Predominantly Extraaxial Astroblastoma: Imaging and Proton MR Spectroscopy Features

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Summary: The CT, angiographic, MR, and proton MR spectroscopy findings in a case of astroblastoma, a rare neoplasm of glial cell origin, are presented. Of particular interest is the predominantly extraaxial location of the tumor. CT and MR demonstrated a complex mass consisting of a solid nodule and a peripheral septated cystic component. The extraaxial nature of the mass was suggested on MR.

Index terms: Brain neoplasms, computed tomography; Brain neoplasms, magnetic resonance; Brain neoplasms, angiography; Magnetic resonance, spectroscopy

Astroblastoma is a well known but uncommon tumor of glial origin (1). Although astroblastic elements are occasionally identified in specimens of glioblastoma and astrocytoma, particularly those of the gemistocytic variety, pure astroblastomas are rare (1–3). A few series and case reports (1–7) have appeared in the neurosurgery, pathology, and oncology literature. This report describes the imaging features of a rare presentation of this uncommon tumor.

Case Report

A 20-year-old woman presented with a 1-month history of sharp left-sided headaches severe enough to wake her from sleep. Her only other complaint was difficulty reading, primarily an interpretive problem rather than a visual one; she stated that it took her an hour to read and comprehend a single written page. Physical examination revealed a homonymous right superior quadrantanopsia. Her neurologic examination was otherwise normal.

Precontrast and postcontrast computed tomographic (CT) scans of the brain (Fig. 1) revealed a 7 × 5-cm complex left medial occipital mass. Its superomedial component, adjacent to the falx and tentorium, consisted of a 3-cm enhancing solid nodule, which was sharply demarcated from the anterior, lateral, and posterior cystic component of cerebrospinal fluid density. There was enhancement of the cyst wall and of septations radiating from the solid nodule to the cyst wall. There was no visible calcification or surrounding vasogenic edema.

Left vertebral angiography (Fig. 2A and B) demonstrated a 3-cm tumor stain with neovascularity and arteriovenous shunting in the left occipital region. The blood supply was of pial origin and derived from the posterior and middle temporal branches of the left posterior cerebral artery, which were draped around a large, avascular component of the mass.

Magnetic resonance (MR) (Fig. 3A–C) indicated a probable extraaxial mass that was in contact with the superior aspect of the tentorium. The solid portion of the mass was hypointense to gray matter on T1-weighted spin-echo and inversion recovery sequences and on proton density-weighted spin-echo images and became hyperintense on T2-weighted spin-echo images. Tumor vessels produced punctate signal voids. The signal intensity of the cystic component paralleled the cerebrospinal fluid on all pulse sequences.

One-dimensional proton (1H) chemical shift imaging (CSI) was performed in the region of the tumor (Fig. 4A and B). The spectra displayed an increased choline (Cho) to-phosphocreatine/creatinine (PCr/Cr) ratio, decreased N-acetyl aspartate (NAA), increased lactate/lipid, and increased myo-inositol (Ins) in those voxels containing primarily solid tumor, as compared with those containing primarily cyst and normal brain.

Gross total resection of the mass was performed. At surgery, the tumor appeared extraparenchymal and was easily separated from the underlying brain. The cyst wall was hypervascular, and the peripherally displaced vessels seen on angiography were embedded in the cyst wall. The solid component of the tumor was adherent to the tentorium.

The frozen section diagnosis was “extraparenchymous neoplasm, probably glial,” although atypical meningioma and hemangiopericytoma were also considered. The final pathologic diagnosis, however, was malignant astroblastoma (Fig. 5). This was confirmed by positive immunoperoxidase staining of the neoplastic cells for glial fibrillary acidic protein, as well as by electron microscopy. Continuity between the solid nodule and the underlying cortex was demonstrated on only one slide, on which the interface
Fig. 1. Contrast-enhanced CT scan demonstrates a mixed cystic and solid left-occipital-region mass. The extraaxial location of the mass is not readily appreciated. Note the enhancement of both the solid tumor nodule and the cyst wall and radiating septations. Also note the normal calcified pineal gland (the pineal gland is known to be a site of origin of astroblastomas).

between tumor and parenchyma was sharp, and no dural infiltration was seen. Tumor cells were present in the cyst wall and septations as well as in the solid nodule.

