

Contents

Introduction to the BCSC	xv
Objectives	1
PART I Ophthalmic Pathology	3
1 Introduction to Part I.	5
Highlights	5
Overview	5
Organizational Framework and Basic Pathologic Concepts	6
Topography	6
Disease Process	8
Differential Diagnosis	18
2 Quick-Start Guide: Specimen Handling and Special Tests and Procedures in Pathology	19
Highlights	19
Glossary of Select Pathology Terminology	19
Introduction	21
Communication With the Pathologist	21
Handling and Transfer of Tissue	22
Fixatives	22
Tissue Submission.	24
Gross (Macroscopic) Examination and Dissection.	24
Enucleation Specimens	24
Eyelid and Conjunctival Biopsies	26
Temporal Artery Biopsies	29
Tissue Processing and Staining	29
Tissue Processing	29
Slide Preparation and Tissue Staining	29
Special Techniques and Procedures	32
Fine-Needle Aspiration Biopsy	32
Frozen Section	34
Special Diagnostic Testing	36
Immunohistochemistry	36
Molecular Pathology.	39
Diagnostic Electron Microscopy.	42
3 Wound Repair	43
Highlights	43
General Aspects of Wound Repair	43

Wound Repair in Specific Ocular Tissues	44
Conjunctiva	45
Cornea	45
Sclera	47
Lens	48
Vitreous	48
Retina	48
Optic Nerve	49
Uveal Tract	49
Orbit and Ocular Adnexa	49
Types of Ocular Trauma and Their Sequelae	50
4 Conjunctiva	57
Highlights	57
Topography	57
Developmental Anomalies	59
Choristomas	59
Hamartomas	59
Inflammation	61
Acute or Chronic Conjunctivitis	61
Infectious Conjunctivitis	61
Noninfectious Conjunctivitis	61
Papillary Versus Follicular Conjunctivitis	63
Granulomatous Conjunctivitis	64
Pyogenic Granuloma	65
Degenerations	67
Pinguecula and Pterygium	67
Amyloid Deposits	68
Epithelial Inclusion Cyst	69
Conjunctivochalasis	70
Neoplasia	70
Squamous Epithelial Lesions	70
Melanocytic Lesions	75
Lymphoid Lesions	86
Glandular Neoplasms	89
Other Neoplasms	89
5 Cornea	91
Highlights	91
Topography	91
Developmental Anomalies	93
Dermoid	93
Peters Anomaly	93
Inflammation	94
Infectious Keratitis	94
Noninfectious Keratitis	100
Degenerations, Depositions, and Ectasias	100
Degenerations	100
Depositions	105
Ectatic Disorders	106

Dystrophies	108
Epithelial and Subepithelial Dystrophies	109
Epithelial–Stromal <i>TGFBI</i> Dystrophies.	109
Stromal Dystrophies.	112
Descemet Membrane and Endothelial Dystrophies	113
Neoplasia	118
6 Anterior Chamber and Trabecular Meshwork.	119
Highlights	119
Topography	119
Developmental Anomalies	120
Primary Congenital Glaucoma	120
Anterior Segment Dysgenesis.	121
Inflammation	122
Degenerations	122
Iridocorneal Endothelial Syndrome	122
Secondary Open-Angle Glaucoma.	124
Neoplasia	130
7 Sclera	131
Highlights	131
Topography	131
Developmental Anomalies	132
Choristomas and Hamartomas	132
Nanophthalmos.	132
Microphthalmia	133
Inflammation	133
Scleritis	133
Degenerations	136
Age-Related Calcific Plaque	136
Scleral Staphyloma	136
Melanoma-Associated Spongiform Scleropathy	136
Neoplasia	137
8 Lens	139
Highlights	139
Topography	139
Capsule	139
Epithelium	141
Cortex and Nucleus	141
Zonular Fibers	141
Developmental Anomalies	142
Congenital Aphakia	142
Lenticonus and Lentiglobus	142
Inflammation	143
<i>Cutibacterium acnes</i> Endophthalmitis	143
Phacoantigenic Uveitis.	143
Phacolytic Uveitis	144

Degenerations 145
 Cataract and Other Lens Abnormalities 145
 Neoplasia and Associations With Systemic Disorders 149

9 Vitreous 151

Highlights 151
 Topography 151
 Developmental Anomalies 152
 Persistent Fetal Vasculature 152
 Bergmeister Papilla 153
 Mittendorf Dot 153
 Vitreous Cysts 153
 Inflammation 153
 Degenerations 155
 Synchysis, Syneresis, and Aging 155
 Posterior Vitreous Detachment 156
 Hemorrhage 159
 Asteroid Hyalosis 160
 Amyloidosis 160
 Neoplasia 162
 Intraocular Lymphoma 162

10 Retina and Retinal Pigment Epithelium 167

Highlights 167
 Topography 167
 Neurosensory Retina 168
 Retinal Pigment Epithelium 172
 Developmental Anomalies 172
 Albinism. 172
 Myelinated Nerve Fibers 172
 Vascular Anomalies 172
 Congenital Hypertrophy of the RPE 175
 Inflammation 176
 Infectious Etiologies 176
 Noninfectious Etiologies 179
 Degenerations 179
 Typical and Reticular Peripheral Cystoid Degeneration
 and Retinoschisis 179
 Lattice Degeneration. 180
 Sequelae of Retinal Detachment. 181
 Paving-Stone Degeneration. 181
 Ischemia 182
 Abusive Head Trauma 192
 Age-Related Macular Degeneration 194
 Polypoidal Choroidal Vasculopathy 199
 Dystrophies 200
 Macular Dystrophies 200
 Diffuse Photoreceptor Dystrophies 203

