RadioGraphics

Neuronal Tumors of the Central Nervous System: Radiologic Findings and Pathologic Correlation[1]

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LEARNING OBJECTIVES

After reading this article and taking the test, the reader will be able to:

- Discuss the importance of distinguishing neuronal tumors from glial tumors.
- Describe the pathologic classification of neuronal tumors and the pathologic basis of the radiologic findings in neuronal tumors.
- Recognize the characteristic imaging features of many neuronal tumors for accurate diagnosis and preoperative planning.

Pure neuronal and mixed neuronal-glial tumors of the central nervous system are uncommon but fascinating because they are less aggressive than the more common glial tumors and their prognosis is excellent. Neurologic manifestations are varied and include seizures, symptoms of increased intracranial pressure, and neurologic deficits according to tumor location. Many neuronal tumors of the central nervous system demonstrate characteristic radiologic findings. At magnetic resonance (MR) imaging, gangliocytomas demonstrate low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and frequent enhancement on gadolinium-enhanced T1-weighted images. Characteristic MR imaging findings of Lhermitte-Duclos disease are a nonenhancing mass in a cerebellar hemisphere with a striated pattern. Central neurocytomas are typically located in the lateral ventricles near the foramen of Monro with a characteristic attachment to the septum pellucidum. Ganglioneurocytoma is a rare variant of central neurocytoma that is characterized by differentiation toward ganglion cells. In ganglioglioma, a well-defined cystic mass with a solid mural nodule is typically seen. Extension of enhancement to the leptomeninges is characteristic of desmoplastic infantile ganglioglioma and correlates with the firm dural attachment of the solid component. Dysembryoplastic neuroepithelial tumor has a well-demarcated, multilobulated or gyriform appearance.

Index terms: Brain neoplasms, diagnosis, 10.364 • Nervous system, neoplasms, 10.364 • Neurocytoma, 10.364 • Neuroma, 10.364

RadioGraphics 2002; 22:1177–1189

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Introduction

Tumors of the central nervous system that contain abnormal neuronal elements, termed *neuronal tumors*, make up approximately 1% of all brain tumors. Distinguishing neuronal tumors from the more common glial tumors is important because neuronal tumors have favorable clinical outcomes and are generally cured with surgery alone, whereas gliomas typically require radiation therapy or chemotherapy depending on their histologic grade. At histopathologic analysis, neuronal tumors are usually classified as pure neuronal cell tumors (gangliocytoma, Lhermitte-Duclos disease [dysplastic cerebellar gangliocytoma], central neurocytoma) and mixed neuronal-glial tumors (ganglioglioma, desmoplastic infantile ganglioglioma, dysembryoplastic neuroepithelial tumor, ganglioneuroma) (Table 1) (1).

In this article, we review the clinical and pathologic characteristics of the various neuronal tumors of the central nervous system and demonstrate representative characteristic magnetic resonance (MR) imaging findings.

Table 1
Pathologic Classification and Distribution of Neuronal and Neuronal-Glial Cell Tumors in a Series of 33 Cases

Type of Tumor	No. of Cases
Neuronal cell	15
Gangliocytoma	3
Lhermitte-Duclos disease*	2
Central neurocytoma	9
Ganglioneurocytoma	1
Mixed neuronal-glial	18
Ganglioglioma	14
Desmoplastic infantile ganglioglioma	1
Dysembryoplastic neuroepithelial tumor	3
Ganglioneuroma	0

*Dysplastic cerebellar gangliocytoma.

Clinical Presentation

In a retrospective series of 33 patients with pathologically proved neuronal tumors diagnosed between January 1996 and March 2001, seizure was the primary clinical presentation ($n = 19$) followed by symptoms of increased intracranial pressure ($n = 7$), nonspecific headache ($n = 5$), and neurologic deficit ($n = 2$) (Table 2). Nonspecific headache was defined as headache without nausea and vomiting, papilledema, or raised opening or closing pressure at lumbar puncture; thus, it was unrelated to increased intracranial pressure. In gangliocytomas ($n = 3$), the primary clinical presentations varied and included seizure ($n = 1$), nonspecific headache ($n = 1$), and neurologic deficit ($n = 1$). Two patients with Lhermitte-Duclos disease had nonspecific headache. In central neurocytomas ($n = 9$), most patients ($n = 7$) showed symptoms of increased intracranial pressure, such as headache, nausea, and papilledema, whereas two patients had nonspecific headache. In one patient with ganglioneurocytoma, the lesion was in the parietal lobe and seizure was the clinical presentation. In gangliogliomas ($n = 14$), most of the lesions were located in the cortex of the temporal lobe ($n = 13$) and seizure was the main clinical manifestation in all patients. In dysembryoplastic neuroepithelial tumor ($n = 3$), all three lesions were located in the temporal lobe and seizure was also the main clinical manifestation in all three patients. One patient with desmoplastic infantile ganglioglioma showed neurologic deficit as the main clinical presentation.

Therefore, according to the locations of the tumors, neurologic manifestations were varied and included seizure, symptoms of increased intracranial pressure, nonspecific headache, and neurologic deficit. It is interesting that Lhermitte-Duclos disease, central neurocytoma, ganglioglioma, and dysembryoplastic neuroepithelial tumor had only one or two kinds of primary clinical symptoms; thus, their clinical presentations would help narrow the possible differential diagnoses.

Table 2
Histopathologic Distribution of Neuronal and Neuronal-Glial Cell Tumors according to the Primary Clinical Manifestation in a Series of 33 Cases

Primary Clinical Manifestation	No. of Cases by Tumor Type
Seizure ($n = 19$)	1 of 3 gangliocytomas 1 of 1 ganglioneurocytoma 14 of 14 gangliogliomas 3 of 3 dysembryoplastic neuroepithelial tumors
Increased intracranial pressure ($n = 7$)	7 of 9 central neurocytomas
Nonspecific headache ($n = 5$)	1 of 3 gangliocytomas 2 of 2 cases of Lhermitte-Duclos disease 2 of 9 central neurocytomas
Neurologic deficit ($n = 2$)	1 of 3 gangliocytomas 1 of 1 desmoplastic infantile ganglioglioma

Pure Neuronal Cell Tumors

Gangliocytoma

The term *gangliocytoma* denotes a spectrum of rare tumors in which neuronal cell lineages are the sole neoplastic constituents (1). In addition to the neuronal population, there is a highly variable normocellular network of nonneoplastic glial cells (2). This distinguishes gangliocytomas from gangliogliomas, which also contain anaplastic glial cells. But clear-cut separation of gangliocytomas from gangliogliomas is difficult. They represent opposite ends of a spectrum of differentiated ganglion cell tumors. Since these two entities are not always distinct, the term *ganglion cell tumor* is a useful compromise in which the neoplastic nature of the glial component remains in doubt (3). Most lesions appear to become clinically symptomatic in children and young adults. The most common sites are the cerebral hemispheres and the cervicothoracic spinal cord (2). Within the cerebral hemispheres, the temporal lobe, either alone or in combination with the frontal or parietal lobes, appears to be the favored site (1). Other intracranial locations include the cerebellum, the hypothalamus adjacent to the floor of the third ventricle, the pineal region, and the pituitary. Patients typically present with seizures or focal neurologic signs according to the location and the resultant mass effect.

