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Computed Tomography of Intracranial Gangliogliomas

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Thirteen patients with pathologically proven gangliogliomas were studied radiographically. The computed tomographic (CT) features of these 13 lesions and the other 35 cases in the literature were analyzed. Although the CT appearance of gangliogliomas was varied, certain characteristics were noted. The most common location was in the cerebral hemispheres, most often the temporal lobe. At least part of the tumor was low density in 71% of the unenhanced CT examinations. There were focal calcifications in 35% and enhancement with contrast material in 50%.

Ganglioglioma is a rare tumor of the central nervous system. Its radiographic features were reviewed in 1971 [1], but little has been done to accurately document the appearance of gangliogliomas since the advent of computed tomography (CT). The infrequency of gangliogliomas and the availability of only scattered reports in the literature, each including relatively few patients [2-14], has led to uncertainty regarding their CT appearance. Thirteen patients with pathologically proven gangliogliomas were examined with CT at our institutions. This is the largest such series reported to date. We analyzed the CT findings in these 13 patients and the other 35 cases in the literature to try to better define the CT spectrum of gangliogliomas.

Subjects and Methods

From 1977 to 1984, 1243 patients at the Montreal Neurological Institute (MNI) and 81 patients at the Montreal Children's Hospital (MCH) were treated for primary intracranial neoplasms. Of these, 12 patients (0.97%) at the MNI and two patients (2.5%) at the MCH were found to have gangliogliomas. One of the patients at the MNI presented with a recurrence of a ganglioglioma that had been partly resected 12 years before. Because of uncertainty as to the proportion of the CT findings that were caused by the tumor as opposed to postoperative changes, this patient was not included in the study. The other 13 patients were reviewed retrospectively.

Results

The clinical and radiographic features of the 13 patients are summarized in table 1. They ranged in age from 9 to 67 years. Four patients were in their first decade, two in the second, two in the third, four in the fourth, and one patient was over 40 years of age. Eight of the 13 were male. Nine presented with seizures. Ten had had symptoms for 1 year or longer. Ten of the lesions were in the cerebral hemispheres with eight in the temporal lobe. Two were in the cerebellar hemispheres. One was located in the pineal gland.

All of the patients underwent plain skull radiography, CT, and arteriography. The skull examinations were normal in nine and showed evidence of increased intracranial pressure in three. One patient had calcification that was detectable on the plain

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TABLE 1: Clinical and Radiographic Features of Gangliogliomas

Case No. (age, gender)	Clinical Findings	Skull Film	CT Appearance			
			Location	Density	Calcification	Enhancement
1 (22,F)	2 yr absence attacks; three grand-mal seizures	Normal	Temporal lobe	Low	+	-
2 (10,F)	Temporal lobe seizures since infancy	Normal	Temporal lobe	Iso	-	-
3 (39,M)	13 yr generalized headaches; 6 mo of transient vertigo	Normal	Pineal	Iso + low	-	+
4 (32,M)	20 yr partial complex seizures	Normal	Temporal lobe	Low	-	-
5 (23,F)	Single generalized seizure	Normal	Occipital lobe	Low	-	+
6 (67,M)	6 mo upper extremity sensory seizures	Normal	Temporal lobe	?*	-	+
7 (35,M)	3 yr leg pain progressing to paresis and ataxia; many yr impaired intellect	Macrocephaly; sellar enlargement and erosion; large posterior fossa	Cerebellar hemisphere	Low	-	-
8 (33,M)	30 yr partial complex seizures	Normal	Temporal lobe	Low	-	-
9 (9,M)	2 yr absence seizures	Normal	Temporal lobe	Low	-	-
10 (19,M)	18 yr absence salaam-type seizures	Focal thinning of parietal bone	Parietal lobe	Cystic	+	-
11 (13,M)	1 yr incoordination and unilateral nerve deafness	Split sutures; sellar enlargement and erosion	Cerebellar hemisphere	Cystic + iso	+	+
12 (9,M)	4 mo headaches, projectile vomiting, and hemiplegia	Split sutures	Temporo-parietal	Iso	-	-
13 (10,F)	9 yr temporal lobe seizures	Normal	Temporal lobe	Low	+	+

Note.—All 13 lesions were avascular angiographically. All CT densities are relative to normal brain on unenhanced studies. + = present; - = not present; ? = uncertain.
* Only a contrast-enhanced study was performed.

