Meningioangiomatosis.

C R Partington, V B Graves and L R Hegstrand

AJNR Am J Neuroradiol 1991, 12 (3) 549-552
http://www.ajnr.org/content/12/3/549.citation

Meningioangiomatosis

Curtis R. Partington,[1] Virgil B. Graves,[1] and Linda R. Hegstrand[2]

Meningioangiomatosis (MA) is an uncommon cerebral cortical mass lesion characterized histologically by cortical meningiovascular proliferation that may extend to involve the overlying meninges [1]. MA has many of the characteristics of an endotheliomatous meningioma that is located within the cerebral cortex, but it may extend to the leptomeninges along the Virchow-Robin spaces associated with abnormal blood vessels [1–4]. Its pathogenesis is uncertain, but the favored hypothesis is that MA represents either an occult vascular malformation, which is later accompanied by meningioendothelial cell proliferation without evidence of malignancy, or a congenital hamartomatous malformation [2–4]. There is an association of MA with neurofibromatosis, but nearly 50% of the reported cases showed no other stigmata of neurofibromatosis [2–15].

There are two populations of patients with MA: (1) symptomatic children and young adults who present with headaches or seizures and (2) asymptomatic individuals (mostly those with neurofibromatosis) in whom MA is discovered as an incidental finding [2–15]. In symptomatic patients, complete [2, 3, 7, 8, 10, 11] or partial [2] resection is curative.

We report here the CT, MR, and angiographic characteristics of two cases of symptomatic MA and review the previous case reports of imaging characteristics of MA.

Case Reports

Case 1

A 19-year-old right-handed boy had been having seizures characterized by 10-min episodes of staring since the age of 13. These seizures were initially well controlled with medications, but had become refractory, occurring at a rate of three to five per week. A CT scan done at age 13 (Fig. 1A) showed a densely calcified mass in the right frontal lobe with no detectable contrast enhancement. A CT scan done at age 19 showed no significant change in the right frontal mass. MR at that time showed an inhomogeneous mass in the right frontal lobe on T1-weighted images with foci of intense contrast enhancement and areas of nonenhancing low signal intensity on postcontrast MR (Figs. 1B and 1C). The mass appeared to be intraaxial, but had broad extension toward the dura (Fig. 1C). On spin-density and T2-weighted images, the mass was predominantly of high signal intensity, with central areas of low signal intensity corresponding to the calcifications seen on CT (Figs. 1D and 1E). Angiography showed an avascular mass, but no abnormal vessels or tumor blush were identified. EEG showed an epileptogenic focus in the right frontal region.

A right median frontal craniotomy was performed with total resection of the stony-hard tumor that lay within the right superior frontal gyrus. There was no involvement of the overlying dura. Histopathology of the lesion (Fig. 1F) showed meningiovascular proliferation with psammomatous calcifications and areas of ossified fibrous tissue characteristic of MA. The patient has remained seizure-free in the 18 months since surgery, and his seizure medications are currently being tapered.

Case 2

A 17-year-old girl with no known illnesses suffered a single generalized seizure 6 months prior to admission. Treatment was begun with antiseizure medications and she has had no further seizures. MR imaging showed a mass in the medial right frontal lobe, which on T1-weighted images was predominantly isointense with gray matter (Fig. 2A). On spin-density and T2-weighted images (Figs. 2B and 2C), the mass was predominantly of high signal intensity but had a prominent area of signal void medially, which raised the possibility of an arteriovenous malformation. A CT scan (Fig. 2D) showed the mass to have both low- and high-density components, but there were no areas of dense calcification present. Angiography was normal, without evidence of a mass, arteriovenous malformation, or tumor stain. EEG showed a right frontal epileptogenic focus.

A right frontal craniotomy was performed and the stony-hard tumor resected. The tumor was intraaxial with no involvement of the overlying dura or falx. Histopathology of the mass showed meningiovascular proliferation and psammomatous calcifications typical of MA. There were numerous hemosiderin-laden macrophages scattered through the medial section of the mass. The patient remains seizure-free 1 year after surgery, although she is still taking anticonvulsants.

Discussion

MA is a rare benign cerebral mass lesion, with only 27 cases reported in the literature [2–15]. Its pathogenesis is uncertain, although it is generally accepted that it represents either a congenital faulty development of angiomatous and

Received September 5, 1990; revision requested October 15, 1990; revision received November 6, 1990; accepted November 8, 1990.

