Cerebellar Vermian Epidermoid Tumor: A Report of 2 Cases

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Key words
- Chemical meningitis
- Congenital
- Epidermoid tumor
- Vermis

Abbreviations and Acronyms
CPA: Cerebellopontine angle
CT: Computed tomography
FLAIR: Fluid-attenuated inversion recovery
MRI: Magnetic resonance imaging
T1WI: T1-weighted imaging
T2WI: T2-weighted imaging

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BACKGROUND: **Epidermoid tumors are rare, benign slow-growing congenital tumors, most frequently located in the cerebellopontine angle of the intracranial cavity. They usually grow to a large size before patients become symptomatic. Although these tumors are amenable to surgery, their adherence to neurovascular structures poses a surgical challenge that results in subtotal resection, thus increasing the risk of recurrence.**

CASE DESCRIPTION: **We report 2 adult patients whose imaging studies revealed epidermoid tumors located in the cerebellar vermis, an uncommon site for such tumors. The patients presented with variable symptomatology. We highlight the imaging features and challenges of surgery. Both patients had good outcomes, with resolution of symptoms and neurologic deficits.**

CONCLUSIONS: **A safe complete excision of epidermoid tumor and its capsule is possible with a good understanding of their clinical and radiologic features and a high index of suspicion. To the best of our knowledge, this is the first report of cerebellar vermian epidermoid tumors from sub-Saharan Africa.**

INTRODUCTION

Epidermoid tumors, also known as "pearly tumors," are rare congenital intracranial neoplasms accounting for roughly 0.2%–1.8% of all intracranial tumors[1-3] and 7%–9% of all cerebellopontine angle (CPA) tumors,[4,5] representing the third-most common tumor in the CPA after vestibular schwannomas and meningiomas.[6] Midline posterior fossa epidermoid tumors involving the cerebellar vermis or the fourth ventricle are rare, particularly the former type.[7] These tumors are slow-growing, with linear growth, often leading to presentation at large sizes and late symptomatology.[1,8] A high index of suspicion and adequate evaluation of appropriate magnetic resonance imaging (MRI) sequences[6,7,9-11] are key to the diagnosis of these tumors, particularly in the vermian region of the posterior fossa, where it can simulate other tumors.

Complete surgical excision of the tumor, including removal of the capsule when safe, is advocated to reduce the risk of recurrence.[1,8,12] The challenges to complete excision often lie in the intimate relationship of the tumor to neurovascular structures.[1,8,9,12,13] Here we report 2 patients with epidermoid tumors in the rare cerebellar vermian location who presented in the third and sixth decades of life.

CASE PRESENTATIONS

Case 1

This 53-year-old clergyman presented with a history of recurrent headaches and complex partial seizures with secondary generalization of 3 years duration, with ataxia of 2 months duration and blurry vision of 1 month duration. There was no history of head trauma or surgery. Clinical examination revealed a middle-aged man, awake, with a broad-based gait, slow mentation, short-term memory impairment, and bilateral papilloedema. There were no lateralizing signs; power was grade 4 in all muscle groups, and plantar response was extensor bilaterally. Moderate truncal ataxia was present, with a tendency to fall to the side.

A cranial computed tomography (CT) scan revealed an irregular cystic mass in the midline of the cerebellum involving the vermis and the cisterna magna. On CT, the mass showed faint marginal contrast enhancement with some solid areas within it. The dilated fourth ventricle was distinct from the mass. Brain MRI revealed a vermian mass that was hypointense on T1-weighted imaging (T1WI) and hyperintense on T2-weighted imaging (T2WI) without contrast enhancement, seen distinctly from the fourth ventricle, compressing the brainstem anteriorly and related to the posterior inferior cerebellar artery (Figure 1). There was a moderate to severe dilatation of the lateral ventricles with transependymal seepage of cerebrospinal fluid and periventricular flare. A suboccipital craniectomy and near total excision of a pearly tumor was done, leaving a cuff of tumor around the posterior inferior cerebellar artery and a thin film of the capsule adherent to the brainstem. The postoperative course was uneventful, and the patient was discharged on postoperative day 4 with no new neurologic deficits. The histological examination confirmed an epidermoid tumor. At the time of this report, he was 6 months postsurgery

