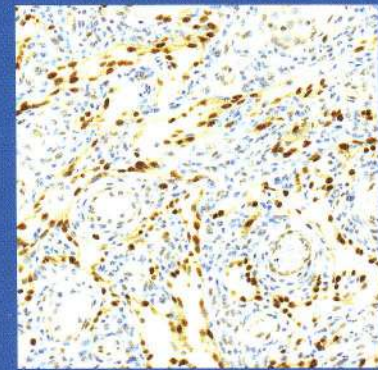
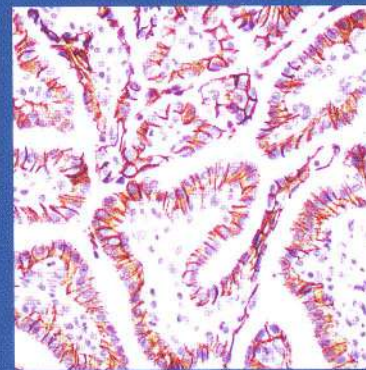
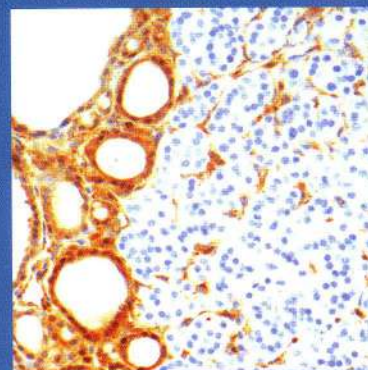
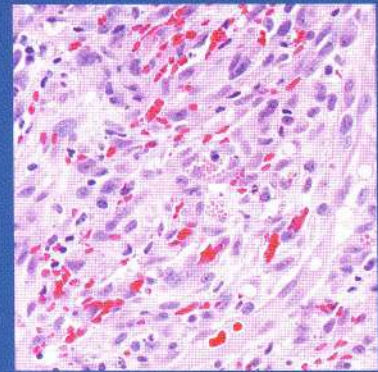
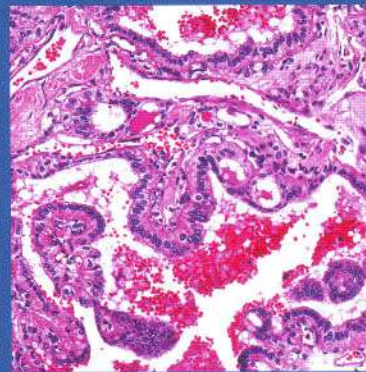
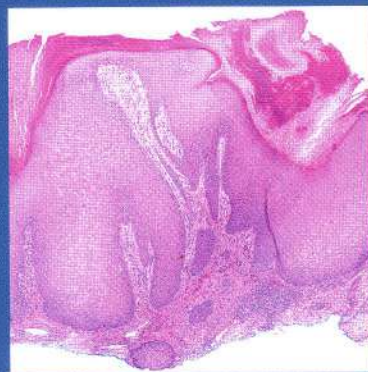
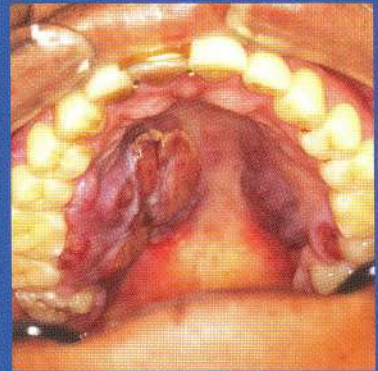
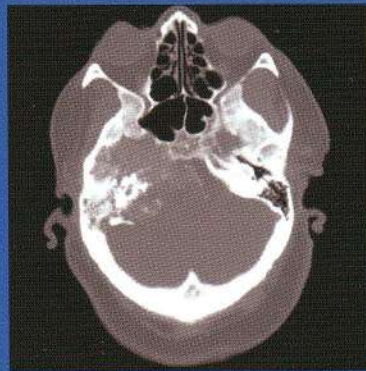


WHO Classification of Tumours • 5th Edition

# Head and Neck Tumours

## Part B

Edited by the WHO Classification of Tumours Editorial Board



International Agency for Research on Cancer





# Contents

Please note that the contents of this volume have been split into two parts for publication in print: Part A (in the accompanying book) and Part B (in this book).