[1] Department of Radiology and Neurosurgery, University of Wisconsin Hospitals and Clinics, 600 Highland Ave., Madison, WI 53792. Address reprint requests to C. R. Partington.

[2] Department of Pathology, University of Wisconsin Hospitals and Clinics, Madison, WI 53792.

AJNR **12:549–552, May/June 1991** 0195–6108/91/1203–0549

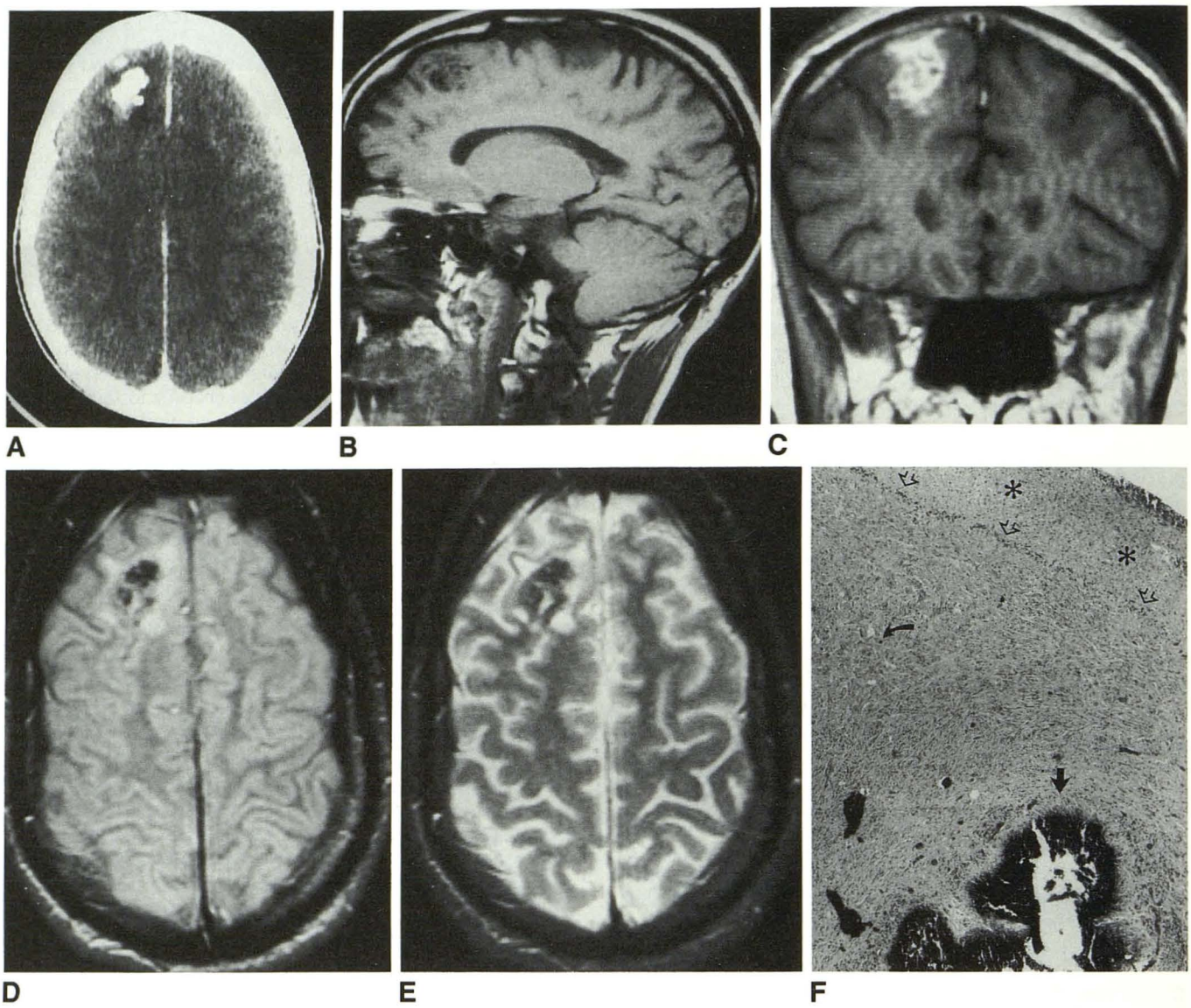

Fig. 1.—Case 1: 19-year-old boy with meningioangiomatosis.

