Supratentorial ependymomas

Neuroimaging and clinicopathological correlation

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In 10 patients with supratentorial ependymomas, the tumors exhibited hyperdensity on computerized tomography (CT) scanning prior to contrast infusion and, with one exception, all tumors were mixed lesions with the low densities suggesting cystic or necrotic portions. Eighty percent of the tumors contained small calcifications. Characteristically, the tumors were well demarcated and demonstrated moderate to marked enhancement after the intravenous administration of contrast material. Angiograms obtained in some patients showed mild hypervascular tumor staining and absence of large feeding arteries. The degree of contrast enhancement, angiographic vascularity, and tumor stain was compared to the pathological anaplasia of the tumors. No correlation was observed. Of four patients who were still alive during a follow-up period of 4 years or longer, three had recurrences with inoperable tumors; the remaining patient is without recurrence after craniospinal radiation. This same patient belonged to a group of five patients with a diagnosis of high-grade ependymoma, four of whom had recurrence. Follow-up CT accurately recorded the clinical course of each patient. Annual routine follow-up examinations are proposed for patients with low-grade ependymomas, and for those with high-grade ependymomas follow-up CT should be performed every 6 months. The characteristic appearance and behavior of these tumors include several distinctive features on angiographic and CT images.

Keywords: ependymoma • supratentorial tumor • brain tumor • computerized tomography

Although the computerized tomography (CT) appearance of intracranial ependymomas has been described previously,21 supratentorial ependymomas have not been emphasized as a group in the recent literature. In spite of their variable clinical course of remission and recurrence, supratentorial ependymomas appear uniform in their neuroimaging and pathological characteristics; because of this, we undertook a study of the ependymomas treated over several years in our two institutions. We present those cases that were examined predominantly with fourth-generation CT scanners and with selective angiography. The results of our study characterize ependymomas sufficiently apart from other tumors in the supratentorial compartment.

The initial surgical findings, and subsequent operations in some patients, confirmed many of the features of ependymomas which we wish to discuss. Pathological presentations, particularly the microscopic findings, are included in each case to allow comparison with the neuroimaging data. The clinical course and treatment for each patient were evaluated to identify significant aspects that may be useful in the management of patients with supratentorial ependymomas.

Clinical Material and Methods

Eleven cases of supratentorial ependymoma, which were encountered between 1977 and 1984 at the medical centers of the University of California at Los Angeles (UCLA) and San Diego (UCSD), comprise this report. The data for two of these patients (Cases 7 and 8) are not complete. The age range of these patients was from 1½ to 43 years, and the mean age of this group was 18.4 years (excluding a patient who originally had an ependymoma of the posterior fossa). Clinical data, including manner of presentation, operative findings, and postoperative treatment and course, were obtained by review of patient charts. Radiological evaluation of
Table 1 summarizes the neuroradiological features in 10 of our 11 patients. Case 8 is omitted because the radiological data were not obtained. All 10 patients presented hyperdense lesions on nonenhanced CT. All lesions were of mixed attenuation except the cystic lesion in Case 2, which contained hyperdense fluid. Eighty percent of patients had calcifications. Contrast enhancement was always present and usually moderate to marked in intensity.

Angiography in five of our patients revealed strikingly similar findings of mild hypervascularity, with moderate tumor stain increasing in density on the serial films and persisting into the venous phase. The stain was nonhomogeneous, correlating with the low-density areas seen on the CT scans. Large feeding arteries were not observed. Only one patient (Case 4) had large draining veins, which probably reflected increased flow through the tumor.

Table 1 also indicates the histopathological diagnosis of the tumors in each patient. In studies of supratentorial astrocytomas, Butler, et al., found that the degree of contrast enhancement correlated with the degree of abnormal microscopic vascularity and considered this feature to be useful in assessing the malignancy of the lesion. We found no correlation between the degree of contrast enhancement, angiographic vascularity, or staining and the degree of anaplasia of the tumors.