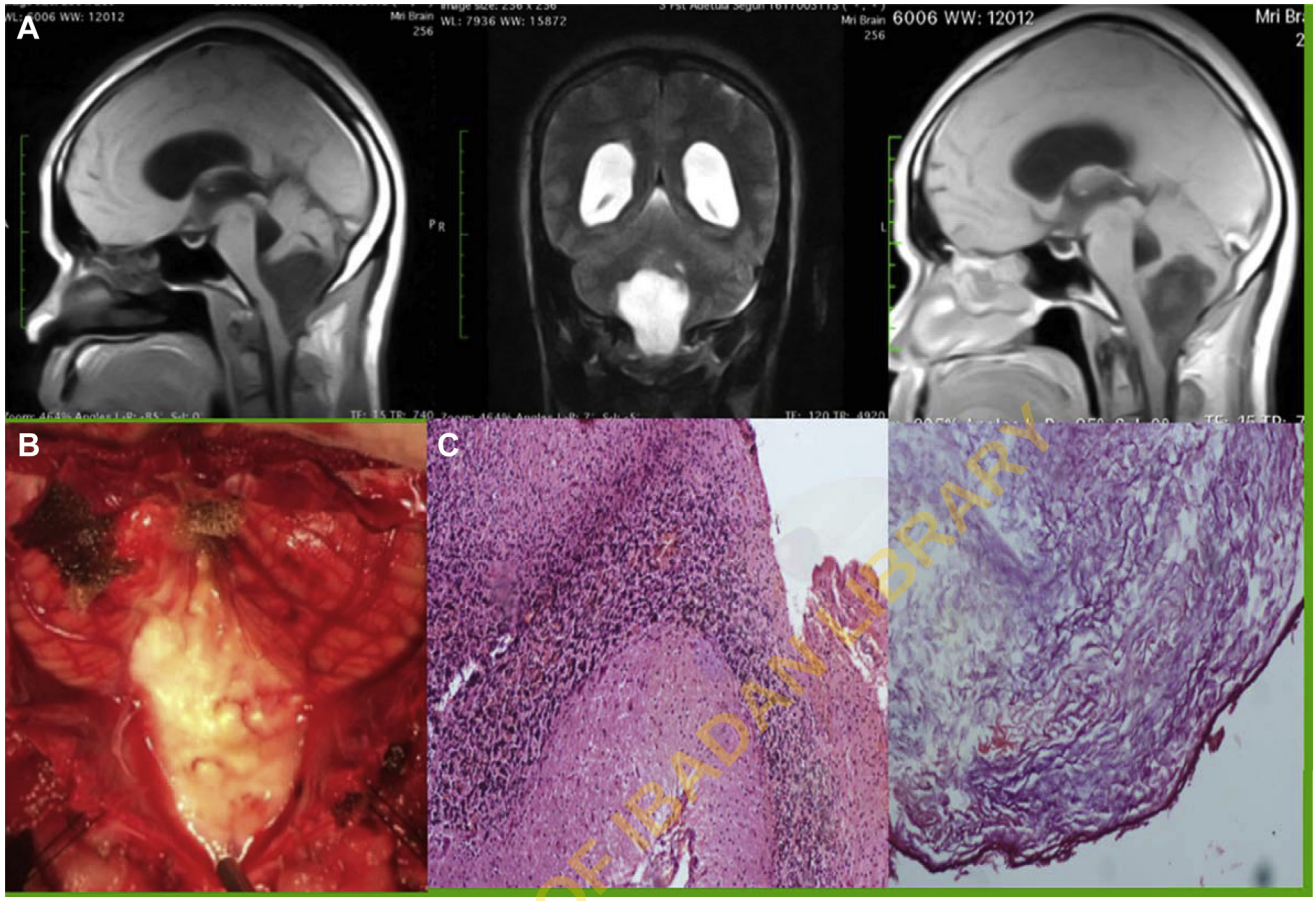

Figure 1. (**A**) Magnetic resonance imaging of the tumor, hypointense on T1-weighted imaging and hyperintense on T2-weighted imaging, with minimal contrast enhancement. (**B**) Intraoperative appearance of the pearly vermian tumor. (**C**) Adjoining normal cerebellar tissue and cysts containing keratin squames.

with complete resolution of seizures, ataxia, and blurred vision. He has since returned to work.

