

MR Imaging and Histopathologic Findings of A Case of Cerebral Ganglioneurocytoma

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We report a case of ganglioneurocytoma manifesting as a complex partial seizure in a young adult male. MR images depicted a well-marginated cystic mass with a heterogeneous solid portion abutting the dura in the parietal lobe. The solid portion showed minimal heterogeneous enhancement, and pressure erosion of the overlying calvarium had occurred. Following gross total resection, the clinical outcome was satisfactory, with no further seizures, and during the five-year follow-up period, the tumor did not recur.

Ganglioneurocytoma is a very rare variant of neuronal tumor and is characterized by differentiation toward neurocytes and ganglion cells. The histopathological characteristics of ganglioneurocytoma match those of central neurocytoma, except that the former shows ganglioid differentiation, frequently forms a cystic lesion, and arises extraventricularly (1). We describe a case of ganglioneurocytoma involving the left parietal lobe and discuss the definition and histogenesis of this rare tumor.

CASE REPORT

A 23-year-old man whose birth and postnatal development were uneventful presented with a longstanding history of complex partial seizures, first experienced at the age of two and subsequently occurring once or twice yearly. He was alert and of normal intelligence, and physical examination revealed no neurologic abnormalities.

MR images indicated that in the left parietal lobe, a large, thin-walled cyst with a solid portion was present. The signals of the cystic portion were hypointense / hyperintense at T1-/ T2-weighted imaging, respectively, similar to those of cerebrospinal fluid (CSF) (Figs. 1A, B). The solid portion demonstrated heterogeneous mixed signal intensity and was located along the cortex, with erosion of the overlying calvarium. At FLAIR (fluid attenuated inversion recovery) imaging (Fig. 1C), the cystic portion also showed low signal intensity, similar to that of CSF, and heterogeneous high signal intensity of the solid portion was observed, with a lobulating contour. Gd-enhanced T1-weighted imaging revealed minimal enhancement of the solid portion (Figs. 1D, E), and there was no evidence of leptomeningeal or intraventricular seeding. The preoperative diagnosis was low-grade glioma, such as oligodendroglioma.

Gross total resection of the mass was performed using stereotactic instruments and intraoperative ultrasonography, and the tumor, together with a minimal amount of surrounding brain tissue, was removed. The solid portion was brownish in color and of relatively firm consistency. The patient's post-operative course was uneventful. Microscopically, the tumor was composed of small round cells with clear cytoplasm,

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which were lobulated by a well-developed vascular network mimicking the histology of central neurocytoma or oligodendroglioma. Ganglioid differentiation was noted throughout the tumor and was represented by scattered individual ganglion cells, or groups of these, among small round cells (Fig. 1F). The tumor cells were embedded in a

neurophil-like fibrillary background which was strongly immunoreactive for synaptophysin (Fig. 1G), and GFAP (glial fibrillary acidic protein) immunostaining disclosed that among them, numerous, large reactive astrocytes with stellate processes were present. All these histopathologic findings indicated the presence of a ganglioneurocytoma.