Neoplasia	204
Retinoblastoma	204
Retinocytoma	209
Medulloepithelioma	209
Nodular Hyperplasia of Ciliary Body Epithelium	211
Combined Hamartoma of the Retina and RPE	212
Adenomas and Adenocarcinomas of the Ciliary Body Epithelium and RPE	213
11 Uveal Tract	215
Highlights	215
Topography	215
Iris	216
Ciliary Body	217
Choroid	218
Developmental Anomalies	218
Aniridia	218
Coloboma	219
Inflammation (Uveitis)	220
Infectious Uveitis	220
Noninfectious Uveitis	221
Degenerations	224
Neovascularization of the Iris (Rubeosis Iridis)	224
Hyalinization of the Ciliary Body	224
Choroidal Neovascularization	224
Neoplasia	225
Iris	225
Choroid and Ciliary Body	226
Metastatic Tumors	234
Other Uveal Tumors	236
12 Eyelids	239
Highlights	239
Topography	239
Developmental Anomalies	241
Distichiasis	241
Phakomatous Choristoma	241
Dermoid Cyst	241
Inflammation	242
Infectious Inflammatory Disorders	242
Noninfectious Inflammatory Disorders	244
Degenerations and Deposits	245
Xanthelasma	245
Amyloidosis	246
Cysts	247
Epidermoid Cysts	247
Ductal Cysts	247

Neoplasia	248
Epidermal Neoplasms	248
Dermal Neoplasms	254
Neoplasms and Proliferations of the Dermal Appendages	255
Merkel Cell Carcinoma	258
Melanocytic Neoplasms	259
13 Orbit and Lacrimal Drainage System	265
Highlights	265
Topography	265
Bony Orbit and Soft Tissues	265
Developmental Anomalies	266
Cysts	266
Inflammation	267
Noninfectious Inflammation	267
Infectious Inflammation	271
Degenerations	273
Amyloid	273
Neoplasia	273
Lacrimal Gland Neoplasia	274
Lymphoproliferative Lesions	277
Soft-Tissue Tumors	280
Vascular Tumors	280
Tumors With Fibrous Differentiation	282
Tumors With Muscle Differentiation	283
Peripheral Nerve Sheath and Central Nervous System Tumors	285
Adipocytic Tumors	286
Bony Lesions of the Orbit	286
Lacrimal Sac Neoplasia	288
Secondary Tumors	288
14 Optic Nerve	289
Highlights	289
Topography	289
Developmental Anomalies	291
Colobomas	291
Optic Nerve Pits	292
Inflammation	292
Infections That Cause Optic Neuropathy	293
Noninfectious Inflammations That Affect the Optic Nerve	295
Degenerations	298
Optic Atrophy	298
Optic Nerve Head Drusen	299
Neoplasia	301
Melanocytoma	301
Glioma	301
Meningioma	303

PART II	Intraocular Tumors: Clinical Aspects	305
15	Introduction to Part II	307
16	Melanocytic Tumors	309
	Highlights	309
	Introduction	309
	Iris Nevus	310
	Iris Melanoma	311
	Ciliary Body and Choroidal Nevi	314
	Melanoma of the Choroid and Ciliary Body	316
	Clinical Characteristics	318
	Diagnostic Evaluation	320
	Differential Diagnosis	324
	Classification	329
	Metastatic Evaluation	329
	Treatment	332
	Prognosis and Prognostic Factors	336
	Melanocytoma of the Iris, Ciliary Body, and Choroid	338
	Epithelial Tumors of the Uveal Tract and Retina	339
	Adenoma and Adenocarcinoma	339
	Simple Hamartoma	340
	Combined Hamartoma	340
17	Vascular Tumors	341
	Highlights	341
	Introduction	341
	Choroidal Vascular Tumors	341
	Choroidal Hemangiomas	341
	Retinal Vascular Tumors	345
	Prenatal Retinal Vascular Tumors: Nonleaking Lesions	345
	Postnatal Retinal Vascular Tumors: Leaking Lesions	347
18	Retinoblastoma	353
	Highlights	353
	Introduction	353
	Diagnostic Evaluation	354
	Clinical Examination	354
	Ancillary Imaging	358
	Differential Diagnosis	360
	Retinoblastoma Classification	365
	The International Intraocular Retinoblastoma Classification	365
	Other Retinoblastoma Classification Systems	365
	Treatment	368
	Enucleation	369
	Chemotherapy	369
	Local Consolidation Therapy	371

Spontaneous Regression 374
Genetic Counseling 374
Associated Conditions 376
 Retinocytoma 376
 Primitive Neuroectodermal Tumor 376
Prognosis 377
 Ocular Prognosis 377
 Life Prognosis 378
 Second Nonocular Tumors 379

19 Ocular Involvement in Systemic Malignancies 381

Highlights 381
Secondary Tumors of the Eye. 381
 Metastatic Carcinoma 381
 Direct Intraocular Extension 390
Lymphoid Tumors 390
 Primary Vitreoretinal Lymphoma 391
 Primary Uveal Lymphoma 393
 Secondary Involvement of Systemic Lymphoma 396
Ocular Manifestations of Leukemia 396

Additional Materials and Resources 399
Requesting Continuing Medical Education Credit 401
Study Questions 403
Answers 411
Index 419