AJNR

Intracranial ganglioglioma: MR, CT, and clinical findings in 18 patients.

M Castillo, P C Davis, Y Takei and J C Hoffman, Jr

AJNR Am J Neuroradiol 1990, 11 (1) 109-114
http://www.ajnr.org/content/11/1/109

Intracranial Ganglioglioma: MR, CT, and Clinical Findings in 18 Patients

Mauricio Castillo[1]
Patricia C. Davis[1]
Yoshii Takei[2]
James C. Hoffman, Jr.[1]

Eighteen cases of pathologically proved intracranial gangliogliomas were reviewed to determine their MR, CT, and clinical characteristics. Seventeen patients were evaluated with contrast-enhanced CT and 14 were studied by MR imaging. Eight tumors were predominantly cystic; half of these demonstrated some contrast enhancement, and five contained calcifications. These cystic gangliogliomas were located, in order of decreasing frequency, in the cerebellum, temporal, frontal, and parietal lobes. Ten tumors were solid; eight of these showed contrast enhancement, and only one contained calcifications. Small cysts were present in one solid mass. Solid gangliogliomas occurred preferentially in the temporal lobes. On MR, the findings were nonspecific and reflected the CT findings. In one patient who received gadolinium-DTPA the lesion did not enhance. Clinically, all patients presented with nonfocal long-standing symptoms and all but three were alive an average of 18 months after the initial diagnosis.

Pathologists are recognizing ganglioglioma with increasing frequency, and although its radiographic characteristics vary, it should be included in the differential diagnosis when the above-described findings are encountered.

***AJNR* 11:109–114, January/February 1990; *AJR* 154: March 1990**

Ganglioglioma is thought to be a rare primary lesion that accounts for only 0.4 to 0.9% of all intracranial tumors [1, 2]. These tumors differ from the most common gliomas in that they contain both glial elements and differentiated nerve cells [3]. These relatively low-grade neoplasms generally behave in a benign fashion and have a favorable prognosis. We present the CT and MR findings correlated with the clinical, surgical, and histological characteristics in 18 patients with proved intracranial gangliogliomas.

Materials and Methods

From 1984 to 1988, 18 patients with intracranial gangliogliomas were evaluated at our institution. The patients were 1 to 70 years old; 13 were male, 5 were female. All cases were histologically proved. All radiographic studies and medical records were reviewed retrospectively. In 16 cases, preoperative CT studies were obtained before and after the IV administration of contrast medium (100 ml of 60% iothalamate meglumine). The CT studies were obtained on a variety of late-generation scanners by means of our routine protocol; that is, axial 5-mm sections through the posterior fossa were followed by 10-mm sections through the remainder of the brain. One patient had only a contrast-enhanced CT study.

MR studies were obtained before surgery in 14 patients by using both spin-echo T1-weighted, 500–800/20–50 (TR/TE), and T2-weighted, 1500–2000/50–100, sequences. Transverse 10-mm images were obtained in all patients, and coronal 10-mm images were acquired in three cases. The MR studies were done on either a 0.5-T (six studies) or a 1.5-T (eight studies) Philips unit. Gadolinium-DTPA (0.1 mmol/kg) was administered intravenously to one patient. One patient was preoperatively evaluated by MR only.

Three patients had selective cerebral angiography with cut film.

The CT and MR studies were reviewed with special attention given to the following

Received April 4, 1989; revision requested May 30, 1989; revision received June 16, 1989; accepted June 16, 1989.

[1] Department of Radiology, Emory University Hospital, 1364 Clifton Rd., N.E., Atlanta, GA 30322. Address reprint requests to P. C. Davis.

[2] Department of Pathology, Emory University Hospital, Atlanta, GA 30322.

0195-6108/90/1101-0109

parameters: location, size, and internal characteristics (solid, cystic, and presence of calcifications) of the lesion; its pattern of enhancement; and the presence of associated abnormalities.

Thirteen patients had postoperative follow-up contrast-enhanced CT studies. Three patients who had recent surgery were followed only with MR. No follow-up studies were available in two instances. The postoperative studies were evaluated for the presence of recurrent tumor (local or disseminated) and the effects of treatment.

