

Symposium Editors: Gregory N. Fuller & Bernd W. Scheithauer

Previous editions of the World Health Organization (WHO) Classification of Tumours of the Nervous System have achieved extensive utilization internationally (1–3, 5). An Editorial and Consensus Conference convened in Heidelberg on November 17–18, 2006 (Figure 1), resulting in the latest iteration of the work (4; Figure 2). Several substantive alterations distinguish the 2007 Classification from the preceding 2000 Classification, including additions, reclassifications, changes in terminology and a number of conceptual modifications. The tumor entities that have been newly codified in the 2007 Classification constitute the focus of this mini-symposium (Table 1). We have chosen to use the term “newly codified” rather than “new” because several of the entities have in fact been recognized by neuropathologists for a considerable time—decades in some cases—but only recently has sufficient international consensus been reached to warrant formal codification. In contrast, other newly codified tumors are of much more recent vintage. Of these, some represent newly recognized prognostically and therapeutically significant subsets of a well-established tumor category, and others are uncommon entities whose formal recognition has awaited accrual of sufficient histopathological data and clinical experience to permit accurate characterization of the clinicopathologic features.

Among organ systems, the diversity and complexity of tumors of the nervous system are unrivaled, and, not unexpectedly, unresolved questions of classification and grading remain and await further elucidation. Some of the more salient of these problematic issues and controversies will be presented separately in a forthcoming overview, and individual topics will subsequently be critically explored in greater detail in future issues of *Brain Pathology* as part of the journal’s *Controversies in Pathology* series.



**Figure 1.** The World Health Organization (WHO) 2007 Editorial and Consensus Conference Working Group. Pictured from left to right: Kenneth D. Aldape, David N. Louis, Webster K. Cavenee, Charles G. Eberhart, Otmar D. Wiestler, Daniel J. Brat, Torsten Pietsch, Hiroko Ohgaki, Paul Kleihues, Peter C. Burger, Guido Reifenberger, Dominique Figarella-Branger, Johann A. Hainfellner, Anne Jouvett, Felice Giangaspero, V. Peter Collins, Arie Perry, Johan M. Kros, Fre T. Bosman, Roger E. McLendon, Yoichi Nakazato, Gregory N. Fuller, Andreas von Deimling, Bernd W. Scheithauer, Werner Paulus, H-K Ng.

## TUMOURS OF NEUROEPITHELIAL TISSUE

### Astrocytic tumours

Pilocytic astrocytoma	9421/1 <sup>1</sup>
Pilomyxoid astrocytoma	9425/3*
Subependymal giant cell astrocytoma	9384/1
Pleomorphic xanthoastrocytoma	9424/3
Diffuse astrocytoma	9400/3
Fibrillary astrocytoma	9420/3
Protoplasmic astrocytoma	9410/3
Gemistocytic astrocytoma	9411/3
Anaplastic astrocytoma	9401/3
Glioblastoma	9440/3
Giant cell glioblastoma	9441/3
Gliosarcoma	9442/3
Gliomatosis cerebri	9381/3

### Oligodendroglial tumours

Oligodendroglioma	9450/3
Anaplastic oligodendroglioma	9451/3

### Oligoastrocytic tumours

Oligoastrocytoma	9382/3
Anaplastic oligoastrocytoma	9382/3

### Ependymal tumours

Subependymoma	9383/1
Myxopapillary ependymoma	9394/1
Ependymoma	9391/3
Cellular	9391/3
Papillary	9393/3
Clear cell	9391/3
Tanycytic	9391/3
Anaplastic ependymoma	9392/3

### Choroid plexus tumours

Choroid plexus papilloma	9390/0
Atypical choroid plexus papilloma	9390/1*
Choroid plexus carcinoma	9390/3

### Other neuroepithelial tumours

Astroblastoma	9430/3
Chordoid glioma of the third ventricle	9444/1
Angiocentric glioma	9431/1*

<sup>1</sup> Morphology code of the International Classification of Diseases for Oncology (ICD-O) {616A} and the Systematized Nomenclature of Medicine (<http://snomed.org>). Behaviour is coded /0 for benign tumours, /3 for malignant tumours and /1 for borderline or uncertain behaviour.

\* The italicised numbers are provisional codes proposed for the 4th edition of ICD-O. While they are expected to be incorporated into the next ICD-O edition, they currently remain subject to change.

