

## The 2007 WHO Classification of Nervous System Tumors: Newly Recognized Members of the Mixed Glioneuronal Group

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**The clinical and pathologic features of two glioneuronal neoplasms newly incorporated in the 2007 revision of the WHO classification of nervous system tumors are reviewed. These are the papillary glioneuronal tumor and the rosette-forming glioneuronal tumor of the fourth ventricle.**

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### INTRODUCTION

Two unusual neoplasms of mixed glioneuronal nature are now recognized as novel, distinct entities in the newly updated WHO taxonomy of tumors of the central nervous system. These are the papillary glioneuronal tumor (27) and the rosette-forming glioneuronal tumor (RGNT) of the fourth ventricle (14).

### PAPILLARY GLIONEURONAL TUMOR

This morphologically distinctive neoplasm was originally described by Komori et al. (20) in 1996 as “pseudopapillary ganglioglioneurocytoma” and thereafter designated “papillary glioneuronal tumor” (PGNT) in an expanded 1998 series detailing the clinical and pathologic features of nine cases (21). An intercurrent report from Kim and Suh (19) in 1997 communicated one example as a “pseudopapillary neurocytoma of temporal lobe with glial differentiation.” Subsequent publications have consistently referred to this entity as PGNT and it is under this designation that the lesion appears in the 2007 WHO classification (27).

**Demographic and clinical features.** The actual (population-based) incidence of PGNT is unknown, but the tumor is certainly rare. To date, 35 examples have been described and illustrated in full-length case report or series format (3–13, 15, 17, 19, 21, 23, 24, 26, 28, 30–34). These have

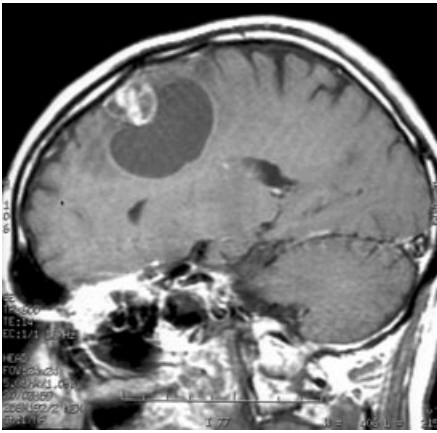
affected 19 females and 16 males, aged 4–75 years (mean, 26 years), being most prevalent in the second and third decades of life. Only four cases have occurred in adults older than 40 years of age. Except for one-third ventricular example (31), all have been situated in the cerebral hemispheres, particularly the temporal and frontal lobes where they comprise 13 and 11 examples, respectively. Headache and seizures are the most common clinical manifestations. Other presenting complaints include nausea, vertigo, visual and gait disturbances, focal sensorimotor deficits, syncope, memory loss, mood changes and difficulties in verbal comprehension or expression. Headache of sudden onset with rapid progression to aphasia and coma has been described as a consequence of tumoral hemorrhage (8). Isolated examples of PGNT have been reported in patients with cleft lip and orbital schwannoma (21), but none have occurred in association with a dysgenetic syndrome or in a familial setting.

**Neuroradiologic appearances.** Both CT and MR studies show PGNTs to be circumscribed supratentorial lesions that may span the cortical ribbon and underlying white matter or be more deeply situated, abutting the lateral ventricular system. Cystic alterations are frequent. Solid components are T<sub>1</sub>-iso/hypointense, T<sub>2</sub>-iso/hyperintense, usually contrast-enhancing

in a diffuse, patchy or rimming fashion, and may present as mural nodules or plaques within otherwise cystic masses (Figure 1). Calcifications may be seen. Significant tumoral hemorrhage is exceptional, but on record (8), as is an example associated with superficial siderosis of the neuraxis (23). Edema and conspicuous mass effect are generally limited to larger lesions. Reported tumor dimensions range from 1–9 cm (mean, 4.5 cm).