A, Axial noncontrast CT scan at age 13 years shows low-density mass with central calcification in right frontal lobe.

B, Parasagittal T1-weighted (600/20/2) MR image at age 19 years shows nonhomogeneous intraaxial frontal lobe mass isointense with scattered areas of low signal intensity.

C, Coronal contrast-enhanced T1-weighted (600/20) MR image (gadopentetate dimeglumine 0.1 mmol/kg IV) shows intense, nonhomogeneous enhancement of intraaxial right frontal lobe mass. Note broad base directed toward dural surface.

D and *E*, Axial spin-density (2000/30/1) (*C*) and T2-weighted (2000/80/1) (*D*) MR images show right frontal lobe mass with central low signal intensity and surrounding high signal intensity.

F, Histopathologic specimen shows areas of dense calcification (*straight solid arrow*) and psammomatous calcification (*curved arrow*). The mass is composed primarily of meningioendothelial cells and has a sharply defined border (*open arrows*) with overlying cerebral cortex (*asterisks*). (H and E)

meningeal tissues (a true hamartoma) or an occult vascular malformation with the meningioendothelial elements proliferating at a later date along the endothelial linings of the occult vascular malformation [3, 6]. The histologic hallmarks of MA are leptomeningeal calcification and meningiovascular proliferation interwoven with fibrous connective tissue bands [1, 2]. There were hemosiderin-laden macrophages scattered throughout the mass in each of our cases, although there was no evidence of true hemorrhage from MA. There are no data available regarding the rate of growth of MA, but in case 1 there was a 6-year interval between identification of the tumor and its resection, during which time there was no perceptible change in the size of the mass on the CT scan. The patient's symptoms did worsen during this time, indicating that some change had taken place in the tumor. This implies that if MA is an actively growing lesion, the growth rate is very slow.