Case 2

This 29-year-old, right-handed female primary school teacher presented with staggering gait and occipital headaches of 2 months duration, along with left visual impairment associated with double vision of 6 weeks duration. There was no antecedent trauma or fever. The most significant clinical finding was truncal ataxia. Brain MRI revealed a cerebellar vermian lesion of mixed intensity on T1WI and hyperintense on T2WI, with heterogenous enhancement and mildly dilated lateral and third ventricles. The lesion was distinct from the fourth ventricle. The lesion was also of mixed intensity on a fluid-attenuated inversion recovery (FLAIR) MRI sequence (Figure 2). Further evaluation did not reveal any other primary lesion or evidence of metastasis.

The patient subsequently underwent a suboccipital craniectomy and complete excision of an encapsulated cerebellar vermian tumor, which appeared pearly with a cheesy intracapsular content. There was some adherence of the capsule to the suboccipital surface of the cerebellum and brainstem from which it was removed by microsurgical dissection. The patient had a transient postoperative cerebrospinal fluid leak, which was successfully managed nonoperatively. She was discharged on postoperative day 8 due to her preference to remain in the hospital until after suture removal. There were no complications. Histological analysis revealed an epidermoid tumor. Her symptoms remained resolved at 6 months postsurgery.

DISCUSSION

Epidermoid tumors are benign congenital tumors that grow slowly in a linear manner as opposed to the exponential growth seen in other neoplastic lesions, with patients presenting with symptoms over a long period.[9,13-15] These tumors are thought to result from the entrapment of aberrant ectodermal cells during the process of neurulation, which normally occurs between the third and fifth gestational weeks.[16] Epidermoid tumors tend to spread across the basal surface of the brain with a lateral preference, such as the CPA, where it forms 7% of the

