

USCAP Neuropathology

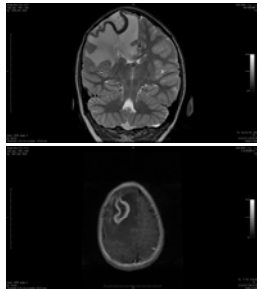
Case No. 3
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Clinical history

- The patient is a 9 year-old boy who has had seizures since age 2, at which time he was discovered to have a right frontal mass. The seizures were controlled with anticonvulsant medication until age 8, when the frequency increased to several per week. The patient's mother also reported behavioral disturbances that paralleled the increased number of seizures.

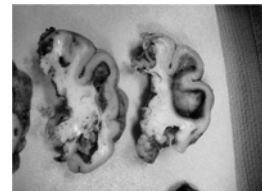
Imaging features

- Coronal T2 weighted image demonstrates low signal along the cortex of the right frontal lobe and the morphology of the cortex is abnormal. The subjacent white matter reveals T2 hyperintensity.
- On the axial post-contrast T1 weighted image, contrast enhancement is noted in the region of the abnormal cortical signal on T2.

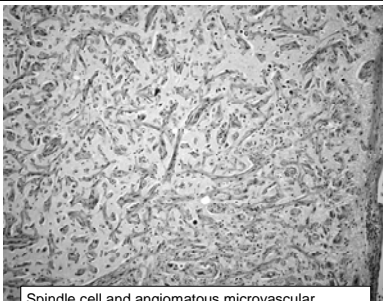


Surgery

- Intraoperative findings are that of a firm, discrete mass that follows the contours of the cortical ribbon.

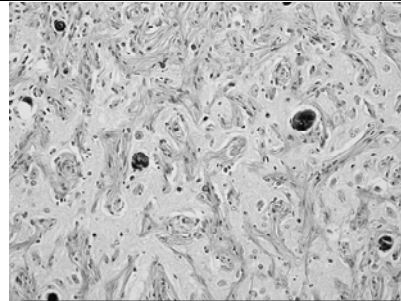


Case 3



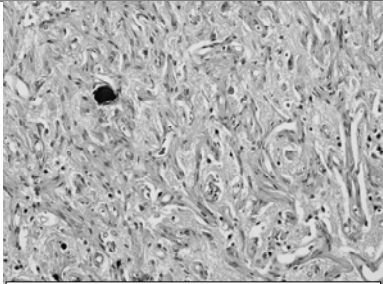
Spindle cell and angiomatous microvascular proliferation permeate the cortex

Case 3



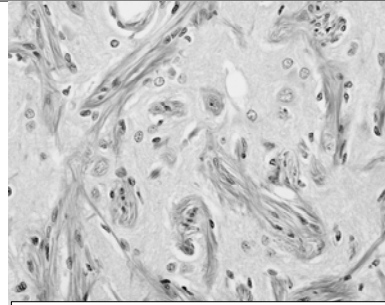
Spindle cells ensheath blood vessels; psammoma bodies are present.

Case 3



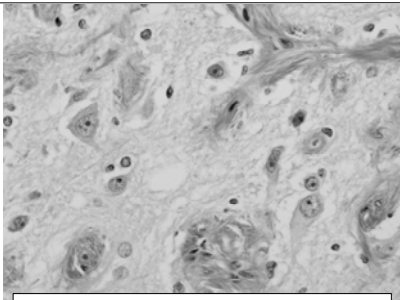
In some areas, only small islands of uninvolved cortex remain

Case 3



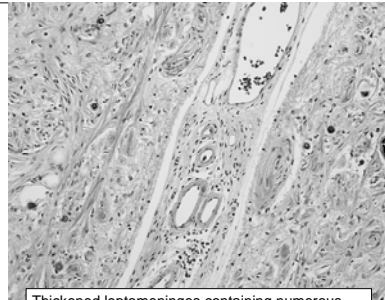
Neurons and gliosis are seen in the intervening cortex

Case 3



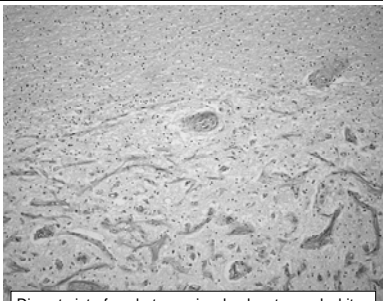
Disorganized cortex with dysplastic-appearing neurons.

Case 3



Thickened leptomeninges containing numerous blood vessels are flanked by gyri with exuberant spindle-cell proliferation and psammoma bodies

Case 3



Discrete interface between involved cortex and white matter

WHAT IS THE DIAGNOSIS?



