Contents

Pease 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).

List of abbreviations Myelodysplastic/myeloproliferative neoplasm with xi SF3B1 mutation and thrombocytosis 105 Foreword xii Myelodysplastic/myeloproliferative neoplasm NOS (unclassifiable) 108 FARTA (IN ACCOMPANYING BOOK) Acute myeloid leukaemia Introduction 110 CO-O topographical coding Acute myeloid leukaemia with defining genetic abnormalities CD-O morphological coding Acute promyelocytic leukaemia with PML::RARA eloid proliferations and neoplasms 2 112 fusion shonory

-stocytic/dendritic cell neoplasms	5		Acute myeloid leukaemia with RUNX1::RUNX1T1	
E-cellymphoid proliferations and lymphomas	6		fusion	115
T-cell and NK-cell lymphoid proliferations and lymphomas	10		Acute myeloid leukaemia with CBFB:: MYH11 fusion	118
Stora-derived neoplasms of lymphoid tissues	12		Acute myeloid leukaemia with DEK::NUP214 fusion	121
			Acute myeloid leukaemia with RBM15::MRTFA fusion	123
General introduction to haematolymphoid tumours	13		Acute myeloid leukaemia with BCR:: ABL1 fusion	125
			Acute myeloid leukaemia with KMT2A rearrangement	127
Intelloid proliferations and neoplasms	17		Acute myeloid leukaemia with MECOM	
eloid precursor lesions			rearrangement	130
Cional haematopoiesis			Acute myeloid leukaemia with NUP98 rearrangement	133
Introduction	19		Acute myeloid leukaemia with NPM1 mutation	135
Cional haematopoiesis	20		Acute myeloid leukaemia with CEBPA mutation	138
Cional cytopenia of undetermined significance	24		Acute myeloid leukaemia, myelodysplasia-related	140
e coroliferative neoplasms			Acute myeloid leukaemia with other defined	
Released iferative neoplasms			genetic alterations	144
Introduction	26		Acute myeloid leukaemia defined by differentiation	
Chronic myeloid leukaemia	28		Acute myeloid leukaemia with minimal differentiation	147
Chronic neutrophilic leukaemia	34		Acute myeloid leukaemia without maturation	149
Chronic eosinophilic leukaemia	37		Acute myeloid leukaemia with maturation	151
Polycythaemia vera	40		Acute basophilic leukaemia	153
Essential thrombocythaemia	44		Acute myelomonocytic leukaemia	155
Primary myelofibrosis	47		Acute monocytic leukaemia	157
Juvenile myelomonocytic leukaemia	53		Acute erythroid leukaemia	160
Meloproliferative neoplasm NOS (unclassifiable)	57	250	Acute megakaryoblastic leukaemia	162
Folloularivervologia			Myeloid sarcoma	
Introduction	60		Myeloid sarcoma	165
Outaneous mastocytosis	61	Se	econdary myeloid neoplasms	
Systemic mastocytosis	64		Myeloid neoplasms and proliferations associated with	
Mast cell sarcoma	71		antecedent or predisposing conditions	
e consplastic neoplasms		285	Introduction	169
Cutements indicte matrix i antiser and an anti-	73		Myeloid neoplasm post cytotoxic therapy	170
le cousplastic neoplasms with defining genetic			Myeloid neoplasms associated with germline	
abnormalities	508		predisposition	173
line odysplastic neoplasm with low blasts and			Myeloid proliferations associated with Down	
5q deletion	76		syndrome	182
Melodysplastic neoplasm with low blasts and		My	/eloid/lymphoid neoplasms	
SF3B1 mutation	78		Myeloid/lymphoid neoplasms with eosinophilia and	
Melocysplastic neoplasm with biallelic TP53			tyrosine kinase gene fusions	
inactivation	80		Introduction	184
e consplastic neoplasms defined morphologically			Myeloid/lymphoid neoplasm with PDGFRA	
le covsplastic neoplasm with low blasts	83		rearrangement	185
lecorsplastic neoplasm, hypoplastic	85		Myeloid/lymphoid neoplasm with PDGFRB	
seconsplastic neoplasm with increased blasts	87		rearrangement	188
eccusolastic neoplasms of childhood			Myeloid/lymphoid neoplasm with FGFR1	
Charles and myelodysplastic neoplasm with low blasts	90		rearrangement	191

