Malignant Fibrous Histiocytoma of the Temporal Bone with Endocranial Extension

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Summary: Cranial malignant fibrous histiocytomas are rare tumors. Most are hypervascular, destructive masses that are similar to other malignant lesions and to malignant fibrous histiocytomas found elsewhere in the body. We describe a myxoid malignant fibrous histiocytoma of the temporal bone, possibly of dural origin, with features that more closely resembled a meningioma at CT, MR imaging, and angiography.

Index terms: Temporal bone, neoplasms; Meninges, neoplasms

Although malignant fibrous histiocytoma is the most common malignant tumor of the soft tissue in adults, it is extremely rare in the skull. Up to one quarter of malignant fibrous histiocytomas of bone involve previously abnormal bone. Malignant fibrous histiocytoma has been reported in areas of radiated or traumatized bone, in bone infarction, or in association with Paget disease and fibrous dysplasia (1). Most of these tumors that arise from the skull appear as transcranial destructive masses similar to other malignant neoplasms.

Case Report

A 19-year-old woman was admitted to our hospital because of a progressive headache and tenderness in the right temporooccipital region. She had no history of irradiation or trauma. Neurologic examination was unremarkable, and findings on a chest radiograph were normal.

A plain skull radiograph showed a poorly defined lytic defect, measuring 2.5 cm in diameter, in the right temporal area. Noncontrast computed tomography (CT) revealed a well-defined, hemispherical hypodense mass involving the mastoid and squama portion of the right temporal bone (Fig 1A). The lesion extended endocranially. The skull adjacent to the mass appeared somewhat thicker than the opposite side. The mass measured 3.2 × 2.5 cm and contained several small calcifications centrally and at its margin (Fig 1A). Inhomogeneous enhancement was noted after intravenous administration of an iodinated contrast medium (Fig 1B). The mass displaced the adjacent brain parenchyma inward, without midline shift. High-resolution CT with bone algorithm revealed bony destruction of the inner table (Fig 1C and D). The outer table of the involved mastoid and temporal squama was intact. At the anterior edge of the lesion, a periosteal reaction was seen (Fig 1D).

T1-weighted MR images obtained after intravenous administration of contrast material showed a well-defined large mass with marked enhancement and irregular, poorly enhanced areas. The adjacent dura mater and the right cerebellar tentorium showed focal, linear enhancement (Fig 1H).

At the beginning of the work-up of the patient, meningioma was a concern, since the mass was hemispherical with a broad base against the temporal squama, contained several areas of calcification, and enhanced after intravenous injection of contrast material. MR findings were compatible with meningioma, although the tumor’s intensity and degree of enhancement were not typical of that found with common meningiomas. Although meningioma was the initial impression, primary skull tumors such as metastasis, osteosarcoma, Ewing sarcoma, and chondrosarcoma were included in the differential diagnosis. The possibility of malignant fibrous histiocytoma was not considered before surgery. The patient’s age as well as the findings at physical examination and whole-body surveys, including gastrointestinal series, abdominal CT, gallium scan, and bone scintigraphy, made the possibility of a metastatic tumor less likely.
Fig 1. A 19-year-old woman with malignant fibrous histiocytoma.

A, Noncontrast CT scan shows a hemispherical, hypodense mass against the right temporal squama containing small areas of calcification. The skull adjacent to the tumor appears to be thicker than the opposite side.

B, CT scan obtained after administration of iodinated contrast medium shows heterogeneous enhancement.

C and D, High-resolution CT scan shows the tumor contains speckled calcifications; the bony destruction is evident in the right temporal squama and mastoid, sparing the outer table. At the anterior edge of the mass, a periosteal reaction (arrow, D) is noted.

E and F, On noncontrast T1-weighted axial MR images, the tumor is well-defined with mixed hypointensity.

G, On T2-weighted coronal MR image, the tumor is well defined with mixed hyperintensity and fewer areas of hypointensity.

H, On contrast-enhanced T1-weighted coronal MR image, the tumor shows heterogeneous enhancement, with suggestions of necrosis or myxomatous changes. Contrast enhancement is observed in the adjacent thickened dura matter and the cerebellar tentorium.

I, Right external carotid arteriogram shows a faint vascular stain, which is fed by the middle meningeal, ascending pharyngeal, and occipital arteries.

J and K, Photomicrographs of the specimen show the tumor is abundant in myxomatous changes. There are partially fibrous structures, in which a storiform pattern (arrows) is seen at low-power magnification (J). At higher magnification, the tumor cells are predominantly spindle shaped, with a few round or oval cells with well-defined, abundant cytoplasm (arrows), indicating histiocytic differentiation (K) (hematoxylin-eosin; original magnification ×25 [J] and ×80 [K]).
An open biopsy of the lesion disclosed malignant fibrous histiocytoma. Extensive, en bloc resection of the temporal region infiltrated by tumor was performed with tumor-free margins, because incomplete resection increases the possibility of local recurrence or distant metastasis. Resected structures included skin, auricle, parotid gland, temporomandibular joint, temporal bone, parietal bone, dura, and transverse sinuses. The facial nerve was repaired primarily with sural nerve grafting between the intracranial and extracranial portions. Intensive adjuvant chemotherapy with doxorubicin hydrochloride and ifosfamide followed.

