Angiographic features of gliosarcoma.

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Angiographic Features of Gliosarcoma

Gliosarcoma is a brain neoplasm that is being recognized with increasing frequency. We discuss the radiographic findings in 14 pathologically proven cases. At angiography in nine cases, four showed mixed dural and pial vascular supply to the lesion. Early cortical venous drainage, irregular tumor vessels, and a prominent vascular stain with well-defined tumor margins were seen in the majority of cases. CT showed an irregular enhancing rim surrounding a necrotic center in 14 cases. Most lesions were peripherally located and invaded dura.

MATERIALS AND METHODS

Fourteen patients (eight women and six men ranging in age from 45–75 years, mean age 59 years) with histologically proven gliosarcoma were seen at Henry Ford Hospital between 1977 and 1986. All pathologic material was rereviewed and histologically verified for purposes of this study. Common short-term symptoms included headache, personality changes, confusion, weakness, and seizures. Two patients had undergone long-term treatment for seizures. In none were extraneural metastases found.

Nine patients underwent preoperative angiography. Each tumor was evaluated for the type of vascular supply, morphology and distribution of tumor vessels, appearance and duration of tumor stain, definition of tumor margin, and pattern of venous drainage (Table 1). All studies were subtracted prior to analysis.

CT scans were performed on all 14 patients, with and without contrast, using an EMI Mark I, a Picker Synerview 1200, or a GE 9800. CT scans were evaluated for tumor location, size, and enhancement.

Histologic criteria for diagnosis were as follows: both gliomatous and sarcomatous elements were present; the glial component was positive for glial fibrillary acid protein stain; and the sarcomatous portion was positive with stains for reticulin and fibrous connective tissue [4].

Results

CT

Fifteen tumors were seen in 14 patients. One patient had both a left thalamic lesion and a lesion in the left frontal horn due to subependymal spread. All were supratentorial with a mean size of 4.1 cm (range, 2.5–7 cm). Eleven tumors were situated in the cortical and subcortical area residing predominately in the temporal
### TABLE 1: Angiographic Findings of Gliosarcomas

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Location</th>
<th>Tumor Vessels</th>
<th>Tumor Stain</th>
<th>Definition of Tumor Drainage</th>
<th>Early-Draining Veins</th>
<th>Angiographic Category</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td>Vessel Morphology</td>
<td>Distribution</td>
<td>Time of Appearance</td>
<td>Duration</td>
<td>Source</td>
</tr>
<tr>
<td>1</td>
<td>Temporal</td>
<td>None</td>
<td>Late arterial</td>
<td>Mid-venous</td>
<td>Pial</td>
<td>Faint</td>
</tr>
<tr>
<td>2</td>
<td>Frontoparietal</td>
<td>None</td>
<td>Late arterial</td>
<td>Late venous</td>
<td>Mixed</td>
<td>Faint</td>
</tr>
<tr>
<td>3</td>
<td>Temporal</td>
<td>None</td>
<td>Late arterial</td>
<td>Early venous</td>
<td>Pial</td>
<td>Prominent nonhomogeneous</td>
</tr>
<tr>
<td>4</td>
<td>Frontal</td>
<td>Irregular</td>
<td>Late arterial</td>
<td>Mid-venous</td>
<td>Predominately dural</td>
<td>Prominent nonhomogeneous</td>
</tr>
<tr>
<td>5</td>
<td>Frontotemporal</td>
<td>Irregular</td>
<td>Early arterial</td>
<td>Mid-venous</td>
<td>Pial</td>
<td>Prominent ring</td>
</tr>
<tr>
<td>6</td>
<td>Frontal-corpus callosum</td>
<td>Irregular</td>
<td>Irregular</td>
<td>Early arterial</td>
<td>Early venous</td>
<td>Predominately pial</td>
</tr>
<tr>
<td>7</td>
<td>Thalamus &amp; lenticular nuclei</td>
<td>Irregular</td>
<td>Ring</td>
<td>Early arterial</td>
<td>Early venous</td>
<td>Pial</td>
</tr>
<tr>
<td>8</td>
<td>Occipital</td>
<td>Irregular</td>
<td>Late arterial</td>
<td>Early venous</td>
<td>Mixed</td>
<td>Prominent part of ring</td>
</tr>
<tr>
<td>9</td>
<td>Temporal</td>
<td>Irregular</td>
<td>Early arterial</td>
<td>Early venous</td>
<td>Pial</td>
<td>Prominent ring</td>
</tr>
<tr>
<td>Summary</td>
<td></td>
<td>Irregular-6</td>
<td>Early-4</td>
<td>Early-5</td>
<td>Pure pial-4</td>
<td>Prominent-6</td>
</tr>
<tr>
<td></td>
<td></td>
<td>None-3</td>
<td>Late-4</td>
<td>Mid-2</td>
<td>Pure dural-0</td>
<td>Faint-2</td>
</tr>
<tr>
<td></td>
<td></td>
<td>Sunburst-1</td>
<td>Late-1</td>
<td>Late-1</td>
<td>Mixed-4</td>
<td>Avascular-1</td>
</tr>
<tr>
<td></td>
<td></td>
<td>None-3</td>
<td>None-1</td>
<td>None-1</td>
<td>None-1</td>
<td></td>
</tr>
</tbody>
</table>
lobe in six patients, frontal lobe in three, parietal lobe in one, and occipital lobe in one. At surgery, nine of these had invaded dura, but none had invaded bone. Four tumors were deeply located, two in the thalamus, one subependymally in the frontal horn, and one in the medial right frontal lobe invading the corpus colosum.