The histological diagnosis of ganglioglioma was made in accordance with the following criteria: (1) lesions were composed of neoplastic astrocytes, and (2) lesions demonstrated the presence of neoplastic ganglion cells, which often exhibited Nissl substance or gave origin to neuronal processes as demonstrated by silver impregnation methods (six cases).

Results

The clinical findings at presentation for the 18 patients studied, the postoperative diagnoses, location of tumors, method of treatment, and follow up are summarized in Table 1.

On CT, eight tumors were primarily cystic (Fig. 1). Four cystic lesions demonstrated laminar (two cases) or nodular (two cases) areas of contrast enhancement along their margins (Fig. 2). Four cystic neoplasms did not enhance. Calcifications were present in five cases. On MR, the cystic components of these lesions appeared heterogeneous and mainly of low signal intensity with respect to the normal CSF on the T1-weighted images. On T2-weighted images, these tumors were of increased signal intensity (Fig. 3). The eight cystic lesions were located in the cerebellum (three cases) temporal lobes (two cases), frontal lobes (two cases), and parietal lobe (one case) (Table 2).

CT showed 10 tumors to be predominantly solid. These tumors were either hypodense (two tumors), isodense (four tumors), or slightly hyperdense (four tumors) with respect to the adjacent normal parenchyma (Fig. 4) (Table 3). All but two of the masses showed some degree of contrast enhancement (Fig. 5). Calcification was detected in one solid tumor (Fig. 6). Small cysts were present in one lesion. On MR, the appearance of these solid tumors varied widely on the T1-weighted images: three were slightly hypointense, three were isointense, and three were slightly hyperintense relative to normal gray matter (Fig. 7) (Table 4). On the T2-weighted images, all but one of these lesions were of increased signal intensity. The solid tumors were found in the cerebellum (one lesion), temporal lobes (five lesions), suprasellar region (one lesion), and basal ganglia (three lesions). In one patient who received gadolinium-DTPA (0.1 mmol/kg), the lesion did not enhance (Fig. 8).

All gangliogliomas (cystic and solid) measured between 2 and 10 cm in their greatest dimension.

One patient with a proved ganglioglioma had no recognizable neoplasm by either CT or MR. This patient had undergone partial resection of the right temporal lobe for an astrocytoma, but because of recurrent seizures the patient was reoperated and further resection of the right temporal lobe showed the presence of ganglioglioma.

Histological examination of all lesions was confirmatory for ganglioglioma. In three patients (cases, 2, 3, and 8) anaplastic components were present.

Postoperative studies were available in 16 patients. Thirteen patients had follow-up contrast-enhanced CT studies and three patients were followed only with MR. No follow-up studies were available in two patients who died. All patients were followed an average of 18 months (range, 8–42 months) after their initial surgery, and all follow-up studies demonstrated postsurgical changes (encephalomalacia) or other treatment-related changes (diffuse atrophy, periventricular white matter disease) but no evidence of recurrent tumor or growth of the residual neoplasm. Although one patient died of unrelated causes, the last available follow-up study showed no change in the size and configuration of the tumor.

TABLE 1: Clinical Features of Patients with Ganglioglioma

Case No.	Age (years)	Sex	Clinical Presentation	Preop. Diagnosis	Treatment	Follow up (months)	Outcome
1	21	F	Headaches	Glioma	TR	30	Alive
2	70	M	Confusion	Glioma	PR, CT*	15	Dead
3	17	M	Seizures	Oligodendroglioma	PR, RT*	42	Alive
4	51	M	Headaches	Glioma	PR	24	Alive
5	16	M	Seizures	Glioma	TR, RT	8	Alive
6	62	M	Memory loss	Glioma	PR	–	Dead
7	59	F	Headaches	Glioma	PR	–	Dead
8	24	F	Weakness	Glioma	PR, RT*	24	Alive
9	26	M	Seizures	Glioma	PR, RT	4	Alive
10	8	F	Headaches	Glioma	PR, RT	2	Alive
11	17	M	Seizures	Glioma	PR, RT	6	Alive
12	15	M	Headaches	Glioma	PR, RT	24	Alive
13	34	M	Seizures	Oligodendroglioma	PR, RT	12	Alive
14	7	M	Headaches	Craniopharyngioma	PR, RT	36	Alive
15	36	M	Seizures	Oligodendroglioma	PR, RT	14	Alive
16	55	M	Seizures	Glioma	PR, RT	12	Alive
17	1	F	Headaches	Glioma	PR	24	Alive
18	1	M	Headaches	Neuroblastoma	PR, RT	24	Alive

Note.—TR = total resection, PR = partial resection, RT = radiation therapy, CT = chemotherapy, * = tumor with anaplastic component.