Discussion

Astroblastoma is among the rarest of central nervous system neoplasms (7), representing 0.45% of gliomas (8). Usually a tumor of children, adolescents, and young adults (1,2), it has been reported in a 67-year-old patient (3). There is no sex predilection (2). Patients present with non-specific neurologic symptoms (1, 3). Slow growth of the tumor often produces a long history of symptoms prior to presentation, usually 1 to 6 months, but occasionally 2 to 4 years (1).

The tumor is most often supratentorial and hemispheric (1), although examples have been reported in the cerebellum, brain stem, fourth ventricle, hypothalamus, and pineal region (1, 6, 9). They are often located in the superficial cortical and subcortical tissues (1, 2), and extensions into the subarachnoid space (5), leptomeninges (1, 2, 6), and dura (1) have been described. A predominantly extraparenchymal location appears to be unusual. Microscopic cortical involvement, as demonstrated in the case presented here, precludes the diagnosis of a primary extraparenchymal glial tumor (2). Although most astroblastomas are well circumscribed (1, 2), small areas of extension into adjacent neural parenchyma can be found (1, 7), and poorly defined and infiltrating tumors have been reported (4). Lobulated solid tumors measuring from a few millimeters to 8 cm are most common (1), but small and large cysts are frequent. One of 23 cases reported by Bonnin and Rubinstein had extensive calcification (1).

Microscopically, the tumor consists of characteristic perivascular tumor cells which, when seen in cross-section, produce a typical pseudorosette pattern reminiscent of ependymoma (Fig. 5) and, when sectioned longitudinally, demonstrate a papillary or pseudopapillary arrangement (1-7). Intervascular cells are indistinguishable from the perivascular cells but are rarified, as opposed to the more compact arrangement of the intervas-
Fig. 3. A, T1-weighted spin-echo sagittal image (450/19 TR/TE), B, T1-weighted inversion recovery axial image (1500/20, inversion time = 600) and, C, T2-weighted spin-echo axial image (2500/50) demonstrate solid and cystic components of the tumor. Note the relationship of the mass to the tentorium in (A), suggesting that the tumor is largely extraaxial. Radiating septa are seen extending from the solid central component to the cyst wall (B and C).

papillary cells in ependymoma (2), the tumor with which astroblastoma is most often confused histologically (2,5-7). The tumor cells typically stain positively for glial fibrillary acidic protein, which is indicative of a tumor of glial origin (2,6,7,10). Papillary meningioma and metastatic papillary carcinoma can produce a similar histologic picture (7). Necrosis is present microscopically in 70% of astroblastomas, regardless of tumor grade (1).

The natural history of astroblastoma lies somewhere between that of astrocytoma and glioblastoma (2,3). Two distinct histologic subtypes occur (1,2). Low-grade astroblastomas are associated with a favorable postoperative prognosis, with survival up to 20 years; high-grade astroblastomas are associated with a worse prognosis, characterized by one or more recurrences, often with increasing degrees of anaplasia as well as conversion to glioblastoma (1,2).

Ultrastructural similarities have suggested to some that the astroblastoma is derived from the tanycyte, a cell that is intrinsic to the ependyma of submammalian vertebrates, but that is also found among the ependymal cells of embryonic and neonatal higher mammals, including humans (2,10). The cells are distinguished from typical ependymocytes by long processes that extend to the pial surface of the brain (11). The persistence of tanycytes into adulthood has been demonstrated in the rodent brain (12); in humans and other higher mammals, the tanycyte appears to
Fig. 4. A, voxels used for $[\text{H}]$CSI. A $2 \times 2 \times 8$-cm voxel was selected with a water-suppressed stimulated-echo acquisition mode sequence (2000/20, TM = 11). Phase encoding in the anteroposterior direction yielded seven $2 \times 2 \times 1$-cm subvoxels labeled 10 through 16. The image corresponds to a 3-mm section through the middle of the stimulated-echo acquisition mode voxel and delineates regions occupied by cyst, solid tumor, and normal brain. Adjacent contiguous slices contain variable amounts of these materials and were used to estimate their relative amounts in each voxel.

