AJNR

MR and CT characteristics of gangliocytoma: a rare cause of epilepsy in children.

N R Altman

AJNR Am J Neuroradiol 1988, 9 (5) 917-921
http://www.ajnr.org/content/9/5/917

MR and CT Characteristics of Gangliocytoma: A Rare Cause of Epilepsy in Children

Nolan R. Altman[1]

Three children were examined who had surgery to remove gangliocytomas in order to control seizures that were refractory to medical therapy. The CT characteristics of gangliocytomas are those of a hyperdense lesion that does not demonstrate contrast enhancement. The MR findings show a region of mixed signal intensity on T1-weighted and proton-density images, and decreased signal intensity on T2-weighted images. Little mass effect, if any, is seen with these lesions.

The ganglion cell tumors in the CNS have been referred to as gangliogliomas, ganglioneuromas, gangliocytomas, ganglioneuroblastomas, and paragangliomas. Of these tumors, considerable overlap in definition and nomenclature occurs between the ganglioglioma and the gangliocytoma [1]. These tumors are not all the same. The term ganglioglioma was introduced by Courville in 1930 [2] and refers to a tumor that has a glial component that can show neoplastic differentiation. The gangliocytoma is a purely neuronal tumor with no glial component. No malignant change has been noted in the neurons of these tumors [3].

With the resurgence of surgical treatment for refractory epilepsy, these rare neoplasms may be encountered more frequently. We believe the MR and CT characteristics of the gangliocytoma are quite typical and may offer a preoperative diagnosis.

Materials and Methods

The MR and CT examinations of three patients at Miami Children's Hospital were studied to demonstrate the similarity between gangliocytomas and gangliogliomas. The patients (age range, 8 months to 15 years) were referred from the Department of Neurology with intractable seizures.

The CT examinations were performed with a GE 9800 scanner with and without IV contrast. MR examinations were performed on a 0.3-T Fonar B 3000 M MR scanner with a proton resonance frequency of 12.9 MHz. The images were acquired using a spin-echo (SE) multislice technique with 300–500/16, 28/4 (TR range/TE/excitations). This resulted in T1-weighted images in the sagittal, axial, and/or coronal planes. The T2-weighted images were obtained with 2000/42, 84/2. The slice thickness was from 5–7 mm with a gap of 2–3 mm. The acquisition matrix was 512 along the frequency-encoding gradient and 384 extrapolated to 512 for T1-weighed images and 310 extrapolated to 512 for T2-weighted images along the phase-encoding gradient.

Case Report

Case 1

An 8-month-old boy fell from a height of 2½ feet and struck the left frontal area of his head. There was no loss of consciousness, but 3 days after the incident his babysitter noticed that he was somewhat lethargic. A physical examination was unremarkable except for a

Received October 19, 1987; accepted after revision February 23, 1988.

[1] Department of Radiology, Miami Children's Hospital, 6125 SE 31st St., Miami, FL 33155. Address reprint requests to N. R. Altman.

