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## Multifocal Meningioangiomas: Case report of a 3 year old patient.

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### Abstract

Meningioangiomas consist of benign hamartomatous lesions of the brain and the leptomeninges, which typically present with seizure. Management is predicated on resection and control of seizures with medication. Lesions are typically solitary. Multifocal meningioangiomas are extremely rare, with only 2 cases reported in adults and none in children. The authors report the first case, to their knowledge, of multifocal meningioangiomas in a child. This unique case highlights therapeutic challenges associated with these lesions and demonstrates that multifocality is possible in the pediatric population. This finding has implications for diagnosis and follow-up for children afflicted with these tumors.

### Keywords

multifocal; meningioangiomas; multiple; solitary; meningioma

## INTRODUCTION

Meningioangiomas consist of benign hamartomatous lesions of the brain and the leptomeninges that typically present with seizure. Found both sporadically and in association with NF2, they have nearly exclusively been described as single lesions, with only 2 reported cases of multiple tumors. Both of these multifocal cases were present in adult patients. In this report we describe a case of pathologically confirmed multifocal meningioangiomas in a child, with the lesions separated in both location and time of presentation.

## HISTORY, PHYSICAL EXAM AND IMAGING

This previously healthy 3-year-old girl presented with right-sided focal motor seizures. She had no other known medical conditions, previous operations, allergies, or relevant family

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### DISCLOSURES

The authors report no conflict of interest concerning the materials or methods used in this study or the findings specified in this paper. Author contributions to the study and manuscript preparation include the following. Conception and design: Smith, Jamil. Acquisition of data: all authors. Drafting the article: Jamil. Critically revising the article: all authors. Reviewed submitted version of manuscript: all authors. Approved the final version of the manuscript on behalf of all authors: Smith.

history. Specifically, there was no known history of NF2, other tumors, or seizures. On examination at the time of presentation, the child was neurologically intact, with age appropriate fluent speech, full strength bilaterally, and normal sensation in all extremities. The findings of a general physical examination were unremarkable. As part of her seizure evaluation, MRI studies of the brain and spine were obtained. These studies revealed a lesion of the left sylvian fissure, with avid Gd enhancement (Fig. 1). No other abnormalities were noted in the brain or spine, including no evident lesion in the region of the gyrus rectus. **First Operation.** Following multidisciplinary review of the case, the child was taken to the operating room for resection of the lesion via a left frontotemporal craniotomy. At surgery, a subtotal resection was performed, with complete removal thwarted by extensive encasement of branches of the middle cerebral artery by the tumor. The child did well, with no morbidity related to the operation and with subsequent control of her seizures (including the use of antiepileptic medication). **Pathological Findings.** Pathological analysis of the lesion confirmed the diagnosis of meningioangiomas. Dispersed throughout the specimen was a network of mildly to moderately hyalinized blood vessels associated with perivascular meningothelial cells and rare microcalcifications (Fig. 2A). Adjacent to the foci of meningioangiomas were distinct regions of meningioma (WHO Grade I) characterized by moderate cellularity and composed of cells with ovoid- to spindle-shaped nuclei and moderate to abundant amounts of eosinophilic cytoplasm (Fig. 2C). The tumor showed no evidence of mitotic activity, sheetlike growth, prominent nucleoli, or necrosis, which was consistent with the assigned low histological grade. **Postoperative Course.** The child was followed with annual clinic visits and MRI sessions. Four years after the resection, routine follow-up imaging revealed a new lesion in the region of the gyrus rectus, distant from the site of the original tumor (Fig. 1). The characteristics were identical to the original sylvian fissure lesion. On retrospective review, it may have been evident on the scan obtained 3 years after surgery, but was not detectable on any other pre- or postoperative images. **Second Operation and Outcome.** Given this finding (albeit without clinical symptoms), the child was taken for resection of the lesion via a left frontal craniotomy. A gross-total resection was achieved, with pathological findings identical to those in the original lesion (Fig. 2).

The child had no morbidity from the surgery. After 2 years of follow-up from the second operation, the child remains symptom free on antiepileptic medication. Radiologically, the residual lesion in the sylvian fissure has minimally increased in size, whereas the area of the gyrus rectus remains disease free. No other lesions have been identified in the CNS. Genetic testing yielded negative results for NF2.

