



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

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Abstract

Keywords

- ▶ epidermoid cyst
- ▶ hemorrhagic epidermoid cyst
- ▶ vermian epidermoid cyst
- ▶ midline posterior fossa

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.

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

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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

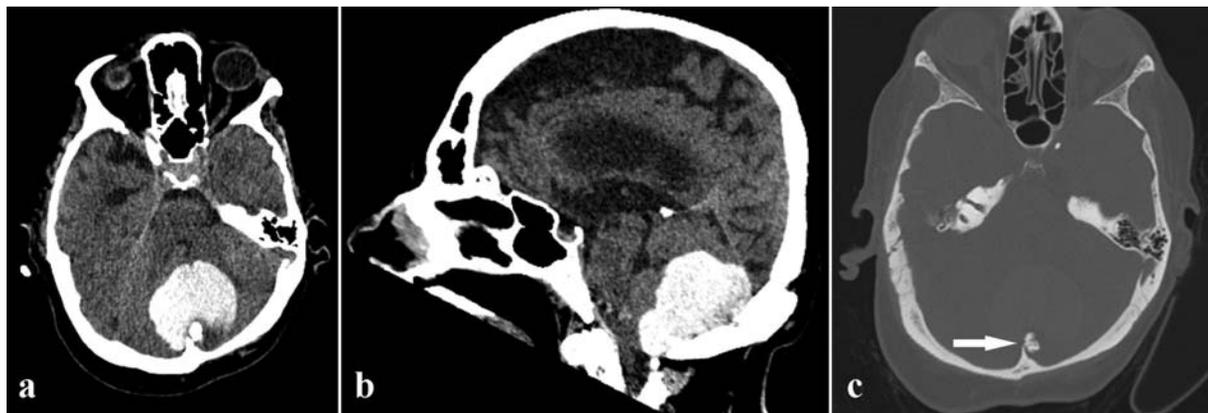


Fig. 1 CT scan of hemorrhagic vermian EC. Native CT scan in axial plane (a) and sagittal plane (b) revealed a midline infratentorial hyperdense lesion causing compression of the fourth ventricle and brainstem. CT scan with bone setting (c) revealed a small, calcified spot (white arrow). CT, computed tomography; EC, epidermoid cyst.

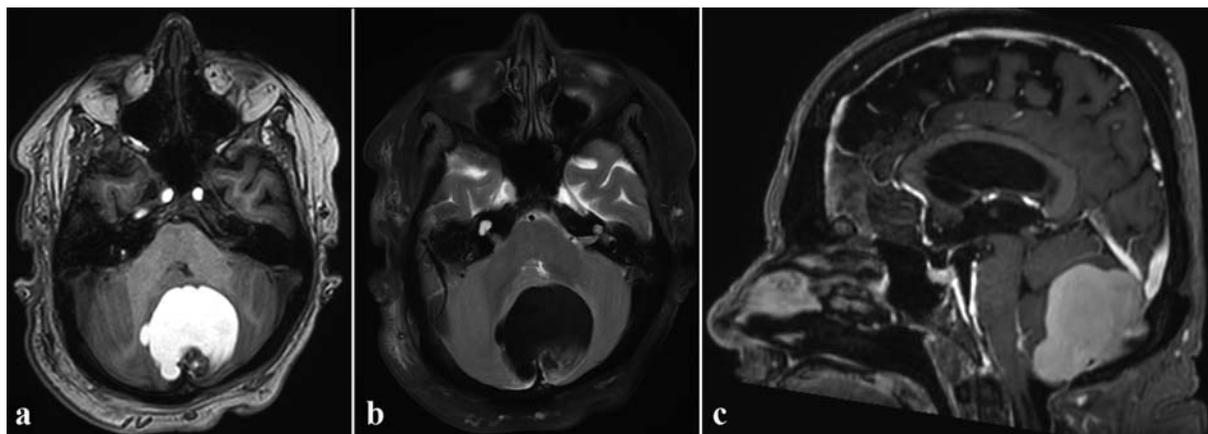


Fig. 2 MRI of hemorrhagic vermian EC. MRI showed a lesion located in cerebellar vermis—hyperintense on T1-weighted images (a) and hypointense on T2-weighted images (b). On T1-weighted imaging with gadolinium in sagittal plane (c) the tumor did not show any contrast enhancement. EC, epidermoid cyst; MRI, magnetic resonance imaging.