MR imaging of gangliocytomas reveals masses of low signal intensity on T1-weighted images, high signal intensity on T2-weighted images, and frequent enhancement on gadolinium-enhanced T1-weighted images (Figs 1, 2) (3–6). However, Altman et al (7) reported that gangliocytomas showed mixed signal intensity on T1-weighted images and low signal intensity on T2-weighted images. Calcification and cyst formation are frequently revealed on CT or MR images (3,5). Therefore, as with the histologic findings, imaging findings of gangliocytomas can mimic those of ganglioglioma. Some cases are cortically located, are accompanied by the dural tail sign, and can mimic meningioma (Fig 2) (3).

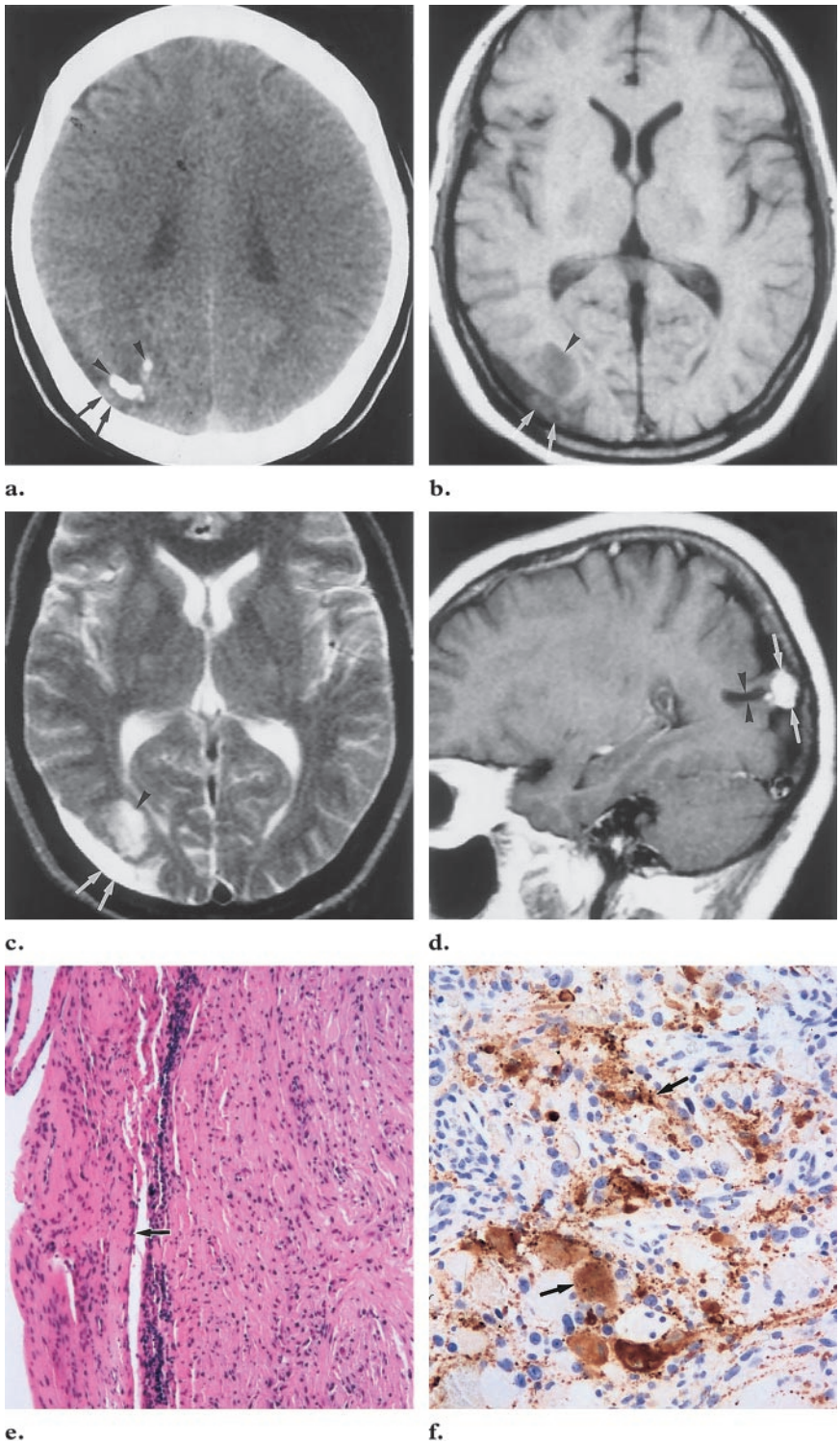
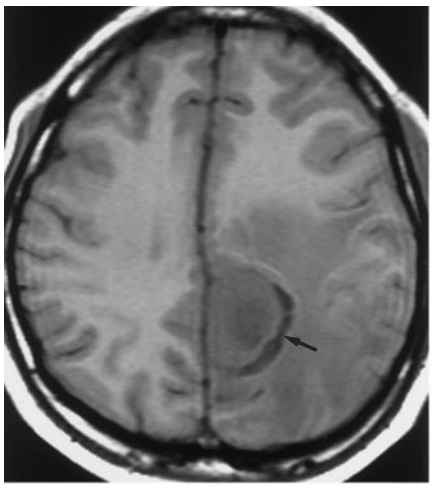


Figure 1. Gangliocytoma involving the right occipital lobe in a 59-year-old woman with headaches and dizziness. **(a)** Nonenhanced computed tomographic (CT) image shows focal calcification (arrowheads) in a tumor located in the cortex with an exophytic component (arrows). **(b, c)** Axial T1-weighted **(b)** and T2-weighted **(c)** MR images show that the tumor (arrows) is small and exophytic with slightly low signal intensity on the T1-weighted image **(b)** and high signal intensity on the T2-weighted image **(c)**. Discrimination between the exophytic component and surrounding cerebrospinal fluid is difficult on the T2-weighted image **(c)**. Widening of the adjacent cisternal space is noted. There is an associated cyst (arrowhead), which has low signal intensity on the T1-weighted image **(b)** and high signal intensity on the T2-weighted image **(c)**. **(d)** Sagittal gadolinium-enhanced T1-weighted MR image shows that the tumor (arrows) is located in the cortex, has an exophytic growth pattern, and demonstrates homogeneous marked enhancement. The cyst (arrowheads) is located medial to the tumor. **(e)** Photomicrograph (original magnification, ×40; hematoxylin-eosin stain) shows that the tumor is attached to the dura mater (arrow). **(f)** Photomicrograph (original magnification, ×200; synaptophysin stain) shows positive staining of the cytoplasm and the process of some ganglion cells with synaptophysin (arrows).