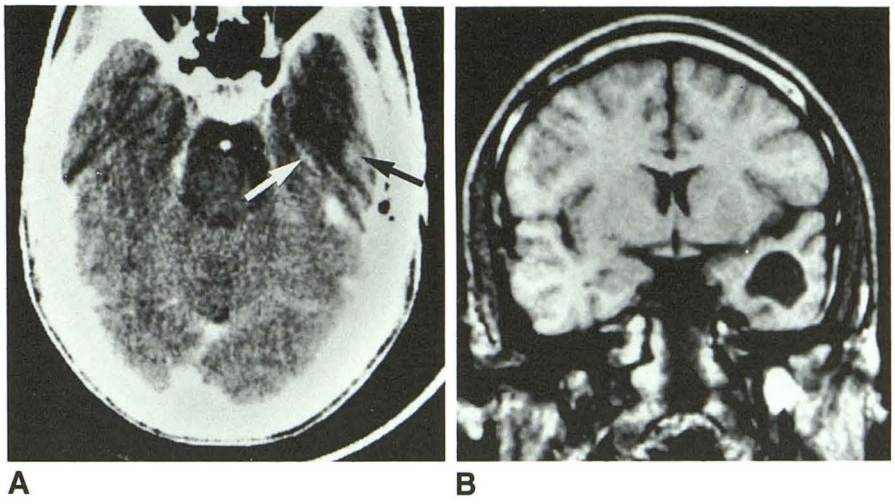

Fig. 1.—Case 1.
A, Contrast-enhanced CT scan shows area of low attenuation in left temporal lobe. Note subtle enhancement of posterior margins of lesion (*arrows*).
B, On this coronal MR image (500/30) through mid temporal lobes, lesion appears hypointense and of similar signal intensity to that of CSF. Note well-defined margins. On T2-weighted image (not shown), lesion was of increased signal intensity.

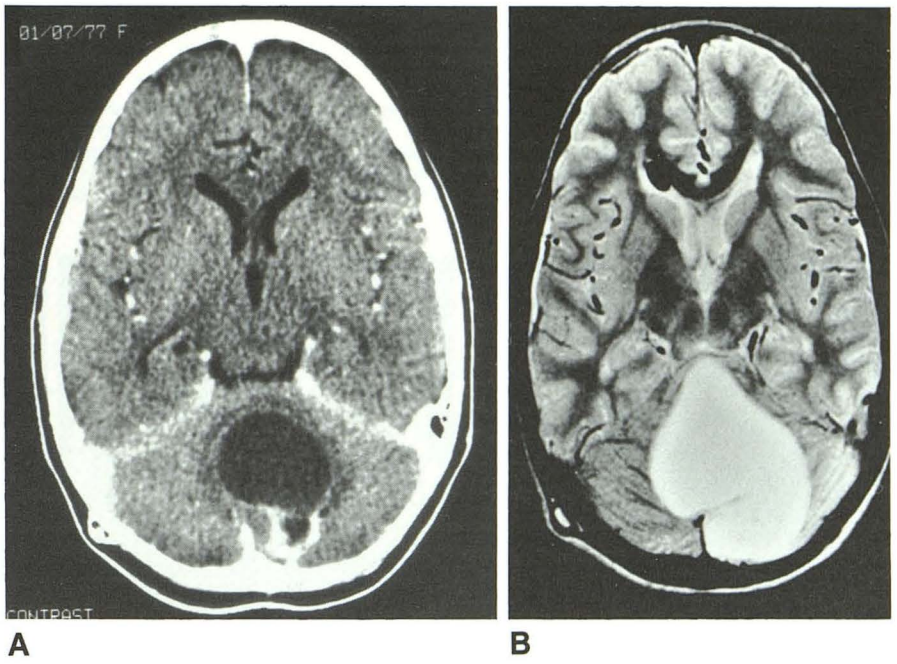

Fig. 2.—Case 17.
A, Contrast-enhanced CT scan shows cystic ganglioglioma with minimal enhancement of posterior margin of cyst present within cerebellum. On the basis of radiographic findings, this lesion cannot be differentiated from a cystic astrocytoma.
B, Axial MR section (2000/80) at same level shows lesion to be of increased signal intensity with well-defined margins. A shunt catheter is present in region of frontal horn of right lateral ventricle.