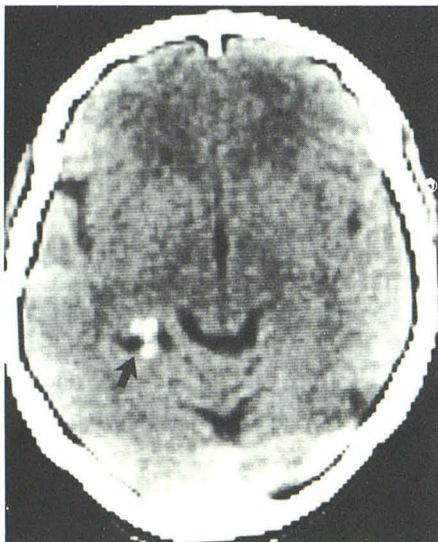


Fig. 1.—Case 1. Unenhanced CT scan. Hypodense lesion (arrow) with calcification in posteromedial aspect of temporal lobe.

films. The lesions were uniformly avascular arteriographically. CT without contrast material showed that 10 of the lesions had decreased attenuation coefficients compared with normal brain. Of the 10 low-density lesions, two appeared cystic and one was of mixed density with isodense and low-density

components. Two lesions were isodense. One patient had only a contrast-enhanced CT study. His lesion was hyperdense, probably because of enhancement. Four lesions showed areas of focal calcification demonstrable by CT. Four of the 12 lesions that were examined with CT both before and after contrast infusion showed enhancement. Two CT examinations in case 2 were completely normal, even in retrospect. Representative cases are illustrated (figs. 1-7).

Discussion

The name *ganglioglioma* was first suggested by Ewing in 1928 and first formally introduced into the literature by Courville [15] in 1930. It represents a slow-growing, usually benign neoplasm of the central nervous system.

Gangliogliomas occur in all age groups, but are most common in children and young adults. They have been reported to constitute from 0.4% to 7.6% of primary brain neoplasms [1, 7, 16, 17], with the higher figures derived from predominantly pediatric populations. Sixty percent are said to be found in patients under 30 years of age [18]. Some investigators suggest that gangliogliomas occur more often in males [11, 15], while others report the male-female incidence to be equal [3].

Most patients present clinically with a long-standing history of symptoms, usually seizures or headaches [19]. Our observations support this concept. Sutton et al. [12] emphasize the indolent nature of gangliogliomas and have found that

