

Ocular Adnexal Lesions

A Clinical, Radiological and Pathological
Correlation

Shantha Amrith
Gangadhara Sundar
Stephanie Ming Young
Editors

Ocular Adnexal Lesions

Shantha Amrith
Gangadhara Sundar • Stephanie Ming Young
Editors

Ocular Adnexal Lesions

A Clinical, Radiological and Pathological
Correlation

Editors

Shantha Amrith
National University Hospital
National University of Singapore
Singapore

Gangadhara Sundar
National University Hospital
National University of Singapore
Singapore

Stephanie Ming Young
National University Hospital
National University of Singapore
Singapore

ISBN 978-981-13-3797-0 ISBN 978-981-13-3798-7 (eBook)
<https://doi.org/10.1007/978-981-13-3798-7>

© Springer Nature Singapore Pte Ltd. 2019

This work is subject to copyright. All rights are reserved by the Publisher, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, reuse of illustrations, recitation, broadcasting, reproduction on microfilms or in any other physical way, and transmission or information storage and retrieval, electronic adaptation, computer software, or by similar or dissimilar methodology now known or hereafter developed.

The use of general descriptive names, registered names, trademarks, service marks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.

The publisher, the authors, and the editors are safe to assume that the advice and information in this book are believed to be true and accurate at the date of publication. Neither the publisher nor the authors or the editors give a warranty, express or implied, with respect to the material contained herein or for any errors or omissions that may have been made. The publisher remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

This Springer imprint is published by the registered company Springer Nature Singapore Pte Ltd.
The registered company address is: 152 Beach Road, #21-01/04 Gateway East, Singapore 189721, Singapore

Foreword

One of the great privileges and obligations in the practice of medicine is the opportunity to engage in lifelong learning. This is particularly significant for the analysis and elucidation of complex disorders. An important vehicle for this is a multidisciplinary approach to the diagnosis and management of orbital and adnexal disorders. I learned this early in my career devoted to pathology, orbital disease, and ocular oncology. In each of these disciplines, one had to interact with colleagues in related disciplines and collate the clinical observations, imaging, and pathology to arrive at a diagnosis and treatment plan in the context of evolving knowledge and techniques. In addition, since the gamut of possible diagnoses may also include such disciplines as rheumatology, infectious disease, immunology, and oncology, it is frequently necessary to involve specialist in these disciplines. From my experience of collating disorders clinically, for diagnosis, management, teaching, presentation, and publication, I found it useful to analyze diverse findings in a disciplined order. This led to the development of the “CLOSE” technique, which has been used as a framework in this book by Professor Shantha Amrith, her coeditors Gangadhara Sundar and Stephanie Ming Young, and colleagues at NUH.

An important and valuable feature of this book is a commitment to organization and documentation of personal experience in developing knowledge and diagnostic tools. This is also a patient-centric book, which focuses on a multidisciplinary approach to diagnosis and management. Too often in our time, students and practitioners are distracted by using digital devices while trying to make a diagnosis in the presence of a patient when they should be focused on what the patient is remembering, describing, feeling, and demonstrating. Direct personal interaction is the framework for the multidisciplinary analysis elucidated in this book.

In an era where we have been increasingly dissociated from personal shared medical knowledge and memory in the interest of the patient with complex problems, it is a delight to share and benefit from their interactive experience and discipline. I am particularly impressed by the quality of discourse and illustrations provided from the clinical, imaging, pathology, and laboratory contributed by the authors. This provides and illustrates a pathway to develop engrams that are so important to the diagnosis of the wide range of disorders presented. It will surely be useful to developing your own visual and experiential memory bank.

The chapters include basic anatomy, contemporary imaging, and pathology. In addition, the authors cover the essential gamut of orbital disorders that we are likely to encounter in clinical practice including structural lesions, inflammations, vascular anomalies and lesions, benign and malignant neoplasms both regional and metastatic, and lymphoproliferative disorders. All are presented with their own case examples, in the context of interdisciplinary rounds. In addition, they provide a review of the literature pertinent to the diagnosis as well as treatment plans and outcomes. The reviews of the disorders also include recent literature as well as regional differences.