<b>List of abbreviations</b>	<b>xi</b>	Adenocarcinomas	
		Intestinal-type sinonasal adenocarcinoma	81
		Non-intestinal-type sinonasal adenocarcinoma	84
<b>Foreword</b>	<b>xii</b>	Mesenchymal tumours of the sinonasal tract	
		Sinonasal tract angiofibroma	87
		Sinonasal glomangiopericytoma	90
		Biphenotypic sinonasal sarcoma	93
		Chordoma	96
<b>PART A (IN ACCOMPANYING BOOK)</b>		Other sinonasal tumours	
		Sinonasal ameloblastoma	99
		Adamantinomatous craniopharyngioma	101
		Meningioma of the sinonasal tract, ear, and temporal bone	103
		Olfactory neuroblastoma	107
<b>ICD-O topographical coding</b>	<b>1</b>	<b>2 Nasopharyngeal tumours</b>	<b>111</b>
<b>ICD-O morphological coding</b>	<b>2</b>	Introduction	112
Nasal, paranasal, and skull base tumours	3	Hamartomas	
Nasopharyngeal tumours	5	Hairy polyp	113
Hypopharyngeal, laryngeal, tracheal, and parapharyngeal tumours	6	Salivary gland anlage tumour	115
Salivary gland tumours	7	Carcinomas	
Oral cavity and mobile tongue tumours	9	Low-grade nasopharyngeal papillary adenocarcinoma	117
Oropharyngeal tumours	10	Nasopharyngeal carcinoma	119
Odontogenic and maxillofacial bone tumours	11	<b>3 Hypopharyngeal, laryngeal, tracheal, and parapharyngeal tumours</b>	<b>127</b>
Ear tumours	14	Introduction	128
Soft tissue tumours	15	Precursor lesions	
Haematolymphoid proliferations and neoplasia	18	Squamous papilloma and papillomatosis	129
Melanocytic tumours	20	Laryngeal and hypopharyngeal epithelial dysplasia	132
Tumours and tumour-like lesions of the neck and lymph nodes	21	Squamous cell carcinomas	
Germ cell tumours	22	Conventional squamous cell carcinoma	135
Neuroendocrine neoplasms and paraganglioma	23	Verrucous carcinoma	141
		Basaloid squamous cell carcinoma	144
		Papillary squamous cell carcinoma	146
		Spindle cell squamous cell carcinoma	148
		Adenosquamous carcinoma	151
		Lymphoepithelial carcinoma of the larynx	153
		Mesenchymal tumours unique to the hypopharynx, larynx, trachea, and parapharyngeal space	
		Laryngeal cartilaginous tumours	155
		<b>4 Salivary gland tumours</b>	<b>159</b>
		Introduction	160
		Non-neoplastic epithelial lesions	
		Nodular oncocytic hyperplasia	163
		Lymphoepithelial sialadenitis	165
		Benign epithelial tumours	
		Pleomorphic adenoma	167
		Basal cell adenoma	171
		Warthin tumour	173
		Oncocytoma	176
		Salivary gland myoepithelioma	178
		Canalicular adenoma	180
		Cystadenoma of the salivary glands	182
		Ductal papillomas	184
		Sialadenoma papilliferum	186
		Lymphadenoma	188
		Sebaceous adenoma	190
		Intercalated duct adenoma and hyperplasia	192
<b>TNM staging of head and neck tumours</b>	<b>24</b>		
Carcinomas of the lip and oral cavity	25		
Carcinomas of the pharynx (oropharynx, nasopharynx, hypopharynx)	27		
Carcinomas of the larynx	30		
Carcinomas of the nasal cavity and paranasal sinuses	32		
Unknown primary carcinoma – cervical nodes	34		
Mucosal melanoma of the upper aerodigestive tract	36		
Carcinomas of the major salivary glands	37		
<b>TNM staging of skin carcinoma of the head and neck</b>	<b>39</b>		
<b>TNM staging of melanoma of the skin</b>	<b>41</b>		
<b>TNM staging of Merkel cell carcinoma of the skin</b>	<b>43</b>		
<b>1 Nasal, paranasal, and skull base tumours</b>	<b>45</b>		
Introduction	46		
Hamartomas			
Respiratory epithelial adenomatoid hamartoma	47		
Seromucinous hamartoma	49		
Nasal chondromesenchymal hamartoma	51		
Respiratory epithelial lesions			
Sinonasal papillomas			
Sinonasal papilloma, inverted	53		
Sinonasal papilloma, oncocytic	56		
Sinonasal papilloma, exophytic	58		
Carcinomas			
Keratinizing squamous cell carcinoma	60		
Non-keratinizing squamous cell carcinoma	62		
NUT carcinoma	65		
SWI/SNF complex-deficient sinonasal carcinoma	68		
Sinonasal lymphoepithelial carcinoma	71		
Sinonasal undifferentiated carcinoma	73		
Teratocarcinosarcoma	76		
HPV-related multiphenotypic sinonasal carcinoma	79		

