Hemorrhagic Epidermoid Cyst in Cerebellar Vermis: Case Report and Review of the Literature

Ján Kozák¹ Jozef Šurkala¹ Martin Novotný¹ Marián Švajdler²,³

¹Department of Neurosurgery, Faculty of Medicine Comenius University and University Hospital Bratislava, Slovakia
²Biopťicka Laborator s.r.o., Pilsen, Czech Republic
³Šílk’s Department of Pathology, Charles University, Faculty of Medicine and Faculty Hospital in Pilsen, Pilsen, Czech Republic

Abstract

Intracranial epidermoid cysts are slow growing congenital avascular neoplasms that spread across the basal surface of the brain. They most commonly occur in the paramedial region in the cerebellopontine angle and the parasellar region. Despite its generally benign nature, sporadically they can be accompanied with hemorrhage or very rarely undergo malignant transformation. The authors present a case report of a patient with a hemorrhagic vermian epidermoid cyst and a review of all published similar cases.

Keywords
► epidermoid cyst
► hemorrhagic epidermoid cyst
► vermian epidermoid cyst
► midline posterior fossa

Introduction

Intracranial epidermoid cysts (ECs) are rare congenital intracranial neoplasms accounting for approximately 1.5% of all intracranial tumors. Pathophysiologically, ECs arise from inclusion of ectodermal epithelial elements and grow by the accumulation of keratin and cholesterol crystals, which are breakdown products created by the desquamation of epithelial cells.¹ ECs tend to spread across the basal surface of the brain with a paramedial preference such as the cerebellopontine angle and the parasellar region, and usually reach large size before patients become symptomatic.¹,² Midline posterior fossa ECs involving the cerebellar vermis are rare and have only been reported in isolated reports when accompanied with hemorrhage. When hemorrhage occurs in EC, the diagnosis is challenging and there is a potential for misdiagnosis. Total resection of EC is usually the definitive treatment with very low recurrence rate.

In this case report we describe the clinical characteristics of a patient with a hemorrhagic vermian EC. In addition, an overview of all similar published cases is presented. We systematically reviewed English written literature using PubMed database to search for relevant English language articles published up to November 2019. All eligible studies were analyzed, and the references were checked for additional relevant publications. Information and data were extracted from all included literature. To the best of our knowledge, our case represents only the sixth reported case of hemorrhagic vermian EC in the English written literature.

Case Report

A 57-year-old deaf-mute female was admitted to our department with a short history of repeated loss of consciousness. Neurologic examination revealed global passivity with inability to walk. Computed tomography (CT) revealed a midline...
infratentorial hyperdense lesion located in the cerebellar vermis causing compression of the fourth ventricle with no surrounding edema. On axial CT scan obtained with a bone setting, a small-calciﬁed spot was present at the border of the lesion, but no hyperplasia or erosion of the adjacent bone was seen. Obstructive hydrocephalus was present (►Fig. 1). Magnetic resonance imaging (MRI) showed atypical cystic tumor with hemorrhagic/high-protein content and vascularized nodus corresponding to calciﬁed lesion on CT. On T1-weighted and T2-weighted images the lesion was hyperintense and hypointense, respectively and demonstrated no contrast enhancement. Due to tumorous expansion, syrinx of cervical spinal cord was present (►Fig. 2). The patient was operated on under general anesthesia. At ﬁrst, external ventricular drain was inserted, with intraventricular pressure been only 60 mm of water column. Then, wide suboccipital craniotomy and laminectomy of the ﬁrst cervical vertebra were performed in a semi sitting position. Dark cyst with a greenish mud-like debris content was identiﬁed (►Fig. 3), and the entire cyst wall with its content was completely removed. Grossly, no hair or yellow cheesy sebaceous material was present. Histological examination revealed organizing blood clot and lightly basophilic lamellar keratin with dystrophic calciﬁcations and heterotopic ossiﬁcation. Few strips of squamous epithelium immunohistochemically positive for CK5/6 and P40 were identiﬁed. No dermal skin appendages, such as hair follicles or sebaceous glands were present (►Fig. 4). According to imaging studies, intraoperative characteristics, and histological examination of the diagnosis of hemorrhagic EC were established. Postoperatively, there was a rapid improvement of neurological status and the patient was immediately alert and able to walk. There were no signs of chemical meningitis in the postoperative period. Control MRI scan showed radical resection of the tumor.

Discussion

Intracranial ECs, also known as pearly tumors are congenital avascular intracranial neoplasms that account for approximately 1.5% of all intracranial tumors and 7 to 9% of all cerebellopontine angle tumors.1–4 ECs are thought to result from aberrantly located ectodermal cells, that have been entrapped during the process of neurulation in the period between the third and ﬁfth gestational weeks. Typical slow and
**Fig. 3** Intraoperative microscopic view of hemorrhagic vermian EC. Via midline suboccipital craniotomy we identified a dark cyst with a greenish mud-like debris. (a,b) The supposed calcified fragment (white arrow, a). During the procedure, a few pearl-like particles were found on the periphery of the lesion (black asterisks, c). During the procedure, a gross total resection was achieved.