93

95

96

Choosed myelodysplastic neoplasm with increased blasts meeloproliferative neoplasms moduction Concorne myelomonocytic leukaemia ecc.solastic/myeloproliferative neoplasm with neutrophilia 102

Myeloid/lymphoid neoplasm with JAK2 194 rearrangement Myeloid/lymphoid neoplasm with FLT3 rearrangement 197 Myeloid/lymphoid neoplasm with ETV6:: ABL1 fusion 199 Myeloid/lymphoid neoplasms with other tyrosine kinase gene fusions 201

4.0

	Acute leukaemias of mixed or ambiguous lineage		B-lymphoblastic leukaemia/lymphoma with	
	Introduction	203	BCR:: ABL1 fusion	342
	Acute leukaemia of ambiguous lineage with defining		B-lymphoblastic leukaemia/lymphoma with	
	genetic abnormalities		BCR:: ABL1-like features	345
	Mixed-phenotype acute leukaemia with		B-lymphoblastic leukaemia/lymphoma with	BA LITER
	BCR:: ABL1 fusion	206	KMT2A rearrangement	348
	Mixed-phenotype acute leukaemia with KMT2A		B-lymphoblastic leukaemia/lymphoma with	
	rearrangement	208	ETV6::RUNX1 fusion	350
	Acute leukaemia of ambiguous lineage with other		B-lymphoblastic leukaemia/lymphoma with	000
	defined genetic alterations	210	ETV6::RUNX1-like features	352
	Acute leukaemia of ambiguous lineage defined		B-lymphoblastic leukaemia/lymphoma with	OOL
	immunophenotypically		TCF3::PBX1 fusion	354
	Mixed-phenotype acute leukaemia, B/myeloid	212	B-lymphoblastic leukaemia/lymphoma with	001
	Mixed-phenotype acute leukaemia, T/myeloid	214	IGH::/L3 fusion	356
	Mixed-phenotype acute leukaemia, rare types	216	B-lymphoblastic leukaemia/lymphoma with	000
	Acute leukaemia of ambiguous lineage NOS	218	TCF3::HLF fusion	358
	Acute undifferentiated leukaemia	219	B-lymphoblastic leukaemia/lymphoma with other	550
	Acute unumerentiateu ieukaenna	219		260
0	Listicautio/dondritio call noonlooma	001	defined genetic alterations	360
3	Histiocytic/dendritic cell neoplasms	221	B-lymphoblastic leukaemia/lymphoma NOS	364
	Introduction	222	Mature B-cell neoplasms	
	Plasmacytoid dendritic cell neoplasms		Preneoplastic and neoplastic small lymphocytic prolife	
	Plasmacytoid dendritic cell neoplasms		Introduction	365
	Mature plasmacytoid dendritic cell proliferation		Monoclonal B-cell lymphocytosis	366
	associated with myeloid neoplasm	224	Chronic lymphocytic leukaemia / small	
	Blastic plasmacytoid dendritic cell neoplasm	226	lymphocytic lymphoma	368
	Langerhans cell and other dendritic cell neoplasms		Splenic B-cell lymphomas and leukaemias	
