Giant Cavernous Malformation of the Occipital Lobe

Carlos Filipe Chicani, MD, Neil R. Miller, MD, and Rafael J. Tamargo, MD

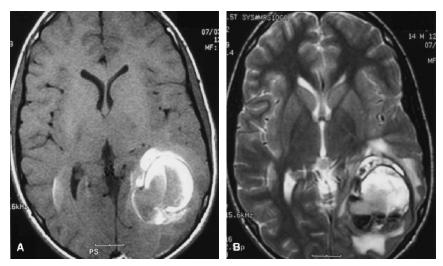


FIG. 1. Magnetic resonance imaging at initial presentation with severe headache. **A**: Axial T1 shows 4×5 cm bloodcontaining cystic mass in the left parietal-occipital region. **B**: Axial T2 shows the lesion's multicystic nature and surrounding edema.

Abstract: A 15-year-old boy who developed severe headaches and an incomplete homonymous hemianopia was found to have a large, well-circumscribed, multilobulated intracranial mass in the contralateral occipital lobe. The initial impression was that of a low-grade glioma or a vascular malformation. When the lesion increased in size and complexity, concern arose about the possibility of a malignant glioma. Upon craniotomy, it proved to be a giant cerebral cavernous malformation. This case is remarkable in that most cavernous malformations do not become symptomatic before the third decade of life and rarely attain such a large size.

(J Neuro-Ophthalmol 2003;23: 151–153)

A 15-year-old white boy was in his usual excellent health until May 1999, when he began to experience severe left-sided headaches accompanied by nausea and vomiting. When the headaches did not respond to antimigraine therapy, he underwent magnetic resonance imaging (MRI) that showed a well-circumscribed, round, left-sided 4×5 cm-diameter parieto-occipital region mass containing blood and blood products (Fig. 1). Neurologic and ophthalmologic examinations revealed no abnormalities except for an incomplete right homonymous hemianopia (Fig. 2). A cerebral angiogram showed evidence of a solid mass without any vascular abnormalities. As the mass was considered probably benign, no intervention took place.

In June 2001, 21 months later, the patient had a tonicclonic seizure followed by loss of consciousness for 30 minutes. A computed tomographic (CT) scan and repeat MRI showed that the hemorrhagic mass had increased in size to 7×5 cm (Fig. 3). Neurologic and ocular examinations revealed no new abnormalities; the right homonymous field defect had improved. Because of its large size and apparent growth, the lesion was suspected of being a glial malignancy, and a left occipital craniotomy was performed.

Copyright © Lippincott Williams & Wilkins. Unauthorized reproduction of this article is prohibited. J Neuro-Ophthalmol, Vol. 23, No. 2, 2003 151

Department of Ophthalmology (CFC and NRM), and Department of Neurosurgery (NRM and RJT), The Johns Hopkins Hospital, Baltimore, Maryland, USA. Dr. Chicani is now with the Department of Ophthalmology, Federal University of Sao Paulo, Brazil.

Address correspondence to Neil R. Miller, MD, Maumenee B-109, Wilmer Eye Institute, Johns Hopkins Hospital, 600 North Wolfe Street, Baltimore, MD 21287, USA; E-mail: nrmiller@jhmi.edu.

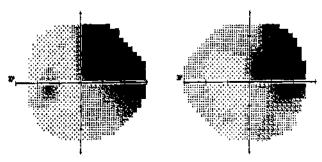


FIG. 2. Static perimetry at initial presentation (Humphrey 24–2) shows an incomplete right homonymous hemianopia.

During surgery, a large, well-circumscribed, multilobulated encapsulated lesion was encountered that contained multiple lobules filled with blood and blood products of different ages. It was completed resected.

Histopathologic examination revealed tortuous and disorganized vascular channels containing thrombi of varying ages. The vessels were separated by fibrotic tissue containing foci of calcification and hemosiderin deposition (Fig. 4). These findings were consistent with a cerebral cavernous malformation (CCM).

After surgery, the patient had no new neurologic deficits, although his visual field defect had worsened, becoming a dense right superior homonymous quadrantanopia. MRI performed 2 months after surgery showed only postsurgical changes in the left-temporo-occipital region.

Cerebral cavernous malformations consist of bloodfilled cavities lined by a single layer of endothelium and separated by neuroglia but not neural tissue (1). Calcification and areas of hemorrhage are commonly present within the lesions (1,2). CCMs are usually intraparenchymal, but extraparenchymal lesions are well described, particularly in the region of the cavernous sinus (3–5). Although most are sporadic occurrences, some are transmitted as an autosomal-dominant trait (3). Most lesions are solitary, but multiple lesions may be present, particularly in hereditary cases.

Cerebral cavernous malformations vary in size from a few millimeters to a few centimeters in diameter. Russell and Rubinstein (1) indicated that the largest CCM was $4 \times 3 \times 2$ cm. Our patient had a CCM that was substantially larger, measuring 5×7 cm. Although such cases have been reported, they are quite rare. The mechanism by which such lesions enlarge is recurrent bleeding, followed by organization of the clot, pseudocapsule formation, and secondary expansion. We presume that recurrent hemorrhage produced both the enlargement and the multicystic appearance in our case.