AJNR 9:917–921, September/October 1988
0195–6108/88/0905–0917

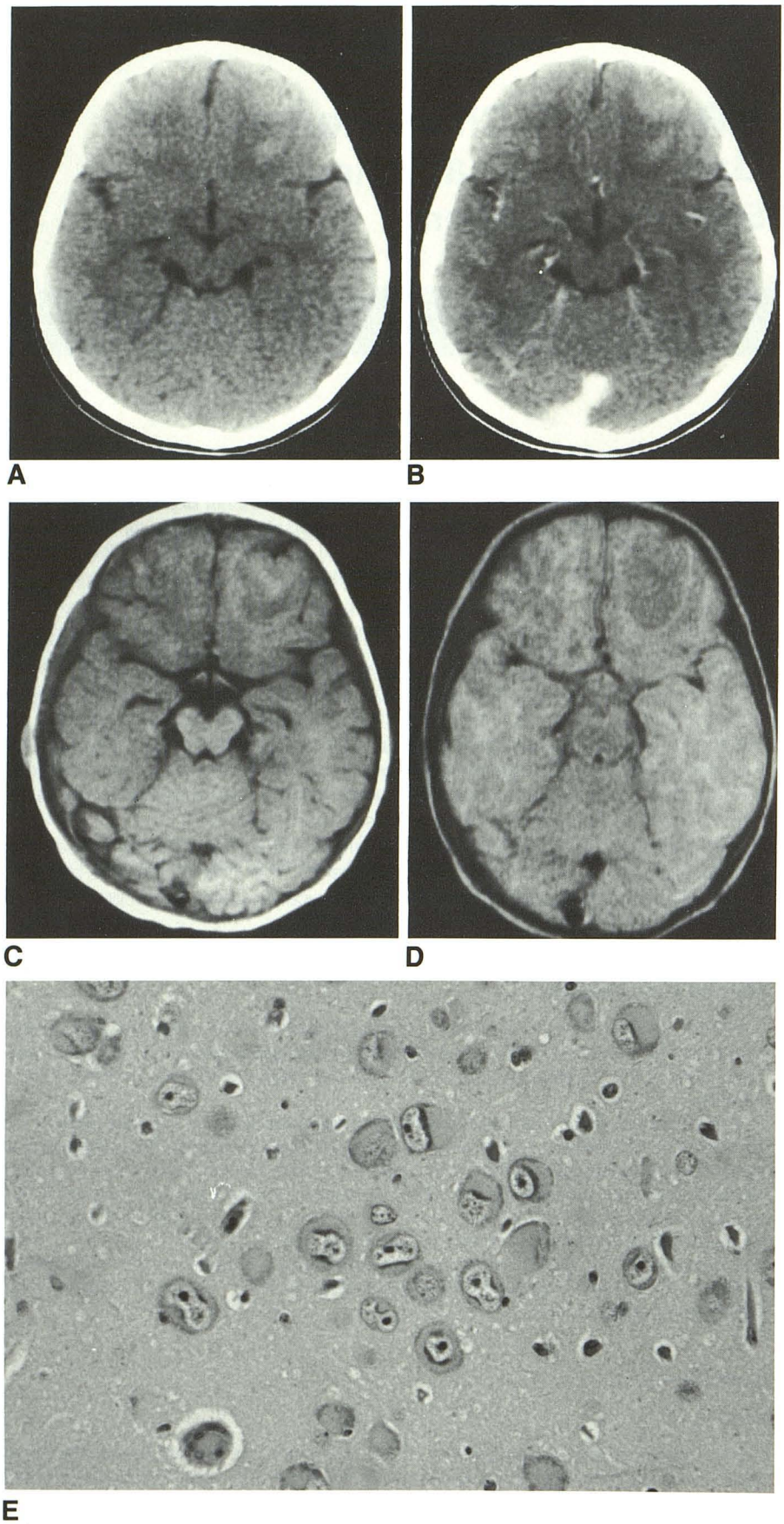

Fig. 1.—Case 1.

A, Unenhanced CT scan demonstrates area of hyperdensity in left frontal lobe. It has no significant mass effect and is surrounded by a region of density similar to that of the anterior forceps.

B, Enhanced CT scan shows no significant contrast enhancement of left frontal lobe lesion.

C, T1-weighted axial MR image, SE 438/16/4, shows left frontal lobe region with central region of intermediate signal intensity and surrounding area of decreased signal intensity. No mass effect is identified.

D, T2-weighted axial image, SE 2000/84/2, shows mass in left frontal lobe with short T2 relaxation characteristics. Incidentally noted is increased signal intensity of white matter, reflecting incomplete myelinization of immature brain, in keeping with patient's age.