## DISCUSSION

Meningioangiomas was initially described in 1915 and remains a rare entity, with fewer than 100 cases described in the literature. 2,16 The pathognomonic finding of this tumor is its invasive nature, characterized pathologically by invasive meningiovascular proliferation, perivascular cuffs of spindle cell proliferation, perivascular connective tissue proliferation, and neurofibrillary tangles, often interspersed with more discrete meningiomas (3,4,9,10,12,16–21,24,26).

Radiographically, the lesions of meningioangiomas are characterized by calcifications on CT scans, with MRI studies revealing low intensity on T1-weighted imaging, high intensity on T2-weighted imaging, and avid enhancement with administration of Gd. The differential diagnosis on imaging includes meningioma, oligodendroglioma, granulomatous meningitis, parasitic diseases, and calcified vascular malformation (1,5,10,17,22,23). Although the majority of cases of meningioangiomas are sporadic, particular note should be made of the association between meningioangiomas and NF2. Sporadic and NF2-associated

lesions are histologically identical, but differ in their clinical presentation. Sporadic cases are usually symptomatic, almost exclusively with seizures. In contrast, NF2-associated lesions are most commonly detected as incidental findings (6,7,10,13,16). Given the identical pathological characteristics, it is reasonable to presume that the asymptomatic nature of the NF2 lesions is probably secondary to incidental discovery on screening imaging studies. Sporadic, non-NF2 cases of meningoangiomas have also been found in association with other types of brain tumors, including astrocytomas, ependymomas, oligodendrogliomas, primitive neuroectodermal tumors, schwannomas, and hamartomas (10,11,14). Individual case reports have also cited the presence of meningoangiomas with anterior cerebral aneurysms, venous angiomas, and encephaloceles (11,25). Meningoangiomas occur in patients of all ages, although more commonly in adults than in children. Sporadic meningoangiomas rarely occur in children younger than 3 years of age. Anatomically, lesions are most frequently identified in the temporal lobe, followed by the frontal, parietal, and occipital lobes in descending order (4,16,21). Clinically, meningoangiomas usually presents with seizures and headaches. 1,3,8,10,11,17 Seizures are usually simple or complex partial, and are notably and remarkably refractory to treatment (1,3,8,10,11). This condition is benign and treatable surgically, although resection can be complicated by the invasive nature of the tumor (1,3,8,10,11,17). When possible, complete resection can result in both cure of the tumor and resolution of seizures. However, careful follow-up of treated patients is necessary because the biology of these lesions increases the risk of local recurrence. Literature on this tumor is limited, with the largest series to date composed of only 6 patients, reported by Jallo et al. (7) in 2005. All 6 patients presented with seizure: 4 with simple partial seizures, 1 with complex partial seizures, and 1 with a generalized tonic-clonic seizure. Contrary to other studies, which usually cite temporal lesions, in this series there were 4 frontal and 2 parietal tumors. With resection, patients were seizure free, citing an average of more than 6 years of follow-up. The next largest series, by Kim et al. (10) reviewed 5 patients who also presented with seizure. In a PubMed review of meningoangiomas only 2 cases of multifocal disease were identified, both of which were in adult patients. 17 Both presented with headaches and seizures. In one patient the lesions were left frontoparietal and right parietal, and the other patient was found to have left frontal and parietal lobe lesions. In both cases the tumors were initially identified as contemporaneous lesions.

We report on a case of meningoangiomas that is unique in being a pediatric report of multifocal disease and also in being a case of 2 lesions growing at different times. The 2 lesions arose in completely different locations and were documented radiographically and confirmed pathologically. This is important because it suggests that these tumors can be multifocal, even in children—a finding that has immediate implications for follow-up in affected patients. In addition, this report offers proof of the principle that subtypes of this tumor with different biological behaviors may exist. Although acknowledging that this is a rare entity in any case, we hope that our reporting of this heretofore unwitnessed phenomenon may serve to inform treating physicians about the potential of this tumor.

## CONCLUSION

Meningoangiomas is a rare, benign, surgically correctible cause of seizures. This report demonstrates the presence of 2 anatomically and temporally distinct lesions in a child with meningoangiomas. This finding is important because it reveals the capacity of these tumors to be multifocal, and it underscores the need for continued long-term follow-up in affected patients.