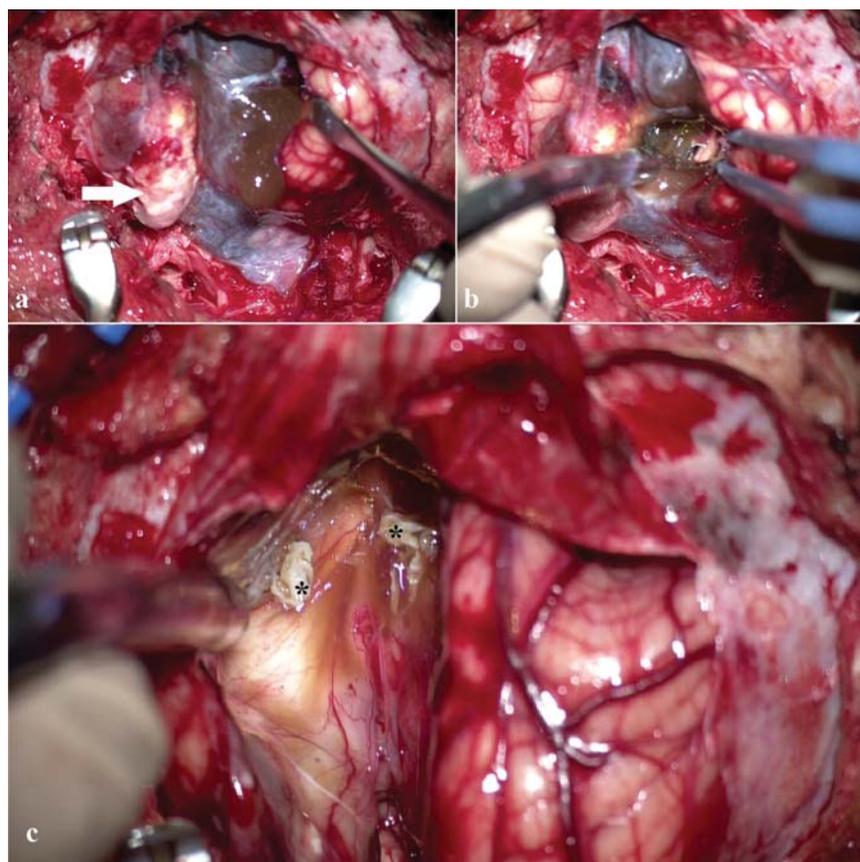


Fig. 3 Intraoperative microscopic view of hemorrhagic vermian EC. Via midline suboccipital craniotomy we identified a dark cyst with a greenish mud-like debris. (a,b) The supposed calcified fragment (*white arrow*, a). During the procedure, a few pearl-like particles were found on the periphery of the lesion (*black asterisks*, c). During the procedure, a gross total resection was achieved.

