

Occipital meningioma mimicking an arachnoid cyst: Case report and review of the literature

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Abstract

Objective: To present an unusual case of a meningothelial meningioma mimicking a growing symptomatic arachnoid cyst on conventional MRI.

Case Presentation: A 47-year-old female patient complained about dizziness over three years. Cranial MRI revealed a right-sided occipital cystic lesion adjacent to the superior sagittal sinus with a slow progression in follow-up MRI. Surgery was performed under suspicion of a symptomatic slowly growing arachnoid cyst. The occipital, gel-like, brown-green lesion was microsurgically extirpated without visual remnants. Histopathologic examination showed a meningothelial meningioma WHO grade I according to the current classification of the World Health Organization (WHO). Follow-up MRI with and without contrast enhancement twelve months after surgery shows no residual or recurrent tumor.

Conclusion: Meningiomas are benign lesion, but may be misdiagnosed if characteristic radiographic findings are missing. Surgery for treatment of mass effect and for final histopathological diagnosis is recommended in growing symptomatic cerebral cystic lesions.

Keywords

meningothelial meningioma; cystic meningioma; surgery.

Abbreviations

CT: Computed tomography; MRI: Magnetic resonance imaging; WHO: World Health Organization

Introduction

Meningiomas are the most common benign intracranial tumors, accounting for approximately 30% of cerebral neoplasms according to autopsy-based studies. Population-based studies indicate an overall incidence of 2.3/100,000. The peak in disease onset is between 40–60 years of age with a female-male ratio of 2:1 [1]. The majority of meningiomas are benign (~90%), atypical (~6%) and anaplastic (~2%) meningiomas are relatively rare [2]. Despite the benign histopathological character, meningiomas located in eloquent areas or the cavernous sinus may lead to neurological deficits. Most meningiomas present as solid, highly cellularized and well vascularized neoplasms with a characteristic appearance on Computed Tomography (CT) and Magnetic Resonance Imaging (MRI). The lesions are typically depicted hyperdense on a non-contrast enhanced CT scans with calcifications and hyperostosis of the surrounding

bone in approximately 25%. After intravenous contrast application meningiomas show an intense homogeneous enhancement on CT scans. Typically, MRI reveals a dural-based tumor, isointense with gray matter and strong homogenous contrast enhancement [2]. Additionally, a dural tail sign is present in approximately 60–72% of meningiomas. It represents either direct tumor invasion or reactive changes surrounding the tumor itself [3]. Nevertheless, 10-15% of meningiomas show an atypical appearance on MRI with cystic parts mimicking metastases or malignant gliomas [4].

In this report, we present an unusual case of a meningothelial meningioma (WHO grade I) mimicking a growing symptomatic arachnoid cyst. A review of the English literature was performed via PubMed, Scopus, Cochrane Collaboration Library and Google Scholar using the keywords: “atypical meningioma”, “cystic meningioma”, “meningioma with cystic lesion”. Only two comparable cases could be found [5,6].

Case Presentation

Case

A 47-year-old female patient presented to our department with a history of dizziness for three years. Cranial MRI revealed a right sided occipital cystic lesion adjacent to the superior sagittal sinus without contrast enhancement. Follow-up MRI showed a slow growth (initially: 25 x 10mm / follow-up: 37 x 29 mm). The suspected preoperative diagnosis was symptomatic growing arachnoid cyst. Surgery was indicated to remove the lesion and to confirm the diagnosis.

Radiological findings

Cranial MRI showed a slightly hypointense, well demarcated lesion on T1-weighted imaging and a hyperintensity on T2-weighted imaging. On fluid-attenuated inversion recovery (FLAIR) imaging an isointensity is observed. On Diffusion-Weighted Images (DWI), the lesion and cerebrospinal fluid had similar signal intensities. A light diffuse contrast enhancement can be seen after administration of contrast agent (Dotarem®, Guerbet, Sulzbach/Taunus, Germany) on T1-weighted imaging (Figure 1).

Surgical treatment

The patient was positioned in left sided park bench position and craniotomy was performed aided by neuronavigation. After incision of the dura mater and preparation towards the superior sagittal sinus, the gel-like, brown-green lesion was microsurgically extirpated piecemeal without residual tumor visible (Figure 2).

Histopathology

Histopathologic examination revealed a meningothelial cell neoplasm composed of tumor cells with round to oval nuclei arranged in lobules. Within those lobules tumor cells appeared to form a syncytium. Solitary mitoses were detected, but they were counting less than 2 mitoses per 10 high power fields. Even though single foci of necrosis were found, the tumor lacked additional features of atypia so that it was diagnosed as meningothelial meningioma (WHO Grade I) according to the current WHO classification [7] (Figure 2).

Postoperative course and follow-up

The postoperative course was uneventful and the patient recovered well after surgery without any postoperative neurological deficit. Follow up examination three months after surgery showed no neurological deficit, MRI twelve months after surgery revealed no tumor recurrence (Figure 2).

Discussion

Meningiomas are typically benign, extra-axial, solid tumors originating from the meningotheelial cells with a distinct appearance in CT and MRI. Atypical radiographic characteristics with cystic changes broaden the variety of differential diagnosis. Primary and secondary malignant tumors, hemangiopericytomas, previous strokes, subdural hematomas, abscesses and even hydrated cysts are possible differential diagnosis [8]. About 8% of all cystic meningiomas are malignant and around 12% are angioblastic [9].