E, Pathology section shows tumor composed of ganglion cells that contain large nuclei and prominent nucleoli. Some of the nuclei have an irregular dumbbell shape. (H and E ×400)

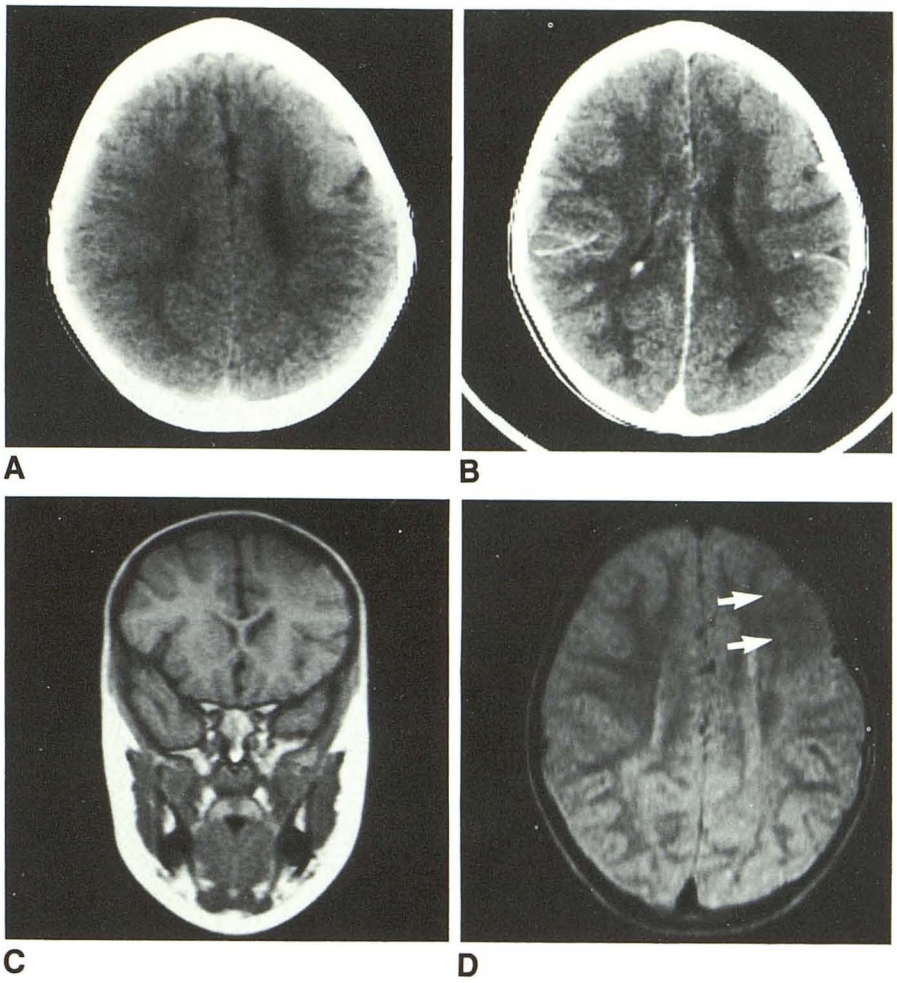

Fig. 2.—Case 2.

***A*, Unenhanced CT scan demonstrates a region of hyperdensity in left frontal lobe without significant mass effect.**

***B*, Enhanced CT scan shows no appreciable contrast enhancement.**

***C*, Coronal MR image, SE 400/16/6, demonstrates loss of normal gyration on the left. No signal abnormalities are identified.**

***D*, Axial MR image, SE 2000/84/2, identifies area of relatively short T2 relaxation in left frontal region (*arrows*).**

slight ecchymosis about the left forehead. While in the hospital, the patient developed seizures, characterized by a blank stare and eye deviation to the left, lasting approximately 30 sec. Seizure control was only minimal despite medical therapy. The EEG showed left frontal polymorphic slowing waves. CT examination revealed a hyperdense, nonenhancing left frontal lobe lesion that did not demonstrate appreciable mass effect (Figs. 1A and 1B). The MR examination revealed a left frontal lobe lesion that showed mixed T1 and short T2 relaxation characteristics (Figs. 1C and 1D). Significant mass effect was not identified. The lesion was removed surgically, and a pathologic diagnosis of gangliocytoma was made (Fig. 1E).