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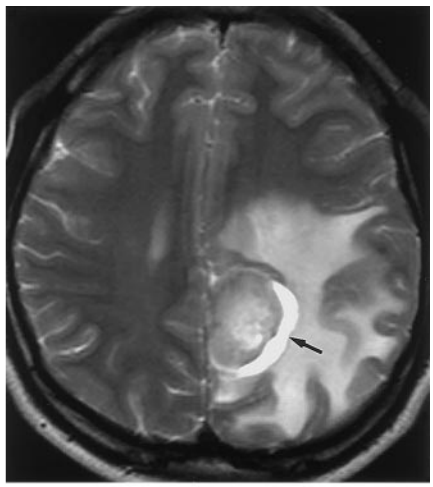

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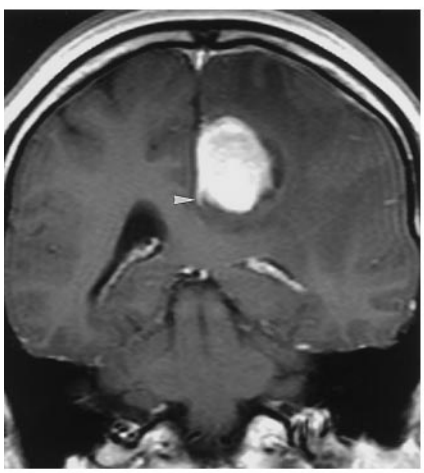

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Figure 2. Gangliocytoma involving the left parasagittal area in a 22-year-old woman with right hemiparesis. **(a)** Axial T1-weighted MR image shows a well-demarcated tumor in the left parasagittal area. The tumor is of slightly low signal intensity and has a broad dural base on the falx cerebri. There is a crescentic peritumoral cyst (arrow). **(b)** Axial T2-weighted MR image shows that the tumor has high signal intensity with marked peritumoral edema. The cyst is seen as high signal intensity (arrow). **(c)** Coronal gadolinium-enhanced T1-weighted MR image shows homogeneous marked enhancement with the dural tail sign (arrowhead).

Lhermitte-Duclos Disease

Lhermitte-Duclos disease (dysplastic cerebellar gangliocytoma) is a rare disorder of uncertain pathogenesis characterized by distortion of the normal cerebellar laminar cytoarchitecture (8). Gross autopsy specimens show expanded cerebellar folia. Microscopic findings include thickening and hypermyelination of the molecular (outermost) layer and large pleomorphic cells that replace the Purkinje (middle) and granular (innermost) layers (1,8). The association of Lhermitte-Duclos disease and Cowden syndrome, an autosomal dominant phakomatosis, has been described, suggesting that Lhermitte-Duclos disease fits the concept of Cowden syndrome as a hamartoma-neoplasia syndrome (9). Most cases of Lhermitte-Duclos disease occur in adults, usually in the third and fourth decades. There is no definite sex preference.

Characteristic MR imaging findings are a nonenhancing mass in the cerebellar hemisphere with an alternating striated pattern of corresponding low signal intensity on T1-weighted images and high signal intensity on T2-weighted images (Fig 3) (8,10,11). It is usually located in the cerebellar hemisphere with special preference for the left side and occasionally extends to the vermis. Hydrocephalus or syrinx may be present due to mass effect along with compression of the fourth ventricle. CT demonstrates a noncalcified low-attenuation cerebellar mass and occasionally a mixed-attenuation cerebellar mass with occasionally scattered calcification. However, CT findings lack specific features to suggest the diagnosis. Correlation of the MR imaging findings with histologic and gross pathologic features of Lhermitte-Duclos disease shows that the characteristic striations demonstrated on MR images represent the abnormally thickened cerebellar folia (8,11).

