

CASE OF THE MONTH

January 1997 - 7 Year Old Girl with Seizures

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Clinical history

This seven year old, right handed, white female presented with a 6-month history of partial complex seizures of increasing frequency, which were eventually controlled with Tegretol. The typical seizure episode was short, lasting only 20-30 seconds, but was followed by a prolonged postictal state. Physical examination was essentially normal and there were no focal neurological deficits. An MRI scan was obtained.

Neuroimaging

T-1 weighted MRI images demonstrated hypointense white matter signal in the left temporal lobe. The overlying cortex was distinct, but thinned and slightly hyperintense when compared to nearby cortical grey matter. T-2 weighted images showed this area to be hyperintense and the lesion was quite distinct on coronal and horizontal cuts (Fig 2). The lesion showed no contrast enhancement.

Gross

The specimen from the left partial temporal lobectomy had slightly thickened and very firm gyri (Fig 2). Sectioning revealed a well demarcated cortical gray ribbon which was slightly gritty. The subcortical white



Figure 2.

matter was soft and contained small cystic areas, often at the gray-white junction.

Microscopic

All sections showed a perivascular proliferation of spindle-shaped cells ensheathing numerous blood vessels present in the leptomeninges and the entire cortex (Fig 3a, 3b), but not in the white matter, which exhibited cystic changes. Many of these vessels were thickened in a concentric fashion. Gomori trichrome (Fig 3c) revealed dense collagen deposition around the vessels. Meningothelial cells were scanty and EMA immunohistochemistry showed only occasional positive cells within the cortical lesion. The intervening parenchyma con-



Figure 1.

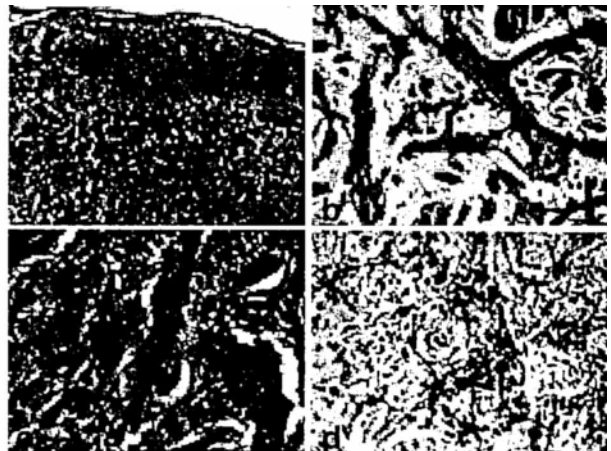


Figure 3. A+B, H=E; c, trichrome; d, GFAP

tained many pyramidal neurons, most of which were morphologically normal, however, there was considerable gliosis (Fig 3d). An occasional neuron contained a neurofibrillary tangle, but none showed granulovacuolar degeneration. Scattered psammoma bodies were present in some areas, especially in the superficial cortex.

Diagnosis

Meningioangiomas

Discussion

Meningioangiomas (MA) is a rare hamartomatous lesion of the cerebral cortex and sometimes the overlying leptomeninges. Approximately 50 cases are now described in the English language literature. Cases associated with NF comprise less than one-third of the reported cases (where commented upon) and all were discovered at autopsy (1, 6). These patients have a mean age of 32 years (range: 15-70), and male: female of 2: 1. About one-third of these cases had multiple lesions (1, 6). The exact incidence of MA in NF is unknown, but it was seen in 4 of 11 patients with "central" NF at autopsy (6), and in 1 of 135 patients with von Recklinghausen's disease in a clinical study. It appears to be far more common in NF-2 than in NF-1 (4). Most cases of MA, however, are not associated with NF (2).

There are clinical differences between the sporadic cases and those cases of MA associated with NF. In the sporadic cases the ratio of male to female patients is about even and the mean patient age is younger (17 years, range: 11 months-55 years). Also, these patients typically present with intractable seizures (5). Duration of seizures range from 2 months to 30 years before diagnosis, and are occasionally preceded by febrile convulsions in infancy (1). Headache is an uncommon presenting symptom. Rarely, MA is found at surgery for other lesions.

Topographically, MA affects the cerebral cortex in 90% of the cases and is usually in the frontal or temporal lobes. Other locations include the third ventricle, thalamus and cerebral peduncles (1), and the brain stem (3). Interestingly, about 80% of the cases are on the right side. Computerized tomography reveals a variable amount of calcification with little or no contrast enhancement. By magnetic resonance imaging, the abnormality is confined to the cortex and consists of isointensity or hypointensity on T1-weighted images. The T2-weighted images reveal a heterogeneous cortical mass surrounded by an area of increased signal intensity.

The gross appearance of MA is usually a sharply demarcated lesion with psammoma bodies and varying degrees of calcification. Abnormally prominent blood vessels may be seen and an associated arteriovenous malformation was seen in one instance (6).

The pathogenesis of MA is unclear. It is currently classified as a hamartomatous lesion, but its association with NF-2 is intriguing. While MA is obviously an abnormal proliferation involving vessels and meningotheial cells, it is uncertain if it is a static lesion or a very gradual process. It has a very low proliferation index (5). Total surgical resection is the treatment of choice for MA and the prognosis following surgery is excellent with cure of seizures in almost all cases (1, 2, 5).

References

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Case Abstract

A seven year old girl presented with six month history of seizures. An MRI scan showed a cortical lesion in the left temporal lobe which was resected. Neuropathologic examination demonstrated meningioangiomas, an unusual hamartomatous condition sometimes associated with neurofibromatosis 2. Approximately 50 cases of meningioangiomas have been reported in the literature. These are reviewed and compared to the current case.

For a more complete discussion of this case and additional micrographs please access this case on the WWW at: <http://path.upmc.edu/divisions/neuropath/bpath/cases/case10.html>. We welcome comments about these or similar cases our readers may have encountered.
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