**Fig. 4** Histological features of hemorrhagic EC. Microscopic examination revealed organizing blood clot (a, hematoxylin-eosin), and lightly basophilic keratin lamellae (b, hematoxylin-eosin), with dystrophic calcifications and heterotopic ossification (c, hematoxylin-eosin). Only few strips of benign squamous epithelium were found, with no adnexal structures (d, hematoxylin-eosin). The epithelium was immunohistochemically positive for squamous markers CK5/6 (d, inset) and P40. EC, epidermoid cyst.
linear rate of growth of EC is caused by accumulation of desquamated keratin, cellular debris, and cholesterol crystals. Slow progression of EC permits presentation of EC at large sizes and prolonged symptomatology. Symptoms are generally attributed to effects of a space-occupying lesion. Infrequently, EC may present with a history of recurring aseptic (chemical) meningitis caused by spontaneous rupture of the highly irritative cyst contents into the subarachnoid space. The resulting response to the leakage of the irritative cyst content and resultant inflammation is responsible for the firm adherence of the capsule to the surrounding neurovascular structures. Occasional, granulation tissue is responsible for the firm adherence of the capsule to the surrounding neurovascular structures. Occasionally, granulation tissue is responsible for the firm adherence of the capsule to the surrounding neurovascular structures. It was proposed that hemorrhage within an EC may arise from the richly vascularized granulation tissue, that forms in response to the leakage of the irritative cyst content and resultant inflammation. Occasionally, granulation tissue is responsible for the firm adherence of the capsule to the surrounding neurovascular structures. Similarly, Hasegawa et al. surmised that bleeding into the cyst originated in the fibrous nodules of the cyst wall, which contained vessels. Clotted blood, hemosiderin-laden macrophages, and neurovascularity in the cyst wall granulation tissue were also found in a case reported by Hsieh. According to Xiaohui et al, the pathogenesis of hemorrhage in the granulation tissue has two explanations. The first explanation is that the hemorrhage results from the mechanical tearing of the vascularized granulation tissue patches due to the outgrowth of the cyst contents. The second explanation is that hemorrhage results from the erosive degeneration of the vessels caused by the contents of the cyst.

### Table 1 Review of published hemorrhagic vermian ET

| Case no. (author) | Age (years), Sex | CT/MRI findings | Cyst contents | Extent of resection | Outcome/F-U |
|------------------|-----------------|-----------------|--------------|-------------------|-------------|
| 1.5              | 24, F           | Diameter: 4 cm  | Old bleeding, yellow cheese-like | GTR               | No neurologic deficit, F-U not mentioned. |
|                  |                 | CT: not specified. MRI: not specified. Preoperative dg.: epidermoid. |              |                   |             |
| 2.5              | 43, F           | Diameter: 5 cm, | Old bleeding, yellow cheese-like | GTR               | No neurologic deficit, F-U not mentioned. |
|                  |                 | CT: not specified. MRI: not specified. Preoperative dg.: epidermoid. |              |                   |             |
| 3.9              | 21, M           | Diameter: not noticed. CT: hyperdense lesion. MRI: T1 hypointense + hyperintense nodule. T2 hypointense + hyperintense nodule. CE: no preoperative dg.: not mentioned. | Mud-like, cheesy debris | GTR | No neurologic deficit, 1 y. |
| 4.5              | 30, F           | Diameter: not specified. CT: hyperdense lesion. MRI: not noticed. Preoperative dg.: hemorrhage to tumor. | Brownish cheesy | GTR | No neurologic deficit, 4 y. |
| 5.4              | 19, F           | Diameter: 6 cm, | Greenish tumor | GTR               | No neurologic deficit, F-U not mentioned. |
|                  |                 | CT: hyperdense lesion with small-calciﬁed spot. MRI: T1 hypointense + hyperintense nodule. T2 hypointense + isointense nodule. CE: nodule enhancement Preoperative dg.: not mentioned. |              |                   |             |
| 6.                | 57, F           | Diameter 5 cm,  | Greenish mud-like debris, crystals | GTR | No neurologic deficit, 6 mo. |
|                  |                 | CT: hyperdense lesion with small-calciﬁed nodule. MRI: T1 hyperintensive T2 hypointensive CE: no preoperative dg.: vascular lesion (AVM, cavernoma). |              |                   |             |

