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Case Report

Posterior fossa microcystic meningioma mimicking an arachnoid cyst ☆

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ABSTRACT

Microcystic meningioma is an uncommon subtype of World Health Organization grade 1 meningiomas often associated with a shorter progression-free survival. Diagnosis through imaging alone can often be challenging due to atypical characteristics, especially when found in unexpected locations. Here, we present a 55-year-old woman who was diagnosed, based on imaging, with a posterior fossa arachnoid cyst 5 years prior after complaints of headaches and gait imbalance. After surgical resection of the "arachnoid cyst," the diagnosis of microcystic meningioma was made. This case report emphasizes the clinical importance and challenges associated with diagnosing microcystic meningiomas.

Introduction

Microcystic meningiomas occur rarely, accounting for only 1.6% of all intracranial meningiomas (Paek et al. [1]). Due to their atypical imaging features, including faint or absent enhancement, microcystic meningiomas pose a diagnostic challenge, especially when they present in uncharacteristic locations [2,3]. Despite being classified as World Health Organization (WHO) grade 1, microcystic meningiomas have a worse prognosis than other WHO grade 1 meningioma subtypes, emphasizing the importance of early identification and appropriate treatment [3–5]. We present a case of a posterior fossa microcystic meningioma which was initially diagnosed as arachnoid cyst.

Case description

A 55-year-old woman with a past medical history of hypertension and a benign phyllodes tumor of the breast presented to the emergency department with headaches and gait imbalance. She had a similar episode previously and was

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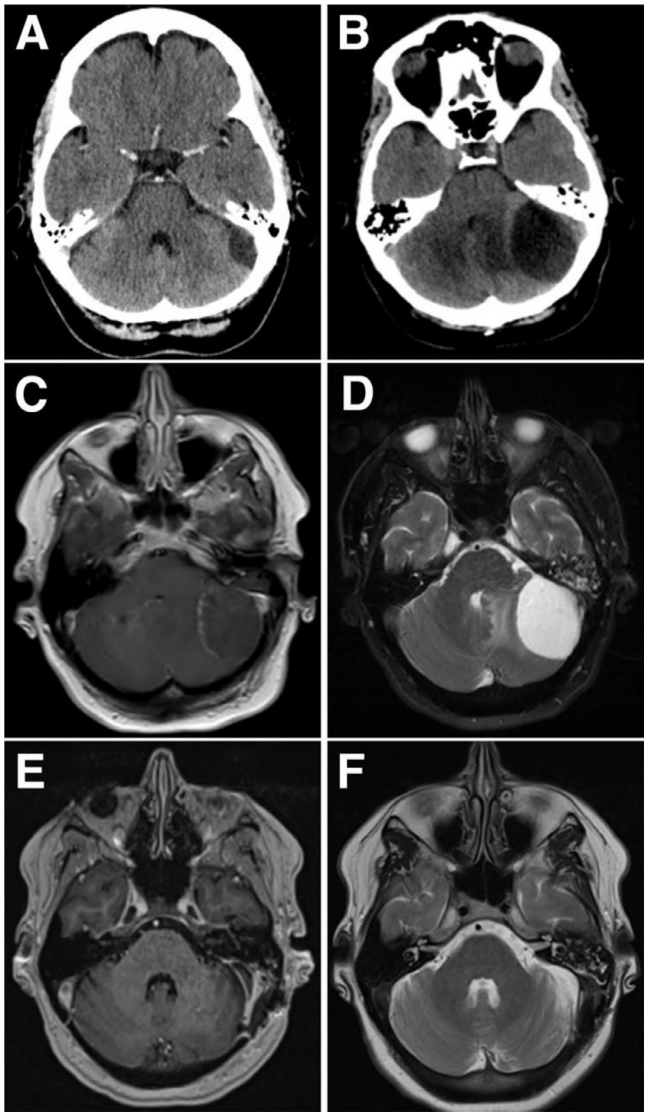

Fig. 1 – Computed tomography (CT) and brain magnetic resonance imaging (MRI) findings of a 55-year old female with a posterior fossa microcystic meningioma. Initial axial CT head demonstrates a nonenhancing cystic lesion in the left posterior fossa (A) with subsequent enlargement, fourth ventricular effacement, and perilesional edema 5 years later (B). Axial MRI demonstrates a T1-weighted hypointense (C) and T2-weighter hyperintense (D) extra-axial mass attached to the left tentorium. Axial MRI completed after resection demonstrates no mass on T1-weighted imaging (E) and complete resolution of perilesional edema on T2-weighted imaging (F).

diagnosed with a lateral cerebellar arachnoid cyst based on imaging. On examination, she did not exhibit any focal neurological deficits. Review of imaging revealed a nonenhancing 1.4 × 2.5 × 1.8 cm cystic lesion in the left posterior fossa on a computerized tomography (CT) head 5 years prior (Fig. 1A), followed by enlargement to 4.9 × 3.5 × 2.4 cm at current presentation with fourth ventricular effacement and perilesional edema (Fig. 1B). Magnetic Resonance Imaging (MRI) demonstrated a T1 hypointense, T2 hyperintense extra-axial mass based on the left tentorium with faint reticular pattern of enhancement (Figs. 1C and D). The mass demonstrated no diffusion restriction. CT chest, abdomen, and pelvis revealed a solitary benign-appearing liver lesion.