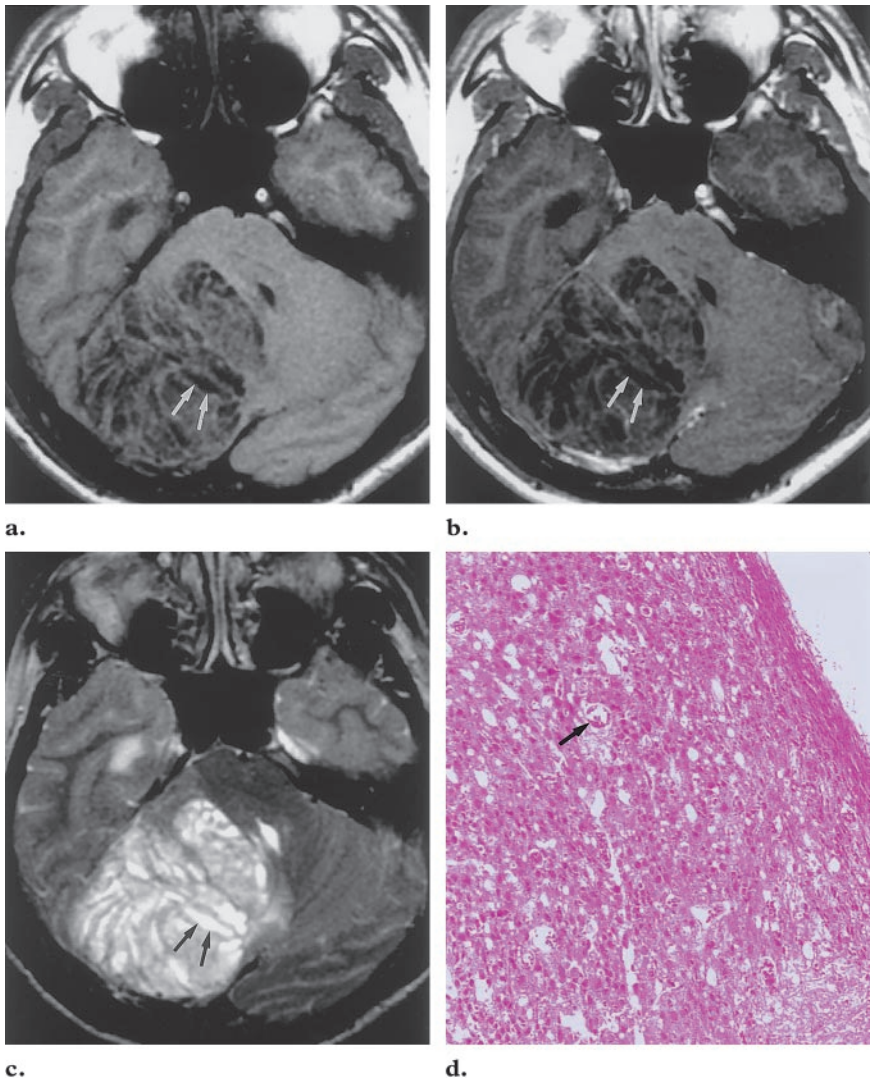


Figure 3. Lhermitte-Duclos disease involving the right cerebellum in a 24-year-old man with headaches. **(a, b)** Axial T1-weighted **(a)** and gadolinium-enhanced T1-weighted **(b)** MR images show roughly parallel linear bands of low signal intensity (arrows) without enhancement. **(c)** Axial T2-weighted MR image shows an alternating striated pattern of high signal intensity (arrows), which represents enlarged cerebellar folia. **(d)** Photomicrograph (original magnification, ×100; hematoxylin-eosin stain) shows abnormal, enlarged ganglion cells (arrow) that distort and replace the normal cerebellar architecture.

Central Neurocytoma and Ganglioneurocytoma

Histologically, central neurocytoma is characterized by a small, uniform neoplastic cell population with features of neuronal differentiation (12). Its characteristic feature is artificial perinuclear halo formation. This feature creates a remarkable likeness to oligodendrogliomas. Before the neuronal immunohistochemical and ultrastructural characteristics of central neurocytoma were recognized, it was usually termed an *intraventricular oligodendroglioma* or an *ependymoma of the foramen of Monro* (13). Immunohistochemical studies show consistent and uniform expression of the neuronal marker proteins, neuron-specific enolase, and synaptophysin. Ultrastructural examination shows neuronal characteristics such as microtubules, neurosecretory granules, clear vesicles, and sometimes even synapses. This tumor affects mainly young adults and has a favorable prognosis. It is typically located in the lateral ventricles near the foramen of Monro with a characteristic attachment to the septum pellucidum (14). Its clinical history is short, and the majority of patients present with symptoms of increased intracranial pressure secondary to obstructive hydrocephalus.

At MR imaging, central neurocytoma is inhomogeneously isointense on T1-weighted images (Fig 4). Signal intensity on T2-weighted images is variable but is mostly isointense to hyperintense with the cerebral cortex (12,14,15). Areas of low signal intensity or absent signal on both T1- and T2-weighted images can represent calcification, cyst, hemorrhage, and tumor vessels (12,14,15). At CT, the lesion is isoattenuating or slightly hyperattenuating and is a well-demarcated, lobulated mass with mild to moderate, inhomogeneous enhancement. Calcification is seen in about

