



Case Report

Journal of Clinical Imaging Science



Meningioangiomatosis: Clinical, Imaging, and Histopathologic Characteristics

Mina S. Makary¹, Peter Kobalka², Pierre Giglio³, H. Wayne Slone¹

¹Department of Radiology, Division of Neuroradiology, ²Department of Pathology, Division of Neuropathology, ³Department of Neurology, Division of Neuro-Oncology, The Ohio State University Wexner Medical Center, Columbus, Ohio, United States.



*Corresponding author: Mina S. Makary, Department of Radiology, Division of Neuroradiology, The Ohio State University Wexner Medical Center, Columbus, Ohio, United States.

mina.makary@osumc.edu

Received : 19 March 2020 Accepted : 26 May 2020 Published : 12 June 2020

DOI 10.25259/JCIS_39_2020

Quick Response Code:



ABSTRACT

Meningioangiomatosis is a rare benign lesion involving the central nervous system. Radiographic appearance can be highly variable which makes pre-operative diagnosis difficult. In this report, we describe meningioangiomatosis in a previously healthy 17-year-old woman who presented with seizures and continued headache and dizziness. This patient presented with a predominately calcified lesion on imaging and eventually underwent near total resection. Meningioangiomatosis is difficult to preoperatively identify, but is an important consideration as prognosis with surgical resection is typically good.

Keywords: Meningioangiomatosis, Hamartomatous lesion, Neural calcifications, Central nervous system, Seizures

INTRODUCTION

Meningioangiomatosis is a rare hamartomatous or meningiovascular lesion involving the central nervous system.^[1] There is often a predilection for the frontal and temporal lobes.^[2] The most common clinical presentation is headache and seizures.^[1,2] Radiographic findings of meningioangiomatosis are variable depending on the histologic components of the mass.^[1] Meningioangiomatosis is typically cortical based and can present as a solitary or multiple lesions. It demonstrates variable degrees of calcification, cystic degeneration, and enhancement.^[1,3] These variable appearances can make radiologic diagnosis a challenge. In this report, we present a case which demonstrates a predominately calcified mass which is one of the many variable imaging findings and subsequent positive response following surgical resection.

CASE REPORT

A 17-year-old woman with no significant medical history originally presented with a generalized tonic-clonic seizure episode which lasted for approximately 1 min. She also reported headache and dizziness. After initial seizure event subsided, physical examination was unremarkable without focal deficits. Laboratory findings were unremarkable. An EEG was performed during wakefulness and sleep and was normal.

At time of initial presentation, a non-contrast CT of the head found a densely calcified mass in the left temporal lobe [Figure 1]. MRI demonstrated a hypointense mass on T1- and T2-weighted imaging with some associated heterogeneous contrast enhancement [Figure 2]. The mass appeared to encase the left middle cerebral artery. The patient opted for non-surgical treatment

This is an open-access article distributed under the terms of the Creative Commons Attribution-Non Commercial-Share Alike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as the author is credited and the new creations are licensed under the identical terms. ©2020 Published by Scientific Scholar on behalf of Journal of Clinical Imaging Science

and was closely followed with medical management of her seizures. She continued with non-operative management, and the mass appeared stable on imaging for several years.

Her last known seizure episode was 2 years after the initial presentation, but she had continued intermittent dizziness and headache. Eight years later, the patient opted to undergo left temporal craniotomy for near total resection of the mass. Total resection could not be performed as the mass was found to be interdigitated and intercalated with the middle cerebral artery vessels. Pathology demonstrated innumerable and confluent psammoma bodies intimately associated with extensive fibrotic bundles [Figure 3]. Perivascular proliferation of spindled cells was consistent with a meningothelial origin. These findings are supportive of meningioangiomatosis. MRI demonstrated the expected post-operative changes including evidence of partial

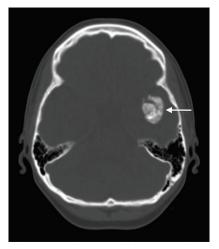


Figure 1: A 17-year-old woman with meningioangiomatosis presented with seizures and persistent headache and dizziness. Non-contrast head CT image demonstrated a densely calcified mass (arrow) in the left temporal lobe.

resection with the residual around the surgical cavity [Figure 4]. Postoperatively, the patient remains seizure free and has noted some subjective improvement in headaches and dizziness symptoms.