After contrast infusion, 14 tumors had intense, thick, irregular ring enhancement surrounding a central hypodensity (Fig. 1). Only the subependymal lesion enhanced homogeneously. Tumor calcification was never present.

Angiography

Angiographic results are found in Table 1. Six of nine tumors were vascular and three were avascular or hypovascular. In the six vascular lesions, tumor vessel walls showed areas of constriction and dilation, and the vessels were distributed in an erratic or ringlike fashion within the tumor. A prominent ringlike or nonhomogeneous stain with a well-defined tumor margin was present. Tumor stain appeared in the early arterial phase but disappeared during the midvenous phase. Early-draining veins were always evident and in all but one case drained superficially (Fig. 2).

The remaining three tumors were either avascular or hypovascular; no tumor vessels or early-draining veins were present. Tumor stain was faint (two cases) or nonexistent (one case).

Three of four tumors with pure pial supply and three of four with mixed dural and pial supply were vascular. Thus, the presence of mixed supply did not predispose to formation of an angiographically vascular lesion. Of the four tumors with mixed dural and pial supply, the supply was predominantly dural in one, predominantly pial in one, and balanced in two (Fig. 3). No difference in the stain appearance is seen when pure pial versus mixed supply is present.

Discussion

The gliosarcoma is usually gray, very firm, lobulated, and contains areas of necrosis (Figs. 4A–4F). The sarcomatous portion is sharply demarcated and often separable from the adjacent brain while the astrocytic component is poorly defined and soft. The microscopic appearance is that of an anaplastic sarcoma, usually of the fibroblastic type, with an interlacing fascicular pattern abruptly alternating or separate from a typical grade 3–4 astrocytoma (glioblastoma multiforme) (Fig. 4G). Within both areas, usually one or more blood vessels show marked neoplastic proliferation of their endothelial cells, from which the sarcomatous component is thought to have arisen (Fig. 4H) [4].

The term gliosarcoma was first used in 1895 by Stroebe [1] to describe a primary brain tumor containing both neoplastic glial and sarcomatous elements. For approximately 30 years after this initial definition, the term gliosarcoma was used erroneously to describe "anaplastic astrocytomas" with associated hyperplasia of vascular endothelial elements, thereby losing its original definition [2]. Endothelial hypertrophy and hyperplasia are seen with most anaplastic astrocytomas and become more marked the higher the grade of astrocytoma [3, 5, 6]. The term gliosarcoma should be reserved for astrocytomas grade 3–4, in which the vascular endothelial changes have progressed beyond hypertrophy.
and hyperplasia to assume the appearance of a spindle-celled fibrosarcoma [3, 5, 6]. The most widely accepted theory as to the genesis of these tumors is attributable to Feigin [7], who believed the sarcoma arises from neoplastic transformation of the vascular elements within a glioblastoma [2, 5, 7].

The term sarcoglioma has also been a source of confusion in the past. This term should be reserved to describe a separate conceptual entity that is essentially the mirror image of gliosarcoma. In sarcoglioma, the sarcoma is believed to be the initial tumor that induced neoplastic transformation of the glial cells at its margin [8].

We found no extraneural metastases in the 14 patients in our series. Extraneural metastases from gliosarcomas are, however, reported to be significantly more common than from glioblastoma multiform. The incidence of extraneural metastases has been reported as 30% by Smith et al. [5] and 15.4% by Cerame et al. [6]. This propensity to metastasize reflects both a different pathogenesis and a different biologic behavior than the usual malignant glial neoplasm. Metastatic dissemination occurs hematogenously to visceral organs. Patient survival time for both gliosarcoma and glioblastoma multiform is approximately 12 to 18 months [6]. The additional burden of metastatic disease does not affect the prognosis in patients with gliosarcoma because the primary cause of death is the intracerebral neoplasm [2]. Nevertheless, with continued growth the sarcomatous component usually outgrows the glial component [3].