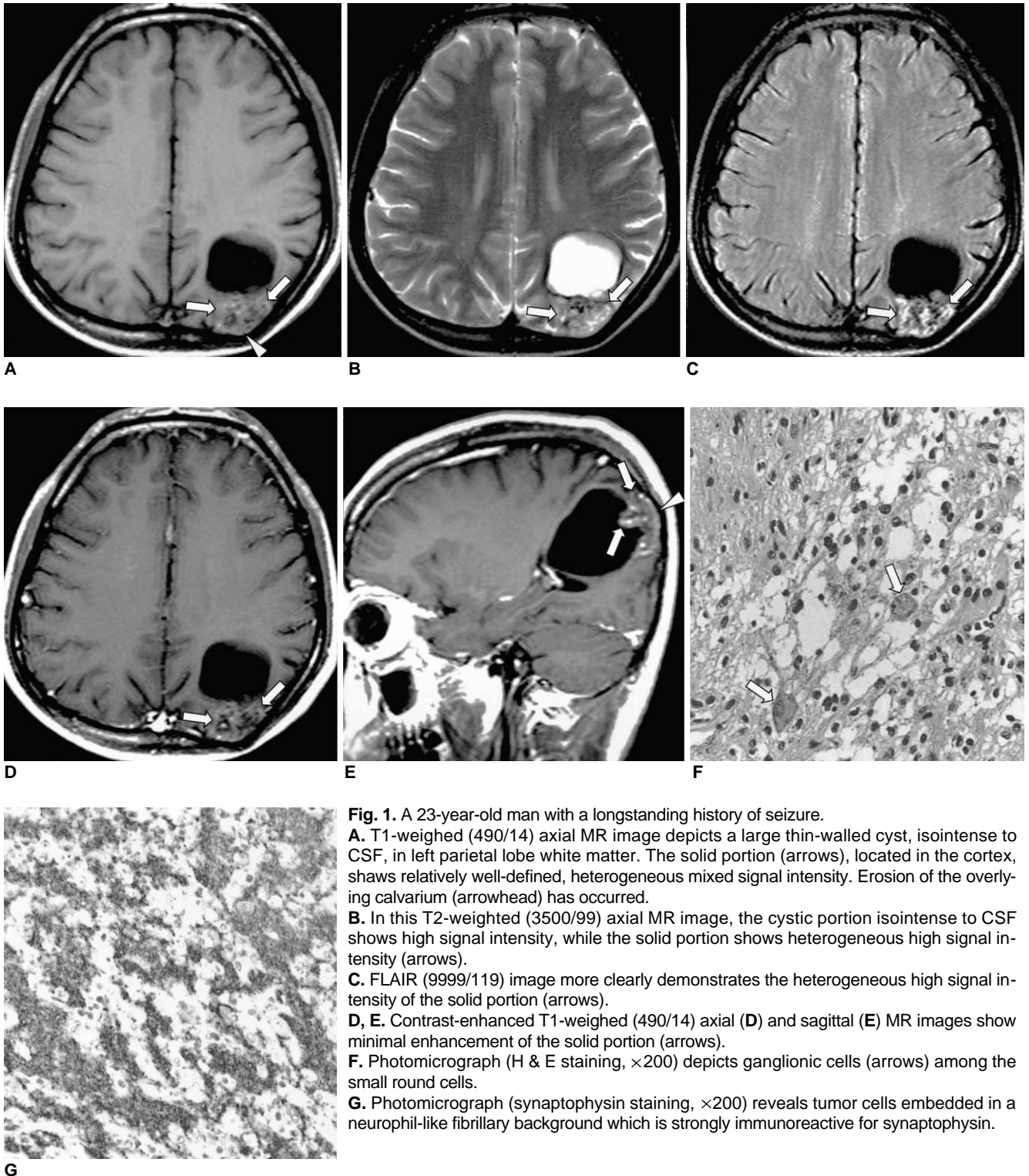


Fig. 1. A 23-year-old man with a longstanding history of seizure.
A. T1-weighted (490/14) axial MR image depicts a large thin-walled cyst, isointense to CSF, in left parietal lobe white matter. The solid portion (arrows), located in the cortex, shows relatively well-defined, heterogeneous mixed signal intensity. Erosion of the overlying calvarium (arrowhead) has occurred.
B. In this T2-weighted (3500/99) axial MR image, the cystic portion isointense to CSF shows high signal intensity, while the solid portion shows heterogeneous high signal intensity (arrows).
C. FLAIR (9999/119) image more clearly demonstrates the heterogeneous high signal intensity of the solid portion (arrows).
D, E. Contrast-enhanced T1-weighted (490/14) axial (**D**) and sagittal (**E**) MR images show minimal enhancement of the solid portion (arrows).
F. Photomicrograph (H & E staining, $\times 200$) depicts ganglionic cells (arrows) among the small round cells.
G. Photomicrograph (synaptophysin staining, $\times 200$) reveals tumor cells embedded in a neurophil-like fibrillary background which is strongly immunoreactive for synaptophysin.

Surgery was performed five years ago, and the seizures have subsequently shown complete remission. MR images obtained one year ago revealed no residual tumor, and at that time, no tumor recurrence was noted.

DISCUSSION

Neuronal tumors of the central nervous system are known to show wide morphologic variability, and hence their histologic diagnosis is based on the degree of differentiation of their neuronal elements as well as the relative proportion of neuronal to glial elements within a single tumor (2, 3). Because of the difficulties in diagnosis due to the variability encountered at pathologic examination, this group of tumors has, however, been poorly understood, and there is still considerable controversy regarding their exact classification and nomenclature. A central neurocytoma is characterized by its intraventricular location, a small uniform neoplastic cell population with features of neuronal differentiation, and the absence of any ganglionic neuronal cells (4, 5). In recent years, however, several cases of central neurocytomas with an extraventricular location and ganglioid differentiation have been reported (1, 5-8). Hence, this tumor is composed at least of two distinctive cell types: large ganglionic and small round cells, both showing the morphologic characteristics of neuronal cells. Cases of ganglioneurocytoma were first reported by Nishio et al. (5), in 1988, and von Deimling et al. (8), in 1990, but few radiologic descriptions of these tumors are available.