Striated duct adenoma	194	Inflammatory collateral cysts	311
Sclerosing polycystic adenoma	196	Surgical ciliated cyst	313
Keratocystoma	198	Nasopalatine duct cyst	315
Malignant epithelial tumours		Gingival cysts	317
Mucoepidermoid carcinoma	200	Dentigerous cyst	319
Adenoid cystic carcinoma	204	Orthokeratinized odontogenic cyst	321
Acinic cell carcinoma	207	Lateral periodontal cyst and botryoid odonto-	
Secretory carcinoma	210	genetic cyst	328
Microsecretory adenocarcinoma	213	Calcifying odontogenic cyst	325
Polymorphous adenocarcinoma	215	Glandular odontogenic cyst	327
Hyalinizing clear cell carcinoma	218	Odontogenic keratocyst	329
Basal cell adenocarcinoma	220	Odontogenic tumours	
Intraductal carcinoma	222	Benign epithelial odontogenic tumours	
Salivary duct carcinoma	225	Adenomatoid odontogenic tumour	331
Myoepithelial carcinoma	228	Squamous odontogenic tumour	333
Epithelial-myoepithelial carcinoma	230	Calcifying epithelial odontogenic tumour	335
Mucinous adenocarcinoma	232	Ameloblastoma, unicystic	337
Sclerosing microcystic adenocarcinoma	234	Ameloblastoma, extraosseous/peripheral	339
Carcinoma ex pleomorphic adenoma	236	Ameloblastoma, conventional	341
Carcinosarcoma of the salivary glands	238	Adenoid ameloblastoma	344
Sebaceous adenocarcinoma	240	Metastasizing ameloblastoma	346
Lymphoepithelial carcinoma	242	Benign mixed epithelial and mesenchymal odontogenic	
Squamous cell carcinoma	244	tumours	
Sialoblastoma	246	Odontoma	348
Salivary gland carcinoma NOS	248	Primordial odontogenic tumour	350
Mesenchymal tumours specific to the salivary glands		Ameloblastic fibroma	352
Sialolipoma	250	Dentinogenic ghost cell tumour	354
<b>5 Oral cavity and mobile tongue tumours</b>	<b>253</b>	Benign mesenchymal odontogenic tumours	
Introduction	254	Odontogenic fibroma	356
Non-neoplastic lesions		Cementoblastoma	358
Necrotizing sialometaplasia	255	Cemento-ossifying fibroma	360
Multifocal epithelial hyperplasia	257	Odontogenic myxoma	362
Oral melanoacanthoma	259	Malignant odontogenic tumours	
Epithelial tumours		Sclerosing odontogenic carcinoma	364
Papillomas		Ameloblastic carcinoma	366
Squamous papilloma	261	Clear cell odontogenic carcinoma	369
Oral potentially malignant disorders and oral epithelial		Ghost cell odontogenic carcinoma	371
dysplasia		Primary intraosseous carcinoma NOS	373
Oral potentially malignant disorders	264	Odontogenic carcinosarcoma	375
Proliferative verrucous leukoplakia	266	Odontogenic sarcomas	377
Submucous fibrosis	269	Giant cell lesions and bone cysts	
Oral epithelial dysplasia	272	Central giant cell granuloma	379
HPV-associated oral epithelial dysplasia	275	Peripheral giant cell granuloma	381
Squamous cell carcinomas		Cherubism	383
Oral squamous cell carcinoma	277	Aneurysmal bone cyst	385
Verrucous carcinoma	282	Simple bone cyst	387
Carcinoma cuniculatum	284	Bone and cartilage tumours	
Tumours of uncertain histogenesis		Fibro-osseous tumours and dysplasias	
Congenital granular cell epulis	286	Cemento-osseous dysplasia	389
Granular cell tumour	288	Segmental odontomaxillary dysplasia	391
Ectomesenchymal chondromyxoid tumour	290	Fibrous dysplasia	393
Melanotic neuroectodermal tumour of infancy	292	Juvenile trabecular ossifying fibroma	395
		Psammomatoid ossifying fibroma	397
		Familial gigantiform cementoma	399
<b>6 Oropharyngeal tumours</b>	<b>295</b>	Benign maxillofacial bone and cartilage tumours	
Introduction	296	Osteoma	401
Benign oropharyngeal lesions		Osteochondroma	403
Hamartomatous polyps	297	Osteoblastoma	405
Epithelial tumours		Chondroblastoma	407
Squamous cell carcinoma		Chondromyxoid fibroma	409
Squamous cell carcinoma, HPV-associated	299	Desmoplastic fibroma of bone	411
Squamous cell carcinoma, HPV-independent	303	Malignant maxillofacial bone and cartilage tumours	
<b>7 Odontogenic and maxillofacial bone tumours</b>	<b>305</b>	Osteosarcoma of the jaw	413
Introduction	306	The chondrosarcoma family of tumours	416
Cysts of the jaws		Mesenchymal chondrosarcoma	418
Radicular cyst	309	Rhabdomyosarcoma with <i>TFCP2</i> rearrangement	420