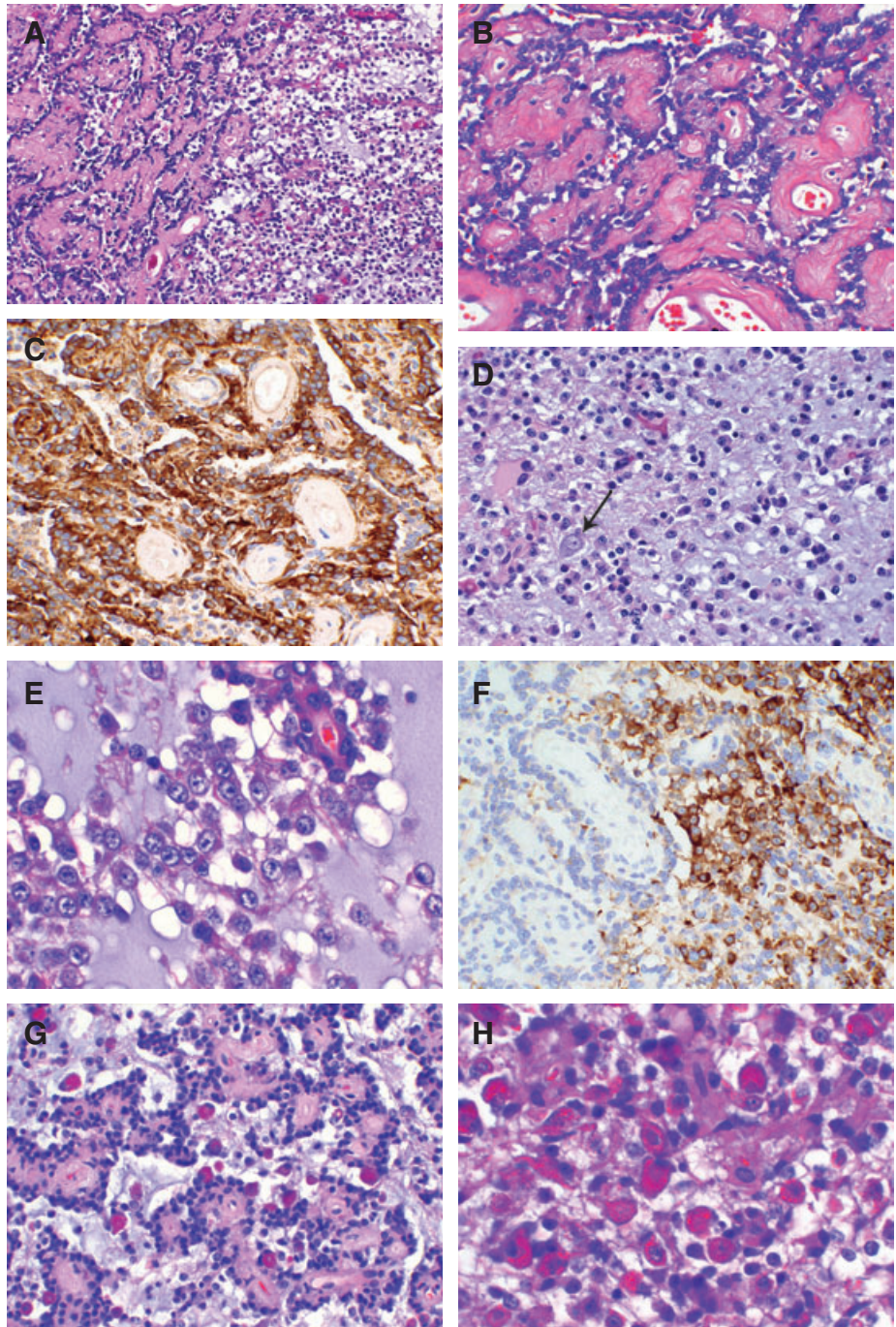
**Morphologic and immunophenotypic features.** Papillary glioneuronal tumors are unique among central nervous system neoplasms in their juxtaposition of (pseudo) papillary gliovascular structures and interposed neurocytic/neuronal tissue (Figure 2A). The former consist of compactly arranged, hyalinized blood vessels of modest caliber surrounded by a monolayer or pseudostratified layer of small, cuboidal glial cells with rounded nuclei and relatively scant, eosinophilic cytoplasm (Figure 2B) that consistently immunolabel for glial fibrillary acidic protein (GFAP; Figure 2C) and S-100 protein (21). These cells do not elaborate the perivascular cytoplasmic processes typical of ependymoma and astroblastoma, but rather sit squarely on their fibrovascular supports and may form solid interpapillary aggregates. Neither mitotic activity nor cytologic atypia is seen. Ultrastructural studies show these cells to contain bundles of intermediate filaments of glial type (6, 19, 21) and to form juxtavascular basal lamina (21).