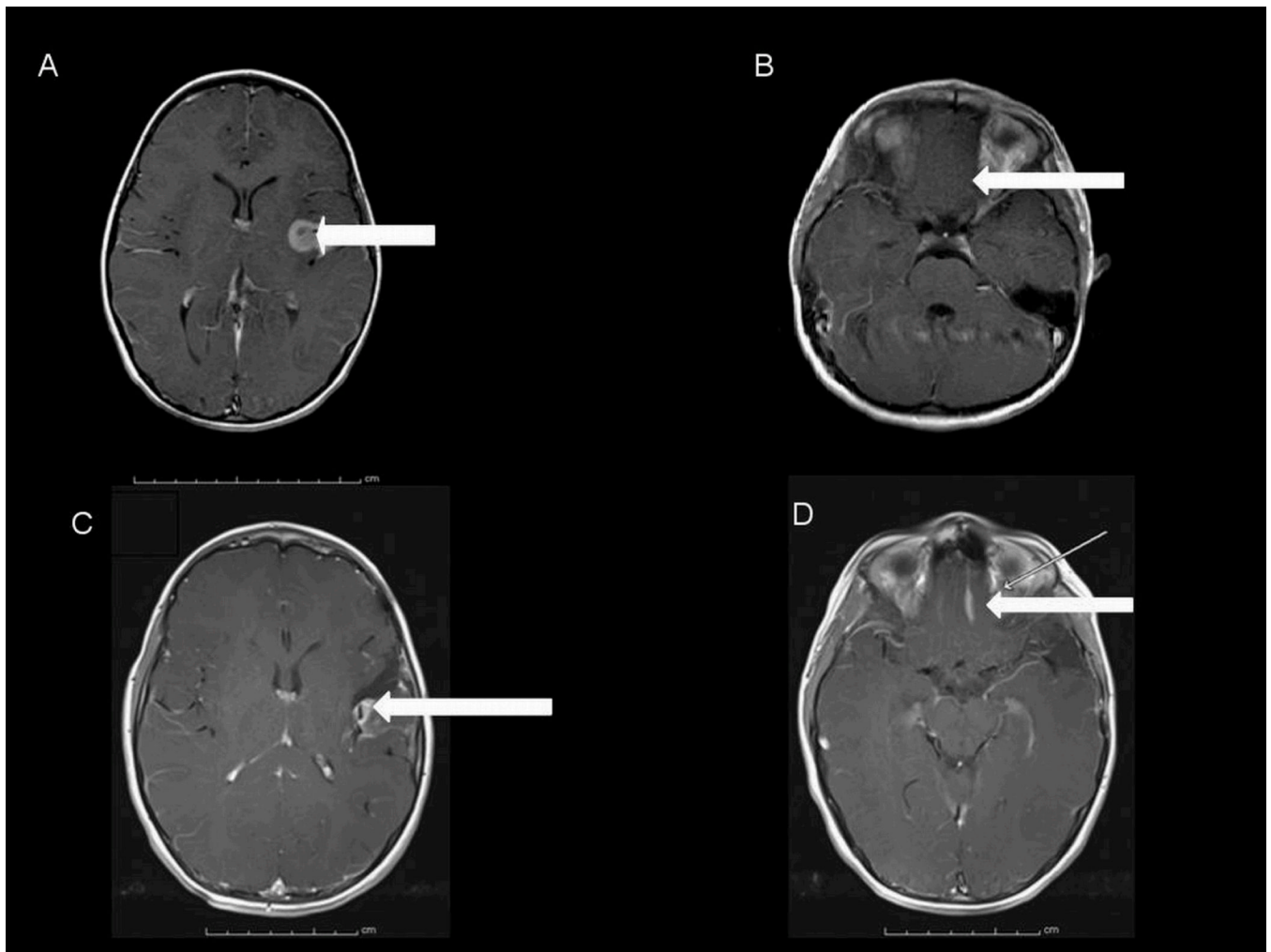
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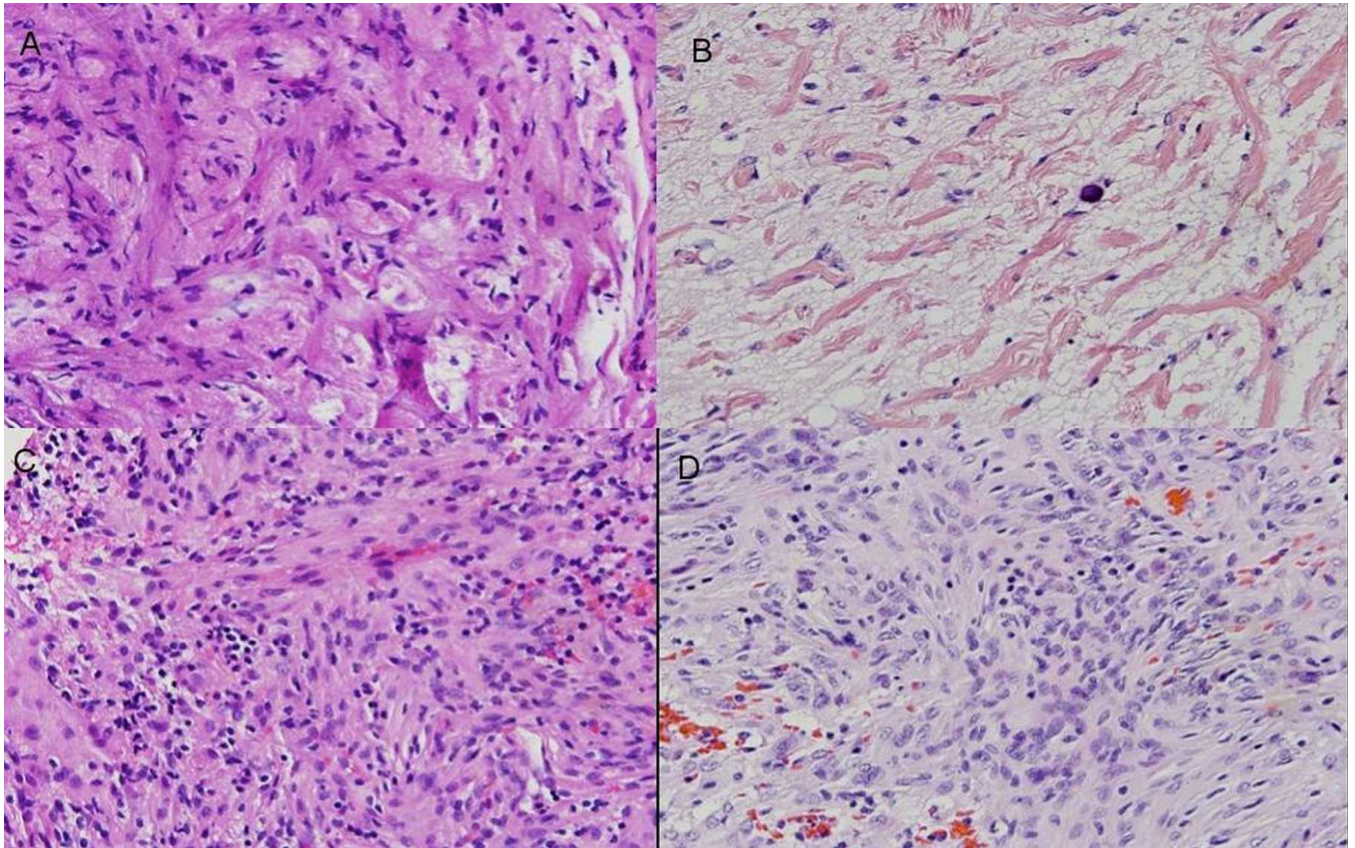
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**Figure 1.** Axial MRI studies obtained with contrast, in 2005, showing an enhancing lesion in the left sylvian fissure (A) and no obvious lesion in the left gyrus rectus (B). Axial postcontrast MRI studies obtained in 2009 showing a partially resected left sylvian fissure lesion (C) and a newly apparent and distinct lesion in the left gyrus rectus (D). The arrows designate the lesions (or in the case of panel B, the lack thereof).





**Figure 2.** Photomicrographs of histopathological specimens obtained during the 2005 and 2009 resections. A: Meningioangiomatosis characterized by mildly to moderately hyalinized blood vessels associated with scattered perivascular meningeothelial cells involving reactive cortex (specimen from the 2005 resection). B: Meningioangiomatosis with prominent vascular hyalinization and infrequent meningeothelial cells in association with rarefied cortex showing neuronal depletion (specimen from the 2009 resection). Moderately cellular meningioma occurring adjacent to meningoangiomatosis in 2005 (C) and in 2009 (D). H & E, original magnification  $\times 40$ .