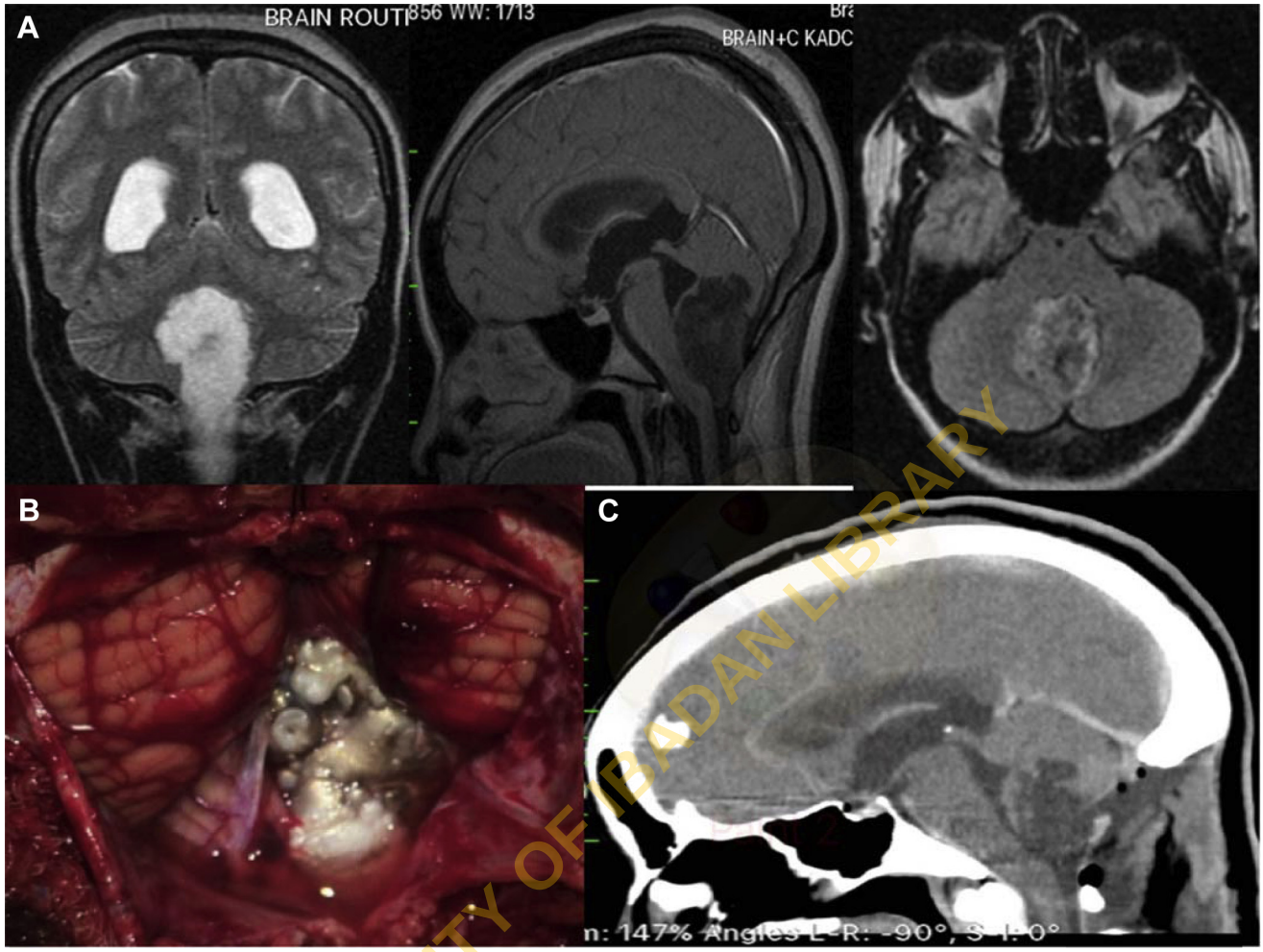

Figure 2. (**A**) A vermian tumor predominantly hypointense on T1-weighted magnetic resonance imaging (MRI) and hyperintense on T2-weighted imaging but heterogenous on fluid-attenuated inversion recovery MRI. (**B**) The tumor intraoperatively. (**C**) Postoperative computed tomography scan showing gross total tumor resection.

tumors in that location, as well as the parasellar region.[8] The preference for a paramedian location tends to differentiate epidermoid tumors from dermoid tumors, which are more often situated along midline structures.[8,17] The lateral preference of epidermoid tumors especially in the CPA is attributed to the possibility of proliferation of multipotential embryonic rests or the transplantation of epithelial rests carried laterally with the migrating otic vesicles or developing neurovasculature.[3,8] Epidermoid tumors grow when keratin and cholesterol from desquamated epithelial cells accumulate. With progressive accumulation, the cyst extends into the subarachnoid space, assuming the shape of it the cavity in which it is found, with a late displacement of adjacent normal neurovascular structures, which accounts for the late symptomatology and large size at presentation.[1,8] These tumors have been described in various locations in the intracranial space, with the CPA the most commonly reported location.[1,5,8,18-20] Midline posterior fossa epidermoid tumors involving the fourth ventricle or the cerebellar vermis are rare, particularly the latter,[7,17,21,22] and especially so in children.[9,23] In our 2 patients, the tumors were located in the cerebellar vermis.

The mean age at presentation of intracranial epidermoid tumors, including those located in the posterior fossa, is 40 years.[18] There appears to be no difference in the mean interval from the onset of symptoms to presentation of posterior fossa epidermoid tumors, which ranges from 2.4 to 3.8 years.[1,12,20] This interval also holds true for the midline posterior fossa tumors,[17] although the mean duration of symptoms in one study was 10 months.[7] Although the patient in our case 1 presented within the average interval, the patient in our case 2 presented within 2 months of symptom onset.