From 30 to 70% of patients with CCMs experience seizures as the first sign of the lesion (3). Others develop slowly progressive neurologic symptoms and signs from

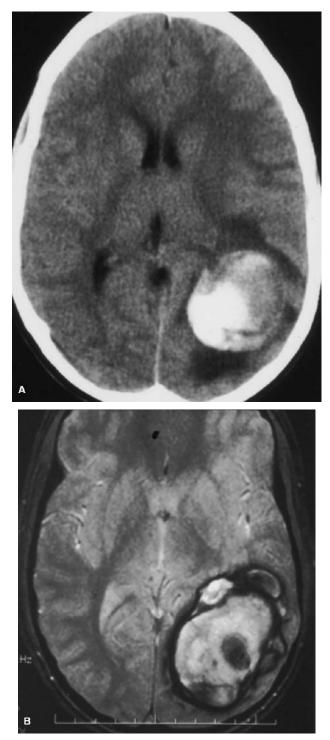


FIG. 3. Neuroimaging at time of seizure, 21 months after initial presentation. **A**: Axial noncontrast computed tomography shows persistent lesion in the left parietal-occipital region with surrounding edema. **B**: Axial proton density magnetic resonance imaging shows that the lesion has increased in size.

Copyright © Lippincott Williams & Wilkins. Unauthorized reproduction of this article is prohibited. 152 © 2003 Lippincott Williams & Wilkins

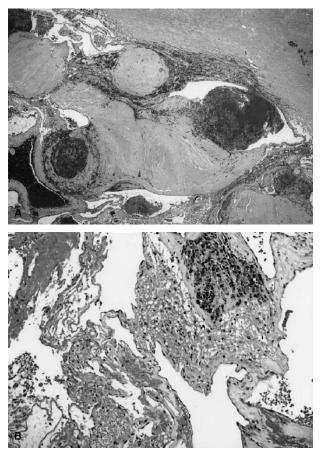


FIG. 4. Histopathologic of the cavernous malformation (**A**: low-power view; **B**: high-power view) shows tortuous and disorganized vascular channels containing thrombi of varying ages. The vessels are separated by fibrotic tissue containing foci of calcification and hemosiderin deposition.

compression, or acute neurologic deterioration from intraparenchymal or subarachnoid hemorrhage.

The neuroimaging appearance of a CCM is often diagnostic. On noncontrast CT, the lesion is typically round and well-circumscribed, with a homogeneous appearance that is isodense to moderately hyperdense with respect to brain parenchyma. There may be changes consistent with calcification, hemorrhage, or both. Enhancement after intravenous injection of iodinated contrast material varies from absent to striking. MRI typically reveals a low-signal hemosiderin rim that completely surrounds the lesion and is particularly prominent on T2 and gradient-echo images. The center of the lesion typically shows a well-delineated reticulated core of mixed-signal intensities caused by different stages of evolution of repeated hemorrhage. Because of their exquisite sensitivity to blood products, gradientecho sequences are particularly good at detecting small hemorrhagic lesions. Patients thought to harbor a solitary CCM often are found to have multiple lesions by this technique (1,3–5). Angiography is typically of little value in assessing CCMs, which show no vascular blush and no feeding arteries or draining veins.

The differential diagnosis of a CCM includes a lowgrade or malignant cystic glioma, a primitive neuroectodermal tumor (PNET), a thrombosed arteriovenous malformation, and an intracerebral hematoma. In many instances, as in our case, the correct diagnosis is not apparent until histopathologic examination of the specimen is made.

The treatment of symptomatic CCMs is controversial. Because of the low likelihood of hemorrhage or growth, most authors recommend observation for patients who are asymptomatic or for those who present only with seizures, particularly if the lesions lie in eloquent brain regions (6). Surgical excision is the treatment of choice for symptomatic CCMs located in accessible, noneloquent cerebral or cerebellar regions (7). Stereotactic radiosurgery is increasingly recommended for the less accessible, deep cerebral, and brainstem lesions (8). Our patient underwent excision of the lesion with an excellent functional result.

REFERENCES

- Bruner JM, Tien RD, McLendon RE. Tumors of vascular origin. In Bigner DD, McLendon RE, Bruner JM, eds. *Russell and Rubinstein's Pathology of Tumors of the Nervous System*, vol 2, edn 6. New York: Arnold, 1998:260–82.
- Muras I, Conforti R, Scuotto A, et al. Les cavernomes cérébraux. Considerations diagnostiques. J Neuroradiol 1993;20:34–41.
- Lee AG. Tumors and hamartomas of the blood vessels. In Miller NR, Newman NJ, eds. *Walsh and Hoyt's Clinical Neuro-Ophthalmology*, vol 2, edn 5. Baltimore: Williams and Wilkins, 1998:2223–96.
- Rigamonti D, Drayer BP, Johnson PC, et al. The MRI appearance of cavernous malformations (angiomas). J Neurosurg 1987;67: 518–24.
- Preul MC, Villemure JG, Leblanc R, et al. MRI diagnosis of brainstem cavernous angiomas presenting as tumours. *Can J Neurol Sci* 1992;19:376–82.
- Zabramski JM, Wascher TM, Spetzler RF, et al. The natural history of familial cavernous malformations: results of an ongoing study. J Neurosurg 1994;80:422–4.
- Churchyard A, Khangure M, Grainger K. Cerebral cavernous angioma: a potentially benign condition? Succesful treatment in 16 cases. *J Neurol Neurosurg Psychiatry* 1992;55:1040–5.
- Pollock BE, Garces YI, Stafford SL, et al. Stereotactic radiosurgery for cavernous malformations. *J Neurosurg* 2000;93:987–91.