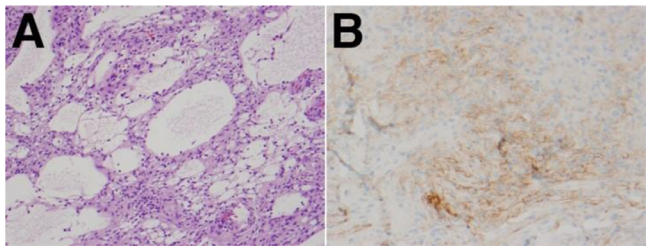

Fig. 2 – Pathological histological slides of a 55-year old female with a posterior fossa microcystic meningioma, demonstrating meningoepithelial cells separated by cystic spaces of varying sizes on hematoxylin and eosin (H&E) stain (A). Staining with epithelial membrane antigen (EMA) was confirmed (B), further supporting the diagnosis of meningioma.

Given the local mass effect on the brainstem and the enlarging size of the mass, the patient was offered a left retrosigmoid craniotomy for resection, to which she agreed. Upon dural opening, a soft, multicystic lesion was encountered with tentorial blood supply and attachments, which was resected completely. The patient recovered well postoperatively. Follow-up imaging at 3 months showed no residual mass (Fig. 1E) and resolution of perilesional edema (Fig. 1F).

Pathology showed meningoepithelial cells interrupted by numerous variably sized cystic spaces that contain granular proteinaceous material on hematoxylin and eosin (H&E) stain (Fig. 2A). The tumor was positive for epithelial membrane antigen (EMA; Fig. 2B). Ki-67 labeling index was elevated at 10%. Altogether, these findings were consistent with WHO grade 1 microcystic meningioma.

Discussion

Traditionally, meningiomas have been classified into 15 subtypes and WHO grades 1-3 based on histologic features [4–6]. WHO grade 1 is the most common grade, constituting 80%-85% of all meningiomas [6]. These are characterized by a low mitotic rate (defined as <4 per 10 high power fields) and an absence of brain invasion [6]. Grade 1 meningiomas are further classified into 9 variants, based off of histological findings: meningothelial, fibroblastic, transitional or mixed, psammomatous, angiomatous, microcystic, secretory, lymphoplasmacyte-rich, and metaplastic [6]. Recent next generation sequencing studies have identified key

driver mutations resulting in genetic instability in meningiomas, which more accurately predict prognosis and grade [7,8]. Nonetheless, in some cases, histology belies genetics, such as the association of KLF4 and TRAF7 mutations with secretory meningiomas [7–9]. Similarly, a characteristic pattern of multiple polysomies in chromosomes 5, 6, 12, 17, 18, and 20 have previously been described in microcystic meningiomas [10].

While imaging is helpful when diagnosing most meningiomas, microcystic meningiomas can have vague and sometimes overlapping characteristics with other cranial pathologies. Typical characteristics include hypointensity on T1-weighted MRI, hyperintensity on T2-weighted MRI, marginal and reticular enhancement, and severe peritumoral brain edema have all been described previously in literature [10–12]. This is unique when compared to other meningiomas, which are commonly associated with strong homogenous contrast enhancement [12]. Chen et al. [11] further investigated the sensitivity and specificity of the above findings, concluding that T1-weighted MRI hypointesity is a particularly useful finding, with an odds ratio of 75. Diagnosis using the above findings are not without difficulties, however, as characteristics are not clear-cut. Around 11% of microcystic meningiomas demonstrate faint or reticular enhancement, leading to a delayed diagnosis [2]. To complicate matters further, posterior fossa is an uncommon location for microcystic meningiomas [2].

In this case, the posterior fossa lesion was initially diagnosed as an incidental arachnoid cyst, based on lack of enhancement and peritumoral edema on CT. Consequently, no follow-up MRI or surveillance imaging was obtained. Indeed, expansion of an arachnoid cyst is a rare phenomenon in adults, and it has not been reported to occur in the posterior fossa [13–16]. Nonetheless, lateral cerebellar location occurs only in 2% of all arachnoid cysts [17], so a follow-up MRI would have been reasonable to exclude other causes. Certainly, in this case, the rapid expansion of the posterior fossa lesion over the space of 5 years precludes the diagnosis of arachnoid cyst.

In summary, this is a case of a posterior fossa microcystic meningioma mimicking an arachnoid cyst. The diagnosis was delayed due to atypical presentation of a rare meningioma variant. We propose that incidentally discovered posterior fossa arachnoid cysts warrant follow-up MRI or surveillance imaging to exclude other etiologies, especially if they present in locations other than the classic retrocerebellar region.

Conclusion

Microcystic meningiomas are an uncommon subtype of WHO grade 1 meningiomas associated with a worse prognosis. Given their atypical imaging characteristics, diagnosis can often become mistaken or delayed.

This case highlights the importance of follow-up MRI or surveillance imaging for arachnoid cysts, especially when located in an uncharacteristic “location.”

Patient consent

Informed consent was obtained from the study patient, and no personal information that could lead to their identification has been included.

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