	Langerhans cell neoplasms		Introduction	378
	Langerhans cell histiocytosis	230	Hairy cell leukaemia	380
	Langerhans cell sarcoma	234	Splenic marginal zone lymphoma	385
		204		389
	Other dendritic cell neoplasms	007	Splenic diffuse red pulp small B-cell lymphoma	309
	Indeterminate dendritic cell tumour	237	Splenic B-cell lymphoma/leukaemia with	004
	Interdigitating dendritic cell sarcoma	240	prominent nucleoli	391
	Histiocyte/macrophage neoplasms		Lymphoplasmacytic lymphoma	
	Histiocytic neoplasms		Lymphoplasmacytic lymphoma	394
	Juvenile xanthogranuloma	242	Marginal zone lymphoma	- +
	Erdheim-Chester disease	245	Introduction	399
	Rosai–Dorfman disease	248	Extranodal marginal zone lymphoma of	
	ALK-positive histiocytosis	252	mucosa-associated lymphoid tissue	401
	Histiocytic sarcoma	255	Primary cutaneous marginal zone lymphoma	409
			Nodal marginal zone lymphoma	412
Soi	urces (Part A)	259	Paediatric nodal marginal zone lymphoma	417
~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~~		200	Follicular lymphoma	111
ont	foronoon (Part A)	263	Introduction	120
161	ferences (Part A)	200		420
			In situ follicular B-cell neoplasm	421
PA	RT B (IN THIS BOOK)		Follicular lymphoma	423
			Paediatric-type follicular lymphoma	434
	B-cell lymphoid proliferations and lymphomas	295	Duodenal-type follicular lymphoma	437
	Introduction to B-cell lymphoproliferative disorders and		Cutaneous follicle centre lymphoma	
	neoplasms	297	Primary cutaneous follicle centre lymphoma	440
	Tumour-like lesions with B-cell predominance		Mantle cell lymphoma	
	Introduction	305	Introduction	443
	Reactive B-cell-rich lymphoid proliferations that	niernanoni	In situ mantle cell neoplasm	444
	can mimic lymphoma	309	Mantle cell lymphoma	446
	IgG4-related disease	315	Leukaemic non-nodal mantle cell lymphoma	453
	•			400
	Unicentric Castleman disease	319	Transformations of indolent B-cell lymphomas	455
	Idiopathic multicentric Castleman disease	322		455
	KSHV/HHV8-associated multicentric Castleman		Transformations of indolent B-cell lymphomas	456
	disease	325	Large B-cell lymphomas	
	Precursor B-cell neoplasms		Introduction	461
	B-lymphoblastic leukaemias/lymphomas		Diffuse large B-cell lymphoma NOS	463
	Introduction	328	T-cell/histiocyte-rich large B-cell lymphoma	473
	B-lymphoblastic leukaemia/lymphoma	330	Diffuse large B-cell lymphoma / high-grade B-cell	