The imaging characteristics of MA have been included for

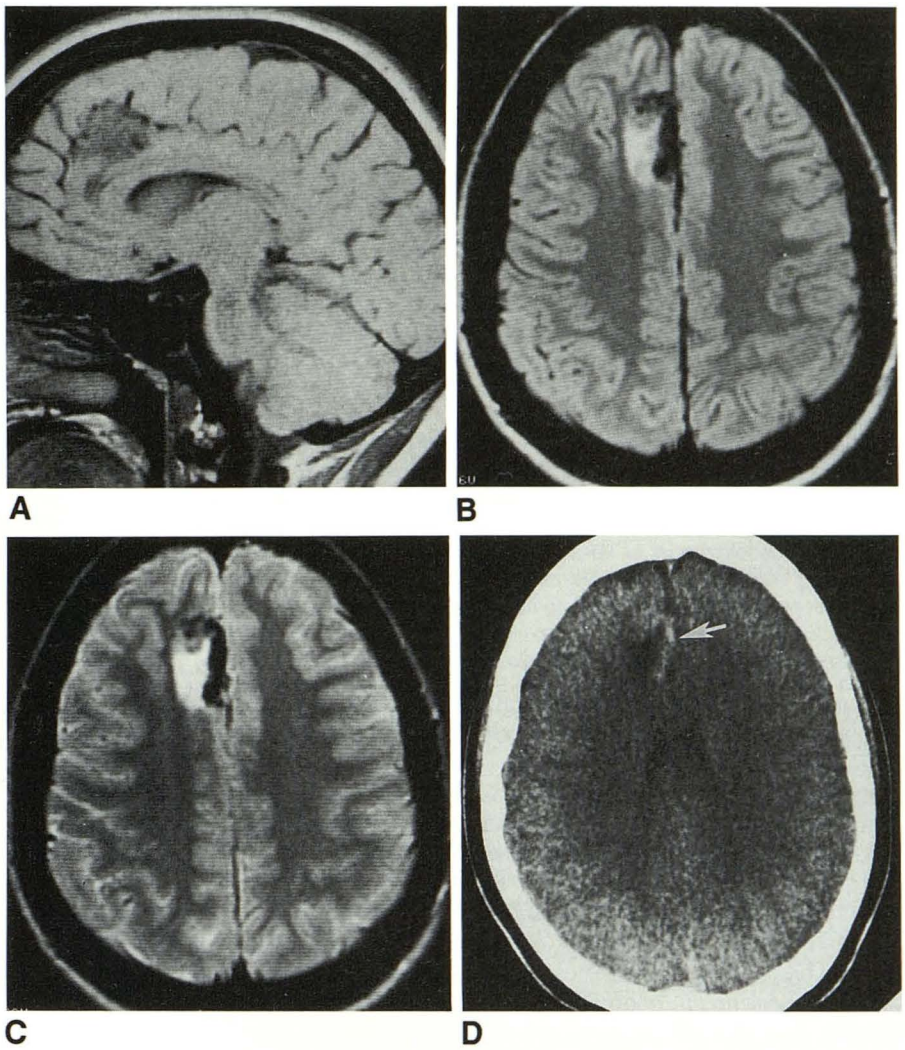

Fig. 2.—Case 2: 17-year-old girl with meningioangiomatosis.

***A*, Parasagittal T1-weighted (600/20/2) MR image shows low signal intensity mass within right frontal lobe. Mass has scattered areas of low signal intensity within it.**

***B* and *C*, Axial spin-density (2000/30/1) (*B*) and T2-weighted (2000/80/1) (*C*) MR images show high signal intensity mass in right medial frontal lobe. Mass has a prominent, very low signal intensity region medially.**

***D*, Axial noncontrast CT scan, obtained 1 month after MR study, shows rounded low-density mass in medial right frontal lobe. Mass has a high-density component medially (*arrow*) in a distribution corresponding to the areas of low signal intensity on MR. At pathology, this region had prominent psammomatous calcification and numerous hemosiderin-laden macrophages.**

15 of the reported cases (Table 1). Preoperative [7] and postbiopsy [5] MR have been reported for two cases. In each of the present cases, T2-weighted MR showed a cortical mass with heterogeneous signal intensity (Figs. 1B–1E, 2A–2C) and a small area of surrounding increased signal intensity representing either edema or gliosis. These findings are identical to those of the previously reported case in which the pathology specimen showed gliosis of the surrounding brain [7] and similar to those reported after a biopsy of MA [5]. The heterogeneity of the signal intensity of MA may be related to its histologic appearance, in which there are areas of dense calcification (expected low signal on T1- and T2-weighted images), loose connective tissue (expected isointense signal on T1-weighted images and high signal on T2-weighted images), and hemosiderin deposition (expected isointense signal on T1-weighted images and low signal on T2-weighted images). The only MR study in which contrast material was administered (Fig. 1C) showed striking enhancement of the mass itself with scattered areas of no enhancement corresponding to the areas of calcification seen on CT and noncontrast MR. In each of our cases and in the previously reported cases [5, 7] MR showed the mass to be intraaxial, but it could be seen to contact a meningeal surface at some point.

The CT findings in MA are those of a low-density mass with varying amounts of calcification. Two cases have been reported as normal on CT [2, 3]. The low density of the mass is presumably related to the loose packing of the cellular matrix with prominent perivascular spaces containing scattered spindle cells [2]. Administration of IV contrast material produced no significant tumor enhancement in our cases nor in five of those in the literature, although slight enhancement has been reported in three cases (Table 1). The calcification pattern seen in the mass on imaging studies and in pathologic specimens follows a spectrum from faint psammomatous calcifications as demonstrated in case 2 (Fig. 2D) to dense osteoid in the connective tissue component as shown by case 1 (Fig. 1A).

Angiography has been reported to show abnormal vessels in only two cases [3] and was normal or showed only signs of an avascular mass in the remaining nine cases in the literature and in both of our present cases (Table 1), even though delayed, magnified, biplane film angiography was done in addition to intraarterial DSA studies. Thus, the abnormal

TABLE 1: Presentation and Imaging Characteristics of Meningioangiomatosis in This Report and in Previously Reported Cases

Presentation	CT	Angiography	MR	Reference No.
Headaches	ND	Abnormal[a]	ND	[10]
Seizures	ND	Abnormal[b]	ND	[3]
Seizures	CM	Normal	ND	[2]
Headaches	CM	ND	ND	[11]
Seizures	Normal	Normal	ND	[3]
Seizures	CM	Normal	ND	[2]
Seizures	Normal	Normal	ND	[2]
Seizures	ND	Normal	ND	[2]
Incidental finding	CM (+E)	AM	ND	[6]
Seizures	CM (+E)	Normal	ND	[8]
Seizures	CM (+E)	Normal	ND	[9]
Seizures	CM	AM	[c]	[5]
Seizures	CM	ND	[c]	[7]
Seizures	CM	AM	[c] (+E)	Case 1
Seizures	CM	Normal	[c]	Case 2

Note.—ND = not done, CM = calcified mass, AM = avascular mass, +E = contrast enhancement.

