Exclusively intracranial cranial fasciitis in a child

Case report

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The authors present a case of an intracranial tumor in a 7-year-old girl leading to increasing hemiparesis. The lesion arose from the dura and consisted of fibroblasts in a myxoid matrix. The diagnosis of cranial fasciitis was made. The histological, immunohistological, and ultrastructural features of the tumor are described and the differential diagnosis is discussed.

KEY WORDS • cranial fasciitis • meningeal tumor • classification

Since Cushing’s3 work in the 1920s, most tumors originating in the meninges have been considered meningiomas. Indeed, the vast majority of meningeal neoplasms arise from or differentiate toward arachnoidal cells.8 There is, however, a high incidence of sometimes rare neoplasms arising from the meninges that have to be distinguished from meningiomas.4,5,13,17 In this report, we describe the histological, immunohistological, and ultrastructural appearance of one such unusual tumor.

Case Report

This 7-year-old girl was admitted to the hospital with a left-sided hemiparesis, which had been developing for more than 3 months, and was particularly pronounced in the arm. Four weeks before admission to our clinic she had become ill with chicken pox and presented with deterioration of the neurological symptoms: her left shoulder was conspicuously lowered, and she complained of blurred vision and pain in her eyes.

Examination. Examination of the family medical history revealed that the patient, her mother, and grandmother all suffered from migraine, although there was no history of tumor. Physical examination revealed papilledema on both sides. A weakness of the left arm was found. The muscle reflexes were found to be symmetrical and normal, but a positive Babinski reflex was evident on both sides.

Imaging Studies. Cerebral computerized tomography revealed a sharply bordered right frontoparietal mass of 8 × 8 × 4 cm, with strong contrast enhancement. It had extensive contact with the dura and the falx (Fig. 1). A subsequent angiogram showed a well-vascularized tumor, supplied by branches of the internal and external carotid arteries (Fig. 2). Preoperatively the branches of the external carotid artery were partly embolized with Ivalon glue.

Operation. Intraoperatively a profusely bleeding, hard, tough tumor arising from the dura was removed. No tumor tissue was found extradurally, and there was no connection with the bone, which was rather thin. The tumor’s borders could be easily traced after coagulation of the vessels feeding the tumor. The dura was removed and replaced by lyodura. After total tumor removal, an atrophic cortex with intact arachnoidal covering was revealed.

Postoperative Course. Postoperatively no further neurological deficits were seen. Clinical neurological control examinations showed a rapid return to normal within 3 months. Postoperative magnetic resonance imaging revealed the unfolded atrophic brain with no evidence of tumor.
Neuropathological Findings. The white-yellowish tumor was hard and elastic with a smooth surface. The cut surface revealed a few gelatinous areas (Fig. 3). Representative specimens were fixed in formaldehyde and embedded in paraffin or resin. The histological examination showed a proliferation of spindle cells of variable cell density. The cells were arranged in a loose, haphazard pattern in a myxoid matrix (Fig. 4). The matrix stained positively with Alcian blue, indicating the presence of mucin. Occasionally, mitotic figures were present, although no atypical figures were seen. Most areas were poorly collagenized, but a few portions of the lesion had abundant collagen fibers. There was a border zone between the dura and the tumor in which collagen fibers from the dura interlinked with the poorly collagenized tumor. The tissue contained abundant thin-walled vessels. Lymphocytic infiltrates were seen in an irregular pattern within the lesion. The embolized portions contained numerous macrophages.

Specimens of the tumor studied under the electron microscope showed the same features. Spindle- and stellate-shaped cells lay in a myxoid matrix. Interstitial collagen fibrils were rarely identified. There were no interdigitations, desmosomes, or basement membranes: structures typical of meningiomas. The cytoplasm of the cells contained abundant rough endoplasmic reticulum (Fig. 5).

Immunohistochemical staining was performed with antibodies against cytokeratin 1, S-100 protein, carcinoembryonic antigen, epithelial membrane antigen, vimentin, desmin, neuron-specific enolase, and α-actin. The results of the immunohistochemical characterization are presented in Table 1.