In 1979, Rengachary et al. [10] classified cystic changes in meningiomas into intratumoral and extratumoral. Nauta et al. [11] described cystic meningiomas and classified them into four subtypes depending on the location of the cyst: Type I: Intratumoral cyst located centrally; Type II: Intratumoral cyst located peripherally; Type III: Peritumoral cyst in the adjacent parenchyma; and Type IV: Peritumoral cyst between the tumor and the adjacent parenchyma.

The cystic meningioma of the presented patient demonstrated according to Type III of the Nauta classification. Cystic meningiomas show an incidence of 2-4% and normally coincidence with a cystic component associated with evident dural contact [5]. The cerebral convexity and the parasagittal region are the most frequent locations of cystic meningiomas [12].

The mechanism of cyst formation is still a controversy. Some authors believe that cyst formation is caused by a central degeneration within the tumor, ischemic necrosis, or hemorrhage [12]. Other authors believe that the demyelination resulting from white matter edema and perfusion deficit causes the cyst formation [13]. An active secretion from tumor cells is also discussed [14]. It is also believed that the production of xanthochromic fluid at the periphery and its coalescence leads to the formation of large cavities resulting in cyst formation [11].

Therapy and long-term outcome depends on the patient's age, presence of comorbidities, tumor location, symptoms and histopathological examinations. Small, asymptomatic, non-growing tumors may be managed conservatively with frequent follow-up images, where as symptomatic, growing tumors should be resected. In cases with inconclusive radiological findings like in the presented patient surgery is recommended.

Conclusion

Meningiomas are benign lesion, but may be misdiagnosed if characteristic radiographic findings are missing. Surgery for treatment of mass effect and for final histopathological diagnosis is recommended in growing symptomatic cerebral cystic lesions.

Authors' contributions

Oliver Gembruch: Conception and design of the study; analysis and interpretation of data; drafting the article; revision and final approval of the version to be submitted.

Mehdi Chihi: Analysis and interpretation of data; drafting the article, revision and final approval of the version to be submitted.

Sarah Teuber-Hanselmann: Histopathological analysis and interpretation of data; drafting the article, revision and final approval of the version to be submitted.

Yahya Ahmadipour: Analysis and interpretation of data; drafting the article, revision and final approval of the version to be submitted.

Ramazan Jabbarli: Analysis and interpretation of data; drafting the article, revision and final approval of the version to be submitted.

Ulrich Sure: Conception and design of the study; analysis and interpretation of data; revision and final approval of the version to be submitted.

Karsten H Wrede: Conception and design of the study; analysis and interpretation of data; revision and final approval of the version to be submitted.

All authors read and approved the final manuscript.

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Figures

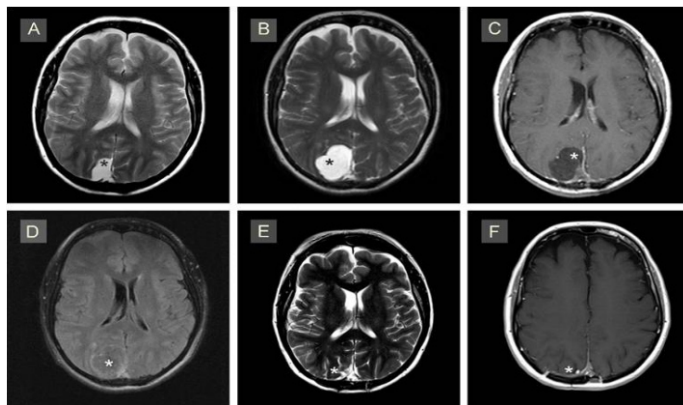


Figure 1: (A,B,C) Preoperative T2-weighted MRI showing 3-years growth of an occipital cystic lesion (*) without contrast enhancement on T1-weighted MRI. (D) FLAIR-sequence showing an isointense lesion suggesting high protein content. (E,F) No residual tumor in T2- and T1 (with contrast) -weighted MRI twelve months after surgery.

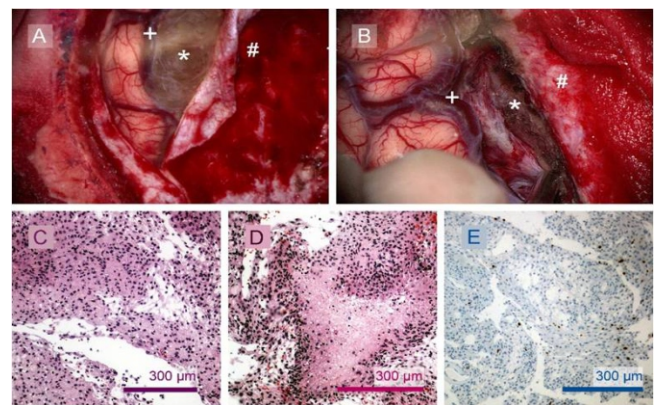


Figure 2: (A,B) Intraoperative view on the meningioma (*) and two cortical veins (+) draining into the superior sagittal sinus (#). HE staining of a meningothelial tumor composed of tumor cells forming a pseudosyncytium arranged in lobules (C) with focal necrosis (D). Ki67 staining showed a proliferation index of 10% (E).

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