Case 2

A 3-year-old girl had a history of intractable seizures, which were initially myoclonic and then changed, at the age of 8 months, to their present pattern, characterized by opisthotonic posturing, blank stare, and eye deviation to the left. The seizures would last approximately 20 sec, and occur 10–50 times daily. Seizure control was dismal despite adequate doses and levels of antiepileptic drugs. The physical examination showed a left hemihypertrophy and left-sided hyperreflexia with incoordination. Routine EEGs showed a large field of interictal left-sided spikes involving the frontal, central, and temporal regions equally. CT examination demonstrated a left frontal hyperdense serpiginous lesion with an abnormal gyral pattern (Fig. 2A). No contrast enhancement was identified (Fig. 2B). MR examination showed a loss of normal gyral pattern on T1-weighted images (Fig. 2C). T2 weighting revealed an area of decreased signal intensity indicating a short T2 relaxation rate (Fig. 2D). No mass effect was identified. Surgery was performed and pathologic evaluation demonstrated a gangliocytoma. No seizures were apparent 1 year after surgery.

Case 3

A 15-year-old boy had a long history of intractable seizures, beginning at approximately 3 years of age. All the seizures were characterized by left-sided sensory abnormalities including pain, paresthesia, and dysesthesia. These were noted predominantly on the left arm and face with occasional involvement of the left leg. He also had tonic stiffening and clonic movements of the left side. There were rare grand mal seizures, and the patient had been on multiple anticonvulsant medications without success. Neurologic examination was normal. The EEG demonstrated paroxysmal discharges in the right parietal region with focal polymorphic slowing. The CT examination revealed a slightly hyperdense region in the posterior parietal region (Fig. 3A), which did not demonstrate contrast enhancement (Fig. 3B). The MR study, obtained at the same time, showed no abnormality on the T1-weighted image (Fig. 3C). An area of signal abnormality was identified on the proton-density images as an area of increased signal intensity (Fig. 3D), and a slight increase in signal was noted on the longer T2-weighted images (Fig. 3E). Surgical removal of the right parietal epileptogenic region revealed a gangliocytoma. To date, there has been no recurrence of the seizures.