Meningioangiomas

- **Definition**
 - Rare, intracortical plaque-like proliferation composed of meningotheelial cells, small blood vessels and fibroblast-like cells
- **Historical note**
 - 1915, Basso and Nuzum described the first case in a 15-year old boy
 - 1937, Worster-Drought coined the term

Meningioangiomas

- **Clinical features**
 - Children and young adults (range, 9 months to 71 years)
 - Sporadic (80%) and in association with NF2
 - Fronto-temporal region, often right hemisphere
 - Other locations include the third ventricle, cerebellum, thalamus and cerebral peduncles, and brain stem
 - Symptomatic cases present with seizures (81%)
 - Cured by excision; seizures persist in a minority

Meningioangiomas

Sporadic	NF2-associated
Solitary	Multifocal
Seizures	Asymptomatic
Mean age, 17	Mean age, 23
M:F=2:1	M:F=1:1
Temporal	Frontal

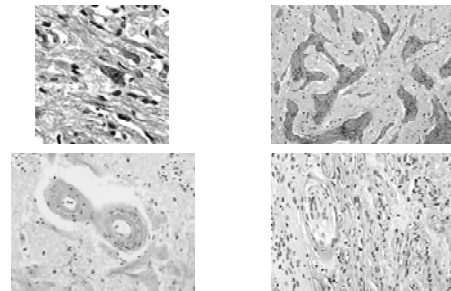
Meningioangiomas

- **Histopathology**
 - Leptomeningeal and intracortical proliferation of small blood vessels with perivascular spindled cells
 - Psammoma bodies, ossification
 - Gliosis and cortical disorganization
 - Entrapped cortical neurons may show neurodegenerative changes, such as neurofibrillary tangles, granulovacuolar degeneration, and Pick-like bodies

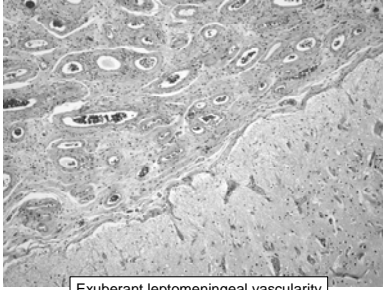
Meningioangiomas

- **Histopathology**
 - Dysplastic nests (microhamartomas), perilesional and remote, NF2 cases*
 - Verocay-like bodies
 - Meningoethelial whorls
 - Extensive hyalinization, NF2 cases*
 - In addition to meningioma, rarely, associated with AVM, oligodendroglioma

Meningioangiomas



Meningioangiomatosis



Exuberant leptomeningeal vascularity

Meningioangiomatosis

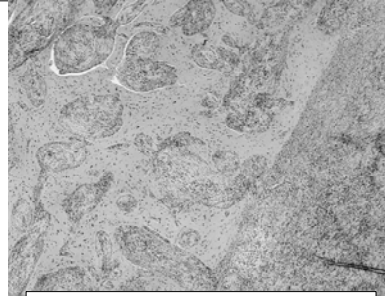
- Immunohistochemistry
 - Vimentin
 - Variable EMA, S100 protein, SMA, CD34
 - Low MIB1 (Ki-67)
- Electron microscopy
 - Proliferating cells contain elongated heterochromatin-rich nuclei and cytoplasm with microfilaments
 - Desmosomal junctions and interdigitating membranes

Meningioangiomatosis-Meningioma

- Share common genetic alterations; therefore, the “MA” component represents spread along the Virchow-Robin spaces
- In most examples, the “MA” spread arises from meningiomas based in the leptomeninges rather than the dura

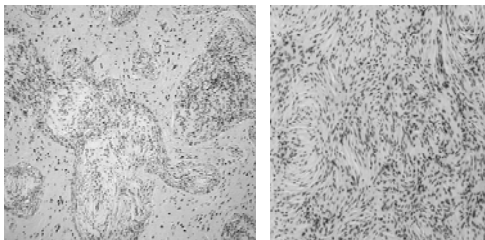
Perry A. et al. Insights into meningioangiomatosis with and without meningioma: a clinicopathologic and genetic series of 24 cases with review of the literature. Brain Pathol. 2005;15:55-65.

Meningioangiomatosis-Meningioma



Leptomeningeal meningioma with extensive infiltration along Virchow-Robin spaces mimicking MA

Meningioangiomatosis-Meningioma



Meningotheelial whorls within Virchow-Robin spaces; morphologically similar to overlying meningioma

Meningioangiomatosis

- Differential diagnosis
 - Vascular malformation
 - Meningioma
 - Schwannoma
 - Glial or mixed glial/neuronal neoplasms
- Associations
 - Meningioma, and rarely, AVM, or other neoplasms
 - Calcifying pseudoneoplasm (Fibro-osseous lesion) of the neuraxis

Meningioangiomas

- Histogenesis is controversial
 - Meningothelial origin:
 - Meningothelial whorls and variable EMA staining
 - Myofibroblastic/fibroblastic origin
 - Paulus et al. described staining for procollagen I and III and lack of collagen IV
 - Pluripotential stem cell origin
- Genetics
 - NF2
 - 15-20% MA cases; however, Rubenstein reported MA in 36% NF2 autopsies

Meningioangiomas

- Take home points
 - Sporadic, isolated MA is a lesion of probable hamartomatous origin that causes seizures
 - Most cases are sporadic
 - Sporadic MA and MA associated with NF2 differ clinically and histologically
 - "MA"-M most likely represents a distinctive pattern of cortical spread that differs from true brain invasion
 - Low-power examination is the key to recognizing the characteristic histopathologic features
 - Immunohistochemical studies contribute little to the practical diagnosis

Joan Miró, La poeta