Interspersed between or regionally separate from the gliovascular papillae described above are monomorphous non-papillary aggregates of neuronal cells in a fine, neuropil-like matrix prone to myxoid



**Figure 1.** As is often the case, the solid component of this papillary glioneuronal tumor is contrast-enhancing on post-gadolinium T<sub>1</sub>-weighted MR imaging. Note its projection into a sizable cyst.

change. These cytologically varied cells include neurocytes with dense round nuclei, well-differentiated neurons of medium-size, large ganglion cells and, prominent in many tumors, transitional forms having vesicular nuclei of intermediate size, relatively small nucleoli and narrow rims of amphophilic or violet-hued cytoplasm (Figure 2D,E). Granular immunoreactivity for synaptophysin (SYN), characteristically strong, is a regular feature of the matrix within which these various cells are disposed. The neurons and transitional forms also exhibit diffuse cytoplasmic as well as punctate surface labeling for this neuronal marker (21) (Figure 2F). Other neuron-associated antigens that may be expressed include neuron-specific enolase (NSE), microtubule-associated protein 2 (MAP2), class III  $\beta$ -tubulin, neural cell adhesion molecule (NCAM) and NeuN (10, 15, 21, 31, 33). Assessment of one PGNT by Western blot analysis revealed a “mature” pattern of neuron-associated adhesion complex expression as evidenced by elaboration of the L1 adhesion molecule and the 180 kd NCAM isoform that is characteristic of fully differentiated neurons, but absence of the polysialylated NCAM isoform elaborated in the developing CNS (6). In most PGNTs, immunolabeling for neurofilament protein (NFP) is restricted to larger neuronal (“ganglioid”) and ganglion cell populations, and absence of chromogranin a (CHR) expression is the rule (21). Not surprisingly, ultrastructural studies have shown that the non-papillary



**Figure 2.** Histologic and immunophenotypic features of papillary glioneuronal tumor. **A.** Papillary gliovascular structures (left) abruptly give way to solid components containing oligodendrocyte-like clear-cell neurocytic elements. **B.** Small, cuboidal cells of uniform appearance cover hyalinized fibrovascular cores in papillary regions. **C.** Cytoplasmic immunolabeling for glial fibrillary acidic protein attests to the glial nature of papillary elements. **D.** Non-papillary components include small round cells, many of oligodendrocyte-like appearance but representing neurocytes, as well as larger neurons (arrow). **E.** Neuronal elements of intermediate type (“ganglioid” cells) exhibit vesicular nuclei and distinct nucleoli, but little rimming cytoplasm. Note myxoid change. **F.** Intense synaptophysin labeling is evidenced in intermediate neuronal elements, while cells surrounding vascular papillae (left) are non-reactive. Deeply eosinophilic cells of minigemistocytic or coarsely granulated appearance may be found adjacent to papillary structures (**G**), or admixed with small, clear-cell neurocytic elements (**H**).

regions within PGNTs contain cells manifesting neuronal specializations (cytoplasmic extensions replete with aligned microtubules, clear vesicles and dense-core

granules in limited numbers, as well as synaptic contacts of varying organization) (6, 19, 21). Hybrid glioneuronal cells have not been identified.