Histologic analysis of a surgical specimen showed that the tumor consisted predominantly of spindle cells. There was also histiocytic differentiation. Tumor cells showed nuclear pleomorphism with abundant myxomatous changes (Fig 1J and K). A storiform pattern was seen in the fibrous component. No lipoblasts or osseous cells were present. The specimen stained positive for alcian-blue, indicating the existence of mucopolysaccharides. Immunohistochemical studies showed positive staining for CD-68, suggesting a histiocytic nature. The specimen stained negative for S-100 antigen and epithelial membrane antigen. These results excluded any unusual variant of meningioma. The pathologic diagnosis was a myxoid type of malignant fibrous histiocytoma.

At 18 months after surgery and chemotherapy, the patient was doing well except for right-sided facial paresis.

Discussion

Malignant fibrous histiocytoma was introduced as a tumor category approximately 25 years ago (2). These are pleomorphic lesions that consist of a predominantly bicellular population of atypical fibroblastic and histiocytic cells in variable proportions. Their histogenesis is unclear, but the mixed-cell population is thought to arise from a common precursor (ie, the primitive mesenchymal cell), which forms both the fibroblastic and histiocytic elements (3–5). Malignant fibrous histiocytoma of the bone is often reported to arise as a consequence of irradiation, bone infarction, trauma, Paget disease, and fibrous dysplasia (1). The prevalence of malignant fibrous histiocytoma involving bone is relatively low, and it is extremely rare in the skull (3). We found 15 cases of malignant fibrous histiocytoma involving the skull in the English-language literature (3, 5–12). In eight of these, patients underwent imaging studies in addition to plain skull radiographs (6, 7–13). These patients ranged in age from 3 months to 72 years (mean, 31 years) and included seven males and one female. The tumor was located in the frontal bone in two patients and in the temporal bone, parietal bone, frontotemporal bone, occipitotemporal bone, frontoparietosphenoidal bone, and clivus in one patient each. Six patients had CT, four had cerebral angiography, and one had MR imaging. CT scans showed a hypodense to heterogeneously hyperdense mass, with destruction of all layers of the involved skull, and inward and outward extension in all but one of the cases (6). None of the eight case reports included descriptions of calcifications in the periphery of the tumor. Five of these patients had contrast-enhanced CT, which showed heterogeneous enhancement with variably sized hypodense areas, probably representing either tumor necrosis or myxomatous changes. Cerebral angiography was performed in four patients, and in three of these the angiograms revealed prominent neovascularity and tumor staining. MR imaging was performed in one case, and heterogeneous enhancement was seen on contrast-enhanced T1-weighted images.

In our patient, some of the imaging findings suggested meningioma, whereas other findings were atypical. Malignant bone tumors originating from the skull, such as metastasis, osteosarcoma, Ewing sarcoma, and chondrosarcoma, were included in the differential diagnosis. Metastasis was less likely, although it was not excluded by normal findings at chest radiography and whole-body surveys. Osteosarcoma and Ewing sarcoma were unlikely, since the skull changes were less aggressive. Chondrosarcoma could not be ruled out, but the patient’s age made that possibility less likely. We believe that this tumor developed from the inner table or diploic space of the skull, but it may have originated from the dura mater.

MR findings of malignant fibrous histiocytoma in other areas of the body have been reported as nonspecific (14). In general, malignant fibrous histiocytoma appears as a hypointense lesion on T1-weighted images, and as a mass of heterogeneous signal intensity on T2-weighted images, except in cases of intratumoral bleeding (14). In cases of abundant fibrous tissue, the lesion appears hypointense on T2-weighted images; alternatively, it is hyperintense on T2-weighted images in cases in which the tumor contains mucinous components (14).

The myxoid type of malignant fibrous histiocytoma is thought to be a variant, featuring less cellularity and a rich matrix; it also tends to be
associated with a better prognosis than its more cellular counterparts (15). The rate of metastases of myxoid types decreases as their myxoid components increase. The significance of the myxomatous changes in these tumors is not clear, but it seems unlikely that this represents merely a degenerative change, as other degenerative changes are lacking. It is regarded as an area in which the tumor cells multiply more slowly but produce an abundant mucoid matrix as a form of differentiation. This may explain the better prognosis (3, 15).

Treatment of malignant fibrous histiocytoma includes surgical removal followed by adjuvant chemotherapy and/or radiation. In a significant number of patients, local recurrence or metastasis develops within 2 years. The mean 5-year survival rate for patients with malignant fibrous histiocytoma originating from bone is 34% to 50% (3, 12).

In conclusion, malignant fibrous histiocytomas rarely arise from the skull. In general, they should be included in the differential diagnosis of any large, aggressive transcranial mass.

References