

THE VASCULAR MASQUERADE: A CASE OF ANGIOMATOUS MENINGIOMA

Dr Priyadarshini S ^{1*}, Dr R Balaji², Dr Sowmiya J³, Dr Muthu S⁴, Dr Anitha⁵

¹ Postgraduate, Department of Pathology, SRM medical college hospital and research centre, Email: priya104darshini@gmail.com, ORCID: 0009-0001-9151-006X

² Associate Professor, Department of Pathology, SRM Medical College Hospital and Research Centre, Kattankulathur, India, ORCID: <https://orcid.org/0009-0004-5032-4561>, email: balajir11@srmist.edu.in,

³ Assistant Professor, Department of pathology, SRM Medical College Hospital and Research centre, Email: sowmiyaj@srmist.edu.in, ORCID: 0009-0001-7945-6259

⁴ Head of Department and Professor, Department of Pathology, SRM Medical College Hospital and Research centre, Email: drmuthus@gmail.com ORCID: 0000-0003-0364-8243

⁵ Assistant Professor, Department of Pathology, SRM Medical College and Reserch Centre, Email: anitharaj.mmc@gmail.com, Orci'd: 0009-0009-7318-4019

ABSTRACT

Angiomatous meningioma is an uncommon, histologically distinct variant of World Health Organization (WHO) CNS Grade 1 meningioma, characterized by a predominant vascular component and a disproportionate tendency to provoke peritumoral brain edema (PTBE) despite its benign nature. We report the case of a 51-year-old woman who presented with headache, visual disturbance, and fatigue. Contrast-enhanced computed tomography of the brain revealed an extra-axial mass in the right frontoparietal region with marked surrounding peritumoral edema, a radiological appearance often mistaken for a more aggressive lesion. The patient underwent craniotomy with gross total surgical excision. Histopathological examination revealed numerous variably sized, hyalinized vascular channels admixed with bland meningothelial cells, comprising more than half of the tumor volume, with immunohistochemistry confirming a diagnosis of angiomatous meningioma. The patient's presenting symptoms resolved in the postoperative period. This case highlights the importance of recognizing angiomatous meningioma as a cause of imaging findings that appear radiologically aggressive despite an entirely benign clinical course, and reinforces surgical excision as the definitive and generally curative treatment.

KEYWORDS: Angiomatous meningioma, Frontoparietal, Peritumoral edema, Vascular, Histopathology

INTRODUCTION

Meningiomas arise from the meningothelial (arachnoid cap) cells of the leptomeninges and account for close to one-third of all primary intracranial tumors in adults, with a marked female predominance.[1,2] Under the current World Health Organization classification, meningiomas are stratified into three CNS grades on the basis of histological features such as mitotic activity, brain invasion, and specific high-risk subtypes; the large majority are CNS WHO Grade 1 lesions with an indolent biological behavior.[1] Risk factors for meningioma development include advancing age, female sex, prior cranial irradiation, and hereditary tumor syndromes such as neurofibromatosis type 2.[2]

Clinically, meningiomas often remain silent for long periods and are frequently discovered incidentally on neuroimaging performed for unrelated reasons. When symptomatic, presentation depends chiefly on tumor location and size, and commonly includes headache, seizures, focal neurological deficits, and visual disturbance when the tumor lies close to the optic pathways or exerts mass effect on the frontal or parietal convexity.[1] A substantial proportion of meningiomas are accompanied by peritumoral brain edema (PTBE), which can itself be responsible for much of the associated morbidity, independent of tumor size. The pathogenesis of PTBE is incompletely understood but is thought to involve secretion of vascular endothelial growth factor (VEGF) by tumor cells, disruption of the arachnoid-pial barrier, venous outflow compromise, and local hypoxia.[4,5] Definitive diagnosis rests on histopathological examination, supplemented where necessary by immunohistochemistry, which remains the gold standard for confirming the meningothelial lineage of the tumor and excluding morphological mimics.[3] Among the recognized histological subtypes, angiomatous meningioma is particularly notable for provoking peritumoral edema disproportionate to its size, occasionally resulting in a radiological appearance that mimics more aggressive pathology despite an entirely benign course.[7,8] We describe a case of an angiomatous meningioma of the right frontoparietal region in a middle-aged woman, presenting with headache, visual disturbance, and fatigue, successfully managed with surgical excision.

