Malignant Fibrous Histiocytoma in the Skull
—Case Report—

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Abstract
A 45-year-old male presented with a large calvarial mass caused by a malignant fibrous histiocytoma arising primarily from the frontal bone and extending into the intracranial space. Magnetic resonance imaging revealed a well-demarcated, mottled enhanced bulky tumor with several low-signal separations. He underwent extensive tumor and bone removal, followed by radiation therapy. Histological examination showed pleomorphic spindle cells in a storiform pattern. The tissue stained positive for alpha-1-antitrypsin and alpha-1-antichymotrypsin by immunohistochemical techniques.

Key words: malignant fibrous histiocytoma, surgery, skull tumor

Introduction
Malignant fibrous histiocytoma is a pleomorphic tumor of the deep soft tissues occurring in the extremities of adults. This tumor is a neoplastic entity of histiocytic origin distinct from osteogenic sarcoma or fibrosarcoma, although it has been confused with other soft tissue sarcomas.

Malignant fibrous histiocytoma also originates from bone, although the incidence is relatively small. In particular, this neoplasm arising primarily within the skull is extremely rare. One series contained only seven of 177 cases with malignant fibrous histiocytoma of bone located in the skull. Recently, six well-documented cases of malignant fibrous histiocytoma arising primarily from the skull have been reported.6,8,9

Here, we report a case of malignant fibrous histiocytoma arising from the right frontal bone, and review the clinical presentation and management strategy of this rare tumor.

Case Report
A 45-year-old male first presented with scalp tenderness in July, 1989. Neurological examination found no abnormality. A plain skull x-ray film revealed a defect in the right frontal bone. He refused any further evaluation until July, 1990, when he presented with a 7-month history of a rapidly enlarging scalp mass in the right frontal area. He had become lethargic and disoriented, but without focal neurological deficits.

Plain skull x-ray films revealed a 6.5 x 7.0 cm defect in the right frontal bone (Fig. 1). Computed tomographic (CT) scans showed a large extracranial tumor involving the right frontal bone and extending into the intracranial space, with postcontrast inhomogeneous enhancement (Fig. 2). T1-weighted magnetic resonance (MR) images after administration of gadolinium-diethylenetriaminepenta-acetic

Fig. 1 Plain skull lateral x-ray film, showing a large defect in the right frontal bone.
acid (Gd-DTPA) showed a well-demarcated, mottled enhanced bulky tumor with several low-signal separations (Fig. 3). External carotid angiograms revealed a right frontal vascular stain (Fig. 4).

Right frontal craniotomy exposed the grayish-brown tumor which had invaded the adjacent temporal muscle, but the dura mater was intact. The tumor was growing out of the frontal bone, and had caused a calvarial defect with a slightly thickened edge. At least 1.5 cm of bone was removed from the periphery of the mass. The tumor was completely removed.

HE staining of surgical specimen revealed very highly cellular and pleomorphic tumor tissue. The tumor contained mainly a storiform pattern with spindle-shaped cells (Fig. 5), consisting of fibroblastic and histiocytic cells with bizarre, pleomorphic nuclei. The immunoperoxidase method showed intracytoplasmic granular staining for alpha-l-antichymotrypsin and alpha-l-antitrypsin (Fig. 6). The histological diagnosis was malignant fibrous histiocytoma.

He became alert and oriented postoperatively. He received a total of 50 Gy irradiation to the tumor bed, and has remained asymptomatic.

**Discussion**

Table 1 summarizes the seven reported cases of malignant fibrous histiocytoma in the skull. Patients' ages ranged from 2.5 to 72 years. There were five males and two females. Six patients presented with a rapidly enlarging mass on the skull, without associ-
ated pain. In two patients, including ours, the tumor was located in the frontal bone, two in the temporal bone, and one in the occipital bone. In one patient, the tumor developed in the frontotemporal bone 9 years after irradiation for a chromophobe adenoma. All patients demonstrated intracranial extension of the tumor in addition to the extracranial mass. In one case, a 2.5-year-old child presented with an osteolytic retropharyngeal mass arising from the clivus. Malignant fibrous histiocytoma arises from the cranial vault and the skull base, and extends into the intracranial space.

