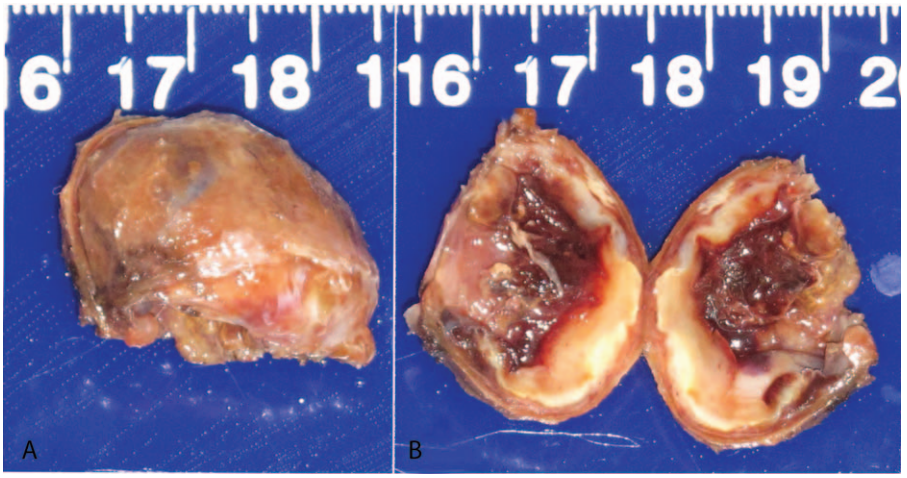

Figure 2. Macroscopic photographs of the appearance (A) and the contents (B) of the cyst. The cyst was about 2.6 × 2 × 2 cm in size.

headache, and motor, visual, and cognitive deficits.[2] Our patient showed a headache and memory deterioration, similar to the patient with intraparenchymal EC reported by Kitamura et al[7] who presented with headache and disorientation. It seems that the symptoms of intraparenchymal ECs are not distinctive from those of other intracranial lesions.

Imaging features of ECs are variable depending on cyst contents. On CT, ECs may appear as hypodense or hyperdense expansive lesions with a smooth margin.[2] On MRI, the signal intensity is typically isointense or slightly hyperintense on T1- and T2-weighted images.[2] The signal intensity reflects protein content in cyst fluid, where a higher proteinaceous cyst fluid often has a higher T1-weighted and lower T2-weighted signal.[8] Generally, ECs cannot be enhanced with contrast on both CT and MRI. Intracystic hemorrhage or fatty components could alter imaging presentation, making diagnosis of EC quite a challenge. In our patient, a higher T1- and T2-weighted signal cyst and slightly enhanced wall was observed, which was suspected as brain abscess based on otitis media history. Nonetheless, the lack of edema surrounding the cyst in the patient was inexplainable.

Diagnosis of EC is based on histological findings. Histologically, ECs are usually lined with pseudostratified or stratified, ciliated or nonciliated, cuboidal or columnar epithelium adhering to a fibrous base membrane and filled with clear, yellowish or brown milky substances. Immunohistochemical staining of epithelium is positive for epithelial membrane antigen, cytokeratin, and carcinoembryonic antigen but negative for neuroectodermal markers such as GFAP, S-100, and vimentin. Sometimes, the cyst wall involves specialized respiratory/gastroenteric mucosa, well-developed muscularis mucosa, or cartilaginous elements. Our patient showed a positive cytokeratin and negative GFAP and S-100, supporting the diagnosis of EC.