DISCUSSION

Meningioangiomatosis is typically divided into a sporadic subtype and a subtype associated with neurofibromatosis Type 2 (NF2).^[1,4,5] One of the most common presentations for the sporadic subtype is headache and seizures and will typically occur in young adults or children.^[1] Some of the most typical locations are the frontal and temporal lobes and typically is cortical based. In one study, it was found to occur sporadically in 75% of patients and was associated with neurofibromatosis in 25% of patients.^[4] In that same study, the mean age of diagnosis in sporadic cases was 28 and in cases associated with neurofibromatosis, the mean age was 21.^[4] Sporadic cases are usually symptomatic and solitary lesions, whereas the NF2 ones are often asymptomatic and multiple lesions with meningioangiomatosis discovered as an incidental finding.^[6]

Imaging findings of meningioangiomatosis are variable.^[7] On CT, it most commonly demonstrates some degree of calcification.^[4,8,9] On MRI, it will most commonly show some degree of calcification and contrast enhancement. One study found that calcification was prevalent in 89.6% of cases.^[8] MRI will typically demonstrate lesion confined to the cortex. One of the most common appearances on MRI with be hypointense on T1, hyperintense on T2 with approximately 80% demonstrating some degree of enhancement.^[8,9] The degree of susceptibility artifact depends on the amount of calcification associated with the lesion.

Due to the variable appearance on imaging, the differential diagnosis is broad and includes meningioma, oligodendroglioma, calcifying pseudoneoplasm of the

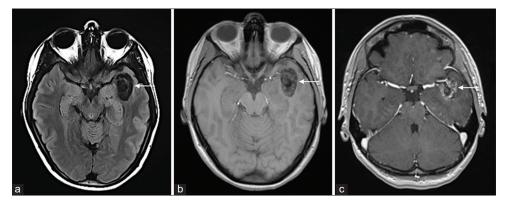


Figure 2: A 17-year-old woman with meningioangiomatosis presented with seizures and persistent headache and dizziness. (a) Brain MRI T2/FLAIR image revealed a hypointense lesion (arrow) in the left temporal lobe. (b) Brain MRI T1-weighted image showed corresponding heterogeneous hypointense signal (arrow). (c) Brain MRI post-contrast T1-weighted image further demonstrated irregular enhancement (arrow) in close association with the left middle cerebral artery.

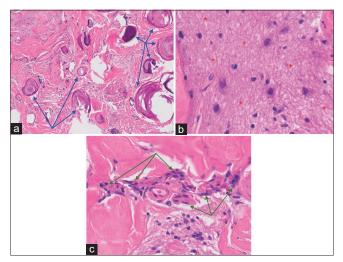


Figure 3: A 17-year-old woman with meningioangiomatosis presented with seizures and persistent headache and dizziness. Hematoxylin and eosin stained tissue sample demonstrated (a) innumerable and confluent psammoma bodies (blue arrows) intimately associated with (b) extensive fibrotic bundles (red asterisks) as well as (c) perivascular proliferation of spindle cells (green arrows), which are characteristic findings of meningioangiomatosis.

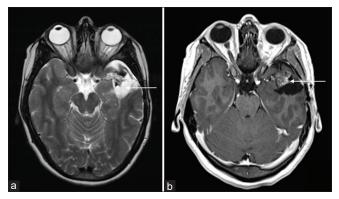


Figure 4: A 17-year-old woman with meningioangiomatosis presented with seizures and persistent headache and dizziness. (a) Brain MRI T2 image revealed a CSF-filled hyperintense resection cavity (arrow) in the anterior left temporal lobe. (b) Brain MRI post-contrast T1-weighted image demonstrated irregular residual enhancement (arrow) intercalated with the middle cerebral artery consistent with residual disease.

cavernous malformation, ganglioglioma, neuraxis, disease, low-grade astrocytoma, or granulomatous meningioangiomatosis, metastasis.^[10] Compared to gliomas typically show surrounding hyperintense FLAIR signal. Meningiomas are typically dural based, whereas meningioangiomatosis is typically cortically based. Cavernous malformations could be distinguished by multiloculated appearance with T2-weighted hypointense rim. Some studies have suggested that gyriform

hyperintensity on FLAIR imaging is one of the main features of meningioangiomatosis.^[10] Rarely, it is possible to see hemorrhage associated with the mass.^[7] This wide differential makes it difficult to diagnose preoperatively, but suggestion of this disease process may be important as meningioangiomatosis is benign without a malignant potential.^[7]

