

#### Illustrated pathology of the spleen

The major aim of this book is to de-mystify splenic pathology for non-specialist consultants and trainees in histopathology. The interpretation of pathological changes in the human spleen provides a significant challenge to the skills of even the most highly trained histopathologists. This comprehensive account of the most important and frequently encountered pathological conditions affecting the spleen is highly illustrated in colour throughout, with numerous detailed macroscopic and microscopic specimens. Emphasis is placed upon the importance of adequate clinical information and technical preparation of splenectomy specimens in obtaining the best possible histopathological assessment of the tissue. A systematic, analytical approach to interpretation of pathological changes is used throughout the book. This should enable readers to feel confident in their assessment of significant processes within the spleen

This volume serves as an illustrated atlas, as a bench manual, as a text and as a source of selected reference. It will be an indispensable guide.

Dennis Wright was Professor of Pathology at Southampton University from 1972, when the medical school was founded, until his retirement in 1995. Since then he has held the title of Emeritus Professor. Previously, he had been a Reader in Pathology at Birmingham University, preceded by a Readership at Makerere University in Uganda. He is a past President of the European Haematopathology Society, his election to this post being a reflection of his international reputation in the field of lympho-reticular pathology. He has maintained an interest in splenic pathology, particularly lymphomas involving the spleen, for many

Bridget Wilkins is a Clinical Senior Lecturer in Pathology at Southampton University and an Honorary Consultant Histopathologist for the Southampton University Hospitals NHS Trust. Previously, she has held posts as a Clinical Lecturer and a Clinical Research Fellow at Southampton University. She specializes in haematopathology, with particular interests in pathology of the spleen and bone marrow. She teaches regularly throughout the UK on these topics, emphasizing an analytical and functional approach to histological interpretation.



# Illustrated pathology of

# THE SPLEEN

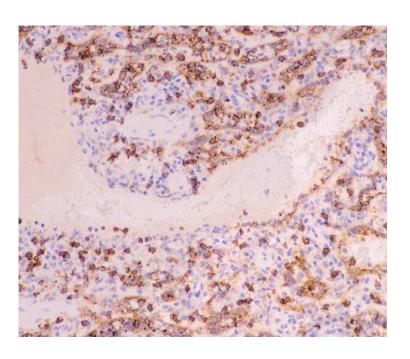
### Bridget S. Wilkins

Senior Lecturer Department of Microbiology and Pathology University of Southampton, UK

#### and

Dennis H. Wright

Emeritus Professor of Pathology University of Southampton, UK







 ${\tt PUBLISHED~BY~THE~PRESS~SYNDICATE~OF~THE~UNIVERSITY~OF~CAMBRIDGE~The~Pitt~Building,} Trumpington~Street, Cambridge, United~Kingdom$ 

CAMBRIDGE UNIVERSITY PRESS

The Edinburgh Building, Cambridge CB2 2RU, UK http://www.cup.cam.ac.uk 40 West 20th Street, New York, NY 10011-4211, USA http://www.cup.org 10 Stamford Road, Oakleigh, Melbourne 3166, Australia Ruiz do Alarcón 13, 28014 Madrid, Spain

© Bridget S. Wilkins & Dennis H. Wright 2000

This book is in copyright. Subject to statutory exception and to the provisions of relevant collective licensing agreements, no reproduction of any part may take place without the written permission of Cambridge University Press.

First published 2000

Printed in the United Kingdom at the University Press, Cambridge

Typeface Utopia 9/13pt. System QuarkXPress [SE]

A catalogue record for this book is available from the British Library

Libary of Congress Cataloguing in Publication data

ISBN 0 521 62227 1 hardback

Every effort has been made in preparing this book to provide accurate and up-to-date information which is in accord with accepted standards and practice at the time of publication. Nevertheless, the authors, editors and publisher can make no warranties that the information contained herein is totally free from error, not least because clinical standards are constantly changing through research and regulation. The authors, editors and publisher therefore disclaim all liability for direct or consequential damages resulting from the use of material contained in this book. The reader is strongly advised to pay careful attention to information provided by the manufacturer of any drugs or equipment that they plan to use