**PART B (IN THIS BOOK)**

<b>8 Ear tumours</b>	<b>423</b>	<i>GLI1</i> -altered soft tissue tumour	530
Introduction	424	Undifferentiated sarcoma	532
Tumours of the external auditory canal		Undifferentiated small round cell sarcomas of bone and soft tissue	
Chondrodermatitis nodularis chronica helioides	425	Ewing sarcoma	534
Cystic chondromalacia	427		
Exostosis (osteoma) of the ear	429	<b>10 Haematolymphoid proliferations and neoplasia</b>	<b>537</b>
Ceruminous adenoma	431	Introduction	538
Ceruminous adenocarcinoma	434	Reactive haematolymphoid and related lesions	
Squamous cell carcinoma of the external auditory canal	437	Reactive lymphoid hyperplasia	539
Tumours of the middle and inner ear		EBV-positive mucocutaneous ulcer	544
Otosclerosis	439	IgG4-related disease	546
Cholesteatoma	441	Myeloid tumours	
Middle ear papilloma	443	Myeloid sarcoma	551
Vestibular schwannoma	445	B-cell lymphomas	
Middle ear neuroendocrine tumour	448	Extranodal marginal zone lymphoma of mucosa-associated lymphoid tissue	553
Endolymphatic sac tumour	451	Mantle cell lymphoma	556
Middle ear squamous cell carcinoma	454	Follicular lymphoma	558
Middle ear adenocarcinoma	457	Paediatric-type follicular lymphoma	561
		Large B-cell lymphoma with <i>IRF4</i> rearrangement	563
		Diffuse large B-cell lymphoma	565
<b>9 Soft tissue tumours</b>	<b>459</b>	Burkitt lymphoma	567
Introduction	460	Plasmacytoma	570
Adipocytic tumours		Plasmablastic lymphoma	573
Lipomas	461	T/NK-cell tumours	
Liposarcomas	464	T-lymphoblastic leukaemia/lymphoma	575
Fibroblastic and myofibroblastic tumours		Primary mucosal CD30-positive T-cell lymphoproliferative disorder	578
Nodular fasciitis	466	Extranodal NK/T-cell lymphoma	580
Desmoid fibromatosis	468	Hodgkin lymphoma	
Solitary fibrous tumour	470	Hodgkin lymphoma	583
Low-grade myofibroblastic sarcoma	473	Histiocytic and dendritic cell tumours	
Inflammatory myofibroblastic tumour	475	Juvenile xanthogranuloma	586
Vascular tumours		Erdheim-Chester disease	588
Haemangioma	478	Rosai-Dorfman disease	592
Epithelioid haemangioma	480	Langerhans cell histiocytosis	594
Lymphangioma	482	Follicular dendritic cell sarcoma	596
Epithelioid haemangioendothelioma	484		
Kaposi sarcoma	486	<b>11 Melanocytic tumours</b>	<b>599</b>
Angiosarcoma	489	Introduction	600
Pericytic (perivascular) tumours		Mucosal melanoma	601
Myofibroma	491		