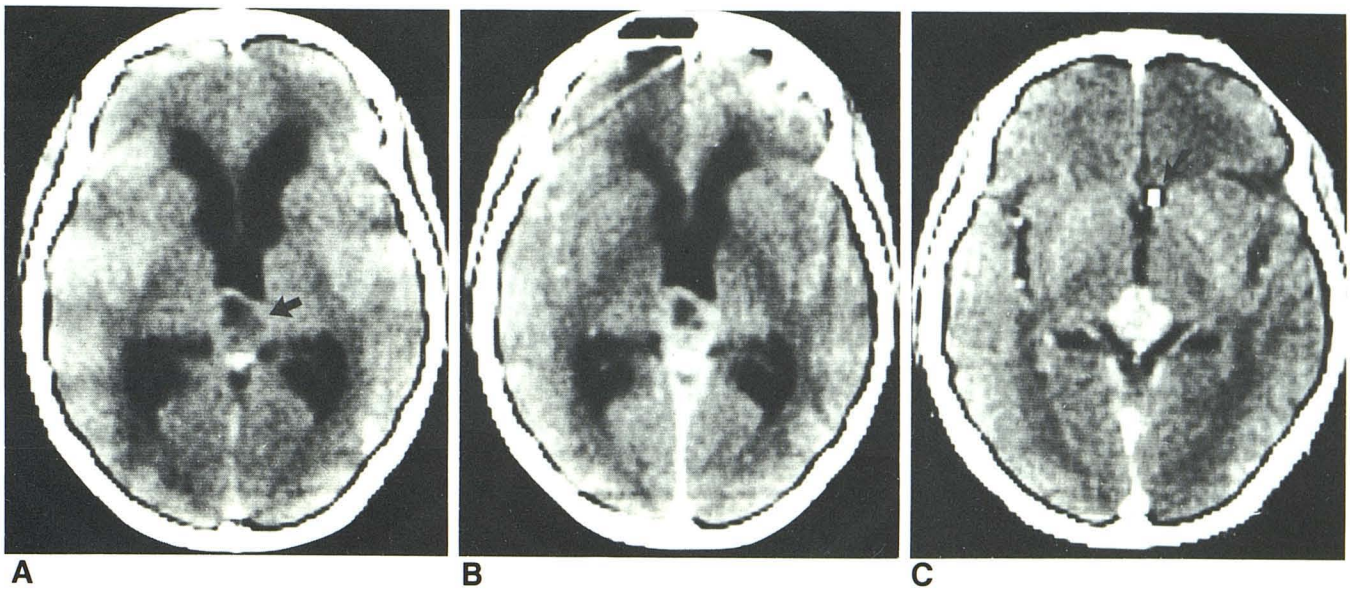


Fig. 2.—Case 3. **A**, Unenhanced CT scan. Hydrocephalus caused by pineal mass with hypodense center and isodense rim (*arrow*). **B**, After contrast infusion. Rim is seen to enhance. **C**, Contrast-enhanced study 2 years after surgical biopsy and radiation therapy of ganglioglioma. Resolution of hydrocephalus due to shunt (*arrow*), but persistence of enhancing pineal tumor.

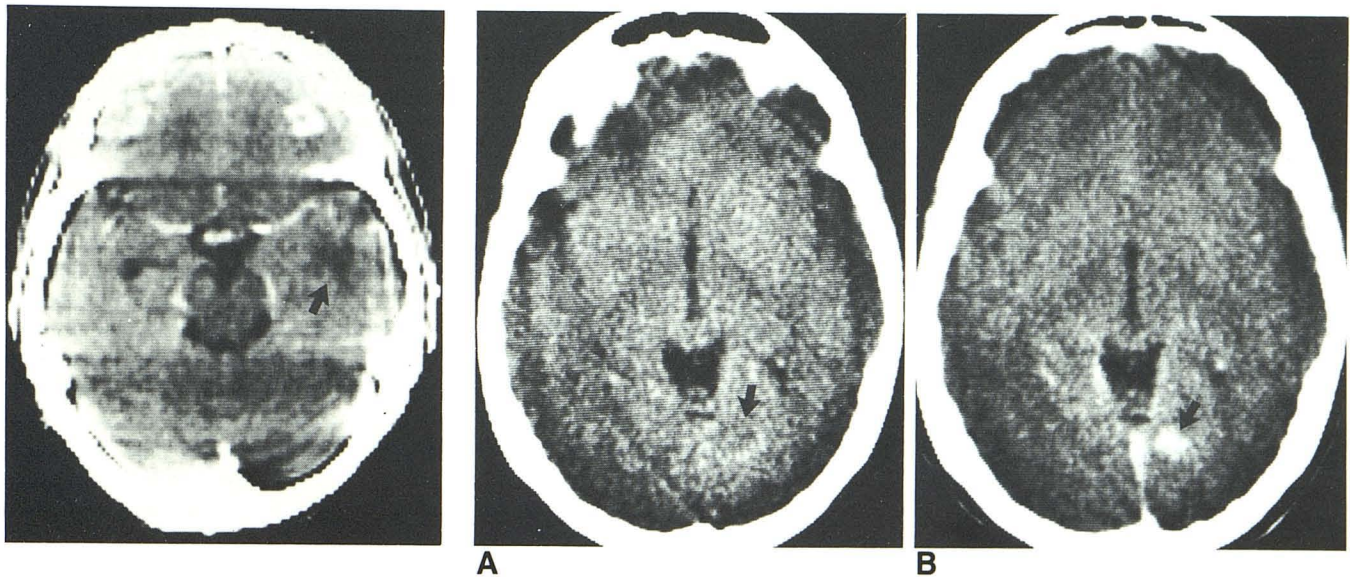


Fig. 3.—Case 4. CT scan after contrast infusion. Subtle, nonenhancing low-density lesion in temporal lobe (*arrow*).