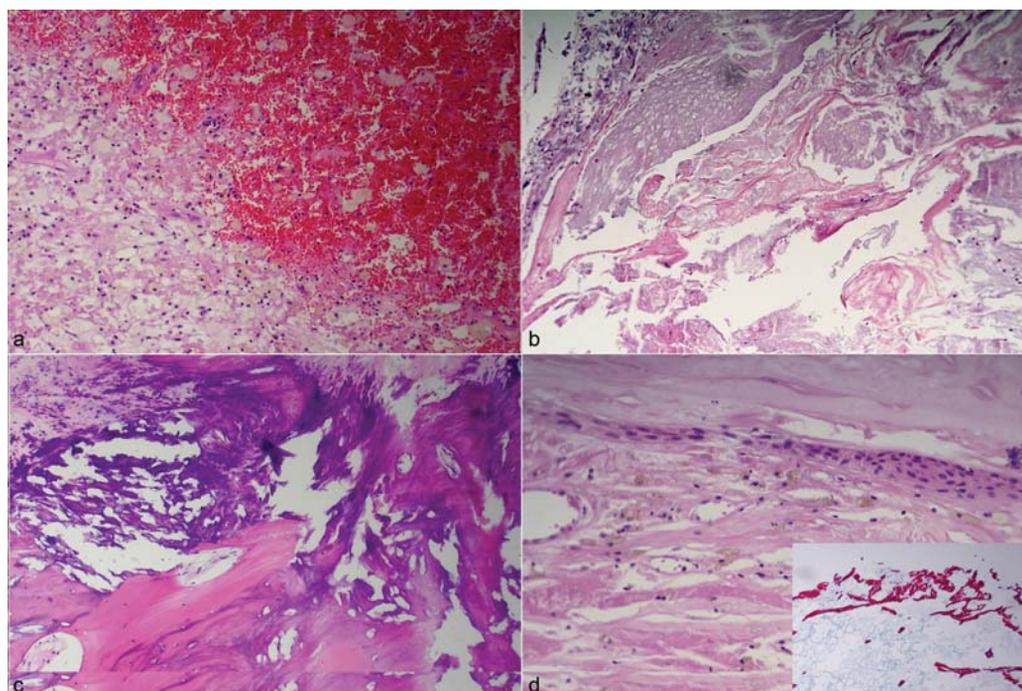


Fig. 4 Histological features of hemorrhagic EC. Microscopic examination revealed organizing blood clot (a, hematoxylin-eosin), and lightly basophilic keratin lamellae (b, hematoxylin-eosin), with dystrophic calcifications and heterotopic ossification (c, hematoxylin-eosin). Only few strips of benign 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.² 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 preference for paramedian location and histological constituents differentiates epidermoid from dermoid cysts, which are more likely to be situated along midline structures and microscopically contain dermal appendage.^{1,2,5} Consequently midline posterior fossa EC involving the cerebellar vermis are uncommon.^{2,6} Treatment of EC consists of complete microsurgical removal of cyst lining and its content. However, complete excision may be challenging, especially when the tumor and neurovascular structures are closely apposed. In such cases, subtotal removal of the cyst is advocated.^{1,2,5}

On CT imaging, typical EC is described as hypodense and well demarcated lesion. Calcifications are rarely described, and when present, usually marginally located. On MRI ECs usually present as hypointense and hyperintense on T1 and T2-weighted images, respectively without contrast enhancement due to the absence of a well-developed vascular network,^{1,2,5,7,8} although minor rim enhancement has been reported.³ Diffusion-weighted imaging shows intense signal as a result of restriction of water movement and this diffusion restriction differentiates EC from other cystic lesions.⁸ Intra-

operatively ECs are typically well-defined lesions with an irregular nodular outer surface and a shiny mother of pearl appearance. Microscopically, the wall of the EC consists of a layer of stratified squamous epithelium without vascularity.⁷ Due to avascular nature of EC, intralesional hemorrhage is extremely rare and has only been reported a few times.^{3-7,9-11} In our literature review, we found five cases of vermian hemorrhagic EC with our case to be the sixth (► **Table 1**).

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.¹⁰ Occasionally, granulation tissue is responsible for the firm adherence of the capsule to the surrounding neurovascular structures.¹ Ren et al found hemorrhage in granulation tissue in 90.5% of cases in their series of EC with hemorrhage.⁵ Similarly Hasegawa et al surmised that bleeding into the cyst originated in the fibrous nodules of the cyst wall, which contained vessels.¹² Clotted blood, hemosiderin-laden macrophages, and neurovascularity in the cyst wall granulation tissue were also found in a case reported by Hsieh.¹¹ 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.⁵

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 CT: not specified MRI: not specified preoperative dg.: epidermoid.	Old bleeding, yellow cheese-like	GTR	No neurologic deficit, F-U not mentioned.
2. ⁵	43, F	Diameter: 5 cm. CT: not specified. MRI: not specified. preoperative dg.: epidermoid.	Old bleeding, yellow cheese-like	GTR	No neurologic deficit, F-U not mentioned.
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. ⁶	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 CT: hyperdense lesion with small-calcified spot. MRI: T1 hypointense + hyperintense nodule. T2 hypointense + isointense nodule. CE: nodule enhancement preoperative dg.: not mentioned.	Greenish tumor	GTR	No neurologic deficit, F-U not mentioned.
6. Presented case	57, F	Diameter 5 cm CT: hyperdense lesion with small-calcified nodule. MRI: T1 hyperintense T2 hypointense CE: no preoperative dg.: vascular lesion (AVM, cavernoma).	Greenish mud-like debris, crystals	GTR	No neurologic deficit, 6 mo.

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.^{3,4,6,13} 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.⁴ 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.^{3,5} 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.^{2,4,14–17}

Although extremely rare, malignant transformation into squamous cell carcinoma confers poor prognosis.¹⁶ 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.⁹

Conclusion

Hemorrhagic vermian 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.

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