A subset of PGNTs contain minigemistocytes or coarsely granulated cells (both GFAP-labeling) of the sort encountered in oligodendrogliomas, these lying between papillary structures or nestled among neuronal cell types in non-papillary areas (Figure 2G,H). Tanaka et al (31) have suggested that PGNTs are capable of oligodendroglial differentiation, identifying within these lesions a population of small cells that fail to label for GFAP or NeuN but express the Olig2 transcription factor. Distributed principally in interpapillary areas surrounding GFAP-positive perivascular elements, these may correspond to cells identified by Komori et al (21) as having non-specific fine structural appearances indicative of neither astrocytic nor neuronal differentiation. In the cases studied by Tanaka et al (31), minigemistocytes tended to an intimate association with these Olig2-labeling forms and were thought to derive from them. Although Olig2 cannot be regarded as an oligodendrocyte-specific “marker” in the neoplastic context, the notion that at least some of the putative neurocytes are cells differentiated along oligodendroglial lines merits further investigation.

Secondary changes that may be seen in PGNTs include foam cell infiltration, hemosiderin deposition, hyaline stromal fibroplasia and dystrophic calcification. Specimens containing adjacent brain parenchyma typically show only a narrow zone of tumor infiltration or a discrete, “pushing” border that provokes chronic piloid astrogliosis with Rosenthal fiber formation. Mitotic figures are typically absent to rare, if demonstrable at all, and MIB-1 labeling indices usually do not exceed 2%–3%. In the series of Komori et al (21), the mean MIB-1 index was 1.3% (range, 0.5%–2.5%). Papillary glioneuronal tumors exhibiting more conspicuous proliferative activity have been reported (see Clinical Biology and Grading below). Similar to conventional PGNTs, however, such atypical examples exhibit neither complex microvascular hyperplasia nor necrosis.

**Genetic features.** Chromosome 1p was found to be intact in six PGNTs examined by fluorescence *in situ* hybridization (31). One example studied by the author showed no deletions of chromosomes 1p

or 19q by loss of heterozygosity analysis (unpublished observation).

**Clinical biology and grading.** Papillary glioneuronal tumors are not accorded a precise grade in the 2007 WHO monograph (27), which, based on experience reported to date, states that they generally behave in grade I fashion and are amenable to control by surgical means. A survey of 24 cases with follow-up (range 2–84 months; mean, 22 months) reveals no instance of tumor regrowth when treated by gross total resection alone. Two additional patients treated by gross total excision and radiotherapy are free of tumor at 5 years (34) and 19 years (13), with the former patient having had an “atypical” PGNT with occasional mitoses and a MIB-1 labeling index of 15%. On the other hand, one abbreviated report (17) (abstract only), mentions multifocal recurrence 4 years after an ostensibly complete resection of one PGNT with a MIB-1 index of 5% (17). One typical PGNT exhibited no regrowth 3 years following subtotal resection (28), while another experienced progression of a subtotally resected tumor with a MIB-1 index of 4% only 3 months after surgery (17). One example of progression following 10 years of tumor stability in a patient managed by observation alone has been communicated (15); in this case, subtotal resection revealed a PGNT with a prominent minigemistocytic component and a MIB-1 index of 5%–10%. This atypical lesion exhibited regrowth 6 months following combined radiation and chemotherapy. Yet another atypical PGNT that exhibited increased mitotic activity and a 12% MIB-1 labeling index was stable 1 year after incomplete excision and chemotherapy (3).

**Histogenesis.** Papillary glioneuronal tumors are presumed to have their cellular origins in multipotential precursor cells. Paraventricular examples could derive from elements of the subependymal matrix, while more superficially positioned cases might descend from cortical components of the secondary germinal layer.

#### ROSETTE-FORMING GLIONEURONAL TUMOR OF THE FOURTH VENTRICLE

The RGNT of the fourth ventricle seems initially to have been described in a

1995 report as a cerebellar form of dysembryoplastic neuroepithelial tumor (25), but was recognized as an entity *sui generis* and characterized in a 2002 study of 11 cases by Komori et al (22).