Case Report

A 51-year-old woman presented to the outpatient department with complaints of persistent headache, disturbance of vision, and generalized fatigue of several weeks' duration. There was no history of seizures, vomiting, or loss of consciousness. General physical examination was unremarkable, and systemic examination did not reveal any focal

neurological deficit apart from the visual symptoms reported by the patient. Baseline hematological and biochemical investigations were within normal limits.

Contrast-enhanced computed tomography (CT) of the brain demonstrated a well-defined, homogeneously enhancing extra-axial mass in the right frontoparietal convexity region, arising from the dura and associated with significant surrounding peritumoral edema, features radiologically suggestive of a convexity meningioma. In view of the mass effect and symptomatic presentation, the patient was referred to neurosurgery, where she underwent a right frontoparietal craniotomy with gross total excision of the tumor. The intraoperative course was uneventful, and the excised tissue was submitted to the department of pathology for histopathological evaluation.

On gross examination, the specimen consisted of multiple grey-white to grey-brown, firm tissue fragments, the largest measuring approximately $2.5 \times 2.0 \times 1.5$ cm, with a smooth external surface corresponding to the dural attachment. The tissue was fixed in 10% neutral buffered formalin, routinely processed, and embedded in paraffin; sections of 4-micron thickness were cut and stained with hematoxylin and eosin (H&E).

Microscopic examination revealed a richly vascular tumor composed of numerous small to medium-sized, thin- to thick-walled, hyalinized blood vessels closely admixed with intervening sheets of bland meningotheial cells; the vascular component was estimated to comprise more than half of the overall tumor volume. The meningotheial cells showed oval nuclei with finely dispersed chromatin, inconspicuous nucleoli, and syncytial eosinophilic cytoplasm, without significant nuclear pleomorphism. Mitotic activity was low (fewer than 4 mitoses per 10 high-power fields), psammoma bodies and calcification were sparse, and there was no evidence of brain invasion, necrosis, or high-grade cytological atypia. These histomorphological features were consistent with an angiomatous meningioma, CNS WHO Grade 1.