The main differential diagnoses from a pathology standpoint include meningioma and vascular malformation.^[6] In general, vascular malformations lack the perivascular proliferations of meningothelial cells and also lack psammoma bodies. Meningiomas consist of masses of syncytial cells arranged in lobules, fascicles, or whorls. Brain tissue is not intermixed, unless the meningioma is cortically invasive – in which case, the meningioma surrounds reactive brain parenchyma. Rarely, meningioangiomatosis can be associated with meningioma.^[3,6,11] In these cases, the meningioma is broadly attached to the cortical surface (en plaque), with a transitional zone with the underlying meningioangiomatosis.^[3]

Although in many cases, surgical resection is the typical treatment strategy, in some cases, adjuvant radiation therapy has been used when an incomplete excision has been performed.^[3] Meningioangiomatosis is a slow-growing tumor without malignant potential; therefore, prognosis is typically excellent following surgical excision.^[7] When patients do undergo surgical resection, one of the main goals is to achieve seizure control for the patient.^[1]

CONCLUSION

We present a 17-year-old woman who presented with seizures and headaches and was found to have a mass in the left temporal lobe. Imaging findings demonstrated a densely calcified mass on CT and heterogeneous predominately hypointense signal on T1-weighted images. After experiencing progressive headaches, the mass was partially resected and was pathology proven meningioangiomatosis. The patient did well postoperatively with improvement in symptoms. Meningioangiomatosis is an important differential consideration as imaging findings can be variable and overlap with other diagnoses, but the treatment and outcomes are unique compared to other similar etiologies.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

- 1. Sun Z, Jin F, Zhang J, Fu Y, Li W, Guo H, *et al.* Three cases of sporadic meningioangiomatosis with different imaging appearances: Case report and review of the literature. World J Surg Oncol 2015;13:89.
- 2. Perry A, Kurtkaya-Yapicier O, Scheithauer BW, Robinson S, Prayson RA, Kleinschmidt-DeMasters BK, *et al.* Insights into meningioangiomatosis with and without meningioma: A clinicopathologic and genetic series of 24 cases with review of the literature. Brain Pathol 2005;15:55-65.
- 3. Cui H, Shi H, Chen X, Wang W, Lai R, Han A. Clinicopathological features of meningioangiomatosis associated with meningioma: A case report with literature review. Case Rep Oncol Med 2012;2012:296286.
- Wiebe S, Munoz DG, Smith S, Lee DH. Meningioangiomatosis. A comprehensive analysis of clinical and loboratory features. Brain 1999;122:709-26.
- Takeshima Y, Amatya VJ, Nakayori F, Nakano T, Sugiyama K, Inai K. Meningioangiomatosis occurring in a young male without neurofibromatosis: With special reference to its histiogenesis and loss of heterozygosity in the NF2 gene region.

Am J Surg Pathol 2002;26:125-9.

- 6. Burger P, Scheithauer B. Diagnostic Pathology: Neuropathology. Salt Lake: Amirsys Publishing, Inc.; 2012.
- 7. Kim WY, Kim IO, Kim S, Cheon JE, Yeon M. Meningioangiomatosis: MR imaging and pathological correlation in two cases. Pediatr Radiol 2002;32:96-8.
- Kashlan ON, LaBorde DV, Davison L, Saindane AM, Brat D, Hudgins PA, *et al.* Meningioangiomatosis: A case report and literature review emphasizing diverse appearance on different imaging modalities. Case Rep Neurol Med 2011;2011:361203.
- 9. Bulut E, Mut M, Soylemezoglu F, Oguz KK. Meningioangiomatosis of the cerebellum: Radiopathologic characteristics of a case. Acta Neurochir (Wien) 2015;157:1371-2.
- Yao Z, Wang Y, Zee C, Feng X, Sun H. Computed tomography and magnetic resonance appearance of sporadic meningioangiomatosis correlated with pathological findings. J Comput Assist Tomogr 2009;33:799-804.
- 11. Deb P, Gupta A, Sharma MC, Gaikwad S, Singh VP, Sarkar C. Meningioangiomatosis with meningioma: An uncommon association of a rare entity--report of a case and review of the literature. Childs Nerv Syst 2006;22:78-83.

How to cite this article: Makary MS, Kobalka P, Giglio P, Slone HW. Meningioangiomatosis: Clinical, imaging, and histopathologic characteristics. J Clin Imaging Sci 2020;10:36.