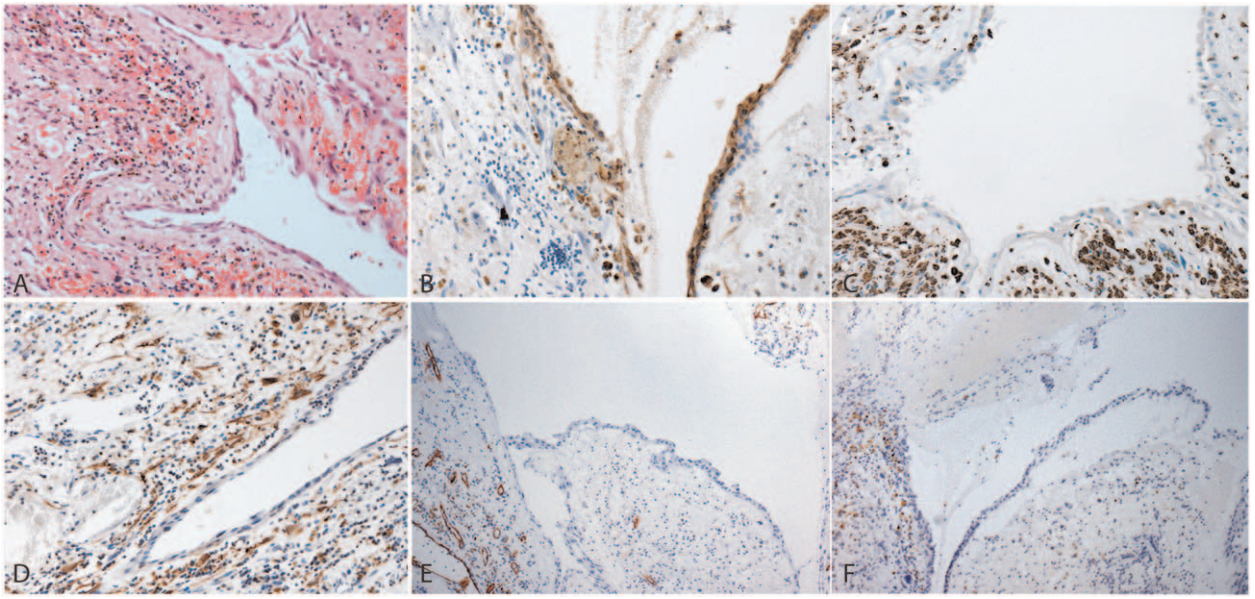

Figure 3. Histology of the cyst wall: (A) HE and (B–F) IHC. (B) AE1/AE3, (C) CD68, (D) SMA, (E) CD34, and (F) Ki-67 (original magnification: A–D, ×100; E and F, ×40). HE = hematoxylin-eosin, IHC, immunohistochemical, SMA = smooth muscle actin.

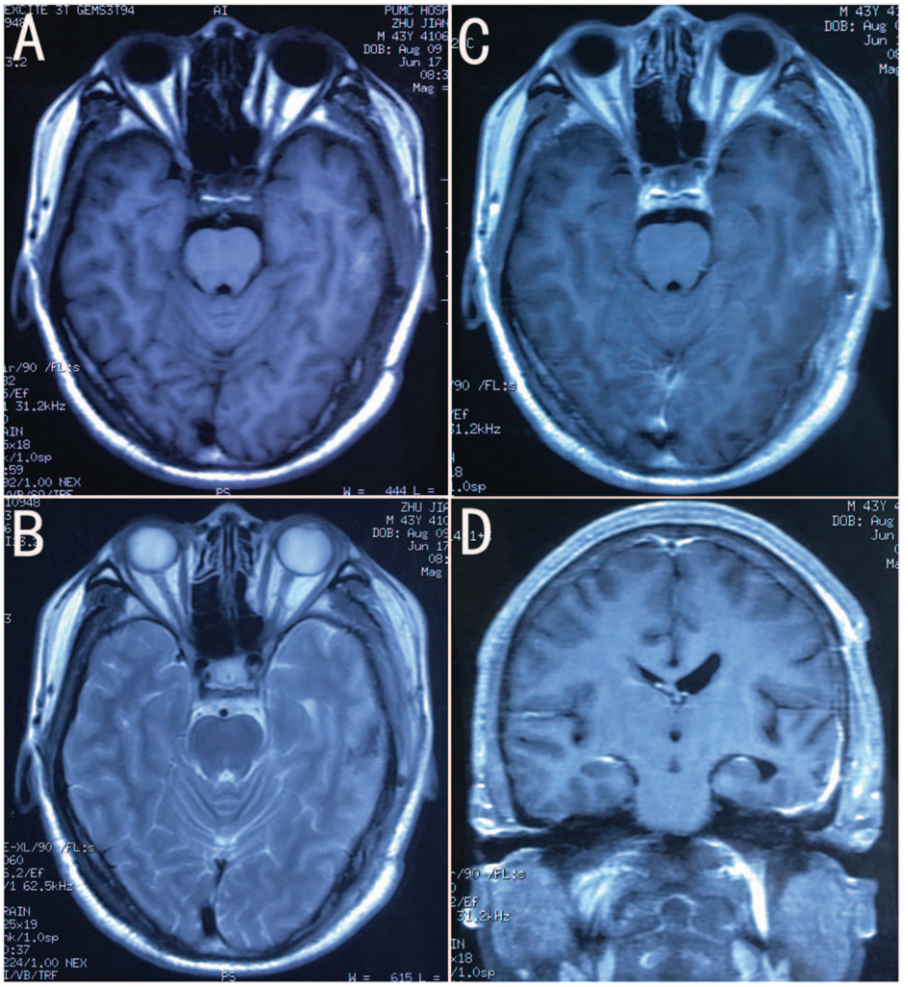

Figure 4. Three-month follow-up MRI. Axial T1-weighted (A) and T2-weighted (B) MRI; axial (C) and coronal (D) enhanced T1-MRI. MRI = magnetic resonance imaging.