B-cell lymphoid proliferations and lymphomas	2
Introduction to B-cell lymphoproliferative disorders and	
neoplasms	7-13
Tumour-like lesions with B-cell predominance	
Introduction	
Reactive B-cell-rich lymphoid proliferations that	
can mimic lymphoma	:
IgG4-related disease	
Unicentric Castleman disease	
Idiopathic multicentric Castleman disease	:
KSHV/HHV8-associated multicentric Castleman	
disease	
Precursor B-cell neoplasms	
B-lymphoblastic leukaemias/lymphomas	
Introduction	
B-lymphoblastic leukaemia/lymphoma	
Digriphobiastic icultaerina/igriphoma	

B-lymphoblastic leukaemia/lymphoma with high hyperdiploidy 336 B-lymphoblastic leukaemia/lymphoma with hypodiploidy 338 B-lymphoblastic leukaemia/lymphoma with iAMP21 340

lymphoma with MYC and BCL2 rearrangements 476 ALK-positive large B-cell lymphoma 481 Large B-cell lymphoma with IRF4 rearrangement 484 High-grade B-cell lymphoma with 11q aberration 486 Lymphomatoid granulomatosis 489 EBV-positive diffuse large B-cell lymphoma 493

Diffuse large B-cell lymphoma associated with chronic inflammation 497 Fiorin-associated large B-cell lymphoma 500 Fuid overload-associated large B-cell lymphoma 503 Plasmablastic lymphoma 506 Primary large B-cell lymphoma of immune-privileged sites 509 Primary cutaneous diffuse large B-cell lymphoma, 517 leg type Intravascular large B-cell lymphoma 520 Primary mediastinal large B-cell lymphoma 523 Mediastinal grey zone lymphoma 527 High-grade B-cell lymphoma NOS 530 Burkitt lymphoma Burkitt lymphoma 532 

Indolent T-lymphoblastic proliferation	649
Precursor T-cell neoplasms	
T-lymphoblastic leukaemia/lymphoma	
T-lymphoblastic leukaemia/lymphoma NOS	651
Early T-precursor lymphoblastic	6 Strong
leukaemia/lymphoma	656
	0.00
Mature T-cell and NK-cell neoplasms	
Mature T-cell and NK-cell leukaemias	0.5.0
Introduction	658
T-prolymphocytic leukaemia	659
T-large granular lymphocytic leukaemia	662
NK-large granular lymphocytic leukaemia	665
Adult T-cell leukaemia/lymphoma	667
Sézary syndrome	674
Aggressive NK-cell leukaemia	677
Primary cutaneous T-cell lymphoid proliferations a	
	lindende
lymphomas	000
Introduction to primary cutaneous T-cell neople	
Primary cutaneous CD4-positive small or medi	
T-cell lymphoproliferative disorder	682
Primary cutaneous acral CD8-positive T-cell	
lymphoproliferative disorder	685
Mycosis fungoides	688
Primary cutaneous CD30-positive T-cell	
lymphoproliferative disorder: Lymphomatoid	
papulosis	694
	034
Primary cutaneous CD30-positive T-cell	
lymphoproliferative disorder: Primary cutane	
anaplastic large cell lymphoma	698
Subcutaneous panniculitis-like T-cell lymphom	na 700
Primary cutaneous gamma-delta T-cell lympho	oma 702
Primary cutaneous CD8-positive aggressive	
epidermotropic cytotoxic T-cell lymphoma	705
Primary cutaneous peripheral T-cell lymphoma	a NOS 708
Intestinal T-cell and NK-cell lymphoid proliferation	
lymphomas	ound
Introduction	710
Indolent T-cell lymphoma of the gastrointestina	
Indolent NK-cell lymphoproliferative disorder of	
the gastrointestinal tract	715
Enteropathy-associated T-cell lymphoma	717
Monomorphic epitheliotropic intestinal T-cell	
lymphoma	722
Intestinal T-cell lymphoma NOS	725
Hepatosplenic T-cell lymphoma	
Hepatosplenic T-cell lymphoma	727
	121
Anaplastic large cell lymphoma	700
Introduction	730
ALK-positive anaplastic large cell lymphoma	731
ALK-negative anaplastic large cell lymphoma	734
Breast implant-associated anaplastic large ce	
lymphoma	738
Nodal T follicular helper cell lymphoma	
Introduction	742
Nodal T follicular helper cell lymphoma,	
angioimmunoblastic type	744
	744
Nodal T follicular helper cell lymphoma,	750
follicular type	752
Nodal T follicular helper cell lymphoma NOS	755
Other peripheral T-cell lymphomas	
Peripheral T-cell lymphoma NOS	757
EBV-positive T-cell and NK-cell lymphomas	

