Fibroma of the meninges in a child: immunohistological and ultrastructural study

Case report

MIGUEL REYES-MUGICA, M.D., PAULINE CHOU, M.D., FRANK GONZALEZ-CRUSSI, M.D., AND TADANORI TOMITA, M.D.
Departments of Pathology and Neurosurgery, Children's Memorial Hospital, Northwestern University, Chicago, Illinois

A case of meningeal fibroma in a 5-year-old girl is described. The lesion presented as a benign intracranial tumor, eroding the frontal bone and protruding under the skin. It was composed of fibroblasts and collagen, embedded in a loose background with focal myxoid changes. The authors describe the patient's clinical presentation and the tumor's histological, immunohistochemical, and ultrastructural features, and discuss its differential diagnosis. It is concluded that fibromas of the meninges should be distinguished from fibroblastic meningiomas.

KEY WORDS • meningeal tumor • fibroma • ultrastructural study

Tumors arising in the meninges are almost invariably classified as meningiomas. The term “meningioma” was coined by Cushing in 1922 in reference to a variety of tumoral lesions that arise in the meninges. However, only those tumors with unequivocal evidence of meningothelial differentiation should be included in this category. On the other hand, diagnostic assignment on the basis of histogenesis is difficult in meningeal tumors because of the well-known complexity of meningeal embryology and the multiplicity of patterns described in meningiomas. Although these tumors are usually easy to identify, some primary meningeal tumors present uncommon histological appearances that challenge the diagnostic skill of the pathologist. In this report, we describe an unusual meningeal tumor that illustrates such difficulties, and we analyze its histological, immunophenotypic, and ultrastructural features.

Case Report

This 5-year-old girl was admitted to the Children's Memorial Hospital in Chicago with a small protuberance on the forehead and a 3-month history of headaches.

Examination. The physical examination showed a firm mass, 1.5 cm in diameter, located in the forehead and covered by unremarkable skin. The neurological evaluation was normal except for papilledema. Cranial computerized tomography (CT) and magnetic resonance imaging revealed a large solid mass in the right frontal lobe, 9 cm in diameter, eroding the frontal bone and compressing the clearly demarcated adjacent brain (Fig. 1). Cerebral angiography revealed a very vascular tumor supplied by a hypertrophic meningeal artery and branches of the anterior cerebral artery (Fig. 2).

Operation. A right frontoparietal craniotomy was performed. There was an extremely vascular tumor protruding through a bone defect 2 cm in diameter. Numerous enlarged diploic vascular channels were noted in the frontal bone. Despite identification and transection of the main trunk of the middle meningeal artery through a burr hole, the vascular appearance of the tumor did not change. After frontoparietal craniotomy, it was noted that the dura mater was tense, and a well-demarcated tumor was seen protruding into the dura. The tumor was not adherent to the inner surface of the dura except for the portion at the cranial defect. Branches of the anterior cerebral artery located in the medial posterior portion of the tumor were identified, secured, and transected, and the mass was removed in toto. The subjacent frontal lobe was covered by arachnoid membrane, and no invasion by the tumor was noted. The inner surface of the frontal bone flap was...
FIG. 1. Sagittal T1-weighted magnetic resonance image revealing a large frontal neoplasm with prominent vascularity. The lesion is eroding the frontal bone and protruding under the skin.

Free of tumor except for the thin indented surface around the bone defect, which was removed. A wide margin of dura around the involved bone defect was also excised. A pericranial graft was used for dural closure. The craniotomy was replaced and the bone defect was closed using the inner table of the bone flap.

Postoperative Course. The postoperative course was uneventful. Papilledema subsided over the ensuing 2 weeks. A postoperative CT scan showed complete disappearance of tumor.