[a] Angioma.

[b] Neovascularity.

[c] See text.

vascular component (vascular malformation) of MA seems to behave angiographically as an occult cerebral vascular malformation [16].

In conclusion, MA is a benign cerebral lesion that appears as a low-density mass with a varying amount of calcification on CT scans. In the few reported cases, MR imaging shows MA as a mass that is isointense with gray matter on T1-weighted images and hyperintense on spin-density and T2-weighted images, with areas of decreased signal intensity presumably representing calcification and hemosiderin deposition. The lesions are generally angiographically occult, and show no or slight contrast enhancement on CT, although the one case in which MR was performed with contrast administration showed intense enhancement. An extension of the mass to the leptomeninges may be demonstrated, although the dura may not be involved at surgery.

It will not always be possible to differentiate MA from glioma, oligodendroglioma, ganglioglioma, or meningioma. Identification of a broad base toward the dura will support the diagnosis of meningioma rather than MA; but in the absence of this, it may not be possible to further narrow the differential.

REFERENCES

1. Rubinstein LJ. Tumors of the central nervous system. In: *Atlas of tumor pathology*, second series, fascicle 6. Washington DC: Armed Forces Institute of Pathology, **1972**:252–307
2. Halper J, Scheithauer BW, Okazaki H, Laws ER. Meningio-angiomatosis: a report of six cases with special reference to the occurrence of neurofibrillary tangles. *J Neuropathol Exp Neurol* **1986**;45:426–446
3. Kasantikul V, Brown WJ. Meningio-angiomatosis in the absence of von Recklinghausen's disease. *Surg Neurol* **1981**;15:71–75
4. Paulus W, Peiffer J, Roggendorf W, Schuppan D. Meningio-angiomatosis. *Pathol Res Pract* **1989**;184:446–452
5. Ogilvy CS, Chapman PH, Gray M, de la Monte SM. Meningioangiomatosis in a patient without von Recklinghausen's disease. *J Neurosurg* **1989**; 70:483–485
6. Kunishio K, Yamamoto Y, Sunani N, et al. Histopathologic investigation of a case of meningioangiomatosis not associated with von Recklinghausen's disease. *Surg Neurol* **1987**;27:575–579
7. Kuzniecky R, Melanson D, Robitaille Y, Olivier A. Magnetic resonance imaging of meningio-angiomatosis. *Can J Neurol Sci* **1988**;15:161–164
8. Liu SS, Johnson PC, Sonntug VKH. Meningioangiomatosis: a case report. *Surg Neurol* **1989**;31:376–380
9. Sakaki S, Nakagawa K, Nakamura K, Takeda S. Meningioangiomatosis not associated with von Recklinghausen's disease. *Neurosurgery* **1987**;20:797–801
10. Rhodes RH, Davids RL. An unusual fibro-osseus component in intracranial lesions. *Hum Pathol* **1978**;9:309–319
11. Jun C, Burdick B. An unusual fibro-osseus lesion of the brain. *J Neurosurg* **1984**;60:1308–1311
12. Willson N, Kaufman MA, Bodansky SM. An unusual intracerebral connective tissue mass. *J Neuropathol Exp Neurol* **1977**;36:373–378
13. Bassoe P, Nuzum F. Report of a case of central and peripheral neurofibromatosis. *J Nerv Ment Dis* **1915**;42:785–796
14. Worster-Drought C, Dickson WEC, McMenemey WH. Multiple meningeal and perineural tumors with analogous changes in the glia and ependyma. *Brain* **1937**;60:85–117
15. Hozay J. Une angioneuromatose meningo-encephalgue diffuse. *Rev Neurol* (Paris) **1953**;89:222–236
16. Gomori J, Grossman R, Goldberg H. Occult cerebral vascular malformations: high field MR imaging. *Radiology* **1986**;158:707–713