TABLE 2: Location of Cystic and Solid Lesions in 18 Patients with Ganglioglioma*

	Cystic	Solid	Total
Temporal	2	5	7
Frontal	2	–	2
Parietal	1	–	1
Suprasellar	–	1	1
Basal ganglia	–	3	3
Cerebellum	3	1	4
Total	8	10	18

* One lesion was not identified by either MR or CT.

TABLE 3: CT Findings in 17 Patients with Ganglioglioma*

	Hypodense	Isodense	Hyperdense	+Enhancement	Ca^{++}
Cystic	8	–	–	4	5
Solid	2	4	4	8	1

* One lesion was not identified by either MR or CT.

Discussion

The term ganglioglioma was initially proposed to describe CNS tumors containing both glial and neuronal elements [4, 5]. Gangliogliomas differ from gangliocytomas in that the latter are composed purely of neuronal elements and contain no glial components [6, 7]. Ganglion neoplasms are classified according to their stages of differentiation and to the relative proportion of neuronal to glial elements, as follows: gangliocytoma, ganglioglioma, ganglioneuroblastoma, anaplastic ganglioglioma, and neuroblastoma [8, 9].

Gangliogliomas can occur at any age, and the pediatric and adult populations are believed to be equally affected [1, 3, 10, 11]. Clinically, both the age and gender distribution of our patients reflect that reported in the literature.