## **Contents**

ion enectomy performed? enic pathology perceived to lt? the surgeon or physician in the pathologist? emation does the pathologist exation and processing be lt to give the best possible envestigations	5 6 7 9
enectomy performed? enic pathology perceived to lt? the surgeon or physician in the pathologist? emation does the pathologist exation and processing be d to give the best possible envestigations	11 33 55 66
enectomy performed? enic pathology perceived to lt? the surgeon or physician in the pathologist? emation does the pathologist exation and processing be d to give the best possible envestigations	11 33 55 66
enic pathology perceived to lt? the surgeon or physician in the pathologist? mation does the pathologist exation and processing be d to give the best possible envestigations	3 5 6
enic pathology perceived to lt? the surgeon or physician in the pathologist? mation does the pathologist exation and processing be d to give the best possible envestigations	3 5 6
the surgeon or physician the pathologist? mation does the pathologist xation and processing be d to give the best possible?	5 6 7 9
n the pathologist? mation does the pathologist  xation and processing be d to give the best possible ? nvestigations	77 99
mation does the pathologist  xation and processing be d to give the best possible  nvestigations	77 99
xation and processing be d to give the best possible ? nvestigations	7 9
d to give the best possible ? nvestigations	7 9
d to give the best possible ? nvestigations	9
? nvestigations	9
nvestigations	9
•	
	1.0
ppsy, fine needle aspiration	10
oscopic splenectomy	10
ns of splenectomy for the	
	11
	12
3	12
tructure, development	
tions of the spleen	13
ınd vascular supply	13
icro-anatomy of the spleen	14
ent of the spleen during ic and fetal life	17
l correlates of red and white	19
	23
cture	24
cture ties of splenic function	25
	aic and fetal life al correlates of red and white acture ities of splenic function nism



Contents	vi	

	An immunohistochemical inventory of the spleen References	27 31
3	Post-traumatic and incidentally removed spleens	35
	Introduction Speens removed incidentally and those removed following trauma are	35
	different An approach to analysis of traumatized	35
	or incidental splenectomy specimens Reactive changes in individual	37
	compartments of the spleen	40
	Spontaneous rupture of the spleen	43
	References	44
4	The spleen in hereditary blood cell abnormalities and auto-immur disorders	1 <b>e</b>
	Introduction	47
	Vaccination before splenectomy	48
	Sickle cell anaemia and thalassaemias	49
	Hereditary spherocytosis and auto-	
	immune blood cell destruction	51
	The nature of HS, AIHA and ITP	52
	Macroscopic appearances of the	<b>5</b> 0
	spleen in HS, AIHA and ITP Red pulp	53 54
	White pulp	55
	Felty's syndrome	56
	Systemic lupus erythematosus	57
	Polyarteritis nodosum	59
	References	59
5	The spleen in immunodeficiency	
	and systemic infections	63
	Inherited immunodeficiency	
	syndromes	63
	Acquired immunodeficiency due to	o=
	drugs or irradiation	65



vii Contents

	Acquired immunodeficiency syndrome		
	and the spleen	66	
	Systemic infections involving the		
	spleen in immunocompetent		
	individuals	70	
	References	74	
	10101011000	• •	
6	Lymphomas involving the		
	spleen	77	
	Introduction	77	
	B cell small lymphocytic lymphoma/		
	chronic lymphocytic leukaemia		
	(B-CLL)	78	
	B-prolymphocytic leukaemia	82	
	Mantle cell lymphoma (MCL)	82	
	Follicle centre cell lymphoma	85	
	Splenic marginal zone lymphoma	87	
	Lymphoplasmacytoid lymphoma/		
	immunocytoma	91	
	Hairy cell leukaemia	92	
	Large B cell lymphomas	95	
	T cell prolymphocytic leukaemia	96	
	Large granular lymphocyte leukaemia,		
	T cell and NK cell types	97	
	Splenic T cell lymphoma with	0.	
	erythrophagocytosis	100	
	Hepatosplenic γδT cell lymphoma	100	
	Hodgkin's disease	101	
	Nodular lymphocyte and histiocyte	101	
	predominant Hodgkin's disease		
	(nodular L&H HD)	105	
	References		
	References	106	
7	The spleen in myeloproliferative		
•	disorders	111	
	disorders	111	
	Splenic extramedullary haemopoiesis	111	
	Systemic mastocytosis	117	
	Transient abnormal myelopoiesis in		
	association with Trisomy 21 (Down's		
	syndrome)	121	
	The spleen in acute myeloid and		
	lymphoblastic leukaemias	122	
	References	122	