Overall, this is a readable handbook for students and clinicians who may encounter similar patients in practice. The authors are to be congratulated for sharing their practical and useful experience for those in the field as well as our students.

Jack Rootman, FRCS
Emeritus Professor Ophthalmology and
Vision Science and Pathology,
University of British Columbia,
Vancouver, BC, Canada

Preface

The Orbit and Oculofacial Service of the Department of Ophthalmology, National University Hospital, Singapore, has been conducting monthly clinical, pathological, and radiological conferences on ocular and ocular adnexal lesions with our colleagues from the Departments of Diagnostic Imaging and Pathology since 2004. The spectrum of diseases covered in these rounds typically included immunological, oncological, and other diseases with multidisciplinary inputs, not only from the radiologist and pathologist but also from specialties such as rheumatology/immunology, endocrinology, adult and pediatric oncology, neurology, and surgical specialties like facial plastic surgery, otorhinolaryngology, neurosurgery, etc. These multidisciplinary sessions contributed immensely to the knowledge and understanding of common disorders, especially for the residents, fellows, and consultants, and aided in the compilation of recent advances in diagnosis and management of rare ocular adnexal disorders.

The cases discussed in the book were carefully chosen from our archives to educate not only the ophthalmology residents and general ophthalmologists but also the residents from the diagnostic imaging and pathology departments.

The term, ocular adnexa, is used in the wider sense in this book, encompassing the eyelids, lacrimal system, and orbit including conditions of the globe with orbital extension. The book begins with a brief description of applied anatomy relevant to clinical diagnosis and surgical principles.

The sections on imaging and pathology were contributed by experts in the field. The coauthors from the Department of Diagnostic Imaging, Poh Sun Goh and Eric Ting, have written a section each on imaging with some basic principles and concepts of computed tomography (CT) and magnetic resonance imaging (MRI), special imaging techniques, and their applications, highlighting some of the indications, contraindications, advantages, and disadvantages of the different forms of imaging. In addition, they have diligently worked to choose representative images for each case with radiologic description of the images.

Similarly, insights into the basic understanding of histopathologic techniques have been provided by Drs. Min En Nga and Bingcheng Wu from the Department of Pathology, National University Hospital. The chapter also provides useful advice regarding the importance of discussing the case with the pathologist, prior to incisional or excisional tissue biopsy. The pathologists have expertly chosen the histopathological figures, along with relevant immunohistochemical stains for each condition, and provided the descriptions and figure legends.

The clinical conditions are classified by pathology rather than the anatomical structures to avoid repetitions. Common eyelid conditions, such as chalazion, papillomas, nevi, and sweat gland cysts, are not included in this book, as most ophthalmic residents and general ophthalmologists are quite conversant with these conditions. Nevertheless, warning signs have been highlighted under different conditions, so that unusual signs of benign-looking lesions could be identified for prompt and early diagnosis.

Each part is divided into chapters representing the anatomical part. All cases are discussed systematically starting with an introduction, clinical scenario, differential diagnoses, imaging features, histopathology including immunohistochemical features, molecular features (where relevant), final diagnosis, outline of management, and a brief discussion and latest evidence-based update of that condition.

After the description of the clinical scenario, there is a CLOSE summary for each case (introduced by Professor Jack Rootman), which is a mnemonic for *C*linical process, *L*ocation, *O*nset, *S*igns and symptoms, and *E*pidemiology. At the end of clinical examination, the summary helps to formulate a differential diagnosis, which in turn guides additional imaging, incisional or excisional biopsy. CLOSE is described in great detail in the introductory chapter of the book, “Orbital Surgery: A Conceptual Approach”. Each case write-up concludes with some learning points and a list of references for further reading.

Our sincere thanks and acknowledgments are due to our neuro-ophthalmology colleagues, Drs. Hazel Lin and Clement Tan for the neuro-ophthalmology section and our hematologist-oncologist Dr. Michelle Poon for the overview of lymphoproliferative disorders. Our sincere thanks also go to all our past fellows and residents, who had researched and presented with fervor in our monthly teaching rounds, most of which has been shared in the book with recent updates.