B, $[\text{H}]$CSI spectra at locations corresponding to subvoxels in (A). Voxels 10, 11, and 15 contain primarily cyst, solid tumor, and normal brain, respectively. Voxels 12, 13, and 14 contain mixtures of all three components with decreasing relative amounts of tumor and increasing amounts of normal brain. Spectral assignments are Ins at 3.5 ppm, Cho at 3.2 ppm, PCR/Cr at 3.0 ppm, NAA at 2.25 and 2.0 ppm, and lactate/lipid (Lac/Lip) at 1.3 ppm. Elevated Cho to PCR/Cr (voxels 11 and 12), decreased NAA (voxel 11), and elevated Ins (voxels 11 and 12) are seen in voxels containing primarily tumor as compared with normal brain (voxels 15 and 16). Note the lactate doublet at 1.33 ppm resolved in voxels 13 and 14 but not in voxels 10 through 12, presumably because of the presence of mobile lipids (e.g., triglycerides with resonances at 0.9, 1.3, and 2.1 ppm appear significantly elevated in these voxels). Interpretation of the NAA resonance at 2.0 ppm is complicated by the presence of some partial volume contamination and an overlapping resonance due to triglyceride at 2.1 ppm.

Fig. 5. Histologic section (hematoxylin and eosin stain, ×620) demonstrates typical perivascular pseudorosettes, reminiscent of an ependymoma. Note the loose stroma between the perivascular rosettes, which is distinct from the typical compact stroma associated with ependymoma, cystic meningioma, and metastatic papillary carcinoma (photomicrograph courtesy of KL Ho, MD, Detroit, MI).

be only a transitory phase (10), although its fate remains undetermined.

Previous reports of the CT appearance of astroblastomas describe a nonspecific solid, isodense mass with marked homogeneous or heterogeneous enhancement (1,3,6). The large cystic area present in our case, although not described on previous CT reports, is consistent with the frequent pathologic demonstration of cystic areas. Previous angiographic descriptions range from an avascular mass to a round tumor stain with moderate vascularity (3–5).

The MR appearance of our case is not entirely unexpected on the basis of the CT and angiographic findings. The hypointensity on T1-weighted image and the hyperintensity on T2-weighted image are typical for glial tumors; the appearance of the cystic portion of the tumor, as expected, parallels the cerebrospinal fluid. MR demonstrated the septations more clearly than
the CT scan. MR also more clearly demonstrated the continuity of the mass with the tentorium, particularly in the sagittal images.

The spectroscopic results observed in this case are consistent with those observed for other central nervous system parenchymal neoplasms and are similar to those of astrocytoma (12-14). Decreased NAA suggests decreased neuronal tissue or displacement of the neuronal tissue by the tumor (12-14). An elevated level of Cho, seen in all central nervous system tumors, is felt to be associated with increased cell membrane turnover during cell proliferation (13,14). The lactate doublet at 1.33 ppm suggests active glycolysis (12-14). The significance of the elevated Ins is not known (15).

Kugel et al. (13) recently described an absent NAA signal on [1H]MR spectroscopy of meningiomas, distinguishing these tumors from gliomas. NAA was apparently decreased but not absent in the case presented here. In addition, Peeling and Sutherland (16) have reported the virtual absence of Ins in the proton spectra of meningioma tissue extracts, inconsistent with the elevated level of inositol seen in this case.

The CT scan suggested an intraaxial lesion, probably a high-grade glioma; MR showed continuity with the tentorium and suggested an extraaxial mass such as a cystic meningioma. The pial blood supply, demonstrated angiographically, would be unusual for a tumor of meningeal origin, thus favoring a glial tumor.

Current therapy for astroblastoma consists of surgical resection and postoperative radiation. Chemotherapy has not been shown to affect prognosis significantly (1). In conclusion, we present a case of an astroblastoma with a large extraparenchymal component, a feature clearly demonstrated only with MR. The pial blood supply, demonstrated angiographically, favored a preoperative diagnosis of a glial tumor as opposed to a cystic meningioma, which would usually have a predominantly dural blood supply. Although rare, astroblastoma should be considered in the differential diagnosis of a well-demarcated solid or complex intracranial mass in a child or young adult.

References