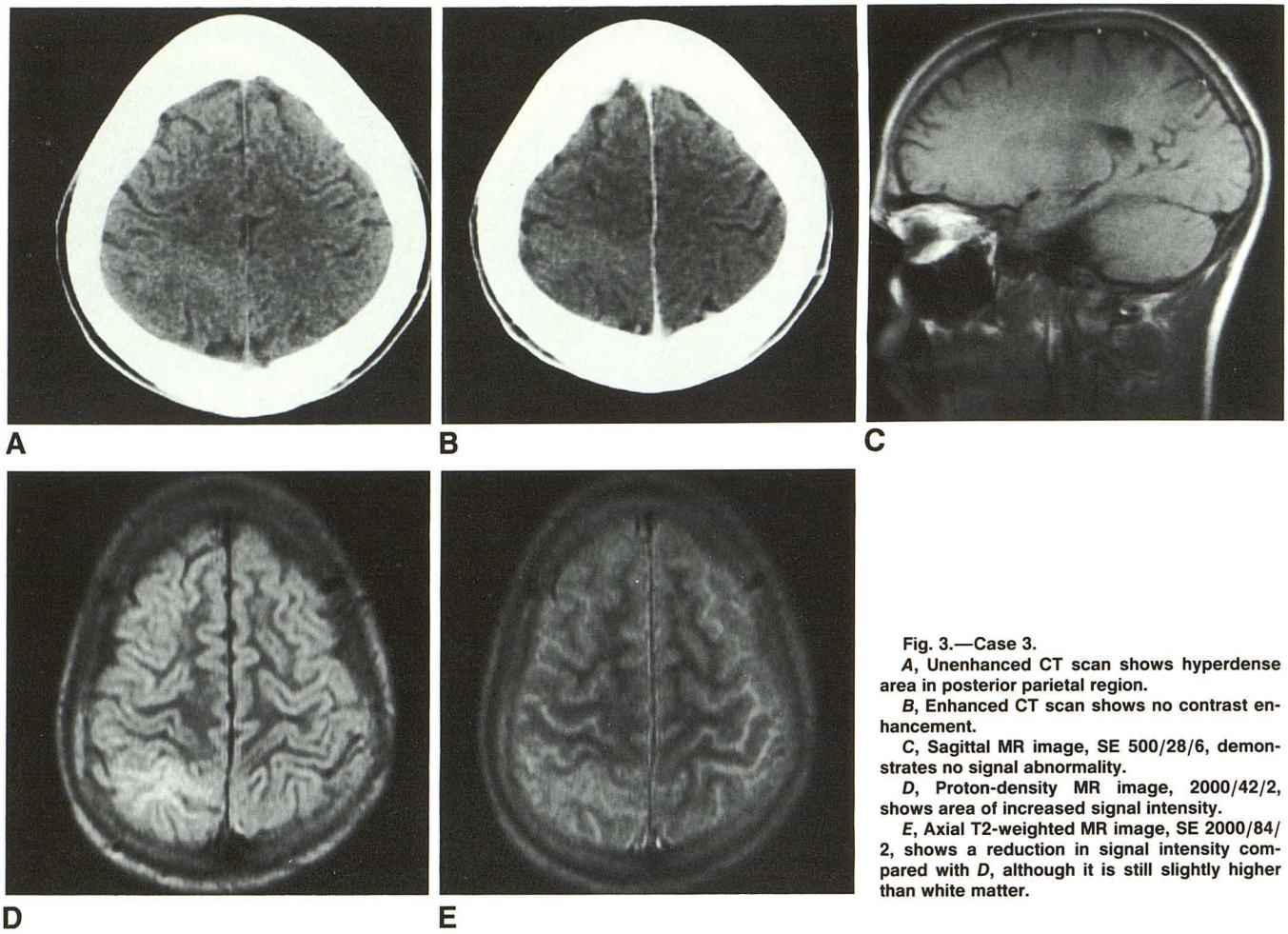

Fig. 3.—Case 3.
***A*, Unenhanced CT scan shows hyperdense area in posterior parietal region.**
***B*, Enhanced CT scan shows no contrast enhancement.**
***C*, Sagittal MR image, SE 500/28/6, demonstrates no signal abnormality.**
***D*, Proton-density MR image, 2000/42/2, shows area of increased signal intensity.**
***E*, Axial T2-weighted MR image, SE 2000/84/2, shows a reduction in signal intensity compared with *D*, although it is still slightly higher than white matter.**

Discussion

In the evaluation of children with intractable seizures, the newer, noninvasive imaging techniques of CT and MR, along with a renewed interest in surgical resection for treatment, has, in certain cases, provided radiologists with the means to discover preoperative clues to the underlying origin of epileptogenic foci. Recently, numerous articles have appeared comparing the ability of these techniques to detect underlying causes of epileptic foci [4, 5].

Rarely, supratentorial gangliocytomas have appeared in the literature as causes of epileptic foci [6–8]. These neuronal tumors have also been observed in the cerebellum, as first described by Lhermitte and Duclos [9] and by others, who have reported this lesion to be a dysplastic gangliocytoma of the cerebellum [10], which may represent a congenital abnormality in granule cell migration [11]. The lesions we described of the cerebrum may also represent some type of focal dysplasia of the cerebral cortex as described by Taylor et al. [12]. Others have reported gangliocytomas of the pineal [13].

These tumors are extremely rare, with a reported frequency of only 0.1% [13]. The tumor is composed of ganglion cells that stain negative with glial fibrillary acetic protein (GFAP). There is a slight male prediliction, as indicated in our cases.

There are only a few reports of CT findings of gangliocytomas [7, 8, 13]. No reports of the MR appearance have been found in the literature to date. Our CT findings in all three cases were quite similar; that is, a slightly hyperdense region with little if any contrast enhancement. No mass effect was identified in any case. This finding should be differentiated from those reported cases of gangliogliomas in which cyst formation, calcifications, contrast enhancement, and mass effect were quite common [14]. Our CT findings are much more "benign" in appearance than those of the gangliogliomas, just as the tumors appeared to be pathologically.