All the patients underwent gross total removal of the tumors. Adjuvant therapy included radiation therapy in three patients, and a combination of radiation therapy and chemotherapy in two. Our patient received extensive surgical removal including resection of the bone edge and temporal muscle, followed by radiation therapy. Three patients survived for 8–12 months after treatment, while our patient is still alive. One patient died from local recurrence 41 months after diagnosis of the primary tumor, and another died of multiple metastases to the mediastinum, thoracic vertebra, and lung. Malignant fibrous histiocytoma can metastasize in the course of the disease, most commonly to the lung, in contrast to fibrosarcoma or osteosarcoma.

Malignant fibrous histiocytoma is considered routinely in the differential diagnosis of a pleomorphic sarcoma. The histological diagnosis of this tumor may be difficult based on only light microscopy and/or electron microscopy as many of the features of these tumors are not entirely specific. Using immunohistochemical techniques, Boulay demonstrated positive staining for alpha-1-antitrypsin and alpha-1-antichymotrypsin in 23 malignant fibrous histiocytomas of the deep-seated soft tissue which was absent in fibrosarcoma, liposarcoma, or

Table 1 Clinical findings in seven cases of malignant fibrous histiocytoma in the skull

<table>
<thead>
<tr>
<th>Author (Year)</th>
<th>Age/Sex</th>
<th>Symptom</th>
<th>Location of lesion</th>
<th>Treatment</th>
<th>Recurrence/Metastasis</th>
<th>Outcome</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chitale et al. (1981)</td>
<td>18/M</td>
<td>mass of the temple</td>
<td>lt temporal bone</td>
<td>surgery, radiation, chemotherapy</td>
<td>yes/no</td>
<td>alive (1 yr)</td>
</tr>
<tr>
<td>Cook et al. (1987)</td>
<td>2.5/M</td>
<td>pain with neck movement</td>
<td>clivus</td>
<td>surgery, radiation, chemotherapy</td>
<td>yes/no</td>
<td>died (3.5 yrs of recurrence)</td>
</tr>
<tr>
<td>Dyck (1987)</td>
<td>41/M</td>
<td>enlarging scalp mass</td>
<td>bifrontal bone (pericranium)</td>
<td>surgery, radiation, chemotherapy</td>
<td>no/no</td>
<td>alive (8 mos)</td>
</tr>
<tr>
<td>Takeda et al. (1987)</td>
<td>59/F</td>
<td>lump in temple</td>
<td>lt temporal bone</td>
<td>surgery, chemotherapy</td>
<td>no/yes</td>
<td>died (1.5 yrs of metastasis) alive</td>
</tr>
<tr>
<td>Romero et al. (1989)</td>
<td>37/F</td>
<td>lump in temple</td>
<td>lt frontotemporal bone</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Matsuura et al. (1991)</td>
<td>72/M</td>
<td>diffuse swelling</td>
<td>lt occipital bone</td>
<td>surgery, chemotherapy</td>
<td>yes/no</td>
<td>died (6 mos of pneumonia) alive (2 yrs)</td>
</tr>
<tr>
<td>Present case</td>
<td>45/M</td>
<td>enlarging scalp mass</td>
<td>rt frontal bone</td>
<td>surgery, radiation</td>
<td>no/no</td>
<td></td>
</tr>
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</table>

Fig 6 Photomicrograph, showing positive staining for alpha-1-antitrypsin (upper, ×120) and alpha-1-antichymotrypsin (lower, ×240) by the immunoperoxidase technique.
spindle-cell squamous carcinoma. He suggested that these are useful specific markers for this tumor. In our case, light microscopy showed pleomorphic spindle cells in a storiform pattern. The positive reaction of antibodies to alpha-1-antitrypsin and alpha-1-antichymotrypsin provided further evidence of the histiocytic nature of the tumor cells. The presence of alpha-1-antitrypsin and alpha-1-antichymotrypsin seems to be a reliable indicator of malignant fibrous histiocytoma occurring in the skull in addition to the deep soft tissues.

References


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