**Demographic and clinical features.** Like PGNT, RGNT is also a decidedly uncommon lesion; its population-based incidence is unknown. Nineteen examples have been reported in full-length study form (1, 2, 16, 18, 22, 25, 29). These affected 12 females and 7 males aged 12–59 years (mean, 31.7 years). Older patients may also be affected; the author has seen two cases in patients over age 65 years. The most common presenting manifestations are headache (secondary to obstructive hydrocephalus) and ataxia, followed by visual disturbances and vertigo. Neck pain and rigidity have also been reported. Incidental neuroradiologic discovery of RGNT has been described. In six reported cases, symptoms referable to the tumor preceded discovery by at least 2 years. A coexistent Chiari type I malformation has been noted in a single case (22), but RGNTs have not been described in familial form or in the setting of any recognized dysgenetic syndrome.

**Localization and neuroradiologic appearances.** Although CT and MRI studies typically show RGNTs to be midline lesions centered in the fourth ventricle, limited extension into the cerebellar vermis, brainstem and cerebral aqueduct may be seen. Tumors primarily situated in the tectal/pineal and aqueductal regions are on record (16, 22), as are multifocal lesions



**Figure 3.** Filling the fourth ventricle, this rosette-forming glioneuronal tumor exhibits linear contrast-enhancement on post-gadolinium T<sub>1</sub>-weighted MR imaging.

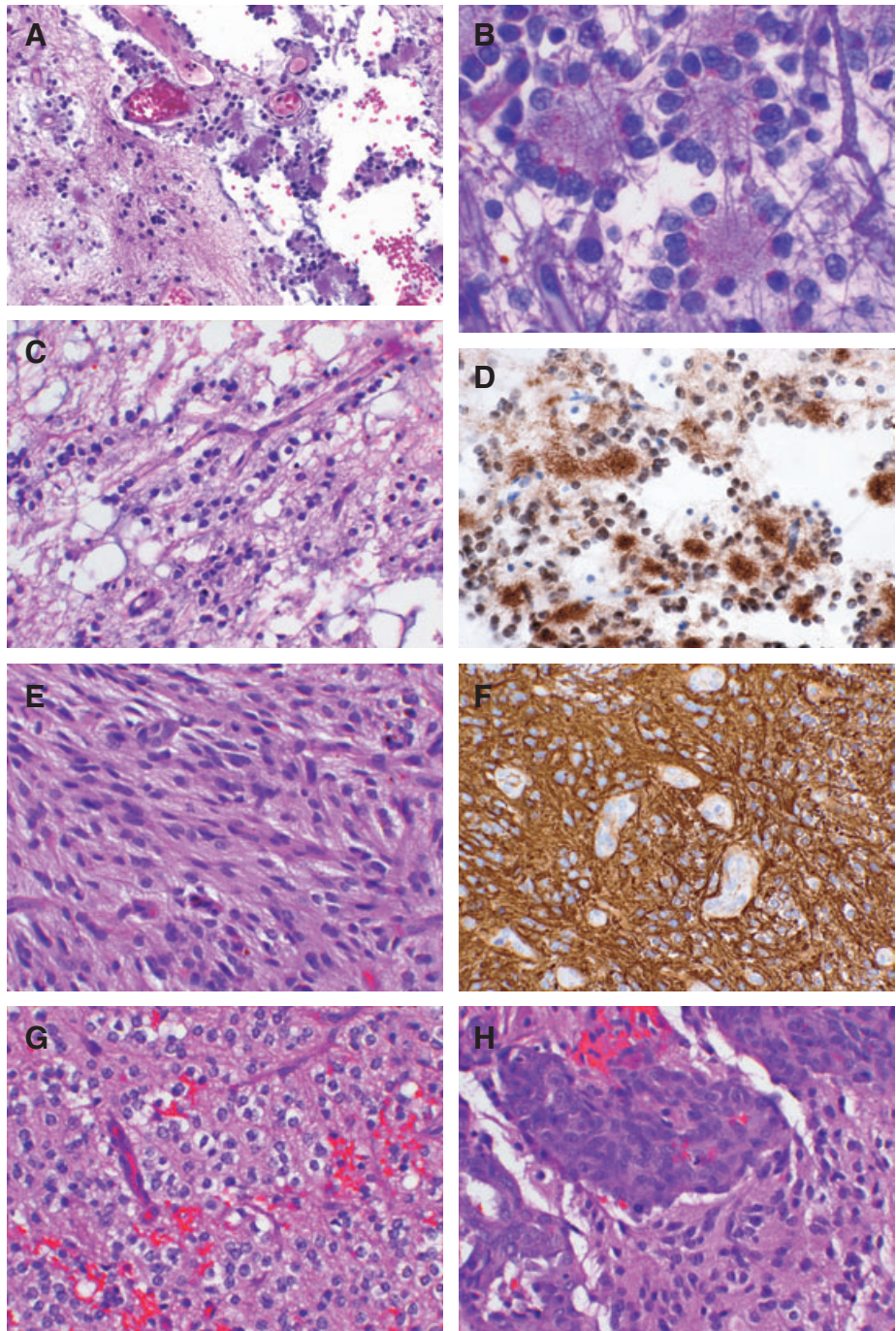
with fourth ventricular, vermian, dorsal pontine and, in one case, mesencephalic and thalamic components (22). RGNTs are relatively circumscribed, may be solid or multicystic, and usually exhibit at least focal contrast-enhancement that may be nodular, linear, ring or spot-like (Figure 3). On MR assessment, T<sub>1</sub>-iso/hypointensity and T<sub>2</sub>-hyperintensity are the rule. Calcification is occasionally seen and may be extensive. There may be associated edema, but this is generally minimal. Tumor dimensions range in size from 1–5 cm (mean, 3.3 cm).