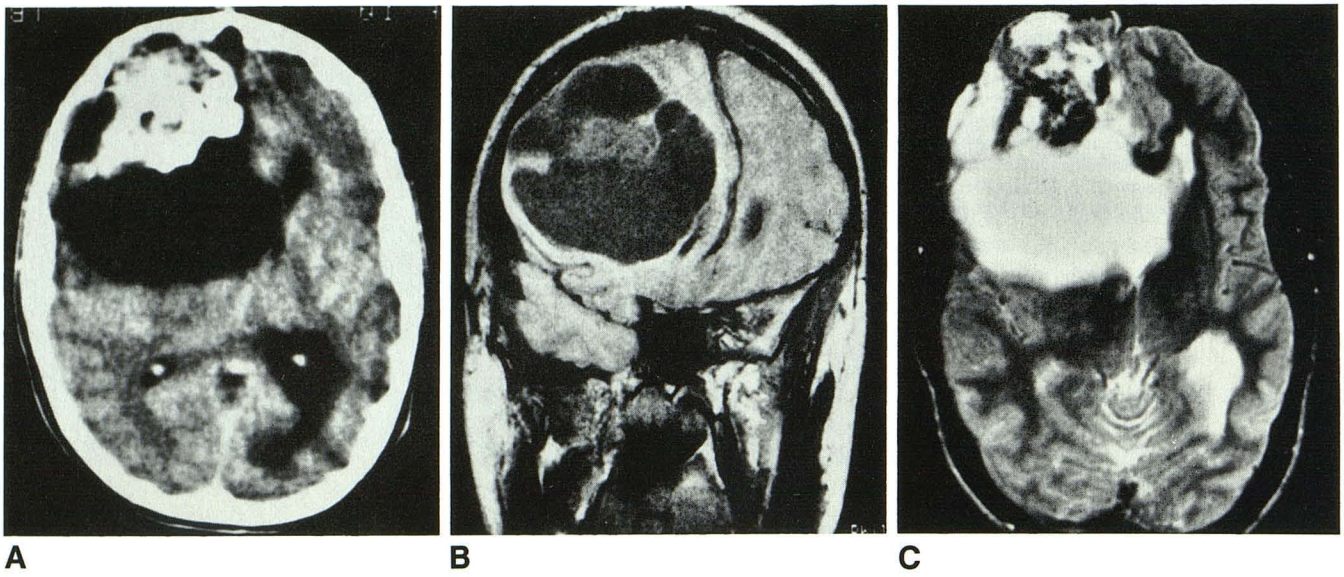

Fig. 3.—Case 3.
A, Contrast-enhanced CT scan shows large cystic ganglioglioma occupying right frontal lobe. Note large areas of calcification and absence of definite enhancement. There is dilatation of left lateral ventricle.
B, Coronal MR image (500/30) shows contents of cystic lesion to be heterogeneous. A rim of slightly increased signal intensity surrounds lesion.
C, Axial MR image (2000/80) at same level as *A* shows fluid within lesion to be hyperintense. Calcifications are seen in areas of signal void in anterior aspect of tumor. Margins of lesion are relatively well defined and no associated edema is present.

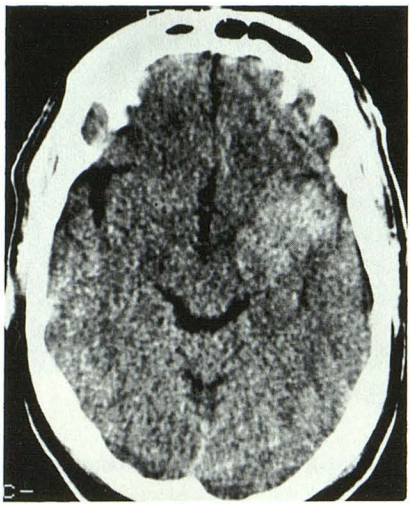

Fig. 4.—Case 4. Noncontrast CT scan shows hyperdense, ill-defined mass in left temporal lobe. There is mass effect upon ipsilateral sylvian fissure. Tumor did not enhance after IV administration of contrast medium.

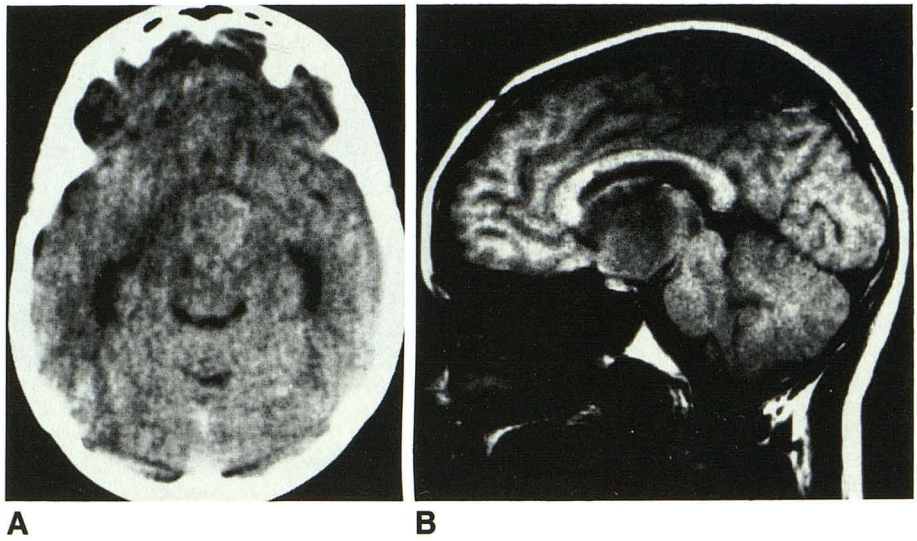

Fig. 5.—Case 14.
A, Noncontrast CT scan shows round, slightly hyperdense mass in suprasellar region. There is dilatation of temporal horns of lateral ventricles.
B, Midline sagittal MR image (550/30) shows suprasellar mass to be heterogeneous but predominantly of intermediate signal intensity. Pituitary gland is seen caudad to mass.

Previously published series agree that the most common location is the temporal lobes [1–3, 11]. However, gangliogliomas can also occur in other parts of the cerebrum as well as in the cerebellum, brainstem, spinal cord, and optic nerves [8, 10, 12–14]. In our series, seven of 16 lesions were found in the temporal lobes. The remainder of the tumors were randomly distributed throughout other parts of the brain. As expected, the clinical presentation of all our cases was related to the presence of nonspecific long-standing symptoms, which probably reflect the slow-growing nature of the lesion (Table 1). Despite the benign nature of the neoplasms, complete resection was accomplished in only two cases, because of the location and size of these lesions. Anaplastic components were found in three of our patients. Of these, one patient died as a result of tumor recurrence; the others were alive without recurrence at 24 and 36 months after diagnosis.

