Intraparenchymal meningioma originating from underlying meningioangiomatosis

Case report and review of the literature

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The authors present the case of a 20-year-old woman with right-sided temporal intraparenchymal meningioma originating from underlying meningioangiomatosis. The patient manifested temporal-type seizures as the initial symptom. She had no stigmata of neurofibromatosis Type 2. Neuroradiological studies revealed an intraxial, contrast-enhancing lesion in the right temporal lobe. After gross-total removal of the tumor, histological examination revealed a transitional meningioma with surrounding neural parenchyma and features of meningioangiomatosis. Although there were no signs of anaplasia, necrosis, or increased mitosis, tumor islands were observed in the adjacent neuropil. The rare association of meningioangiomatosis and meningioma is discussed, along with radiological and surgical findings.

Key Words • meningioma • intracranial–intraparenchymal meningioma • meningioangiomatosis

Meningioangiomatosis is a rare meningovascular lesion characterized by proliferation of meningothelial and fibroblast-like cells encircling small cortical vessels. The histogenesis of these sporadic lesions is unclear. They may present with seizures and persistent headache, and multiple meningioangiomatosis is generally associated with NF2. Unusual associations of meningioma and meningioangiomatosis are reviewed. All of the reported cases are extraaxial meningiomas with meningioangiomatosis in the underlying cortex (Table 1).

Herein we present a unique example of an intraparenchymal meningioma originating from an underlying meningioangiomatosis.

Case Report

History. This 20-year-old woman was admitted to the neurosurgery clinic after experiencing symptoms for 10 months, including transient unresponsiveness to people during conversation, involuntary movements of her hands, and staring steadily with loss of consciousness. Her medical history was normal and there was no family history of NF2.

Examination. The patient was intellectually normal with no stigmata of NF2. Results of her physical and neurological examinations were within normal limits. Results of electroencephalographic studies demonstrated no epileptic abnormality. A lesion was observed in the right temporal lobe that was hypointense on T1-weighted and hyperintense on T2-weighted MR images. After intravenous administration of Gd-DTPA contrast agent, there was an intense enhancement of the lesion. Edema was apparent in the surrounding parenchyma. We observed a regularly contoured 8-mm cyst within the brain parenchyma that was isointense to cerebrospinal fluid and located medial to the described lesion in the right temporal lobe. This lesion was totally intraaxial, and the signal-void tubular structures within the lesion were thought to be blood vessels. There was no calcification (Fig. 1). Possible preoperative diagnoses included a glial tumor.
Operation. A neuronavigation system was used to determine the exact location of the lesion while preserving optimal cortical neuronal protection. After a right temporal craniotomy, the solid, pinkish, firm tumor was removed gross totally via the transsulcal route. The opercular part of one of the middle cerebral artery branches, which bypassed the tumor mass, was saved and the tumor behind it was coagulated.

Postoperative Course. The postoperative course was uneventful. The patient was doing well, with no residual tumor found on a follow-up MR image obtained 12 months after the operation.

Histopathological Findings. The surgical specimen was fixed in 10% formalin, embedded in paraffin, and stained with hematoxylin and eosin and Gomori’s trichrome. For immunohistochemical analysis, the following antibodies were used: polyclonal antibody to S-100 protein (1:300) and monoclonal antisera to glial fibrillary acidic protein (1:300), vimentin (1:100), EMA (1:100), and factor VIII–related antigen (1:40). (All antibodies were purchased from Dako Corp., Carpinteria, CA, with the exception of the factor VIII–related antigen, which was acquired from Signet, Dedham, MA.) Immunohistochemical studies were performed using the avidin-biotin-peroxidase complex method. Diaminobenzidine was used as a substrate.

Histopathological analysis revealed a neoplasm that showed well-defined borders with the adjacent brain parenchyma (Fig. 2 upper left). The tumor was composed of neoplastic syncytial cells that formed characteristic meningeal whorls (Fig. 2 upper right). There was a tendency toward concentric whorl formation, often around a central capillary. Some of the whorls contained hyaline and occasionally psammoma bodies. Tumor cells showed oval nuclei and an even chromatin pattern with eosinophilic cytoplasm. On immunohistochemical studies the neoplastic cells expressed vimentin, EMA, and S-100 protein. There was no mitosis, necrosis, or any other sign of anaplasia; however, several tumor islands were present in the adjacent parenchyma (Fig. 2 center). The surrounding cortex showed perivascular proliferation of spindle-shaped cells ensheathing numerous parenchymal and leptomeningeal vessels (Fig. 2 lower left). The vessels were thickened concentrically with dense collagen deposition, as demonstrated by Gomori’s trichrome stain (Fig. 2 lower right). Immunohistochemical studies revealed that many of the proliferating spindle-shaped cells expressed vimentin, whereas occasional EMA-positive meningothelial cells were present in the perivascular lesion. The parenchyma embedded in the tumor contained enlarged reactive astrocytes that showed strong glial fibrillary acidic protein im-