The symptoms of epidermoid tumors vary depending on tumor location. Most patients present with features of elevated intracranial pressure, including headaches,

vomiting, and visual symptoms. Posterior fossa tumors present with cerebellar signs, including ataxia, dysmetria, dysarthria, and nystagmus.[8,13,24] Some patients may present with symptoms of chemical meningitis, cranial nerve paresis, and long tract involvement.[1,13,24] These features are also invariably seen in the midline posterior fossa lesions,[7] as demonstrated in our 2 cases.

Epidermoid tumors are described as hypodense lesions on CT with no enhancement. They are hypointense and hyperintense on T1WI and T2WI without contrast enhancement.[10,11] Significant enhancement of the capsule may be seen in recurrent tumors[1]; however, they are slightly to moderately hyperintense on FLAIR sequences, and show heterogeneous intensities differentiating them from arachnoid cysts, which are hypodense on FLAIR sequences.[7,9-11] On diffusion-weighted imaging, epidermoid cysts are seen as hyperintense lesions, whereas in contrast, apparent diffusion coefficient mapping shows hypointense lesions in the posterior fossa.[10,11]

Surgery remains the optimal treatment for de novo tumors, most importantly in younger patients.[3] Complete excision of the tumor with removal of the capsule, which contains the stratified squamous epithelium considered the real disease,[1,8] is done using microsurgical techniques and adjuncts, such as intraoperative neurophysiological monitoring, neuronavigation, and endoscope-assisted dissection, to facilitate cure.[1,12] The attachment of the capsule to critical structures, such as perforating arteries, brainstem, and cranial nerves, may limit complete excision, however,[1,8,9,12,13] and predispose to tumor recurrence with the attendant increased risk at repeat surgery, which may have a significant impact on a patient's quality of life.[1,8] It has been suggested that chemical meningitis, which may result from a break in the capsule,[9] may provoke a strong inflammatory reaction, leading to adherence of the capsule to critical structures and resulting in incomplete excision.[12]

Postoperative complications following epidermoid tumor excision, which were absent in the two reported cases, include aseptic meningitis, cranial nerve palsies, communicating hydrocephalus, tumor recurrence,[7-9,12] and neurogenic pulmonary edema.[7] Tumor location in the midline in the posterior fossa may especially predispose to chemical meningitis, which is often associated with incomplete excision of the capsule and spillage of the contents into the subarachnoid space.[5,7,8] Postoperative administration of corticosteroids tends to reduce the risk of chemical meningitis.[5,8,9,13] Clinical recurrence is said to be low when only small fragments of the cyst lining are left behind.[12,25] Therefore, if the assumption that with a single residual tumor cell, the patient is at risk of tumor recurrence for a duration equal to his or her age at diagnosis plus 9 months is correct,[14] then the tumor might not become symptomatic within his or her lifetime. Nonetheless, serial follow-up with neuroimaging is advised to monitor asymptomatic recurrences and to detect occasional earlier symptomatic recurrences.[7]

Malignant degeneration of postoperative residual epidermoid tumors, although rare, can occur and is characterized by early and rapid progression of symptoms with contrast enhancement and aggressive neuroimaging findings with poor prognosis, as opposed to the findings in benign primary lesions.[7,26-28] Malignant transformation has been reported to occur in primary epidermoid cysts that were not surgically removed.[2,27,29]

Although surgery remains the treatment modality for recurrent tumors, there is some evidence supporting the efficacy of radiotherapy for recurrent tumors, particularly for patients who have multiple disease recurrence and those who may be poor candidates for surgery.[30]

CONCLUSION

Intracranial epidermoid tumors are rare, and even rarer are cerebellar vermian epidermoid tumors, which can be a differential diagnosis of other, more aggressive lesions. A high index of suspicion, an accurate evaluation of symptoms in relation to history, and interpretation of appropriate neuroimaging will help clinch the correct diagnosis. Surgery can provide a cure with complete resolution of symptoms, as seen in our present patients, with a low risk of morbidity or mortality. To the best of our knowledge, this is the first report of cerebellar vermian epidermoid tumors from Sub-Saharan Africa.

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