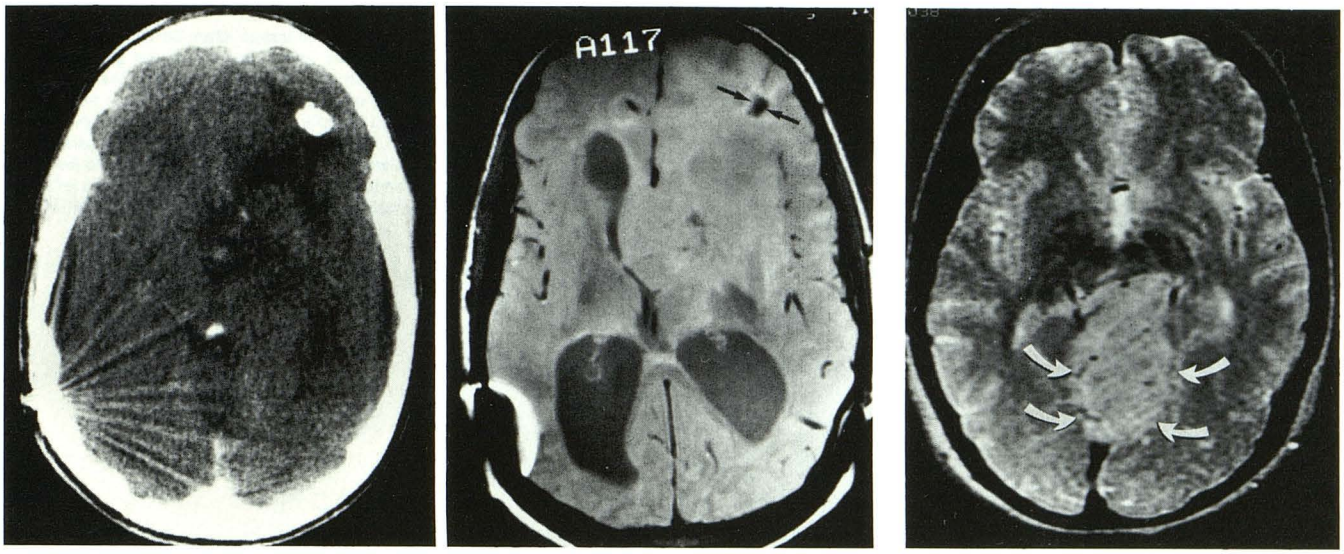

Fig. 6.—Case 15.
***A*, Contrast-enhanced CT scan shows low-attenuation lesion in left frontal lobe that contains a large calcification in its anterior aspect. There is no definite enhancement.**
***B*, MR image (1800/30) obtained 6 weeks after CT study (*A*) shows large tumor to be isointense with gray matter. Areas of calcification are well seen (*arrows*). In the interval, hydrocephalus has developed as a result of shunt malfunction.**

Fig. 7.—Case 10. Axial MR image (2000/80) shows large ganglioglioma arising in cerebellar vermis (*arrows*). Lesion is slightly hyperintense relative to normal brain parenchyma. On T1-weighted MR study (not shown), the lesion was also slightly hyperintense. Radiographically, the lesion is indistinguishable from Lhermitte-Duclos disease.

TABLE 4: MR Findings in 14 Patients with Ganglioglioma*

	T1-Weighted			T2-Weighted		
	Low	Intermediate	High	Low	Intermediate	High
Cystic	7	–	–	–	–	7
Solid	3	3	1	–	1	4

* One lesion was not identified by either MR or CT.

In a review of 48 cases of pathologically proved ganglioglioma compiled from the literature, Dorne et al. [1] found that 38% of the lesions were cystic in nature, the remainder of the patients harbored solid tumors. The solid tumors in that review had a wide variety of appearances on noncontrast CT studies (38% were low density, 15% were isodense, 15% were high density, 32% were mixed density, and 17% were indeterminate density). After contrast administration, enhancement was noted in half the lesions. Although our findings agree with the above, we are uncertain as to the meaning of the term *indeterminate density* used in that article.