Fig. 4.—Case 5. **A**, Unenhanced CT scan. Faint area of hypodensity in occipital lobe (*arrow*), perceptible only in retrospect. **B**, After contrast administration, lesion (*arrow*) becomes more obvious.

increasing seizure frequency is an important clue to their diagnosis. Such a history suggests that the cause of the seizures is not a static process, but a slow-growing neoplasm. Focal neurologic signs, symptoms of increased intracranial pressure, and intellectual impairment are less common manifestations of the tumor [12].

Gangliogliomas may occur anywhere in the central nervous system, both intracranially and extracranially. A patient with a ganglioglioma of the optic nerve has been described [20].

Russell and Rubinstein [18] reported that the most common location is the floor of the third ventricle, although the temporal lobe was the most common site in material referred to them from other centers. Henry et al. [19] found gangliogliomas most often in the temporal lobe, followed by the cerebellum, parietooccipital lobe, frontal lobe, and spinal cord, respectively. The temporal lobe was the most common site in our series. Gangliogliomas are usually single, although multiple tumors have been noted [18].

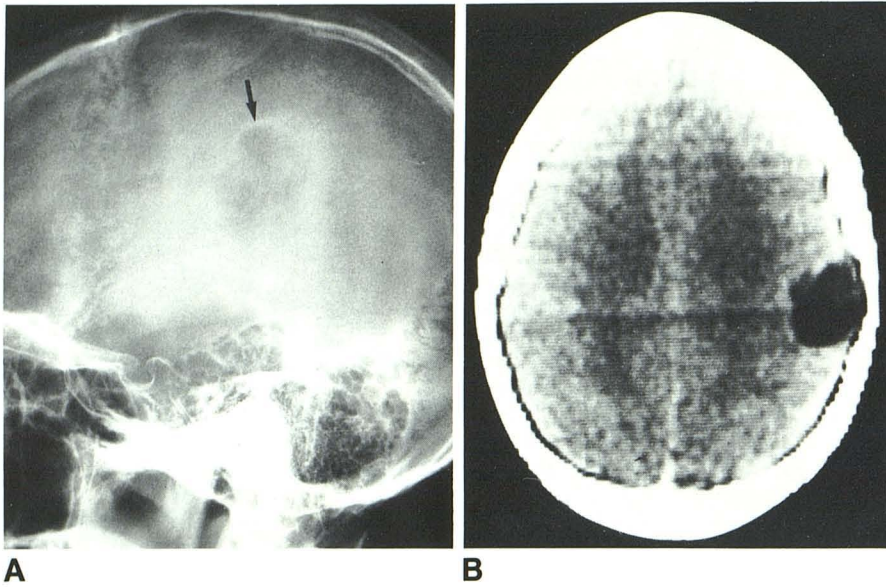


Fig. 5.—Case 10. A, Plain skull film. Lucent region (arrow) in parietal bone. B, Unenhanced CT scan. Cystic mass of parietal lobe. Calcification was seen at other levels.



Fig. 6.—Case 11. Contrast-enhanced CT scan. Large cystic mass with medial calcifications in cerebellar hemisphere (arrows), as well as obstructive hydrocephalus.