Smooth muscle tumours		<b>12 Tumours and tumour-like lesions of the neck and lymph nodes</b>	<b>605</b>
Leiomyoma and angioleiomyoma	493	Introduction	606
EBV-associated smooth muscle tumour	495	Cysts and cyst-like lesions	
Smooth muscle tumour of uncertain malignant potential	497	Ranula	607
Leiomyosarcoma	499	Lymphoepithelial cyst	609
Skeletal muscle tumours		Branchial cleft cyst	611
Rhabdomyoma	502	Thyroglossal duct cyst	613
Rhabdomyosarcomas	505	Dermoid and teratoid cysts	616
Chondro-osseous tumours		Other tumours of the neck and lymph nodes	
Chondroma	509	Branchioma	618
Peripheral nerve sheath tumours		Heterotopia-associated carcinoma	621
Neurofibroma	511	Squamous carcinoma of unknown primary site	624
Schwannoma	513		
Neuromas	516	<b>13 Germ cell tumours</b>	<b>627</b>
Malignant peripheral nerve sheath tumour	518	Germ cell tumours of the head and neck	628
Tumours of uncertain differentiation			
Phosphaturic mesenchymal tumour	520	<b>14 Metastasis</b>	<b>631</b>
Myxoma	523	Metastases to the head and neck region	632
Extraskeletal myxoid chondrosarcoma	525		
Synovial sarcoma	527		

<b>15 Neuroendocrine neoplasms and paraganglioma</b>	<b>637</b>		
Introduction	638		
Neuroendocrine neoplasms			
Neuroendocrine tumours			
Neuroendocrine tumour	640		
Ectopic or invasive PitNET / pituitary adenoma	642		
Neuroendocrine carcinomas			
Small cell neuroendocrine carcinoma	646		
Large cell neuroendocrine carcinoma	649		
Merkel cell carcinoma	652		
Paraganglion tumours			
Head and neck paraganglioma	655		
<b>16 Genetic tumour syndromes involving the head and neck</b>	<b>659</b>		
Introduction	660		
Naevoid basal cell carcinoma syndrome (Gorlin syndrome)	663		
Neurofibromatosis type 1	666		
Familial adenomatous polyposis, extraintestinal manifestations	669		
Brooke–Spiegler syndrome	671		
Cowden syndrome	673		
Familial paraganglioma syndromes	676		
		Multiple endocrine neoplasia type 2	680
		Hyperparathyroidism–jaw tumour syndrome	683
		Li–Fraumeni syndrome	686
		Fanconi anaemia	688
		Dyskeratosis congenita	691
		Ataxia–telangiectasia	693
		Bloom syndrome	695
		Von Hippel–Lindau syndrome	696
		Tuberous sclerosis	699
		<b>Contributors</b>	<b>701</b>
		<b>Declaration of interests</b>	<b>710</b>
		<b>IARC/WHO Committee for ICD-O</b>	<b>711</b>
		<b>Sources</b>	<b>712</b>
		<b>References</b>	<b>721</b>
		<b>Subject index</b>	<b>799</b>
		<b>Previous volumes in the series</b>	<b>809</b>