Abbreviations: AVM, arteriovenous anomaly; CE, contrast enhancement; F-U, follow-up; GTR, gross total removal.
Hemorrhage into EC makes diagnosis quite challenging. Hyperdensity seen CT, that refers to hemorrhage, is not characteristic. Moreover, various authors reported hyperdensity of intracranial EC, caused by high-concentrated protein content of the cyst. In the series by Li et al, hyperdense EC represented 3% of all cases. The authors assumed that recurrent leakage of the cyst contents and a subsequent chemical inflammatory response may be responsible for the high density of some EC on CT. Likewise, causes of atypical signal changes present on MRI, except of hemorrhage, include calcification and increased protein concentration as a result of inflammation or infection. Importantly, sudden development of symptoms and exponential growth of EC with a novel finding of contrast enhancement seen on imaging studies may suggest malignant transformation.

Although extremely rare, malignant transformation into squamous cell carcinoma confers poor prognosis. Careful histological examination of hyperdense EC lesions is essential to differentiate EC from other diseases, and to rule out malignant transformation, which would require adjuvant therapy after surgery.

**Conclusion**

Hemorrhagic vermican ECs are rare and diagnostically challenging lesions. Gross total resection is the goal of surgical treatment and when ECs are located in cerebellar vermis this is often fully achieved. Histological examination is crucial to rule out malignant changes.

**Conflict of Interest**

None declared.

**References**

1. Yaşargil MG, Abernathey CD, Sarioglu AC. Microneurosurgical treatment of intracranial dermoid and epidermoid tumors. Neurosurgery 1989;24(04):561–567
2. Balogun JA, Adeleke NA, Salami AO, Odebode TO. Cerebellar vermican epidermoid tumor: a report of two cases. World Neurosurg 2018;112:153–157
3. Singh N, Symons SP, Montanera W, et al. Hemorrhagic epidermoid cyst in a patient with generalized tonic clonic seizure. J Clin Neuroradiol 2013;20(05):750–752
4. Li F, Zhu S, Liu Y, Chen G, Chi L, Qu F. Hyperdense intracranial epidermoid cysts: a study of 15 cases. Acta Neuroradiol (Wien) 2007;149(01):31–39, discussion 39
5. Ren X, Lin S, Wang Z, et al. Clinical, radiological, and pathological features of 24 atypical intracranial epidermoid cysts. J Neuroradiol 2012;116(03):611–621
6. Gupta S, Vaishya ND, Senger RL. Hyperdense intracranial epidermoid: an uncommon presentation. Neurol India 2000;48(02):158–160
7. Chen CY, Wong JS, Hsieh SC, Chu JS, Chan WP. Intracranial epidermoid cyst with hemorrhage: MR imaging findings. AJNR Am J Neuroradiol 2006;27(02):427–429
8. Greenberg MS. Handbook of Neurosurgery. 8th ed. New York: Thieme; 2016:756–765
9. Tao CY, Wei P, Wang JJ, You C. Hemorrhagic epidermoid cyst in cerebellar vermis. Turk Neurosurg 2015;25(05):828–830
10. Gadgil N, Humphries WE III, Clay Goodman J, Gopinath SP. Hemorrhagic intradiploic epidermoid cyst. Clin Neurol Neurosurg 2013;115(12):2549–2551
11. Hsieh CH, Huang KM, Kao MC, Peng S, Wang CC. Hemorrhage in intracranial epidermoid cyst. J Formos Med Assoc 1996;95(02):173–175
12. Tsurushima H, Kamezaki T, Tomono Y, Nose T. Intracranial epidermoid cyst including elements of old hematoma. Neurol Med Chir (Tokyo) 1997;37(11):861–864
13. Hasegawa H, Bitoh S, Nakata M, Fujiwara M, Yasuda H. Intracranial epidermoid mimicking meningioma. Surg Neurol 1981;15(05):372–374
14. Roh TH, Park YS, Park YG, Kim SH, Chang JH. Intracranial squamous cell carcinoma arising in a cerebellopontine angle epidermoid cyst: a case report and literature review. Medicine (Baltimore) 2017;96(51):e9423
15. Kokkalis P, Chamilos C, Sgouros S. Epidermoid cyst of the fourth ventricle in a child. Childs Nerv Syst 2018;34(01):29–30
16. Cuoco JA, Rogers CM, Busch CM, Apfel LS, Entwistle JJ, Marvin EA. Intracranial squamous cell carcinoma arising from a cerebellopontine angle epidermoid cyst remnant four decades after partial resection. Front Oncol 2019;9:694
17. Muto J, Yoshida K, Momoshima S, Kazuno M, Kazuno T. Intracranial epidermoid tumor with changes in signal intensity on magnetic resonance imaging because of non-hemorrhagic pathology: case report. Neurol Med Chir (Tokyo) 2010;50(10):936–938