The MR characteristics of ganglioglioma have not been well established. In one series, Denierre et al. [2] described four cases. Two solid masses were of increased signal intensity on the T1-weighted images and of low signal intensity on the T2-weighted images. The short T1 relaxation characteristics of these solid masses are hard to explain, as the presence of hemorrhage, cholesterol, or proteins was not mentioned. Two patients had cystic tumors with long relaxation times on both the T1- and T2-weighted images. Our results differ somewhat from those mentioned above. The solid masses in our cases showed a variable and nonspecific appearance on T1-weighted images. On T2-weighted images, all solid lesions showed some degree of increased signal intensity. Four cystic lesions were heterogeneous and slightly hyperintense relative to normal CSF on the T1-weighted images. On the T2-weighted images, these cystic lesions showed increased signal intensity. In their series, Denierre et al. [2] observed that "there was good contrast between the lesion and the normal brain tissue." Although we believe that this observation might be valid for the cystic tumors, we were unable to distinguish well-defined tumor margins from the adjacent brain parenchyma in six cases of solid gangliogliomas examined by MR. It is conceivable that gadolinium-DTPA–enhanced MR studies may be helpful in better delineating the margins in some tumors. In our series only one patient received gadolinium-DTPA; the tumor in this patient did not enhance (Fig. 8).

Calcifications occurred in five (28%) of our 18 patients. It is not surprising that calcifications within gangliogliomas have not previously been detected by MR. We were able to visualize nonspecific areas of signal void by MR in all patients in whom CT showed calcifications. The areas of calcifications in our cases were fairly large, and on MR, with only spin-echo techniques used, they could not be differentiated from vessels with high flow or areas of old hemorrhage without previous knowledge of the CT findings.

Cerebral angiography showed three lesions to be avascular, a radiographic finding that has previously been reported [15].

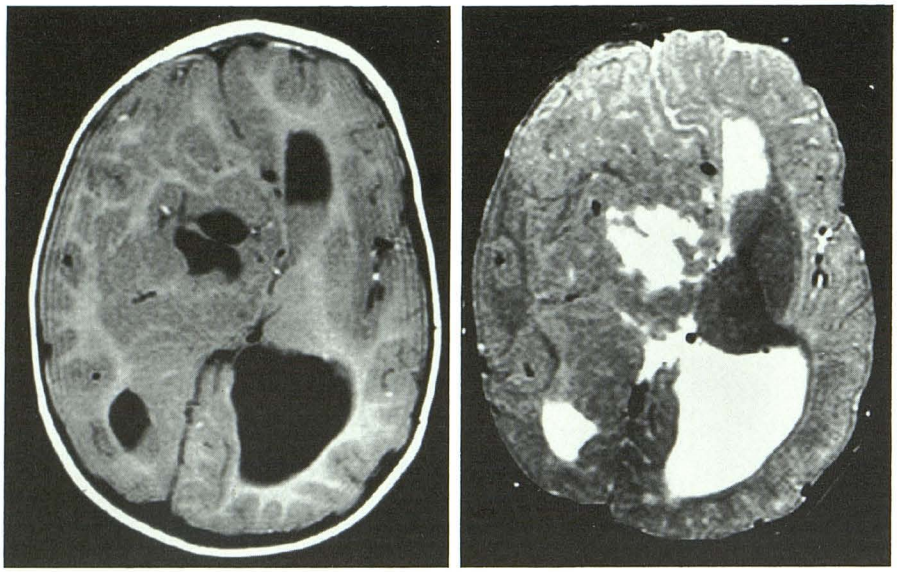

Fig. 8.—Case 18.
***A*, Axial MR image after Gd-DTPA administration shows large, nonenhancing ganglioglioma in region of right basal ganglia. Mass is isointense with gray matter and contains two central cystic components. Hydrocephalus is present.**
***B*, Axial MR section (2000/80) at same level shows cystic components of lesion to be of increased signal intensity. Solid portion of mass remains isointense with normal gray matter.**

From the radiographic and histological viewpoint, an important differential diagnosis when considering ganglioglioma of the cerebellum is that of Lhermitte-Duclos diseases [16, 17]. This lesion contains neuronal elements, and pathologically it is thought to represent a hamartoma. Clinically, this lesion behaves as a true neoplasm: it distorts the normal cerebellar architecture and causes thickening of the overlying folia, a pattern similar to that seen in one of our patients (Fig. 7).