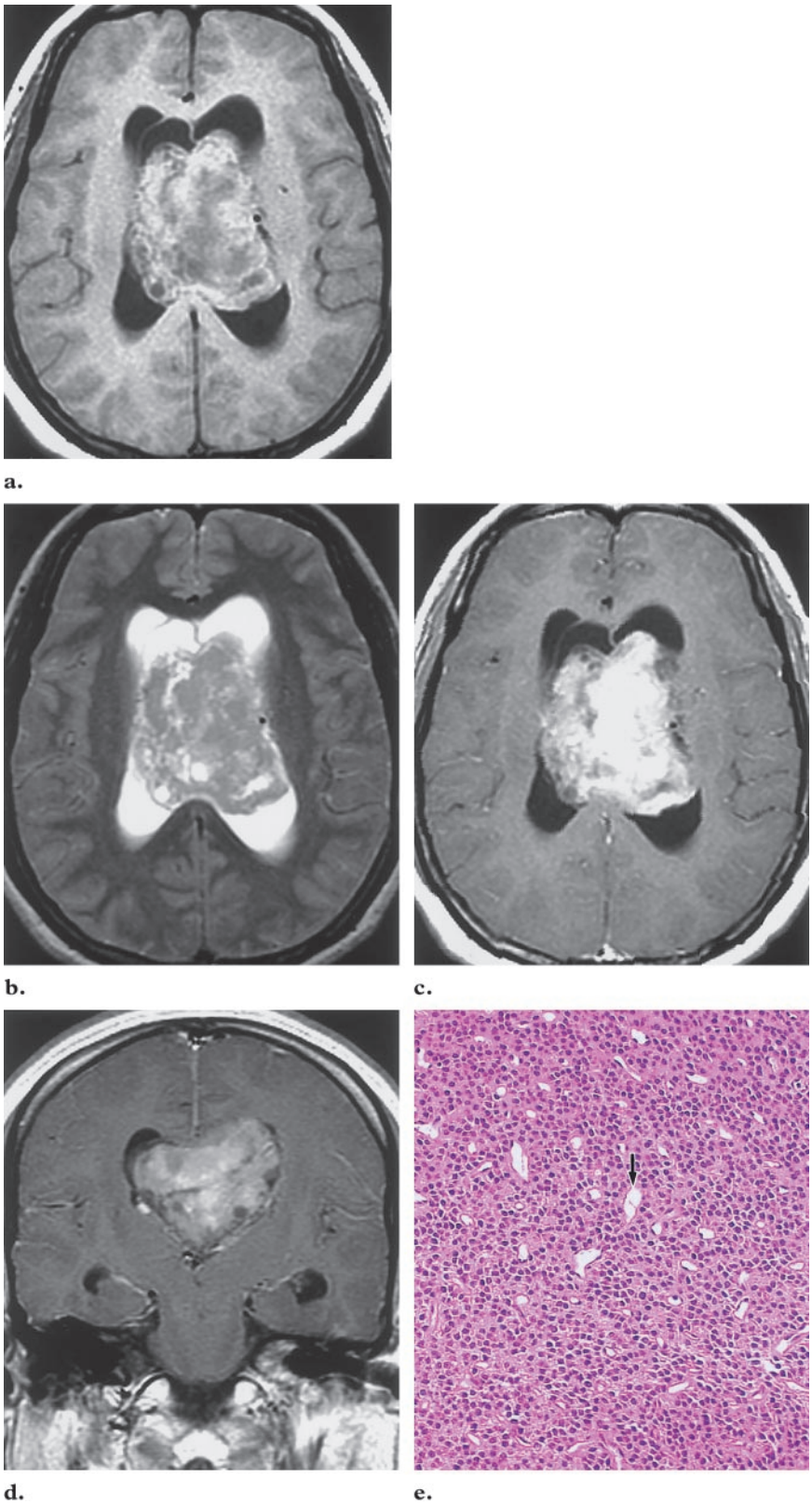


Figure 4. Central neurocytoma involving the lateral ventricles in a 28-year-old woman with headaches and gait disturbance. **(a)** Axial T1-weighted MR image shows a heterogeneous mass with low to intermediate signal intensity and multiple foci of low signal intensity. The mass is located in the lateral ventricles near the foramen of Monro with an attachment to the septum pellucidum. **(b)** Axial T2-weighted MR image shows that the mass has heterogeneous low signal intensity. **(c, d)** Axial **(c)** and coronal **(d)** gadolinium-enhanced T1-weighted MR images show heterogeneous, moderate to marked enhancement. **(e)** Photomicrograph (original magnification, ×100; hematoxylin-eosin stain) shows small, monotonous, round to oval cells with round nuclei and perivascular zones of fibrillarity (arrow).

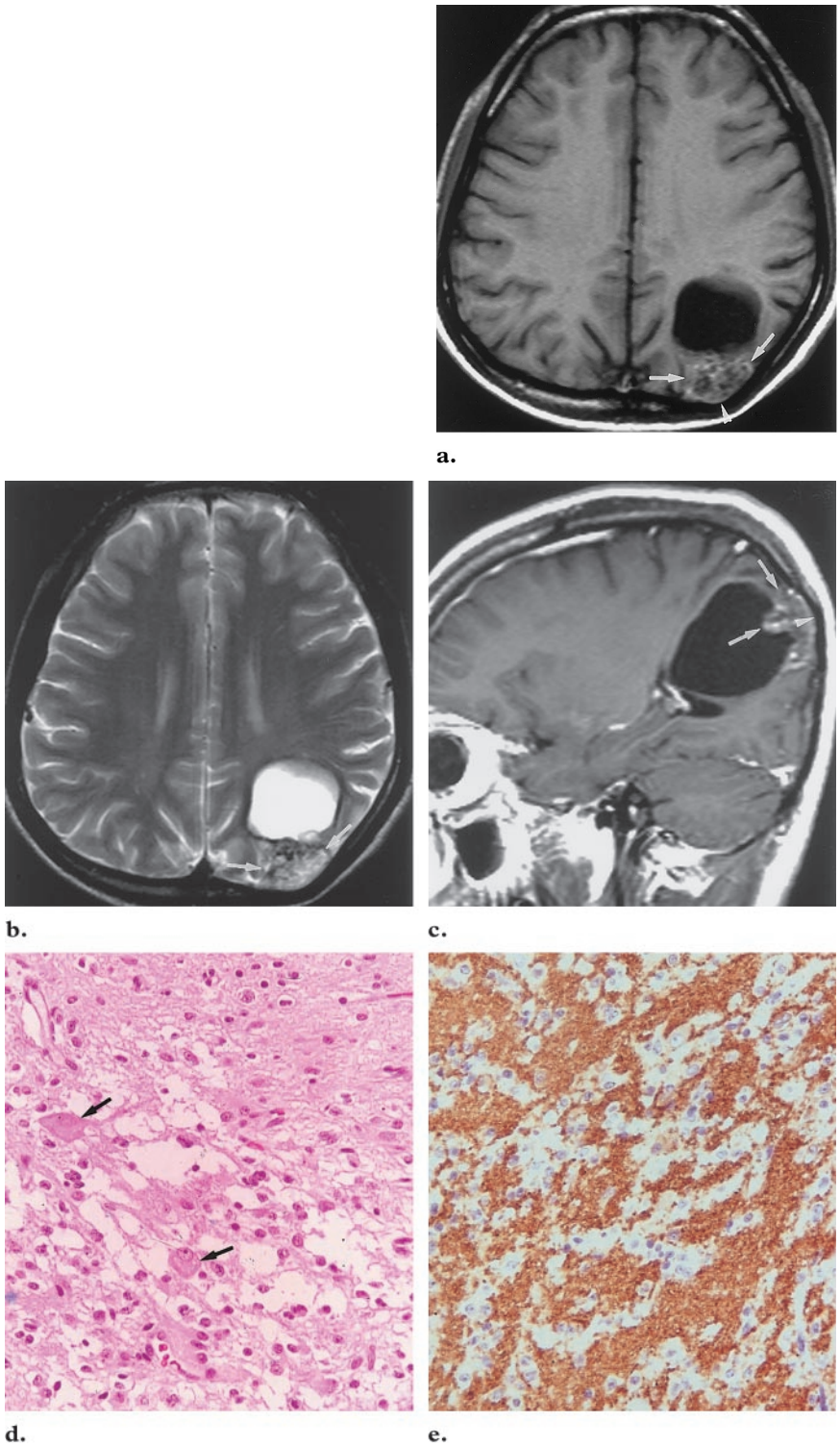
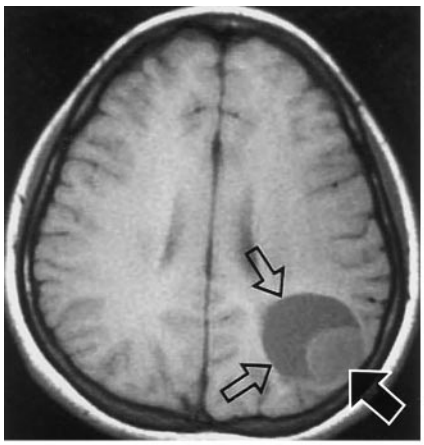


Figure 5. Ganglioneurocytoma in a 23-year-old man with a long history of seizures. **(a)** T1-weighted image shows a large, thin-walled cyst that is isointense to CSF in the white matter of the left parietal lobe. Solid portion (arrows) along the cortex has well-defined, heterogeneous mixed intensity and erodes the overlying calvaria (arrowhead). **(b)** On the T2-weighted image, the cystic portion is isointense to CSF; the solid portion has heterogeneous high intensity (arrows). **(c)** Gadolinium-enhanced T1-weighted image shows lobulated contour of the solid portion with minimal enhancement (arrows) and erosion of the calvaria (arrowhead). **(d)** Photomicrograph (original magnification, ×200; hematoxylin-eosin stain) shows ganglionic cells (arrows) among the small round cells. **(e)** Photomicrograph (original magnification, ×200; synaptophysin stain) shows tumor cells in a neurophil-like fibrillary background, which is strongly immunoreactive for synaptophysin (brown areas).

half of the cases and is usually clumped, amorphous, and globular (12,14). The imaging differential diagnosis includes other intraventricular tumors that occur in young adults (ie, oligodendroglioma, subependymal giant cell astrocytoma, low-grade or pilocytic astrocytoma, and ependymoma).