The uniqueness of this book lies in the contribution of not only the orbital surgeons/clinicians in conventional knowledge and recent advances in diagnosis and management but also the radiologists and pathologists to each case. We sincerely hope that this book will be a good teaching tool for all the aspiring and practicing orbit and oculoplastic surgeons and serve as a reference tool for residents and fellows in training institutions worldwide.

Singapore

Shantha Amrith
Gangadhara Sundar
Stephanie Ming Young

Acknowledgments

Other special thanks to our past fellows, who put in hard work to make stellar presentations during the teaching rounds: Mike Munoz (the Philippines), Passorn Preechawai (Thailand), Cheng Jin Fong (Singapore), Radwan Al Mousa (Syria), Naseem Mansurali (Malaysia), Rosniza Abdul Razak (Malaysia), Shruthi Tara (India), Tricia Morada (the Philippines), Ko Ko Lin (Myanmar), Sharifah Intan (Malaysia), Joy Chan (Singapore), Raghuraj Suresh Hegde (India), Stephanie Ming Young (Singapore), Zin May Htoon (Myanmar), Mariel Angelou Parulan (the Philippines), and Prerana Kansakar (Nepal).

Thanks also to Bay Song Lin, medical illustrator from the Department of Anatomy, National University of Singapore, who made anatomical illustrations for this book.

Contents

Part I Basic Anatomy, Imaging and Pathology

- 1 Anatomy** 3
Shantha Amrith and Stephanie Ming Young
- 2 Imaging: Computerised Tomography** 13
Poh Sun Goh
- 3 Imaging: Magnetic Resonance Imaging** 19
Yun Song Choo and Eric Ting
- 4 Pathology: Principles of Basic Histopathology** 25
Bingcheng Wu and Min En Nga

Part II Structural Lesions

- 5 Dermoid** 33
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 6 Fibro-osseous Lesions: Fibrous Dysplasia** 39
Gangadhara Sundar, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Shantha Amrith

Part III Infections

- 7 Orbital Cellulitis: Bacterial** 45
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 8 Orbital Cellulitis: Invasive Fungal** 49
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar

Part IV Inflammations

- 9 Granulomatosis with Polyangiitis (GPA)** 59
Gangadhara Sundar, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 10 Thyroid Eye Disease** 63
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar

11 IgG4-Related Ophthalmic Disease	67
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
12 Sarcoidosis	73
Gangadhara Sundar, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu, Min En Nga, and Shantha Amrith	
13 Langerhans Cell Histiocytosis	77
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
14 Xanthogranuloma	83
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
15 Kimura Disease	87
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
16 Non-specific Orbital Inflammatory Disease	91
Stephanie Ming Young, Shantha Amrith, Poh Sun Goh, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
17 Nodular Fasciitis	95
Stephanie Ming Young, Shantha Amrith, Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
Part V Vascular Lesions	
18 Vascular Tumours: Capillary Haemangioma	101
Shantha Amrith, Stephanie Ming Young, Eric Ting, and Gangadhara Sundar	
19 Lymphatic and Lymphatic-Venous Malformation	105
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
20 Distensible Venous Malformation	111
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, and Gangadhara Sundar	
21 Non-distensible Cavernous Venous Malformation (Cavernous Haemangioma)	115
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
22 Arteriovenous Malformation: Carotid-Cavernous Fistula	121
Stephanie Ming Young, Shantha Amrith, Eric Ting, and Gangadhara Sundar	
Part VI Benign Neoplasms: Eyelid	
23 Pilomatrixoma	127
Stephanie Ming Young, Shantha Amrith, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
24 Plexiform Neurofibroma	131
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	

Part VII Benign Neoplasms: Orbit

- 25 Schwannoma** 137
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 26 Solitary Fibrous Tumour** 141
Gangadhara Sundar, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 27 Meningioma** 145
Hazel Anne Lin, Shantha Amrith, Clement Tan, Stephanie Ming Young,
Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar
- 28 Glioma** 151
Hazel Anne Lin, Clement Tan, Shantha Amrith, Stephanie Ming Young,
Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar
- 29 Osteoma** 157
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 30 Ossifying Fibromyxoid Tumour** 161
Mariel Angelou Parulan, Shantha Amrith, Stephanie Ming Young,
Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar

Part VIII Benign Neoplasms: Lacrimal System

- 31 Lacrimal Gland: Pleomorphic Adenoma** 169
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 32 Lacrimal Sac Inverted Papilloma** 173
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar

Part IX Lymphoproliferative Disorders

- 33 Overview** 179
Limei Michelle Poon
- 34 Reactive Lymphoid Hyperplasia** 183
Mariel Angelou Parulan, Shantha Amrith, Stephanie Ming Young, Eric Ting,
Bingcheng Wu, Min En Nga, and Gangadhara Sundar
- 35 MALT Lymphoma** 187
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 36 Follicular Lymphoma** 193
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 37 Diffuse Large B-Cell Lymphoma** 201
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar

- 38 Mantle Cell Lymphoma** 205
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 39 T-Cell Lymphoma** 211
Gangadhara Sundar, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 40 Plasmacytoma/Multiple Myeloma** 215
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar

Part X Primary Malignant Neoplasms: Eyelid

- 41 Basal Cell Carcinoma** 221
Stephanie Ming Young, Shantha Amrith, Bingcheng Wu, Min En Nga,
and Gangadhara Sundar
- 42 Squamous Cell Carcinoma** 225
Shantha Amrith, Stephanie Ming Young, Bingcheng Wu, Min En Nga,
and Gangadhara Sundar
- 43 Sebaceous Gland Carcinoma** 229
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 44 Merkel Cell Carcinoma** 235
Gangadhara Sundar, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 45 Mucinous Carcinoma** 239
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 46 Liposarcoma** 243
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 47 Melanoma of Ocular Adnexa** 247
Gangadhara Sundar, Stephanie Ming Young, Bingcheng Wu, Min En Nga,
and Shantha Amrith

Part XI Primary Malignant Neoplasms: Orbit

- 48 Rhabdomyosarcoma** 253
Gangadhara Sundar, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 49 EBV Smooth Muscle Tumour** 257
Stephanie Ming Young, Shantha Amrith, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 50 Alveolar Soft Part Sarcoma** 263
Mariel Angelou Parulan, Shantha Amrith, Stephanie Ming Young, Eric Ting,
Bingcheng Wu, Min En Nga, and Gangadhara Sundar

Part XII Primary Malignant Neoplasms: Lacrimal System

- 51 Lacrimal Gland: Adenoid Cystic Carcinoma** 269
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 52 Lacrimal Gland: Ductal Adenocarcinoma** 275
Gangadhara Sundar, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 53 Lacrimal Sac: Epithelial Tumours** 279
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar

Part XIII Metastatic Tumours

- 54 Overview** 287
Shantha Amrith and Gangadhara Sundar
- 55 Neuroblastoma** 289
Mariel Angelou Parulan, Shantha Amrith, Stephanie Ming Young, Eric Ting,
Bingcheng Wu, Min En Nga, and Gangadhara Sundar
- 56 Carcinoma of Breast** 293
Gangadhara Sundar, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 57 Prostate Adenocarcinoma** 297
Gangadhara Sundar, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 58 Gastric Carcinoma** 301
Gangadhara Sundar, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 59 Ewing Sarcoma** 305
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar
- 60 Renal Cell Carcinoma** 311
Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar

Part XIV Tumours from the Globe

- 61 Retinoblastoma** 319
Gangadhara Sundar, Stephanie Ming Young, Eric Ting, Bingcheng Wu,
Min En Nga, and Shantha Amrith
- 62 Uveal Melanoma** 323
Mariel Angelou Parulan, Shantha Amrith, Stephanie Ming Young, Eric Ting,
Bingcheng Wu, Min En Nga, and Gangadhara Sundar

Part XV Tumours from Paranasal Sinuses and Nasopharynx

- 63 Paranasal Sinus Mucocoele** 331
Shantha Amrith, Stephanie Ming Young, Poh Sun Goh, Bingcheng Wu,
Min En Nga, and Gangadhara Sundar

64	Malignant Epithelial Tumours of Paranasal Sinuses	335
	Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
65	Sino-nasal Lymphomas	341
	Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
66	Nasopharynx: Nasopharyngeal Carcinoma	345
	Shantha Amrith, Stephanie Ming Young, Eric Ting, Bingcheng Wu, Min En Nga, and Gangadhara Sundar	
Index	351