**Morphologic and immunophenotypic features.** The RGNT is distinctive in its juxtaposition of patterned neurocytic and piloid astroglial components. The former, with their small, uniform round nuclei, evenly distributed delicate chromatin and scant cytoplasm form narrow perivascular pseudorosettes as well as diminutive Homer Wright-like rosettes that can be arrayed in cribriform pattern and that sometimes lie unanchored within tumoral microcavities (Figure 4A,B). Sectioning along the vascular axes of pseudorosettes frequently imparts the appearance of columnar organization (Figure 4C), which is often accompanied by myxoid and microcystic alterations, and may therefore bring to mind the specific glioneuronal element of dysembryoplastic neuroepithelial tumor. The latter's complement of mature "floating" neurons, however, is not a feature of RGNT, although a minority of RGNTs have been described as harboring small numbers of dysmorphic ganglion cell-like neuronal forms embedded within otherwise astrocytic regions (22).

Immunohistochemical studies of RGNT show that the delicate matrix of both the Homer Wright-like rosettes and the perivascular pseudorosettes label for SYN in granular fashion (Figure 4D). The fine cytoplasmic processes of these structures are also reactive for NSE and MAP-2, but lack staining for CHR, GFAP and, in most hands, NFP and S-100 protein (16, 22, 25, 29). The nuclei of the rosetted cells have been described in selected cases to manifest reactivity for PGP 9.5 (25), S-100 protein (16), or NeuN (16). In accord with the observations of Preusser et al (29), the author has been unable to demonstrate NeuN immunolabeling of rosetted cells in

five personally examined cases. Ultrastructural studies have confirmed the neuronal nature of these patterned elements, showing them to form cytoplasmic processes

containing parallel microtubules and small numbers of dense-core granules, and potentially forming well-developed synaptic contacts (1, 22).



