

Contents

List of abbreviations	xi	Ependymal tumours	
Foreword	xii	Introduction	159
ICD-O topographical coding	1	Supratentorial ependymoma	161
ICD-O morphological coding	1	Supratentorial ependymoma, <i>ZFTA</i> fusion-positive	164
CNS tumours	2	Supratentorial ependymoma, <i>YAP1</i> fusion-positive	167
1 Introduction to CNS tumours	7	Posterior fossa ependymoma	169
2 Gliomas, glioneuronal tumours, and neuronal tumours	15	Posterior fossa group A (PFA) ependymoma	172
Introduction	16	Posterior fossa group B (PFB) ependymoma	175
Adult-type diffuse gliomas		Spinal ependymoma	177
Astrocytoma, IDH-mutant	19	Spinal ependymoma, <i>MYCN</i> -amplified	180
Oligodendroglioma, IDH-mutant and 1p/19q-codeleted	28	Myxopapillary ependymoma	183
Glioblastoma, IDH-wildtype	39	Subependymoma	186
Paediatric-type diffuse low-grade gliomas		3 Choroid plexus tumours	189
Diffuse astrocytoma, <i>MYB</i> - or <i>MYBL1</i> -altered	56	Choroid plexus papilloma	190
Angiocentric glioma	59	Atypical choroid plexus papilloma	193
Polymorphous low-grade neuroepithelial tumour of the young	62	Choroid plexus carcinoma	195
Diffuse low-grade glioma, MAPK pathway-altered	65	4 Embryonal tumours	199
Paediatric-type diffuse high-grade gliomas		Medulloblastoma	
Diffuse midline glioma, H3 K27-altered	69	Introduction	200
Diffuse hemispheric glioma, H3 G34-mutant	74	Medulloblastomas, molecularly defined	
Diffuse paediatric-type high-grade glioma, H3-wildtype and IDH-wildtype	77	Medulloblastoma, WNT-activated	203
Infant-type hemispheric glioma	81	Medulloblastoma, SHH-activated and <i>TP53</i> -wildtype	205
Circumscribed astrocytic gliomas		Medulloblastoma, SHH-activated and <i>TP53</i> -mutant	208
Pilocytic astrocytoma	83	Medulloblastoma, non-WNT/non-SHH	211
High-grade astrocytoma with piloid features	90	Medulloblastomas, histologically defined	
Pleomorphic xanthoastrocytoma	94	Medulloblastoma, histologically defined	213
Subependymal giant cell astrocytoma	100	Other CNS embryonal tumours	
Chordoid glioma	104	Introduction	220
Astroblastoma, <i>MN1</i> -altered	107	Atypical teratoid/rhabdoid tumour	221
Glioneuronal and neuronal tumours		Cribriform neuroepithelial tumour	226
Ganglioglioma	111	Embryonal tumour with multilayered rosettes	228
Gangliocytoma	116	CNS neuroblastoma, <i>FOXR2</i> -activated	232
Desmoplastic infantile ganglioglioma / desmoplastic infantile astrocytoma	119	CNS tumour with <i>BCOR</i> internal tandem duplication	235
Dysembryoplastic neuroepithelial tumour	123	CNS embryonal tumour NEC/NOS	238
Diffuse glioneuronal tumour with oligodendroglioma-like features and nuclear clusters	127	5 Pineal tumours	241
Papillary glioneuronal tumour	130	Introduction	242
Rosette-forming glioneuronal tumour	133	Pineocytoma	243
Myxoid glioneuronal tumour	136	Pineal parenchymal tumour of intermediate differentiation	246
Diffuse leptomeningeal glioneuronal tumour	139	Pineoblastoma	249
Multinodular and vacuolating neuronal tumour	143	Papillary tumour of the pineal region	253
Dysplastic cerebellar gangliocytoma (Lhermitte-Duclos disease)	146	Desmoplastic myxoid tumour of the pineal region, <i>SMARCB1</i> -mutant	256
Central neurocytoma	149	6 Cranial and paraspinal nerve tumours	259
Extraventricular neurocytoma	153	Introduction	260
Cerebellar liponeurocytoma	156	Schwannoma	261
		Neurofibroma	265
		Perineurioma	269
		Hybrid nerve sheath tumours	271
		Malignant melanotic nerve sheath tumour	273
		Malignant peripheral nerve sheath tumour	275
		Cauda equina neuroendocrine tumour (previously paraganglioma)	279