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Neuropathological Examination. The tumor measured 9 × 7 × 6 cm. The external surface was smooth, with an area of attachment to the frontal bone. The cut surface (Fig. 3) was yellow and homogeneous. No areas of necrosis or hemorrhage were seen, but a central feeding vessel was identified in the tumor. Representative sections were processed for routine histology; frozen material was used for estrogen receptor determinations according to the method of Pensler, et al., and for flow-cytometric deoxyribonucleic acid (DNA) studies, as described by Koss, et al. Glutaraldehyde-fixed 1-cm samples were processed for electron microscopic examination.

The histological examination (Fig. 4) showed a pro-

TABLE 1
Results of immunostaining

<table>
<thead>
<tr>
<th>Antigen</th>
<th>Tumor Cells*</th>
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<tbody>
<tr>
<td>S-100 protein</td>
<td>+++</td>
</tr>
<tr>
<td>carcinoembryonic antigen</td>
<td>+</td>
</tr>
<tr>
<td>vimentin</td>
<td>+</td>
</tr>
<tr>
<td>factor XIIIa</td>
<td>+++</td>
</tr>
<tr>
<td>neuron-specific enolase</td>
<td>+</td>
</tr>
<tr>
<td>glial fibrillary acidic protein</td>
<td>-</td>
</tr>
<tr>
<td>epithelial membrane antigen</td>
<td>-</td>
</tr>
<tr>
<td>cytokeratin</td>
<td>-</td>
</tr>
<tr>
<td>factor VIII-related antigen</td>
<td>-</td>
</tr>
</tbody>
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* Degree of staining: +++ = strongly positive; + = weakly positive; = negative.

Fig. 2. Cerebral angiograms showing a vascularized tumor in the frontal region. a: External carotid angiogram disclosing a hypertrophic middle meningeal artery (arrowhead) and abnormal vascular ramifications. b: Internal carotid angiogram showing a right frontal mass supplied by abnormal neovascularization arising from the anterior cerebral artery.
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FIG. 3. Cut surface of the resected tumor showing a homogeneous surface.

Liferation of spindle cells with centrally located normochromic nuclei and scanty eosinophilic cytoplasm with long thin prolongations. The general appearance of the neoplasm was loose, and a small amount of myxoid background was seen. There were no mitoses, anaplasia, or foci of necrosis. In some areas, a storiform pattern was observed; occasional plasma cells were present. Masson's trichrome, hematoxylin-phloxine-saffron, and reticulum stains demonstrated abundant fine collagen fibers in the intercellular spaces.

The avidin-biotin complex immunostaining procedure, as described by Hsu, et al.,\(^9\) was used to detect expression of the following antigens: glial fibrillary acidic protein (GFAP), neuron-specific enolase, cytokeratin, epithelial membrane antigen, carciNoembryonic antigen (CEA), S-100 protein, vimentin, factor XIIIa, and factor VIII-related antigen. The results of immunostaining are summarized in Table 1.

Three representative fragments were studied under the electron microscope and showed the same morphology (Fig. 5). The tumor cells had a moderate amount of cytoplasm with abundant dilated cisterns of rough endoplasmic reticulum; some lipid vacuoles were also present. No interdigitations, desmosomes, or basement membranes were observed.Interstitial collagen fibrils were frequently identified.

Estrogen receptor determination was negative. Flow-cytometric DNA studies showed a diploid population with a low S-phase, suggesting a slow-growing lesion.

Discussion

Brain tumors are the most frequent solid tumors found in children.\(^13\) Accordingly, pediatric pathologists are generally familiar with these neoplasms. In contrast, meningeal tumors are uncommon in childhood;\(^5,19\) meningoIomas constitute 2% of all intracranial tumors at this age.\(^4,24\) Likewise, only 1.7% of all meningiomas occur in pediatric patients,\(^16\) and may behave differently compared with the same type of tumors in adults.\(^4,5,10\)

Differential Diagnosis

Tumors and tumor-like meningeal conditions that must be considered in the differential diagnosis of our case include: extra-axial glioma, fibroblastic meningioma, juvenile fibromatosis, cranial fasciitis of infancy,
and meningeal myxoma. Astrocytic neoplasms may be found primarily in meningeal localization,26 but are negative for collagen in trichrome stains and the GFAP immunostain should be positive.