Ganglioneurocytoma is a recently described, rare variant of central neurocytoma that is characterized by differentiation toward ganglion cells. The histopathologic characteristics of ganglioneurocytoma are the same as those of a central neurocytoma, except that ganglioneurocytoma shows ganglioid differentiation, frequently forms a cystic lesion, and frequently arises extraventricularly (Fig 5) (13,16,17). This tumor is thought to grow



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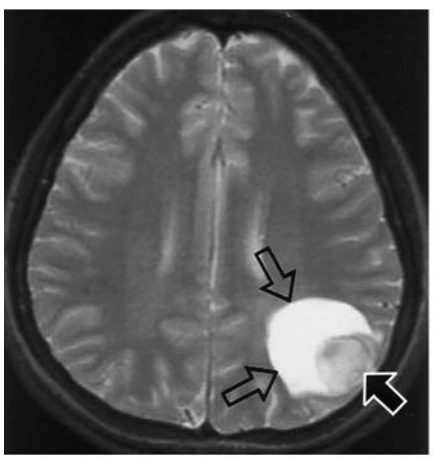

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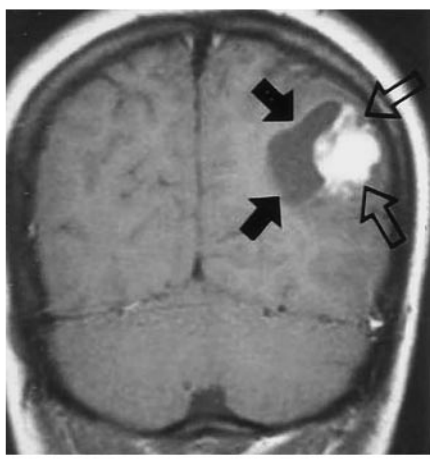

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Figure 6. Ganglioglioma involving the left parietal lobe in a 20-year-old woman with partial complex seizures. **(a, b)** T1-weighted **(a)** and T2-weighted **(b)** images show a large cystic mass (open arrows) with a solid mural nodule (solid arrow) that involves the cortex and white matter of the left parietal lobe. The nodule has intermediate signal intensity on the T1-weighted image **(a)** and high signal intensity on the T2-weighted image **(b)**. **(c)** Gadolinium-enhanced T1-weighted image shows marked enhancement of the nodule (open arrows), which is located eccentrically within the cystic mass (solid arrows).

slowly and have a favorable prognosis, as do most central neurocytomas (16).

Mixed Neuronal-Glial Cell Tumors

Ganglioglioma

Gangliogliomas, although rare, are the most common of the neuronal-glial neoplasms arising within the central nervous system. The essential histopathologic feature of gangliogliomas is the admixture of both atypical ganglion cells and neoplastic glial cells in varying amounts. However, clear-cut separation between ganglioglioma and gangliocytoma is difficult because there are varying degrees of neoplastic evolution of the glial component (3). Classically, ganglioglioma is described as a cystic mass with a mural nodule in approximately 40% of diagnosed cases (18,19). Most gangliogliomas occur in children and young adults. Although any site within the central nervous system may be affected, most tumors develop in the temporal lobe (19,20). Gangliogliomas of the temporal lobe are commonly associated with the clinical presentation of medically refractory seizures, particularly those of the partial complex type.

The solid portion of the tumor appears as varying degrees of attenuation on nonenhanced CT images. Calcification has been noted in about 30% of cases. Peripherally located gangliogliomas may cause scalloped pressure erosion of the overlying calvaria due to their slow-growing nature. At MR imaging, a well-defined cystic mass with a solid mural nodule is typically seen (Fig 6). However, a solid mass showing nonspecific low to intermediate signal intensity on T1-weighted images and high signal intensity on T2-weighted

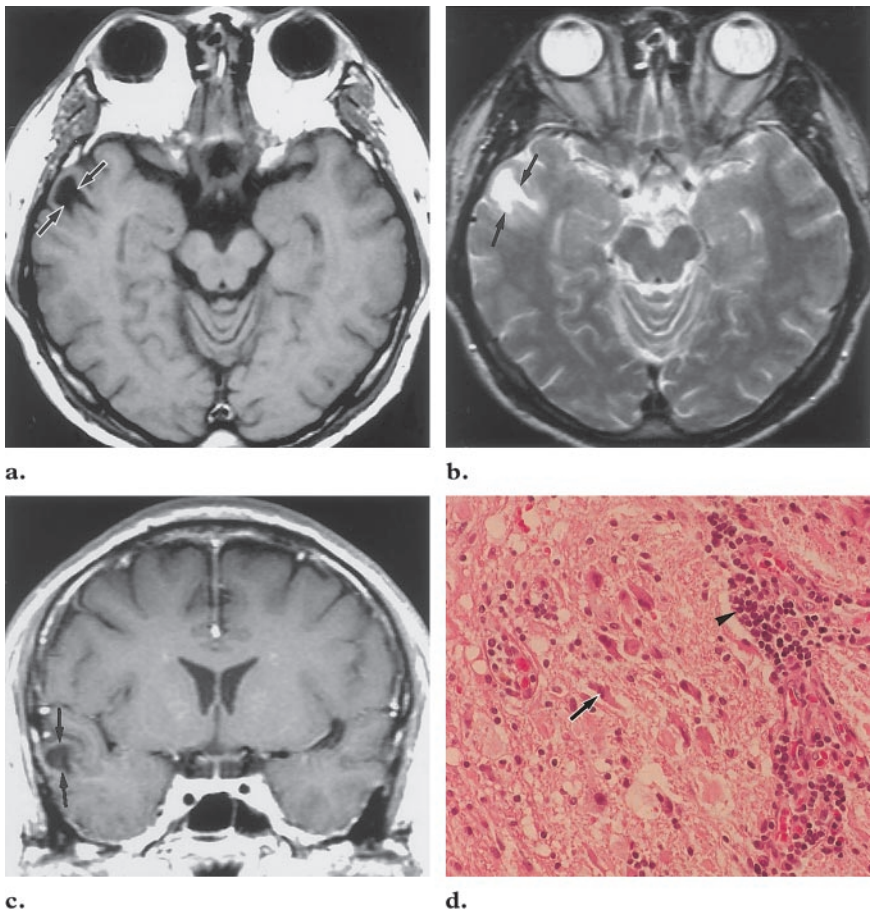

Figure 7. Ganglioglioma involving the right temporal lobe in a 38-year-old man with partial complex seizures. **(a)** Axial T1-weighted MR image shows a cortical and subcortical tumor (arrows) with low signal intensity. **(b)** Axial T2-weighted MR image shows that the tumor (arrows) has high signal intensity. **(c)** Coronal gadolinium-enhanced T1-weighted MR image shows no enhancement of the tumor (arrows). **(d)** Photomicrograph (original magnification, ×200; hematoxylin-eosin stain) shows several pleomorphic neoplastic ganglion cells (arrow) in a fibrillary astrocytic background and perivascular lymphocytic infiltration (arrowhead).

images is also not uncommon (Fig 7) (19,20). Enhancement of the solid portion is highly variable, ranging from nonenhancing to ringlike to intense homogeneity (Figs 6, 7). There is usually little associated mass effect or surrounding vasogenic edema.