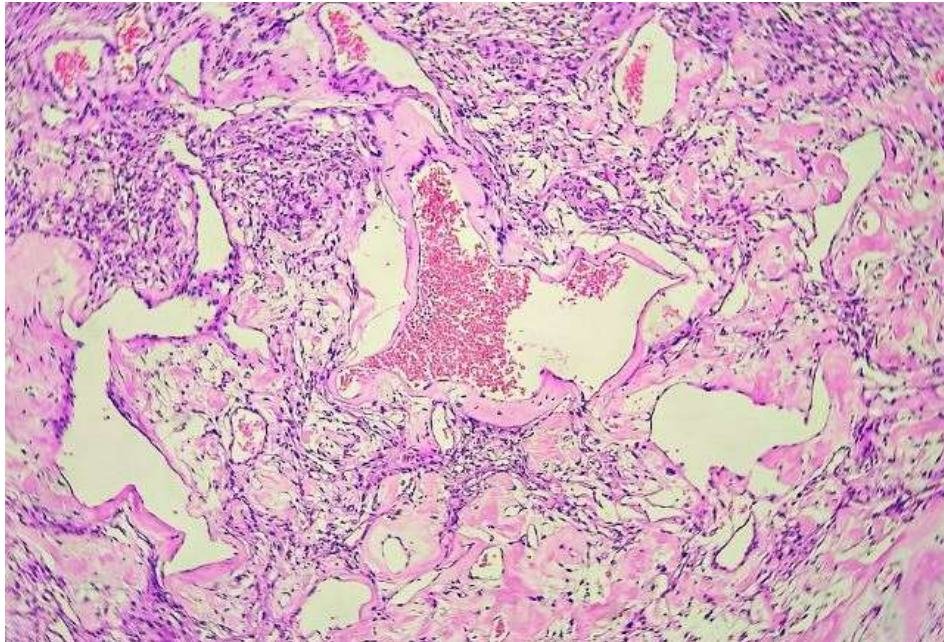


Fig 1. Congested thick walled blood vessels of various caliber

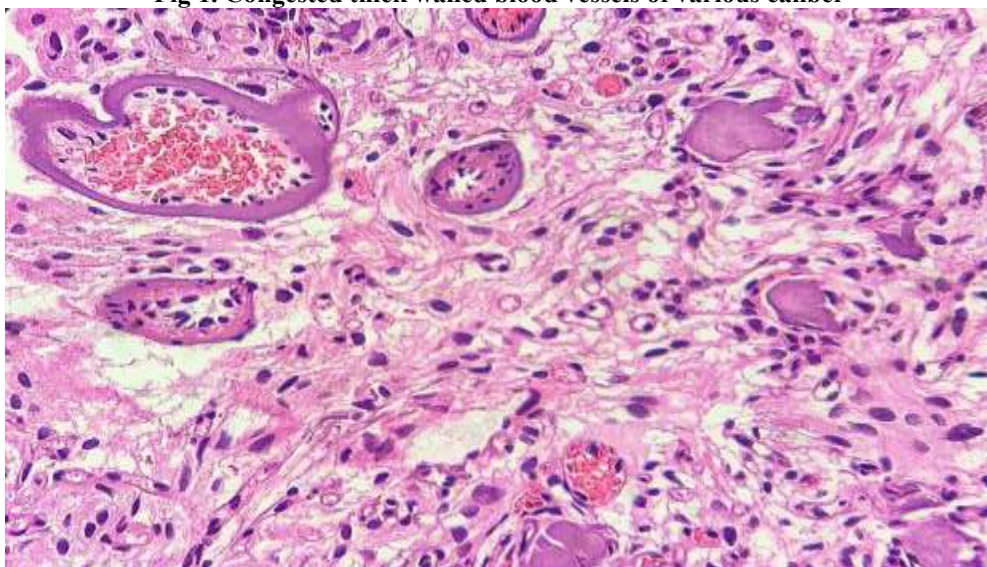


Fig 2. Meningotheial cells, psammoma bodies and multiple congested blood vessels.

The postoperative period was uneventful, without any new neurological deficit. On follow-up examination one week after surgery, the patient's headache and fatigue had resolved substantially, and her visual symptoms showed marked improvement. She was discharged in a stable condition and advised regular clinical and radiological follow-up.

DISCUSSION

Angiomatous meningioma (AM) is an uncommon histological variant of meningioma, accounting for approximately 2.1-2.6% of all meningiomas, and is classified under the current WHO scheme as a CNS Grade 1 tumor with a generally favorable prognosis.[7,8] Unlike the common meningothelial, transitional, and fibrous subtypes, AM is defined histologically by a predominance of blood vessels, which by convention must comprise more than half of the tumor volume, with the intervening meningothelial cells typically appearing cytologically bland.[8] Reported series describe a wide age range at presentation, frequent involvement of the cerebral convexity, as in the present case, and headache as one of the most common presenting symptoms, often accompanied by seizures, focal deficits, or visual disturbance depending on tumor location.[7,8]

The single most distinctive clinical feature of AM is its propensity to provoke peritumoral brain edema that is disproportionately extensive relative to tumor size, a phenomenon well documented across multiple series.[7] Among the recognized WHO Grade 1 subtypes, angiomatous and secretory meningiomas in particular have been associated with peritumoral edema volumes many times greater than those seen with common-type meningiomas.[6] This exaggerated edema response is attributed largely to the tumor's rich vascular stroma: the numerous small, markedly hyalinized vessels seen histologically are thought to secrete vascular endothelial growth factor (VEGF) and other angiogenic mediators that increase capillary permeability in the surrounding brain, compounded by mechanical compression of adjacent parenchyma and draining veins.[6,7] The peritumoral edema noted on CT imaging in the present case, out of proportion to what might be expected for a benign, well-circumscribed extra-axial lesion, is characteristic of this subtype.