In the meningioma group, only the fibroblastic variant is histologically similar to our patient's tumor; however, the latter showed neither a whorled pattern nor meningothezial features, which are usually seen in at least some foci in meningiomas.10,11,15,25 In addition, the immunophenotype in our case was negative for cytokeratin and epithelial membrane antigen, two markers that are reportedly positive in 32% and 100% of these tumors, respectively.23 The only immunoreactive epithelial marker in our case was CEA, which has been reported as positive mainly in secretory meningiomas.16 The positivity for S-100 protein and vimentin suggests an ectomesenchymal nature in our patient's lesion. Ultrastructural features of a meningothezial nature, such as cellular interdigitations and desmosomes, were not found. Since hormone dependence has been described in meningiomas based on epidemiological, clinical, and laboratory evidence and estrogen and progesterone receptors have been found in these tumors,16 we performed estrogen receptor determination, but obtained negative results.

Fibromatoses are pseudotumoral lesions with invasive borders, abundant broad collagen bundles, great cellularity, and compact disposition.5 The distinction of these lesions from fibrosarcomas is established with difficulty, but they lack the loose appearance and myxoid background seen in our patient. To the best of our knowledge, fibromatoses have not been described in the meninges.

Cranial fasciitis of childhood is an entity closely related to nodular fasciitis. Its presentation usually features a rapidly growing mass arising in the deep fascia in the scalp, protruding under the skin, and causing some erosion of the outer table of the skull, but lacking intracranial growth. The mass averages 2.5 cm in size, and attachment to the dura is rare. Histologically, osseous metaplasia has been described in some cases.14,17,21 Ultrastructural examination usually discloses fibroblasts and myofibroblasts.21 No cases with intracranial growth have been reported. Recently, Montgomery and Meis18 have reported 53 cases of nodular fasciitis uniformly negative for S-100 protein immunohistochemical staining, in contrast with the observed results in our case.

Meningeal Fibroma

The diagnosis favored for our case is meningeal fibroma, a lesion known to occur intracranially,1,11,15,20 as well as in extracranial sites,7 and rarely arising as a meningeal primary tumor.16,24 The intracranial cases presumably represent true fibromas, although this has only been substantiated by electron microscopy in rare cases8,20 and in none by immunohistochemical studies. In support of this diagnosis is the immunostaining pattern and the uniformity of the ultrastructural appearance. Vimentin is generally expressed by mesenchymal tumors. Another interesting finding is the positivity of factor XIIIa, which has been found in some fibroblastic cells in the dermis as well as in some other locations. More studies are required to define the significance of this finding.

Meningeal and cerebral fibromas are benign tumors that can be clearly distinguished from fibroblastic meningiomas based on the absence of meningothezial differentiation, negative immunostaining for epithelial membrane antigen, and lack of desmosomes and cellular interdigitations on ultrastructural study. Flowcytometric DNA studies in our case demonstrated a diploid cell population, in keeping with the benign-looking morphological appearance of the lesion. This finding supports the previously described benign clinical course of intracranial fibromas.20 Meningeal and cerebral fibromas usually occur in the first two decades of life, and there is a slight male predominance. Of nine previously reported cases, only one was infratentorial.15,20

Interestingly, some cases histologically similar to ours have been classified as meningeal myxomas.2,22 We suggest that the term "fibroma" is more appropriate because it conveys the presumed histogenesis of the tumor although, in the past, this term was used in reference to the tumors now grouped as "fibroblastic meningiomas."11 The term "myxoma" is at present usually applied to lesions without clear gross demarcation that appear microscopically as locally infiltrative lesions,1 and are thus markedly different from the case described here.

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References

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Address reprint requests to: Miguel Reyes-Mugica, M.D., Department of Pathology, Children’s Memorial Hospital, 2300 Children’s Plaza, Box #17, Chicago, Illinois 60614.