viii
ıts

8	Pathology of the spienic stroma	125
	Splenic granulomas	125
	Storage disorders involving cordal	
	macrophages	126
	Langerhans' cell histiocytosis	136
	Splenic pseudotumours and true	
	stromal tumours	137
	Peliosis of the spleen	141
	Splenic vascular hyalinosis	144
	Portal hypertension and the spleen	144
	Splenic infarction	145
	Gamna–Gandy bodies	147
	References	147
9	Metastases and miscellaneous	
	conditions	151
	Appearances and primary origins of	
	metastatic cancers involving the spleen	ı 151
	Hamartomas	152
	Splenic cysts and false cysts	153
	Amyloidosis	154
	Congenital anomalies of splenic size,	
	number or anatomical position	155
	Splenunculi and splenosis	156
	Splenic atrophy and fibrosis	157
	Peri-splenitis ('sugar-icing' of the spleen)	159
	Autolysis in splenic tissue	160
	References	161
10	Summary: some key points in	
10	splenic differential diagnosis	163
	spienic unferential diagnosis	103
	Introduction	163
	Reactive spleen or neoplastic lymphoid	
	proliferation?	163
	Granulomas and cordal macrophage	
	disorders	173
	Extramedullary haemopoiesis – is it	
	significant or not?	176
	References	177
	Index	181



### **Preface**

The interpretation of splenic pathology is perceived as difficult by many histopathologists. Few diseases arise primarily within the spleen, and most pathology seen at this site represents involvement of the organ by processes originating elsewhere and/or also affecting other tissues in the body. Primary diagnosis has often been made from haematological, microbiological or histopathological investigations of other tissues by the time splenectomy is performed. The role of the histopathologist in most cases is to provide confirmation of the known, or suspected, diagnosis and to exclude unsuspected pathology.

Splenic pathology is an area of shared interest for histopathologists, haematologists, oncologists and surgeons. The key to successful interpretation of splenic pathology lies in obtaining adequate clinical information and in ensuring optimal tissue fixation. Dialogue between the various clinicians involved in diagnosis and care of individual patients is required to achieve these factors. Without this dialogue, no amount of expertise in splenic histology can provide full answers to clinical problems.

In this book, we have aimed to present a succinct, comprehensive account of those aspects of splenic pathology likely to be encountered by diagnostic histopathologists. We hope we have provided useful guidance to permit confident recognition of normal and non-specifically reactive histological appearances in the spleen. We have also attempted to convey principles of

ix



Preface x

systematic analysis which can be applied to achieve diagnoses following recognition of broad categories of abnormality affecting individual splenic compartments. The use of immunohistochemistry, cytogenetic and molecular genetic investigations to supplement morphological analysis is described where appropriate.

This book is aimed primarily for the use of general histopathologists at consultant and trainee levels. We hope that it will also be of value for histopathologists with specialist expertise in haematopathology, and to haematologists with an interest in this area.

We acknowledge that pathology in the spleen remains a challenging area for histopathological interpretation. For the foreseeable future, there will be occasions when no satisfactory, complete diagnosis can be made. None the less, we hope this book will convey some of our enthusiasm for the subject and that it will help make day-to-day practice of splenic diagnosis more rewarding for our fellow histopathologists. We hope also that some of our readers will feel encouraged to investigate further those aspects of splenic involvement by disease which remain unsolved at present.

**Bridget Wilkins** 

Dennis Wright



## **Acknowledgements**

We are grateful to the many colleagues who have shared their interesting and challenging cases of splenic pathology with us in recent years. The opportunity we have gained, through them, to extend our experience of splenic pathology and see examples of rare disorders involving the spleen, has contributed significantly to our interest in the subject and our impetus to write this book.

Thanks are due to Duncan Kempson and, in particular, Stephen Bottoms from the Medical Illustration Department at Southampton General Hospital, who provided invaluable assistance with photography of macroscopic specimens. We should also like to thank Julia Berry for assistance with typing.

хi