List of abbreviations

7 Meningioma	283	11 Germ cell tumours	381
8 Mesenchymal, non-meningothelial tumours involving the CNS	299	12 Tumours of the sellar region	391
Introduction	300	Introduction	392
Soft tissue tumours		Adamantinomatous craniopharyngioma	393
Fibroblastic and myofibroblastic tumours		Papillary craniopharyngioma	397
Solitary fibrous tumour	301	Pituicytoma, granular cell tumour of the sellar region, and spindle cell oncocytoma	401
Vascular tumours		Pituitary adenoma / pituitary neuroendocrine tumour	406
Haemangiomas and vascular malformations	306	Pituitary blastoma	415
Haemangioblastoma	310	13 Metastases to the CNS	417
Skeletal muscle tumours		Metastases to the brain and spinal cord parenchyma	418
Rhabdomyosarcoma	314	Metastases to the meninges	421
Tumours of uncertain differentiation		14 Genetic tumour syndromes involving the CNS	423
Intracranial mesenchymal tumour, FET::CREB fusion-positive	317	Introduction	424
<i>CIC</i> -rearranged sarcoma	320	Neurofibromatosis type 1	426
Primary intracranial sarcoma, <i>DICER1</i> -mutant	323	Neurofibromatosis type 2	429
Ewing sarcoma	326	Schwannomatosis	434
Chondro-osseous tumours		Von Hippel–Lindau syndrome	437
Chondrogenic tumours		Tuberous sclerosis	441
Mesenchymal chondrosarcoma	330	Li–Fraumeni syndrome	446
Chondrosarcoma	332	Cowden syndrome	449
Notochordal tumours		Constitutional mismatch repair deficiency syndrome	452
Chordoma	335	Familial adenomatous polyposis 1	456
9 Melanocytic tumours	339	Naevoid basal cell carcinoma syndrome	458
Introduction	340	Rhabdoid tumour predisposition syndrome	460
Diffuse meningeal melanocytic neoplasms		Carney complex	462
Melanocytosis and melanomatosis	341	<i>DICER1</i> syndrome	464
Circumscribed meningeal melanocytic neoplasms		Familial paraganglioma syndromes	467
Melanocytoma and melanoma	344	Melanoma-astrocytoma syndrome	471
10 Haematolymphoid tumours involving the CNS	349	Familial retinoblastoma	473
Introduction	350	<i>BAP1</i> tumour predisposition syndrome	475
Lymphomas		Fanconi anaemia	478
CNS lymphomas		ELP1-medulloblastoma syndrome	481
Primary diffuse large B-cell lymphoma of the CNS	351	Contributors	483
Immunodeficiency-associated CNS lymphomas	356	Declaration of interests	489
Lymphomatoid granulomatosis	358	IARC/WHO Committee for ICD-O	491
Intravascular large B-cell lymphoma	360	Sources	493
Miscellaneous rare lymphomas in the CNS		References	501
MALT lymphoma of the dura	362	Subject index	557
Other low-grade B-cell lymphomas of the CNS	364	Previous volumes in the series	568
Anaplastic large cell lymphoma (ALK+/ALK-)	366		
T-cell and NK/T-cell lymphomas	368		
Histiocytic tumours			
Erdheim–Chester disease	370		
Rosai–Dorfman disease	372		
Juvenile xanthogranuloma	374		
Langerhans cell histiocytosis	376		
Histiocytic sarcoma	379		