Discussion

The ependyma, a layer of inactive cells that retains its epithelial character, provides lining for various parts of the nervous system such as the ventricles, choroid plexus, central canal of the spinal cord, and filum terminale. Ependymal cells are embryological derivatives of the primitive neuroectodermal cells, and give origin to more mature cells that radially migrate toward the cortex from the periventricular regions. Ependymal cells have also been found in portions of the brain not related to the ventricular lining (cell rests). In these locations, they tend to form rosettes or clusters of cells: a common finding not only among ependymomas but also in normal brains. The normal anatomic location of ependymal cells and the occurrence of cell rests in the parenchyma with varying distances from the ventricular lining are believed to explain the different sites of origin of ependymomas as intraventricular, extraventricular, or both.
Ependymomas are relatively uncommon central nervous system tumors. In a recent series of 26 cases, they occurred both above and below the tentorium, and in infants and children were found most frequently in the posterior fossa. They comprise a small percentage of tumors in the spinal canal, although they have been reported to be the most common form of intramedullary glioma if filum terminale ependymomas are included. In children and adults, ependymomas represent 6% of all intracranial gliomas. In the pediatric age group

![Image of preoperative studies](link_to_image)

**Fig. 1.** Case 6. Preoperative studies in a 1½-year-old baby girl who was noted to have emesis after each meal 2 weeks prior to admission. At surgery, a large biventricular mass containing some cysts and areas of necrosis was found. The capsule of the tumor was noted to be quite vascular. *Left* and *Center:* Plain *(left)* and contrast-enhanced *(center)* computerized tomography scans of an intraventricular mass exhibiting hyperdensity with mixed components and suggestive fine calcification. *Right:* Subtraction image of the right carotid injection, capillary phase, showing a massive intraventricular tumor stain *(arrowheads)*.
FIG. 2. Case 3. This 42-year-old man had a 6-month history of focal seizures and headaches. At craniotomy, a discrete tumor was found which appeared to be invading brain in some areas. Upper: Preoperative studies. Upper Left: Initial plain computerized tomography (CT) scan showing a hyperdense rim of the cystic and solid portions with slightly coarse calcification. Upper Center: Contrast-enhanced CT scan showing a thick irregular cyst wall. A moderate degree of edema surrounds the large mass. Upper Right: Angiogram showing a tumor stain (arrowheads) at the arterial phase. Lower: Photomicrograph showing several calcification deposits (arrows) within the tumor. The histopathological diagnosis was malignant ependymoma. H & E, × 25.

The observed characteristics of supratentorial astrocytomas from grades I to IV vary among different reports. Utilizing an EMI Mark I head scanner, Weisberg\(^2\) reported that 92% of grade I supratentorial astrocytomas were hypodense prior to contrast injection and 8% showed calcification. Hypodensity was observed in 52% and 12% of grade II and III supratentorial astrocytomas, respectively. Glioblastomas (grade IV astrocytomas) presented with decreased density in 38%, but were of mixed density (hypo- or hyperdensity) in 62%; increased densities were sometimes due to hemorrhage (7%) or calcification (4%), but the majority (51%) were considered neither hemorrhagic nor calcific. Although the finding of a mixed but predominantly increased density bears some similarity to our series of ependymomas, Weisberg's finding of calcification in glioblastomas is uncommon. Furthermore, glioblastomas and lower-grade astrocytomas usually do not appear as well demarcated as ependymomas on nonenhanced CT scans. Lastly, we believe that the increased density of ependymomas on nonenhanced CT scans is primarily due to finer calcification, indicating discrete calcifications (Fig. 1). Steinhoff, et al.,\(^1\) reported that 85% of grade I astrocytomas presented with decreased density, 33% of grade II astrocytomas presented similarly, and the corresponding incidence in glioblastomas was 36%. Mixed density was seen in 65% of glioblastomas in their series, a figure similar to Weisberg's finding; however, areas of mixed density due to hemorrhage and calcification were not specified.

Thus, the finding in the supratentorial region on plain CT scans of a mixed-attenuation lesion containing a
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solid component that exhibits hyperdensity and calcification, particularly if well demarcated in appearance, suggests the possibility of ependymoma. Moderate to marked enhancement should be expected on a postcontrast CT study. An appearance similar to glioblastoma was found in Case 3 of our series (Fig. 2), although hyperdensity with some calcification would be unusual for glioblastoma. The appearance of the predominantly intraventricular high-density lesion as seen in Case 4 (Fig. 3) may be seen in grade II and III supratentorial astrocytomas, although in those tumors, a decreased density would be more commonly seen on plain CT scans.

Case 2 deserves separate comment because the apparently purely cystic lesion exhibited a fluid-fluid level on plain CT scans with layering secondary to differences of specific gravity of the fluid content (Fig. 4). There has been no previous report of a fluid-fluid level within a cystic portion of an ependymoma, although there were several reports of this finding in other types of glioma and metastatic tumors. The high-density material may be infarcted tissue, necrosis, or hemorrhage. Surgical aspiration of serous material from the cyst in our Case 2 would favor hemorrhage as the cause for the high-density material. Following contrast administration, there was enhancement of the thin rim of the cyst and accentuation of the fluid-fluid line, presumably because of leakage of contrast material into the cyst.