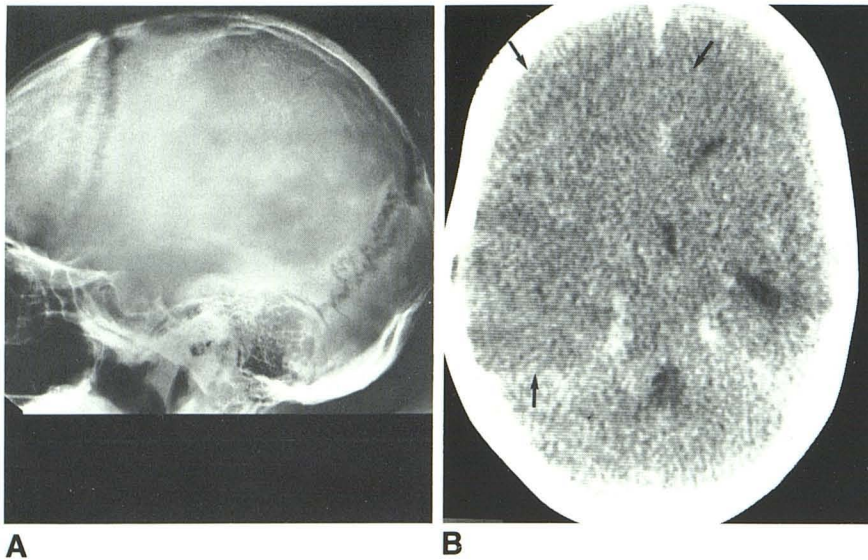


Fig. 7.—Case 12. A, Skull film. Widening of sutures with enlargement and erosion of sella turcica. B, Contrast-enhanced CT scan. Large, isodense, temporoparietal mass (arrows) causing compression and displacement of ventricular system. Lesion was shown to be avascular by cerebral angiography.

Macroscopically, gangliogliomas are usually firm, well circumscribed tumors with gray, finely granular cut surfaces that may be cystic or exhibit small foci of calcification. Occasionally a mural nodule in the wall of a large cyst is found. Rarely, they are diffusely infiltrating. Gangliogliomas histologically contain both mature ganglion cells and supporting stromal elements derived from glial tissue. Malignant transformation is quite unusual [18].

Radiographically, gangliogliomas generally act as avascular space-occupying lesions. Only one patient in our series had calcification visible on plain skull films, in keeping with the figure of 10% reported in the literature [1]. The lack of

arteriographic tumor vascularity in each of our 13 patients is also consistent with previous reports [1].

The CT features of the patients in our series plus those previously reported [2–14] are summarized in table 2. There were 48 patients in all. In some circumstances, the precontrast CT density or the presence or absence of contrast enhancement could not be ascertained from the information provided. These findings were classified as indeterminate and not included in the calculation of percentages.

Most of the 48 gangliogliomas were located in the cerebral hemispheres, with the greatest proportion found in the temporal lobes. Lesions involving at least part of the parietal lobe

TABLE 2: CT Features of Gangliogliomas in 48 Patients

	No.	(%)*
Location:		
Cerebral hemisphere	35	(73)
Posterior fossa	9	(19)
Thalamus	2	(4)
Pineal	2	(4)
Density:		
High density	6	(15)
Isodense	6	(15)
Mixed isodense and low density or cystic	13	(32)
Low density or cystic	15	(38)
Indeterminate	8	(17)
Calcification:		
Present	17	(35)
Absent	31	(65)
Enhancement:		
Present	22	(50)
Absent	22	(50)
Indeterminate	4	(8)

Note.—CT density is relative to normal brain on unenhanced study.
* Percentages exclude indeterminate studies.

were the next most common. A considerably smaller percentage of the tumors was located in the posterior fossa. These were equally divided in location between the brainstem and the cerebellar hemispheres. There were two thalamic lesions, one of which was bilateral. Two patients had tumors of the pineal gland. These two lesions and some of the posterior fossa tumors produced obstructive hydrocephalus.

Most gangliogliomas were either isodense, hypodense, or cystic on noncontrast CT studies. Two-thirds of the lesions had at least some low-density component. Sutton et al. [12] reported that most of the lesions in their series that appeared cystic using CT actually were proven to be entirely solid pathologically. For this reason, the cystic and low-density lesions were grouped together.

Focal areas of calcification detectable by CT were identified in about one-third of the 48 patients. Enhancement with contrast material was seen in about one-half. Neither of these findings was quite as common in our series, with four patients (31%) showing calcification and five (38%) demonstrating contrast enhancement.

The currently recommended therapy of gangliogliomas is surgical excision. This is based on the belief that they are true neoplasms. In most cases, grossly total removal can be performed. Such patients probably do not require postoperative radiotherapy. In those patients in whom only a partial resection or biopsy of the tumor can be accomplished, it has been suggested that therapeutic irradiation be withheld until there is clinical progression of the disease [12, 16, 17]. Depending on the site of the lesion, most patients do well with a decrease or a remission in their symptoms, even after long-term follow-up [12, 16, 17, 19].

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