**Figure 4.** Histologic and immunophenotypic features of rosette-forming glioneuronal tumor (RGNT). **A.** Diminutive rosettes often seem to float in tumoral clefts or cavities (right) that border solid astrocytic components. Perivascular pseudorosetting is also seen (left). **B.** True Homer Wright-like rosettes consist of delicate fibrillar cores rimmed by small, monomorphous, rounded nuclei. **C.** Perivascular pseudorosettes assume columnar configurations when sectioned along vascular axes. Myxoid and microcystic alterations are apparent. **D.** Rosettes consistently manifest granular synaptophysin immunoreactivity of their cores. **E.** Glial components of the RGNT are usually dominated by spindled astroglial cells that may achieve moderate cell density and are immunoreactive for glial fibrillary acidic protein (GFAP) (**F**). **G.** Small oligodendrocyte-like cells may also populate glial regions and are non-reactive for GFAP and neuron-associated antigens. **H.** Complex microvascular proliferation is a feature of some RGNTs.

The often dominant glial component of RGNT tends to exhibit pilocytic astrocytoma-like morphology. Spindled (Figure 4E) and, in some cases, stellate astroglial elements radiate lengthy cell processes that are GFAP and S-100 protein-positive (Figure 4F). Ultrastructurally, they are filled with bundles of intermediate filaments of glial type (22). There is considerable intra- and inter-tumoral variation in the levels of cellularity of the astrocytic constituent, ranging from a cellular density that is unarguably neoplastic to a hyponuclear fibrillary meshwork that is difficult to accept as anything but a glial scar. Strengthening the resemblance to pilocytic astrocytoma is the finding in some cases of Rosenthal fibers, eosinophilic granular bodies, and GFAP-negative/SYN-negative clear cells of oligodendrocyte-like appearance that form microcysts or aggregate in sheets (Figure 4G). The latter may exhibit S-100 protein immunolabeling. As mentioned, dysmorphic neurons of ganglion cell dimension may be found in the glial regions of RGNT but are not seen in conspicuous numbers.

In addition to regions of gliotic-appearing tissue, RGNTs commonly manifest stromal alterations indicative of chronicity and degeneration. These include vasosclerosis and dense collagenization, microcalcifications, hemosiderin deposits indicative of prior hemorrhage and, in an occasional case, focal infarction because of vascular thrombosis. Ectatic and hyalinized blood vessels with fibrin deposition may cluster in malformation-like complexes. RGNTs can exhibit proliferative microvasculature of the complex, glomeruloid type (Figure 4H), but are typically devoid (or very nearly so) of mitotic figures and show relatively low MIB-1 labeling. In the series of Komori et al (22), tumoral MIB-1 indices ranged from 0.35%–3.07% (mean, 1.58%). Subsequent reports have also documented similarly low labeling indices of less than 1% to 2.2% (1, 2, 29). These reassuring features are accompanied by a relatively well-defined tumor-parenchyma interface.

**Clinical biology and grading.** Based on the experience reported to date, RGNTs emerge as generally indolent and have been accorded grade I status in the updated WHO formulation (14). Only one

instance of tumor regrowth after 10 years of postoperative clinical stability was found among 13 evaluable cases treated by subtotal or gross total resection alone (16). The remaining 12 patients so treated remained recurrence-free 2–162 months after operation, with follow-up intervals extending over a period of at least 24 months in half of the cases (1, 2, 16, 22, 29). Two patients in this group had multifocal RGNTs that exhibited no enlargement of untreated nodules through 2 years of post-surgical surveillance (22), although isolated examples of lesion growth over relatively brief (6 month) periods of pre-treatment observation have been described (16, 22). Dissemination via the cerebrospinal fluid has not been reported. The sole tumor-associated fatality on record followed partial excision and irradiation of an RGNT in a 52-year-old woman who subsequently developed progressive bulbar dysfunction related to a new ring-enhancing abnormality near the operative bed; the patient died 45 months after initial treatment (22). As an autopsy was not performed, the cause of death in this case is unclear. While RGNTs are associated with a favorable prognosis in terms of survival, their location complicates surgical removal and a significant risk of neurologic injury, principally ataxia and sixth and seventh cranial nerve palsies, attends attempts at excision (2, 22).

**Histogenesis.** RGNTs are posited to derive from elements capable of divergent neuronal and glial differentiation, and may have their origin in the pluripotential cells of the subependymal plate.

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