TABLE 1

<table>
<thead>
<tr>
<th>Authors &amp; Year</th>
<th>Patient Age (yrs), Sex</th>
<th>NF2 Onset</th>
<th>Localization</th>
<th>Type</th>
<th>Signs of Anaplasia</th>
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<tbody>
<tr>
<td>Auer, et al., 1982</td>
<td>15, M</td>
<td>NM</td>
<td>headache</td>
<td>frontal</td>
<td>fibroblastic</td>
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<tr>
<td>Louw, et al., 1990</td>
<td>33, M</td>
<td>NM</td>
<td>headache</td>
<td>frontotemporal</td>
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<tr>
<td>Wilson, et al., 1991</td>
<td>17, M</td>
<td>none</td>
<td>seizure</td>
<td>frontal</td>
<td>transitional</td>
</tr>
<tr>
<td>Blumenthal, et al., 1993</td>
<td>9/12, M</td>
<td>none</td>
<td>seizure</td>
<td>frontal</td>
<td>transitional</td>
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<tr>
<td>Giangaspero, et al., 1999</td>
<td>28, M</td>
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<td>seizure</td>
<td>temporoparietal</td>
<td>transitional</td>
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<tr>
<td>present study</td>
<td>20, F</td>
<td>none</td>
<td>seizure</td>
<td>temporal</td>
<td>transitional</td>
</tr>
</tbody>
</table>

* NM = not mentioned.

FIG. 1. Upper Left: Coronal T1-weighted MR image revealing an intraaxial mass in the superior part of the right temporal lobe, with a cystic component (arrowheads) located medially; tubular vascular structures are indicated by arrows. Upper Right: Axial T1-weighted MR image demonstrating a hyperintense mass (arrow) with surrounding edema. Lower Left: Axial T1-weighted MR image demonstrating striking enhancement of the lesion (arrowheads) after Gd-DTPA administration. The vascular structure of the tumor is indicated by the arrow. Lower Right: Coronal T1-weighted MR image demonstrating striking enhancement of the lesion (arrowheads) after Gd-DTPA administration. Vascular structure is indicated by a thin arrow and the cyst by a thick arrow.
munoreactivity. There were a few neurofibrillary tangle-bearing neurons, as reported previously.5,16,20,22

Discussion
This case involved an intraparenchymal meningioma that originated from an underlying meningioangiomatosis, which is a rare plaquelike cortical meningo-vascular lesion. Several theories have been proposed to explain the histopathogenesis of meningioangiomatosis, in a spectrum ranging from developmental abnormality to meningioma en plaque.7,17 Although the histogenesis of meningi-
Meningioma associated with meningioangiomatosis

Meningioangiomatosis is still controversial, it is accepted that the proliferating cells are meningothelial. Histologically, this entity is characterized by the proliferation of meningothelial and fibroblast-like cells ensheathing small vessels. Few cases developing outside the cerebral cortex have been reported. Meningioangiomatosis may be single or multifocal, with NF2-associated examples generally multifocal and asymptomatic. Sporadic meningioangiomatosis is a single cortical lesion that usually occurs in children and young adults suffering from epilepsy or persistent headache. There is a single case of concurrence of meningioangiomatosis and oligodendroglioma in the literature; moreover, an association with extraaxial meningioma has been reported occasionally.

Two of the reported meningiomas associated with meningioangiomatosis were fibroblastic, mimicking schwannoma histologically. One of the two was reported as an example of a frontal lobe perivascular schwannoma in a 15-year-old child before the general application of immunohistochemistry. The other four examples were histologically identified as transitional-type meningiomas like the present case. Five of the reported meningiomas lacked notable mitotic activity, necrosis, or any other signs of anaplasia, whereas the sixth tumor contained a central area of necrosis with scattered mitotic figures. In our case, although there was no mitosis, necrosis, or any other signs of anaplasia, there were tumor islands in the surrounding brain parenchyma. It is widely accepted that brain invasion by a meningioma, even in the absence of other signs of malignancy, qualifies the tumor as malignant, a criterion used for a wide-based dural attachment, signal changes in skull due to tumor infiltration, surrounding connective tissue that forms a sharp demarcation between the tumor and the brain, and mass effect on adjacent brain tissue. However, other features of meningiomas were present, such as peritumoral edema, homogeneous enhancement after intravenous administration of a contrast agent, hypointense appearance to gray matter on T1-weighted MR images.

Intraparenchymal meningiomas are usually related to the ventricular system and they are well described. There are meningiomas of the deep Sylvian cleft anchored to the internal carotid artery and its branches. The tumor in our case can be classified in the deep Sylvian category, and it was anchored to the opercular artery. The suspected cells of origin are the stromal cells of the pia-arachnoid that wrap the perforating blood vessels as they enter the surface of the brain.

In our case, we speculate that a clonal proliferation of perivascular meningothelial cells encircling the vessels of the meningioangiomatosis, possibly secondary to some aberrant production of growth factors or loss of inhibitory factors, such as NF2 gene inactivation, could have led to meningioma formation. We assume that meningioangiomatosis may accompany intraparenchymal meningiomas that are not related to the ventricular system more commonly than was previously thought, especially in children and young adults. Biopsy sampling of the surrounding brain parenchyma in cases of intraparenchymal meningioma may help to identify cortical meningioangiomatosis.

References


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