Patients who present with the symptoms of ganglioneurocytoma are mostly children or adults aged less than thirty. As in our case, the symptoms in the reported cases were nonspecific and varied according to the location of the tumor. They included seizure, increased intracranial pressure, neurologic deficit, and headache (1, 5-7). In our patient, the presenting symptom was longstanding partial complex seizure, and in view of his long history and the pressure erosion of the calvarium revealed by MR images, the tumor was presumed to be slow-growing and benign.

The MR imaging findings for this tumor are described in only two case reports. Funato et al. (1) stated that Gd-enhanced T1-weighted MR imaging revealed a large cyst with an enhanced mural nodule, though did not mention T2-weighted MR imaging. The case reported by Chan et al. (6) involved a patient in whom a heterogeneous hyperintense mass was revealed by T2-weighted MR imaging, and extensive, heterogeneous enhancement by Gd-enhanced T1-weighted MR imaging; the moderate mass effect mimicked a malignant tumor. The MR imaging findings in our case included those of both previous reports: the tumor had a large cystic area as well as a heterogeneous solid compo-

nent. The degree of enhancement, however, was less than previously reported, and it thus appears that this varies in both ganglioneurocytoma and other neuronal tumors. The reported CT findings, namely a large cystic mass with an enhancing solid nodule and occasional calcification, did not differ from those of other neuronal tumors (1, 5-7). Although our patient did not undergo CT scanning, we inferred from MR imaging findings that the CT findings would be similar to those of previous reports.

Differentiation between this tumor and low-grade gliomas or ganglion cell tumors is difficult or even impossible. Low-grade gliomas including oligodendroglioma can show similar imaging findings to those of this present case, though in a glioma a large cystic component is uncommon. Ganglion cell tumors such as ganglioglioma or gangliocytoma have been described as cystic masses with a variously enhancing solid portion, the so-called mural nodule, and occasional calcification (9, 10). In addition, it is important to distinguish ganglioneurocytomas from neoplasms containing neuroblastic elements: the latter have a less favorable prognosis, and ganglioneurocytoma can sometimes mimic a malignant neoplasm (6).

Complete surgical excision is the treatment of choice, though post-operative radiotherapy is a possible adjunctive treatment (5, 7). Since, in our patient, there was no evidence of recurrence after four years, nor after seven years in a patient described in a previous report (5), this tumor is thought to have a favorable prognosis.

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References

1. Funato H, Inoshita N, Okeda R, Yamamoto S, Aoyagi M. Cystic ganglioneurocytoma outside the ventricular region. *Acta Neuropathol* 1997;94:95-98
2. Nishio S, Takeshita I, Fukui M. Primary cerebral ganglioneurocytoma in an adult. *Cancer* 1990;66:358-362
3. Shimada H. Transmission and scanning electron microscopic studies on the tumors of neuroblastoma group. *Acta Pathol Jpn* 1982;32:415-426
4. Chang KH, Han MH, Kim DG, et al. MR appearance of central neurocytoma. *Acta Radiol* 1993;34:520-526
5. Nishio S, Tashima T, Takeshita I, Fukui M. Intraventricular neurocytoma: clinicopathological features of six cases. *J Neurosurg* 1988;68:665-670
6. Chan A, McAbee G, Queenan J, Manning A. Ganglioneurocytoma mimicking a malignant tumor: case report with a literature review of the MRI appearance of neurocytomas and gangliogliomas. *J Neuroimaging* 2001;11:47-50
7. Biernat W, Zakrzewski K, Liberski PP. Twelve-year-old boy with recent onset seizures. *Brain Pathol* 2000;10:313-314, 319
8. von Deimling A, Janzer R, Kleihues P, Wiestler OD. Patterns of differentiation in central neurocytoma: an immunohistochemical study of eleven biopsies. *Acta Neuropathol* 1990;79:473-479

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9. Castillo M, Davis PC, Takei Y, Hoffman JC Jr. Intracranial ganglioglioma: MR, CT, and clinical findings in 18 patients. *Am J Neuroradiol* 1990;11:109-114
10. Kim HS, Lee HK, Jeong AK, Shin JH, Choi CG, Khang SK. Supratentorial gangliocytoma mimicking extra-axial tumor: a report of two cases. *Korean J Radiol* 2001;2:108-112