Desmoplastic Infantile Ganglioglioma

Desmoplastic infantile ganglioglioma is an uncommon variety of ganglioglioma. It is usually a very large tumor with two distinct components. There is a predominant cystic component and a solid component representing desmoplastic reaction located adjacent to the meninges and attached to the dura (21–23). In areas of prominent desmoplasia, neoplastic cells are spindle-shaped with elongated, moderately pleomorphic nuclei. Ganglion cells are found within the neoplasm and can be numerous, mimicking a ganglioglioma. Desmoplastic infantile ganglioglioma is an uncommon variety of ganglioglioma that manifests in the first year of life. It invariably arises in the supratentorial region and affects more than one lobe, preferentially the parietal and frontal followed by the temporal. The symptoms and signs are of short duration and include increasing head circumference and tense, bulging fontanelles.

At CT, the lesion has a large well-defined cystic region, similar to cerebrospinal fluid, and an isoattenuating or slightly hyperattenuating superficial solid portion that shows enhancement (21,22). T1-weighted images characteristically show a hypointense cystic mass with a peripheral solid component that enhances intensely following administration of gadolinium contrast media. Extension of the enhancement to the leptomeninges is characteristic and correlates with the firm dural attachment of the solid component (Fig 8). The solid portion is heterogeneous on T2-weighted images (21,22).

Desmoplastic infantile ganglioglioma has distinct difference from classic ganglioglioma. These differences include manifestation in infancy, inclusion of immature neuroepithelial cells, presence of dense desmoplasia, and predominant location in the parietal or frontal lobe (21). Treatment is surgical excision. If the tumor can be removed without compromising normal tissue, the prognosis is good.

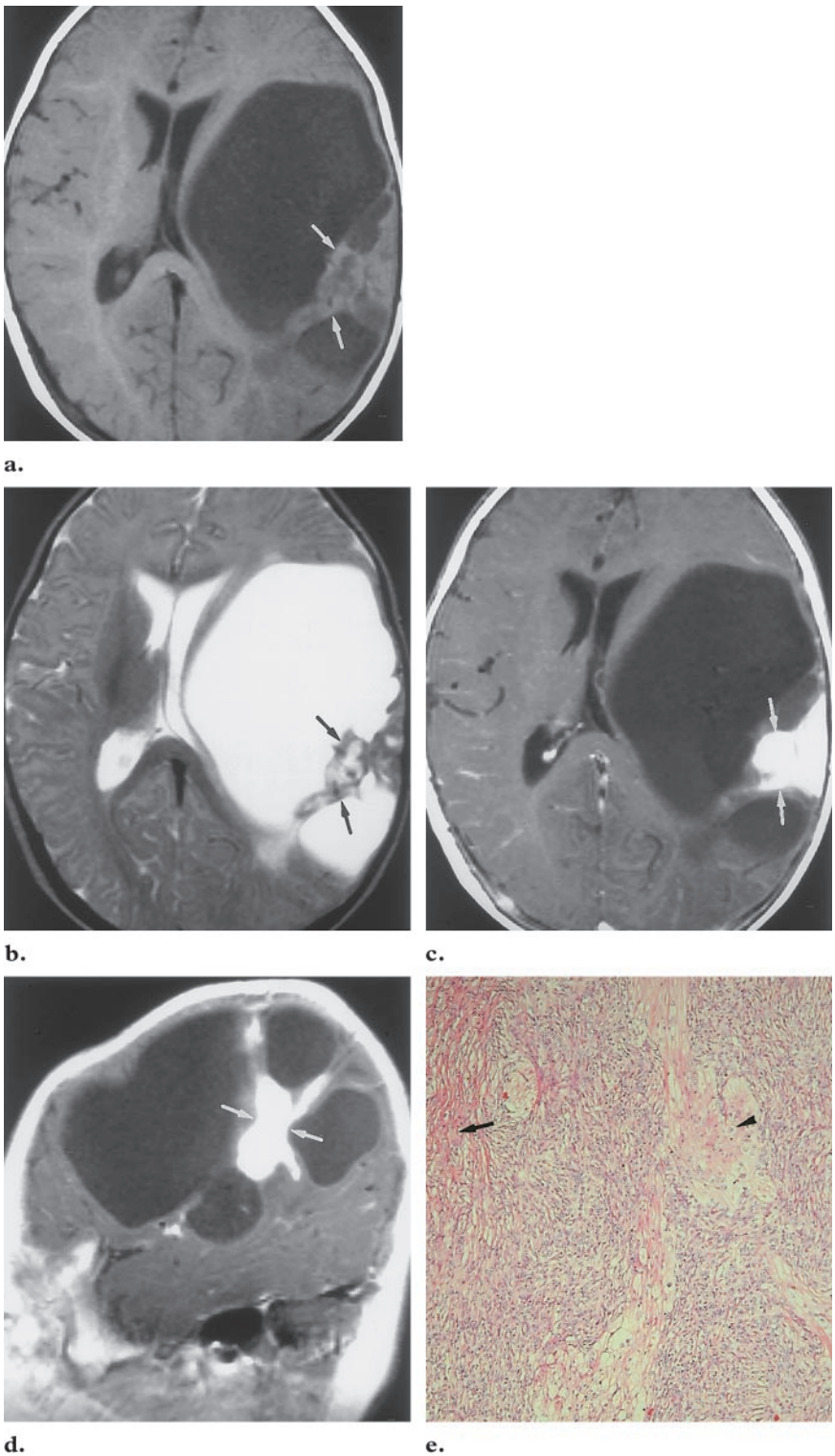

Figure 8. Desmoplastic infantile ganglioglioma involving the left frontal and parietal lobes in a 7-month-old girl with right hemiparesis. **(a)** Axial T1-weighted MR image shows a large cystic mass with an isointense solid portion (arrows) that abuts the dura. **(b)** Axial T2-weighted MR image shows that the cystic portion has high signal intensity and the solid portion (arrows) has heterogeneous low signal intensity. **(c, d)** Axial **(c)** and sagittal **(d)** gadolinium-enhanced T1-weighted MR images show intense enhancement of the solid portion of the tumor (arrows). **(e)** Photomicrograph (original magnification, ×100; hematoxylin-eosin stain) shows that the tumor is mainly composed of spindle cells within dense desmoplastic tissue (arrow) and demonstrates glial and neuronal differentiation (arrowhead).