Intracranial ECs are rarely related to spontaneous hemorrhage, and there are only 4 such cases reported till now.[7,9–11] Of all the reported cases, 1 showed hemorrhage in the subarachnoid space and 3 showed intracystic bleeding. Histological examination of the cyst wall found rich blood vessels, which were thought to be the cause of intracystic hemorrhage. Subarachnoid hemorrhage could be associated with rupture of cyst surface vessels. Both inflammation and leakage of cystic contents could cause rupture of cyst wall vessels.[7] We found a wide staining of vessels in the cyst wall, explaining the intracystic hemorrhage and slight enhancement of cyst wall. However, the hemorrhage confused imaging presentation, leading to misdiagnosis. This case told us that intracystic hemorrhage should be considered in cases of cystic lesions with enhanced walls.

A radical removal of ECs is widely accepted for preventing recurrence.[12] A review of recurrent intracranial ECs showed that cyst walls were left in part in the initial surgery of most cases and recurrence occurs at the same site from 2 months to 32 years.[12,13] Recurrent ECs could proliferate, migrate, or become malignant, and some of them could release cystic contents into subarachnoid space leading to chemical meningoencephalitis.[12,13] In 1 case of frontal lobe EC, Kitamura et al[7] reported that the cyst recurred at 34 months after cyst aspiration and there was occasional intense headache during the disease process. We performed a total cyst excision in the patient and 3-year follow-up found no recurrence of the lesion. Thus, a complete cyst removal is the best surgical strategy for EC.

4. Conclusions

Intraparenchymal EC with intracystic hemorrhage is rare. The EC should be considered in the diagnosis of intracranial cystic lesions. A complete removal is the best strategy of choice.

References

[1] Arai Y, Yamauchi Y, Tsuji T, et al. Spinal neurenteric cyst. Report of two cases and review of forty-one cases reported in Japan. Spine 1992;17:1421–4.
[2] Chakraborty S, Priamo F, Loven T, et al. Supratentorial neurenteric cysts: case series and review of pathology, imaging, and clinical management. World Neurosurg 2016;85:143–52.
[3] Small J. Pre-axial enterogenous cysts. J Neurol Neurosurg Psychiatry 1962;25:184.
[4] Batuk D, Koichiro Y, Tsukasea K, et al. Cerebellopontine angle endodermal cyst: a rare occurrence. Neurol India 2010;58: 676–7.
[5] Mittal S, Petrecca K, Sabbagh AJ, et al. Supratentorial neurenteric cysts—a fascinating entity of uncertain embryopathogenesis. Clin Neurol Neurosurg 2010;112:89–97.
[6] Miyagi A, Katayama Y. Neurenteric cyst arising in the high convexity parietal lesion: case report. Neurosurgery 2007;60:E203–4. discussion E204.

[7] Kitamura Y, Sasaki H, Hashiguchi A, et al. Supratentorial neurenteric cyst with spontaneous repetitive intracystic hemorrhage mimicking brain abscess: a case report. Neurosurg Rev 2014;37:153–9.
[8] Preece MT, Osborn AG, Chin SS, et al. Intracranial neurenteric cysts: imaging and pathology spectrum. AJNR Am J Neuroradiol 2006;27:1211–6.
[9] Khalatbari MR, Moharamzad Y. Spontaneous hemorrhage into a neurenteric cyst of the cerebellar vermis. Neuropediatrics 2011;42:116–8.
[10] Malcolm GP, Symon L, Kendall B, et al. Intracranial neurenteric cysts. Report of two cases. J Neurosurg 1991;75:115–20.
[11] Scarone P, Boissonnet H, Heran F, et al. Neurenteric cyst of the posterior fossa. Case report and review of the literature. Neurochirurgie 2009;55:45–52.
[12] Hashimoto M, Yamamoto J, Takahashi M, et al. Surgical strategy for intracranial endodermal cyst—case report. Neurol Med Chir (Tokyo) 2011;51:531–4.
[13] Kimura H, Nagatomi A, Ochi M, et al. Intracranial neurenteric cyst with recurrence and extensive craniospinal dissemination. Acta Neurochir (Wien) 2006;148:347–52. discussion 352.