## Neuronal and mixed neuronal-glial tumours

Dysplastic gangliocytoma of cerebellum (Lhermitte-Duclos)	9493/0
Desmoplastic infantile astrocytoma/ganglioglioma	9412/1
Dysembryoplastic neuroepithelial tumour	9413/0
Gangliocytoma	9492/0
Ganglioglioma	9505/1
Anaplastic ganglioglioma	9505/3
Papillary glioneuronal tumor	9509/1*
Rosette-forming glioneuronal tumour of the fourth ventricle	9509/1*
Central neurocytoma	9506/1
Extraventricular neurocytoma	9506/1*
Cerebellar liponeurocytoma	9506/1*
Paraganglioma of the filum terminale	8680/1

## Tumours of the pineal region

Pineocytoma	9361/1
Pineal parenchymal tumour of intermediate differentiation	9362/3
Pineoblastoma	9362/3
Papillary tumour of the pineal region	9395/3*

## Embryonal tumours

Medulloblastoma	9470/3
Desmoplastic/nodular medulloblastoma	9471/3
Medulloblastoma with extensive nodularity	9471/3*
Anaplastic medulloblastoma	9474/3*
Large cell medulloblastoma	9474/3
CNS primitive neuroectodermal tumours (PNETs)	
CNS PNET, NOS	9473/3
CNS neuroblastoma	9500/3
CNS ganglioneuroblastoma	9490/3
Medulloepithelioma	9501/3
Ependymoblastoma	9392/3
Atypical teratoid / rhabdoid tumour	9508/3

## TUMOURS OF CRANIAL AND PARASPINAL NERVES

Schwannoma (Neurilemoma, neurinoma)	9560/0
Cellular	9560/0
Plexiform	9560/0
Melanotic	9560/0
Neurofibroma	9540/0
Plexiform	9550/0

**Figure 2.** WHO Classification of Tumours of the Nervous System. Reprinted with permission of the WHO 2007 editors.

Perineurioma	9571/0	Haemangiopericytoma	9150/1
Intraneural perineurioma	9571/0	Angiosarcoma	9120/3
Soft tissue perineurioma	9571/0	Kaposi sarcoma	9140/3
Malignant peripheral nerve sheath tumour (MPNST)	9540/3	<b>Primary melanocytic lesions</b>	
Epithelioid	9540/3	Diffuse melanocytosis	8728/0
MPNST with divergent mesenchymal and / or epithelial differentiation	9540/3	Melanocytoma	8728/1
Melanotic	9540/3	Malignant melanoma	8720/3
		Meningeal melanomatosis	8728/3
<b>TUMOURS OF THE MENINGES</b>		<b>Other neoplasms related to the meninges</b>	
<b>Tumours of meningotheial cells</b>		Haemangioblastoma	9161/1
Meningioma	9530/0	<b>LYMPHOMAS AND HAEMOPOIETIC NEOPLASMS</b>	
Meningothelial	9531/0	Malignant lymphomas	9590/3
Fibrous (fibroblastic)	9532/0	Plasmacytoma	9731/3
Transitional (mixed)	9537/0	Granulocytic sarcoma	9930/3
Psammomatous	9533/0		
Angiomatous	9534/0	<b>GERM CELL TUMOURS</b>	
Microcystic	9530/0	Germinoma	9064/3
Secretory	9530/0	Embryonal carcinoma	9070/3
Lymphoplasmacyte-rich	9530/0	Yolk sac tumour	9071/3
Metaplastic	9530/0	Choriocarcinoma	9100/3
Chordoid	9538/1	Teratoma	9080/1
Clear cell	9538/1	Mature	9080/0
Atypical	9539/1	Immature	9080/3
Papillary	9538/3	Teratoma with malignant transformation	9084/3
Rhabdoid	9538/3	Mixed germ cell tumours	9085/3
Anaplastic (malignant)	9530/3		
<b>Mesenchymal tumours</b>		<b>TUMOURS OF THE SELLAR REGION</b>	
Lipoma	8850/0	Craniopharyngioma	9350/1
Angiolipoma	8861/0	Adamantinomatous	9351/1
Hibernoma	8880/0	Papillary	9352/1
Liposarcoma (intracranial)	8850/3	Granular cell tumour	9582/0
Solitary fibrous tumour	8815/0	Pituicytoma	9432/1*
Fibrosarcoma	8810/3	Spindle cell oncocytoma of the adenohypophysis	8291/0*
Malignant fibrous histiocytoma	8830/3		
Leiomyoma	8890/0	<b>METASTATIC TUMOURS</b>	
Leiomyosarcoma	8890/3		
Rhabdomyoma	8900/0		
Rhabdomyosarcoma	8900/3		
Chondroma	9220/0		
Chondrosarcoma	9220/3		
Osteoma	9180/0		
Osteosarcoma	9180/3		
Osteochondroma	9210/0		
Haemangioma	9120/0		
Epithelioid haemangioendothelioma	9133/1		

Figure 2. Continued

Angiocentric glioma
Pilomyxoid astrocytoma
Papillary glioneuronal tumor (PGNT)
Rosette-forming glioneuronal tumor of the 4th ventricle (RGNT)
Papillary tumor of the pineal region (PTPR)
Spindle cell oncocytoma (SCO)
Pituicytoma

**Table 1.** WHO 2007 classification of tumors of the central nervous system: newly codified entities.

## REFERENCES

1. Kleihues P, Cavenee WK (1997) *Pathology and Genetics of Tumours of the Nervous System*. IARC: Lyon.
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5. Zulch KJ (1979) *Histological Typing of Tumours of the Central Nervous System (International Histological Classification of Tumours, NO. 21)*. World Health Organization: Geneva.