We do not recommend routine angiography in supratentorial gliomas. However, ependymomas can mimic the appearance of truly intraventricular lesions, such as meningiomas. Meningiomas, choroid plexus papillomas, and colloid cysts that are truly intraventricular lesions derive their vascular supply solely from the choroidal arteries. In gliomas, angiography will usually demonstrate either a blood supply from lenticulostriate arteries, thalamoperforating arteries, and other arteries if the tumor is sufficiently large (as in Case 4), or no feeding arteries at all in 50% of cases. An angiographic study, therefore, can distinguish the nature of a tumor occurring within or around the ventricles.

Based on neuroimaging, supratentorial ependymomas may be differentiated from the more common supratentorial astrocytomas, less common oligodendrogliomas, intraventricular meningiomas, and tumors of neural origin (neuroblastomas and gangliogliomas). The last two tumors are not commonly encountered and are found in specific age groups. In addition, neuroblastomas are usually detected within a shorter period of time after onset of symptoms, in contrast to the longer symptom period in gangliogliomas before the diagnosis is obtained. If a suspected ependymoma ap-

FIG. 3. Case 4. This 21-year-old woman presented with a history of progressively severe headaches and several episodes of loss of consciousness. At craniotomy, a predominantly intraventricular tumor was confirmed with extension into the Sylvian fissure. Upper: Preoperative studies, including plain (left) and contrast-enhanced (center) computerized tomography scans showing a hyperdense lesion with small cystic components, and a moderately enhancing predominantly intraventricular mass; and an angiogram, venous phase (right), demonstrating several large draining veins (arrow). Lower: Photomicrograph showing a papillary ependymoma with perivascular disposition of tumor cells. H & E, × 25.
pears to be predominantly within the lateral ventricle, other intraventricular lesions should be considered. The differentiation may be aided by closely spaced axial and coronal CT slices in order to better outline the tumor in relation to the lateral ventricle. Angiographic studies can be of assistance in further delineating the location of the tumor.

Treatment and Outcome

Our cases demonstrate that recurrence appears to be the rule in spite of surgery and radiation therapy. However, the recurrences may be spread out over several years, as in Case 5 (see Table 2). That tumor had originated from the posterior fossa where survival time is reported to be doubled after irradiation compared to supratentorial ependymomas.\(^7\) In a report of a smaller number of patients, however, Dohrmann, et al.,\(^4\) noted a longer period of survival in patients with supratentorial ependymomas than in those with infratentorial lesions 42 months after the diagnosis was made, but there was no difference among children and adults in the supratentorial and infratentorial ependymoma groups during a 5-year follow-up period. In a recent study, Salazar, et al.,\(^{16}\) reported no difference in survival between infratentorial and supratentorial ependymomas in 51 patients after 50 months. There was a difference in survival beyond 5 years in patients with grade I and II tumors compared to grade III and IV tumors. The authors stated that with aggressive postoperative radiotherapy and consideration of tumor grade, location, status of the cerebrospinal fluid, and patient’s age, a 10-year survival rate of 69% was achieved, with a range from 67% (high-grade ependymomas) to 75% (low-grade ependymomas). Of our five patients whom an initial diagnosis of high-grade ependymoma was obtained, only one (Case 8, follow-up period 4 years) is alive without evidence of recurrence. That patient was also the only one who received craniospinal irradiation.

In a report on CT follow-up monitoring of medulloblastomas and ependymomas, Enzmann, et al.,\(^6\) indicated that there was poor correlation between the clinical course and CT evidence of recurrence, although it is to be noted that there were 29 patients with medulloblastoma and seven patients with ependymoma, and in only one patient was the tumor located in the supratentorial compartment. In that series, there was a high incidence of intraventricular metastases (80%) and involvement of the subarachnoid space (40%). We have not documented such a high incidence on CT scans. Enzmann, et al., examined their patients routinely on a 1- to 2-month basis; however, with regular CT follow-up monitoring (average every 7.4 months) of those patients who remained under our care, there was a close correlation between a patient’s clinical course and the CT appearance. We believe that asymptomatic patients with a treated low-grade tumor should have annual follow-up CT scanning, but patients with high-grade tumors and those with positive cerebrospinal fluid cytology should be examined every 6 months. In two of our patients with tumor recurrence there was a more marked appearance of edema surrounding the lesion compared with the initial CT; one of these patients had a benign histological diagnosis and the other had malignant changes. Both exhibited marked mass effect and edema on recurrence, probably related to several factors caused by the primary neoplastic tissue.\(^{18,19}\)

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References

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