This tendency toward marked edema and intense contrast enhancement means that AM can closely mimic a more aggressive intracranial lesion on imaging, including atypical or anaplastic meningioma, solitary fibrous tumor/hemangiopericytoma, or even a vascular malformation, despite its entirely benign biological behavior.[8] Preoperative recognition of this radiological pattern is clinically important, both because AM is markedly hypervascular and carries a higher risk of intraoperative bleeding than common meningiomas, and because failure to anticipate this vascularity can complicate surgical planning.[7,8] Histopathology and immunohistochemistry remain the definitive means of diagnosis where the meningothelial component typically shows positivity for epithelial membrane antigen (EMA) and variable progesterone receptor (PR) expression, while CD34 marks the vascular endothelium rather than the tumor cells themselves and STAT6 negativity helps exclude solitary fibrous tumor/hemangiopericytoma, its principal histological mimic.[3]

Despite its radiologically aggressive appearance, AM carries a prognosis similar to that of other WHO Grade 1 meningiomas, with a generally benign clinical course and low recurrence rate following surgical resection, even when peritumoral edema at presentation is extensive.[7] Gross total excision, as achieved in this patient, is considered curative in the majority of cases. Resolution of the presenting symptoms and, over time, of the peritumoral edema following complete tumor removal is the expected postoperative course, though periodic clinical and radiological follow-up remains prudent given the rarity of this subtype and the comparatively limited long-term outcome data available for it.

CONCLUSION

This case illustrates an angiomatous meningioma of the right frontoparietal region presenting with headache, visual disturbance, and fatigue in a 51-year-old woman, in whom marked peritumoral edema on imaging belied an entirely benign tumor that was successfully treated with gross total surgical excision. It underscores the importance of recognizing angiomatous meningioma as a distinct, richly vascular subtype capable of mimicking more aggressive pathology on neuroimaging, and reinforces histopathology and immunohistochemistry as the definitive means of arriving at an accurate diagnosis and guiding appropriate surgical management.

Conflict of Interest

The authors declare that there are no conflicts of interest pertaining to the publication of this case report.

Source of Funding

None.

REFERENCES

1. Yarabarla V, Mylarapu A, Han TJ, McGovern SL, Raza SM, Beckham TH. Intracranial meningiomas: an update of the 2021 World Health Organization classifications and review of management with a focus on radiation therapy. *Front Oncol.* 2023;13:1137849.
2. Ostrom QT, Gittleman H, Truitt G, Boscia A, Kruchko C, Barnholtz-Sloan JS. CBTRUS statistical report: primary brain and other central nervous system tumors diagnosed in the United States in 2011-2015. *Neuro Oncol.* 2018;20(Suppl 4):iv1-iv86.
3. Boulagnon-Rombi C, Fleury C, Fichel C, Lefour S, Marchal Bressenot A, Gauchotte G. Immunohistochemical approach to the differential diagnosis of meningiomas and their mimics. *J Neuropathol Exp Neurol.* 2017;76(4):289-298.
4. Fiani B, Jarrah R, Bhandarkar AR, De Stefano F, Amare A, Aljameey UA, Reardon T. Peritumoral edema in meningiomas: pathophysiology, predictors, and principles for treatment. *Clin Transl Oncol.* 2023;25(4):866-872.

5. Nassehi D, Dyrbye H, Andresen M, Thomsen C, Juhler M, Laursen H, Broholm H. Vascular endothelial growth factor A protein level and gene expression in intracranial meningiomas with brain edema. *APMIS*. 2011;119(12):831-843.
6. Maiuri F, Mariniello G, de Divitiis O, Esposito F, Guadagno E, Teodonno G, Barbato M, Del Basso De Caro M. Progesterone receptor expression in meningiomas: pathological and prognostic implications. *Front Oncol*. 2021;11:611218.
7. Hua L, Luan S, Li H, Zhu H, Tang H, Liu H, Chen X, Bozinov O, Xie Q, Gong Y. Angiomatous meningiomas have a very benign outcome despite frequent peritumoral edema at onset. *World Neurosurg*. 2017;108:465-473.
8. Verma PK, Nangarwal B, Verma J, Dwivedi V, Mehrotra A, Das KK, Maurya VP, Bhaisora KS, Singh J, Sardhara J, Srivastava AK, Behari S, Jaiswal S, Jaiswal AK. A clinico-pathological and neuro-radiological study of angiomatous meningioma: aggressive look with benign behaviour. *J Clin Neurosci*. 2021;83:43-48.