Discussion

In 1980 Lauer and Enzinger described cranial fasciitis as a tumorlike fibroblastic lesion resembling nodular fasciitis. The authors distinguished this new entity from nodular fasciitis by emphasizing its location, microscopic appearance, and characteristic age incidence. Cranial fasciitis is rare. Since its first description in 1980, only 21 cases have been reported. Clinically, the lesion presents as a rapidly growing mass along the cranium. Although the lesion grows rapidly, the expansion seems to be self-limiting. It arises from the deep layers of the scalp or from the periosteum of the skull. In some cases...
cases, the lesion extends through the skull and is attached to the dura. Macroscopically, the whitish-gray tumors are well circumscribed. They are rubbery-to-hard in consistency. Microscopically, these lesions, like those of nodular (pseudosarcomatous) fasciitis, consist of loose, irregular accumulations of fibroblasts set in a myxoid matrix. In some cases the lesions contain densely collagenized areas resembling a fibromatosis. Focal areas of hemorrhage and lymphocytic infiltrations are regularly seen. At the ultrastructural level, nodular fasciitis lesions consist of spindle-shaped cells rich in rough endoplasmic reticulum. The nuclei contain finely dispersed chromatin, usually a single nucleolus. Collagen fibrils are located outside the cells identified as myofibroblasts by Wirman. Wide areas between the cells are empty.

In our case the lesion consisted of spindle- and stellate-shaped cells arranged in an irregular pattern. On the ultrastructural level the cells were equivalent to fibroblasts. Most likely because the fixation with formaldehyde proved to be suboptimal, electron microscopy did not detect myofilaments, although immunohistochemical staining for actin was positive in a few tumor cells. The cells were set in a myxoid matrix with a variable extent of collagenization. In the border zone to the dura, the septa of the collagen fibers interlinked with the myxoid portions of the tumor.

Although the majority of cranial fasciitis lesions develop outside the cranium, most of the proliferations show roentgenographic evidence of bone erosion. In some cases the tumor extends to the dura mater. In one case the lesion originated in the epidural space. In our patient the tumor arose from the dura mater and proliferated in the subdural space, displacing the underlying brain. Computerized tomography scans, as well as the intraoperative findings, revealed no infiltration but showed atrophy of the skull. To our knowledge, this is the first case in which cranial fasciitis developed in the subdural space.

Despite the benign nature of cranial fasciitis, the lesions grow rapidly. Neurological symptoms increased quickly in our patient. The tumor, however, must have been growing for a longer period because the brain showed atrophy due to displacement by the tumor. In contrast to the rapid growth and the malignant appearance, features that in the past frequently resulted in nodular fasciitis being diagnosed as malignant tumor, the prognosis of cranial fasciitis is very good: the vast majority of patients showed no recurrence at follow up. In our patient there was no evidence of recurrence 18 months after excision of the tumor.

**Differential Diagnosis**

Several differential diagnoses must be considered in the

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**Fig. 4.** Photomicrographs showing tumor specimen. H & E. **Left:** The tumor consists of spindle-shaped fibroblasts in a myxoid matrix. Note the thin-walled vessels. Original magnification × 180. **Right:** Higher magnification shows cytological details of the tumor cells. Original magnification × 540.

**Fig. 5.** Electron micrograph showing a tumor cell rich in rough endoplasmic reticulum. Original magnification × 5500. **Inset:** No desmosomes or interdigitations are seen. Original magnification × 28,000.

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**Table 1**

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<thead>
<tr>
<th>Antigen</th>
<th>Expression in Tumor Cells</th>
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<tbody>
<tr>
<td>vimentin</td>
<td>present</td>
</tr>
<tr>
<td>α-actin</td>
<td>present</td>
</tr>
<tr>
<td>MIB (Ki-67)</td>
<td>present</td>
</tr>
<tr>
<td>epithelial membrane antigen</td>
<td>absent</td>
</tr>
<tr>
<td>S-100 protein</td>
<td>absent</td>
</tr>
<tr>
<td>desmin</td>
<td>absent</td>
</tr>
<tr>
<td>carcinoembryonic antigen</td>
<td>absent</td>
</tr>
<tr>
<td>cytokeratin (KL-1)</td>
<td>absent</td>
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<tr>
<td>neuron-specific enolase</td>
<td>absent</td>
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present case. Because of the localization, meningiomas, particularly the myxoid variant, have to be ruled out. The histological appearance of our lesion was not characteristic of a meningioma. Fiber density was very low and the tissue was not pseudosyncytial. On the ultrastructural level, there was a lack of the typical structures such as cellular interdigitations or desmosomes usually present in meningiomas.\textsuperscript{2,18} Finally, with negative staining for epithelial membrane antigen, which is found in all meningiomas, a meningioma could be ruled out.\textsuperscript{16,20} Meningeal fibroma is histologically similar to the tumor presented here, but the distinguishing features of the fibroma include higher cellularity and expression of S-100 protein, which were not present in our case. Furthermore, we also considered juvenile fibromatosis.\textsuperscript{15} This lesion, however, differs from cranial fasciitis in its uniform proliferation pattern, compact appearance, and dense collagenization. Fibrosarcoma is characterized by higher cellularity, and the cells are directed in a more uniform manner. Moreover, both of the latter tumors are distinguishable from the lesion in our patient by their tendency to infiltrate the surrounding tissue.

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References


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