Dysembryoplastic Neuroepithelial Tumor

Dysembryoplastic neuroepithelial tumor is a benign, usually supratentorial, mixed neuronal-glial neoplasm characterized by multinodular architecture and a predominantly intracortical location. Histopathologically, it is composed of a population of heterogeneous cellular components, such as oligodendrocytelike cells with admixtures of mature ganglion cells and astrocytes, which are located in a myxoid or mucinous interstitial matrix (24,25). A background of cortical dysplasia adjacent to a dysembryoplastic neuroepithelial tumor is often noted. This histopathologic

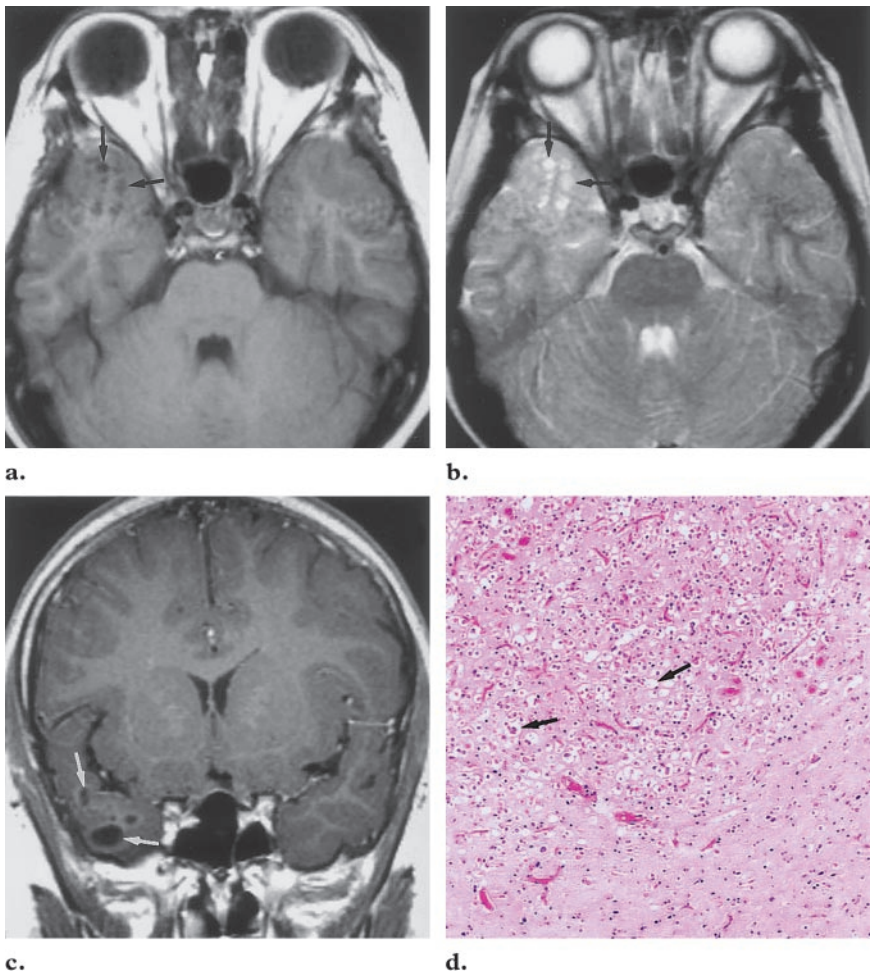

Figure 9. Dysembryoplastic neuroepithelial tumor involving the right temporal lobe in a 10-year-old girl with seizures. **(a)** Axial T1-weighted MR image shows a multinodular growth pattern of multiple, variably sized, low signal intensity lesions (arrows) in the cortical and subcortical areas of the right temporal lobe. **(b)** Axial T2-weighted MR image shows that the lesions (arrows) have high signal intensity. **(c)** Coronal gadolinium-enhanced T1-weighted MR image shows little enhancement of the lesions (arrows). **(d)** Photomicrograph (original magnification, ×100; hematoxylin-eosin stain) shows a cortical nodule composed of cells with small round nuclei and a clear perinuclear halo (arrows) in a loose myxoid matrix.

appearance and the benign clinical course suggest that it may be a malformation rather than a neoplastic lesion (24). A large majority of dysembryoplastic neuroepithelial tumors are located in the cortical region of the temporal lobe, with common extension into adjacent subcortical white matter and preferential involvement of the mesial structures. Therefore, it is usually associated with medically refractory partial seizures in adolescents and young adults and usually causes no neurologic deficit (24).

At CT, it appears as a moderately or markedly hypoattenuating cystlike lesion (24,26). Calcification is reported in less than 20% of cases (24). On MR images, it has a well-demarcated, multilobulated or gyriform appearance and is hypointense on T1-weighted images and markedly hyperintense on T2-weighted images (Fig 9) (24,26,27). There is a high frequency of bone remodeling of the adjacent calvaria at CT or MR imaging. Typically, no surrounding vasogenic edema is present, and enhancement at CT or MR imaging occurs in about one-third of cases but is not a prominent feature (24,26).

Ganglioneuroma

Ganglioneuroma is known as a fully differentiated, circumscribed tumor with no immature elements. It is composed of ganglion cells, Schwann cells accompanying the neuritic processes, and fibrous tissue (28). The presence of any immature elements would negate the diagnosis of ganglioneuroma (28,29). However, because it is histologically similar to and biologically indistinguishable from ganglioglioma, some clinicians do not consider them separate tumors. Involvement of the central nervous system is extremely rare (30).

Conclusions

In neuronal tumors of the central nervous system, neurologic manifestations are varied and include seizures (most common), symptoms of increased intracranial pressure, and neurologic deficits corresponding to the tumor locations. Many neuronal tumors of the central nervous system show characteristic radiologic findings. Understanding the radiologic findings as well as the pathologic background and classification of neuronal tumors

is important because they are generally benign and their prognosis is generally favorable if appropriate treatment, such as resection, is not delayed.

Acknowledgment: We are very grateful to Bonnie Hami, MA, for her editorial assistance.

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