The MR characteristics are also quite interesting. These lesions are difficult to identify on T1-weighted images, as was demonstrated in two of our cases. If any signal abnormality is identified, it is mixed, as in our case 1. On the balanced proton-density images, the signal was from intermediate to high. On T2-weighted images, the signal was intermediate in one case and low in the other two cases. The MR findings relate to the microscopic description of the lesions. They are

composed of ganglion cells on a background of neuropil and can look quite like normal brain or a hamartomatous lesion. This may explain the difficulty in demonstrating these lesions on T1-weighted images. The signal abnormality on T2 may be the result of large nuclei and prominent nucleoli with long-chain nucleic acids, which can increase the T2 relaxation rate if these areas are particularly dense. The two cases in which the T2 relaxation rate was increased occurred in younger children, which may be a factor of the parenchymal lesion relative to the immature brain. Few, if any other, neoplasms that we encountered showed these findings, making these lesions quite distinctive. Confusion can occur with a hemorrhagic contusion, as in case 1, in which there was a presenting history of trauma. However, follow-up examinations have revealed no change in these lesions on CT or MR, which would tend to rule out this differential possibility. Most recent reports on the MR findings in epileptic patients cite the various lesions as having long T2 relaxation times [4, 5], thereby further singling out these lesions.

In summary, we believe that the preoperative diagnosis of gangliocytoma can be made fairly accurately because of the relatively hyperdense, benign CT appearance and the characteristic short T2 relaxation times demonstrated on MR imaging. These extremely rare tumors are benign and should be differentiated from their more aggressive counterparts, the ganglioglioma.

ACKNOWLEDGMENT

I thank Patricia Parker, Department of Pathology, Miami Children's Hospital, for providing the photomicrograph and for helping with the pathologic description.

REFERENCES

1. Kernahan JW, Learmonth JR, Doyle JB. Neuroblastomas and gangliocytomas of the central nervous system. *Brain* **1932**;55:287–310
2. Courville CB. Ganglioglioma tumor of the central nervous system: review of the literature and report of two cases. *Arch Neurol Psychiatr* **1930**;24:439–491
3. Rubinstein LJ, Herman MH. A light and electron microscopic study of a temporal lobe ganglioglioma. *J Neurol Sci* **1972**;16:27–48
4. Schorner W, Meencke HJ, Felix R. Temporal lobe epilepsy: comparison of CT and MR imaging. *AJNR* **1987**;8:773–781
5. Ormson MJ, Kispert DB, Sharbrough FW, et al. Cryptic structural lesions in refractory partial epilepsy: MR imaging and CT studies. *Radiology* **1986**;160:215–219
6. Dom R, Brucher JM. Hamartoblastome (gangliocytome diffus) unilateral de l'ecorce cerebrale associe. A une degenerescence soudanophile de la substance blanche du dote oppose. *Rev Neurol* (*Paris*) **1969**;120:5:307–318
7. Kosaka H, Hidetaka W, Fuma I, Igawa T. Gangliocytoma. Case report and review of the literature. *Neurol Med Chir* (*Tokyo*) **1984**;24:257–264
8. Kawamoto K, Yamanouchi Y, Suwa J, Kurimoto T, Matsumura H. Ultrastructural study of a cerebral gangliocytoma. *Surg Neurol* **1985**;24:541–549
9. Lhermitte J, Duclos P. Sur un ganglioneurone diffus du cortex du cervelet. *Bull Cancer* (*Paris*) **1920**;94(5):99–107
10. Oppenheimer DR. A benign "tumor" of the cerebellum. Report on two cases of diffuse hypertrophy of the cerebellar cortex with a review of nine previously reported cases. *J Neurol Neurosurg Psychiatry* **1955**;18:199–213
11. Roski RA, Roessmann U, Spetzler RF, Kaufman B, Nulsen FE. Clinical and pathological study of dysplastic gangliocytoma. A case report. *J Neurosurg* **1981**;55:318–321
12. Taylor DC, Falcone MA, Bruton CJ, Corsellis JAN. Focal dysplasia of the cerebral cortex in epilepsy. *J Neurol Neurosurg Psychiatry* **1971**;34:369–387
13. Ebina K, Suzuki S, Takahashi T, Iwabuchi T, Takei Y. Gangliocytoma of the pineal body. A case report and review of the literature. *Acta Neurochir* (*Wien*) **1985**;74(3–4):134–140
14. Kazner E, Wende S, Grumme TH. *Computed tomography in intracranial tumors*. New York: Springer-Verlag, **1982**:143–147