Follow-up studies of our patients occurred 2 to 42 months after initial treatment. Fifteen patients were known to be alive at the completion of this study, and CT or MR studies showed no recurrence of the tumor in those patients in whom total resection was accomplished and no significant change in the size of those tumors that were only partially resected. The latter observation could be related to the relatively short follow-up period.

Ganglioglioma was not considered in the preoperative differential diagnosis in any of our patients. However, ganglioglioma is being recognized with increasing frequency by pathologists. In our series, nonenhanced MR images did not provide additional useful information to that obtained from the contrast-enhanced CT scans. Because of its relatively better prognosis as compared with the more usual types of brain neoplasms, radiologists should include ganglioglioma in the differential diagnosis of intracranial masses when (1) a relatively large cystic mass is present (preferentially located in the temporal lobes or the cerebellum) that has little or no discernible solid components, has margins that enhance only slightly, and that at times contains dense and large calcifications; or (2) a solid, poorly defined mass is found in the temporal lobes that is generally hypodense by CT or of increased signal intensity on T2-weighted MR images.

REFERENCES

1. Dorne HL, O'Gorman AM, Melanson D. Computed tomography of intracranial gangliogliomas. *AJNR* **1986**;7:281–285
2. Denierre B, Stinchnoth FA, Hori A, Spoerri O. Intracerebral gangliogliomas. *J Neurosurg* **1986**;65:177–182
3. Johanson JH, Rekate HL, Roesmann U. Gangliogliomas: pathological and clinical correlation. *J Neurosurg* **1981**;54:58–63
4. Perkins OC. Ganglioglioma. *Arch Pathol Lab Med* **1926**;2:11
5. Courville CB. Ganglioglioma: tumors of the central nervous system: review of the literature and report of two cases. *Arch Neurol Psychiatry* **1930**;24:439–491
6. Altman NR. MR and CT characteristics of gangliocytoma: a rare cause of epilepsy in children. *AJNR* **1988**;9:917–921
7. Rommel T, Hamer J. Development of ganglioglioma in computed tomography. *Neuroradiology* **1983**; 24:237–239
8. Mizuno J, Nishio S, Barrow DL, et al. Ganglioglioma of the cerebellum: case report. *Neurosurgery* **1987**;21:584–588
9. Russell DS, Rubinstein LJ. *Pathology of the tumors of the nervous system,* 4th ed. Baltimore: Williams & Wilkins, **1977**:261–282
10. Nass R, Whelan MA. Gangliogliomas. *Neuroradiology* **1988**;22:67–71
11. Zimmerman RA, Bilaniuk LT. Computed tomography of intracerebral gangliogliomas. *J Comput Tomogr* **1979**;3:24–30
12. Garcia CA, McGarry PA, Collada M. Ganglioglioma of the brainstem. *J Neurosurg* **1984**;60:431–434
13. Henry JM, Heffner RR, Earle KM. Gangliogliomas of the central nervous system: clinicopathological study of fifty cases (abstr). *J Neuropathol Exp Neurol* **1978**;37:626
14. Gritzman MC, Snyckers FD, Proctor NS. Ganglioglioma of the optic nerve. *S Afr Med J* **1983**;63:863–865
15. Katz MC, Kier EL, Schecter M. The radiology of gangliogliomas and ganglioneuromas of the central nervous system. *Neuroradiology* **1972**;4:69–73
16. Ambler M, Pagacer S, Sidman R. Lhermitte-Duclos disease (granule cell hypertrophy of the cerebellum): pathological analysis of the first familial cases. *J Neuropathol Exp Neurol* **1969**;28:622–647
17. Oppenheimer DR. A benign "tumor" of the cerebellum: report on two cases of diffuse hypertrophy of the cerebellar cortex with review of nine previously published cases. *J Neurol Neurosurg Psychiatry* **1955**;18:199–203