We found a temporal lobe preference, as did Morantz et al. [2] in their series of 24 patients [2]. The marked tendency toward peripheral location and dural invasion described by others is also evident in this series [2, 5, 6]. There is no apparent cause-and-effect relationship between dural invasion and formation of a gliosarcoma from a glioblastoma. The sarcomatous component arises from vascular elements, not from neoplastic transformation of the dura that has been invaded. The tendency toward dural invasion appears to be due solely to proximity of these central tumors to the dura. Why a glioblastoma located peripherally, particularly in the temporal lobe, would be more prone to the development of a gliosarcoma is unclear.

The only published report describing the CT appearance of gliosarcoma is by Lee et al. [3]. In that report of five cases, the tumor is described as being homogeneously enhancing. In contrast, but one of the tumors in this series had an intensely enhancing irregular rim with a low-density (necrotic) center. Unlike Lee et al. we can only conclude that CT alone is not helpful in distinguishing glioblastoma multiform from gliosarcoma [3].

To our knowledge, the angiographic findings of gliosarcoma have not been previously described in the radiologic literature. Morantz et al. [2] noted a tumor stain and early-draining veins but gave no further details. Not surprisingly, angiographic manifestations of gliosarcoma are similar to those of glioblastoma multiform as they are closely related histologically [9]. Irregular tumor vessels, a transient nonhomogeneous stain, and early-drainage veins are features commonly seen with both tumors. Also, a minority of both tumors are avascular at angiography [10]. However, we commonly found several features in angiographically vascular gliosarcomas that are rare in glioblastoma. (1) In 80% of gliosarcomas venous drainage is peripheral, while glioblastomas typically drain into the deep venous system via medullary veins [10, 11]. (2) A mixed dural and pial vascular supply is present in nearly half of gliosarcomas, and is rare in glioblastoma [10, 11]. Both these findings can be explained by the tendency of gliosarcomas toward peripheral location while glioblastomas typically are located in white matter. (3) The vascular stain of a gliosarcoma is more prone to form a well-defined tumor margin (particularly with a ringlike configuration) because the sarcomatous component of the tumor forms a capsule that is tough and well defined. For this reason, these tumors are often mistaken for...
meningiomas at surgery. In contrast, the capsule of glioblastomas is routinely soft, friable, or nonexistent on gross inspection [3, 6, 9].

Little difficulty should be encountered in differentiating most meningiomas (the most common dural-based lesion) from a gliosarcoma. The typical benign meningioma will have a purely dural supply, no irregular tumor vessels, a homogeneous stain that persists through the venous phase, and no early-draining veins [10]. No gliosarcoma in our series had these findings.

Angiographically, difficulty may arise in differentiating a high-grade gliosarcoma with mixed but predominantly dural vascular supply from an aggressive or malignant meningioma. All are dural based, have mixed vascular supply, irregular tumor vessels, and early-draining veins [10]. A single case in our series illustrates this problem (Fig. 5). In fact, this particular patient was initially thought to have an angioblastic meningioma, and the dural supply was embolized preoperatively. A unique feature seen with this particular tumor, however, is a...
prominent capsular vein. This is not a classic feature of meningioma and could prove to be a helpful differential finding [10]. In addition, deep venous drainage has been described as a sign of an aggressive meningioma [12]. This feature was not seen in any dural-based gliosarcoma. Differentiation of an aggressive/malignant meningioma from a gliosarcoma should be possible by CT criteria. "Mushrooming" (the presence of a prominent pannus of tumor extending well away from the glial mass), described by New et al. [13] in malignant meningiomas, was not seen in any of our cases, nor were lytic changes in the calvarium [12].

Differentiation from other, less common, dural-based lesions should not be difficult. In contrast to our findings in gliosarcoma, in hemangiopericytoma, early venous drainage is reportedly rare and persistent dense vascular stain common [14]. Neurofibromas are typically avascular or hypovascular at angiography and do not present as thick ring-shaped lesions on CT [10]. Hemangioblastomas can invade dura, particularly when recurrent. However, none of our lesions presented infratentorially and none displayed a homogeneously enhancing nodule at angiography, which is characteristic of hemangioblastoma [10]. Olfactory neuroblastoma as well as recurrent ependymoma and medulloblastoma can present as vascular dural-based lesions; however, differentiation should again be possible on the basis of location [15].

We conclude that when a peripheral or dural-based lesion with a CT appearance of glioblastoma (thick ring) combined with superficial venous drainage, mixed dural-pial vascular supply, and a well-defined capsular margin at angiography is evident, the differential diagnosis should include gliosarcoma.

REFERENCES

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