and lymphomas

Introduction	539
Primary effusion lymphoma	541
SHV/HHV8-positive diffuse large B-cell lymphoma	545
SHV/HHV8-positive germinotropic	
In choproliferative disorder	547
Lumehold proliferations and lymphomas associated	
immune deficiency and dysregulation	
Introduction	549
Hyperplasias arising in immune	010
deficiency/dysregulation	555
Polymorphic lymphoproliferative disorders arising	000
in immune deficiency/dysregulation	561
EBV-positive mucocutaneous ulcer	565
	505
Lymphomas arising in immune	ECO
deficiency/dysregulation	568
Inborn error of immunity-associated lymphoid	570
proliferations and lymphomas	573
Hodgkin lymphoma	570
Introduction	579
Classic Hodgkin lymphoma	580
Nodular lymphocyte-predominant Hodgkin	504
lymphoma	591
Plasma cell neoplasms and other diseases with paraproteins	
Introduction	598
Monoclonal gammopathies	
Cold agglutinin disease	599
IgM monoclonal gammopathy of undetermined	
significance	602
Non-IgM monoclonal gammopathy of	
undetermined significance	603
Monoclonal gammopathy of renal significance	605
Diseases with monoclonal immunoglobulin deposition	
Immunoglobulin-related amyloidosis (AL amyloidosis)	608
Monoclonal immunoglobulin deposition disease	611
Heavy chain diseases	
Introduction	613
Mu heavy chain disease	615
Gamma heavy chain disease	617
Alpha heavy chain disease	619
Plasma cell neoplasms	
Introduction	621
Plasmacytoma	622
Plasma cell myeloma / multiple myeloma	625
Plasma cell neoplasms with associated	
paraneoplastic syndrome	631

### T-cell and NK-cell lymphoid proliferations and lymphomas 635 5 637 Immoduction Turnour-like lesions with T-cell predominance Kikuchi-Fujimoto disease 643 Autoimmune lymphoproliferative syndrome 646

EBV-positive nodal T- and NK-cell lymphoma 762 Extranodal NK/T-cell lymphoma 765 EBV-positive T-cell and NK-cell lymphoid proliferations and lymphomas of childhood 770 Introduction Severe mosquito bite allergy 772

	Hydroa vacciniforme lymphoproliferative disorder	77
	Systemic chronic active EBV disease	77
	Systemic EBV-positive T-cell lymphoma of childhood	779
6	Stroma-derived neoplasms of lymphoid tissues	78
	Introduction	78
	Mesenchymal dendritic cell neoplasms	
	Follicular dendritic cell neoplasms	
	Follicular dendritic cell sarcoma	78
	EBV-positive inflammatory follicular dendritic cell	
	sarcoma	79
	Fibroblastic reticular cell tumour	794
	Myofibroblastic tumours	
	Myofibroblastic tumours	
	Intranodal palisaded myofibroblastoma	796
	Spleen-specific vascular-stromal tumours	
	Splenic vascular-stromal tumours	

7 Gene	etic tumour syndromes ass	sociated with	
ha	ematolymphoid tumours		
	Introduction		
	Fanconi anaemia		
	Ataxia-telangiectasia		
Contribu		eu/ismohoaaqehdel	
158			
Declarat			
ARC/W	HO Committee for ICD-O		
sources	(Part B)		
Deferen			
reieren	ces (Part B)		

Littoral cell angioma Splenic hamartoma	798 800
Sclerosing angiomatoid nodular transformation of	
spleen	802
Ivmphopolitentre-moordert-with ant-supportertier	
Frimary cotopies (C2330, positive, Ficelians	

## Subject index

## Previous volumes in the series

prolitionatory: and fyritementar resoon the ubindebaieres/tence dysong failer and taken and the strengthener and streng and the strengthener and signey/dysregulation and the strengthener and the strengthener branch and the str

Classicibledigido (minora manani landoni la Classicibledigido (minora manani landoni la Modula dyangi nocita-nyudominani Modgiani Ilymatiotopia socita-nyudominani Modgiani 930

841

805

806

807

809

810

812

815

828

833

835

ipheral T-cell lymphoma MOS no 20 Lateriqu		
osti vaniseri anno 1000 - Coo-Siddanar (moniseri)		

Additive most internet and With solil Innet one Extranodal NK/T-cell lymphomeses and an all EBV-positive Tradit and NK-cell Tymphomeses and and lymphomas of childhood Savere mosquito bite allergy		
	*	