

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	7
Disease Processes	7
Differential Diagnosis	16
2 Specimen Handling	19
Highlights	19
Overview	19
Communication	20
Fixatives for Tissue Preservation	21
Specimen Orientation	21
Gross Examination and Dissection	24
Processing and Staining	24
Tissue Processing	24
Slide Preparation and Tissue Staining	26
3 Special Testing and Procedures in Pathology	29
Highlights	29
Introduction	29
Immunohistochemistry	29
Flow Cytometry, Molecular Pathology, and	
Diagnostic Electron Microscopy	33
Flow Cytometry.	33
Molecular Pathology.	33
Diagnostic Electron Microscopy.	39
Special Procedures	39
Fine-Needle Aspiration Biopsy	39
Frozen Section	40
4 Wound Repair	43
Highlights	43
General Aspects of Wound Repair	43

Wound Repair in Specific Ocular Tissues	44
Cornea	45
Conjunctiva	45
Sclera	47
Uveal Tract	47
Lens	47
Retina	48
Vitreous	48
Orbit and Ocular Adnexa	48
Optic Nerve	49
Pathologically Apparent Sequelae of Ocular Trauma	49
5 Conjunctiva	55
Highlights	55
Topography	55
Developmental Anomalies	57
Choristomas	57
Hamartomas	57
Inflammation	57
Acute or Chronic Conjunctivitis	59
Infectious Conjunctivitis	59
Noninfectious Conjunctivitis	60
Papillary Versus Follicular Conjunctivitis	61
Granulomatous Conjunctivitis	61
Pyogenic Granuloma	63
Degenerations	64
Pinguecula and Pterygium	64
Amyloid Deposits	67
Epithelial Inclusion Cyst	68
Conjunctivochalasis	68
Neoplasia	69
Squamous Epithelial Lesions	69
Melanocytic Lesions	74
Lymphoid Lesions	85
Glandular Neoplasms	88
Other Neoplasms	88
6 Cornea	89
Highlights	89
Topography	89
Developmental Anomalies	91
Dermoid	91
Peters Anomaly	91
Inflammation	91
Infectious Keratitis	91
Noninfectious Keratitis	98
Degenerations, Depositions, and Ectasias	98

Salzmann Nodular Degeneration	98
Calcific Band Keratopathy	99
Actinic Keratopathy	100
Pinguecula and Pterygium	101
Pannus	101
Bullous Keratopathy	101
Corneal Graft Failure	102
Corneal Pigment Deposits	104
Ectatic Disorders	104
Dystrophies	106
Epithelial and Subepithelial Dystrophies	107
Epithelial–Stromal <i>TGFBI</i> Dystrophies	107
Stromal Dystrophies	110
Descemet Membrane and Endothelial Dystrophies	111
Neoplasia	116
7 Anterior Chamber and Trabecular Meshwork	117
Highlights	117
Topography	117
Developmental Anomalies	118
Primary Congenital Glaucoma	119
Anterior Segment Dysgenesis	119
Inflammation	120
Degenerations	120
Iridocorneal Endothelial Syndrome	120
Secondary Glaucoma	122
Neoplasia	128
8 Sclera	129
Highlights	129
Topography	129
Developmental Anomalies	130
Choristoma	130
Nanophthalmos	130
Microphthalmia	131
Inflammation	131
Episcleritis	131
Scleritis	131
Degenerations	134
Senile Calcific Plaque	134
Scleral Staphyloma	134
Melanoma-Associated Spongiform Scleropathy	134
Neoplasia	135
9 Lens	137
Highlights	137
Topography	137

Capsule	137
Epithelium	137
Cortex and Nucleus	139
Zonular Fibers	139
Developmental Anomalies	140
Congenital Aphakia	140
Anterior Lenticonus and Lentiglobus	140
Posterior Lenticonus and Lentiglobus	140
Inflammation	141
<i>Propionibacterium acnes</i> Endophthalmitis	141
Phacoantigenic Uveitis	141
Phacolytic Uveitis	143
Degenerations	143
Cataract and Other Lens Abnormalities	143
Neoplasia and Associations With Systemic Disorders	147
Pathology in Intraocular Lenses	147

10 Vitreous 149

Highlights	149
Topography	149
Developmental Anomalies	150
Persistent Fetal Vasculature	150
Bergmeister Papilla	151
Mittendorf Dot	151
Vitreous Cysts	151
Inflammation	152
Degenerations	153
Syneresis and Aging	153
Posterior Vitreous Detachment	153
Hemorrhage	156
Asteroid Hyalosis	157
Vitreous Amyloidosis	158
Neoplasia	159
Intraocular Lymphoma	159

11 Retina and Retinal Pigment Epithelium 163

Highlights	163
Topography	163
Neurosensory Retina	164
Retinal Pigment Epithelium	167
Developmental Anomalies	167
Albinism	167
Myelinated Nerve Fibers	168
Vascular Anomalies	169
Congenital Hypertrophy of the RPE	170
Inflammation	171
Infectious Etiologies	171
Noninfectious Etiologies	174

Degenerations	174
Typical and Reticular Peripheral Cystoid Degeneration and Retinoschisis	174
Lattice Degeneration	175
Sequelae of Retinal Detachment	176
Paving-Stone Degeneration	176
Ischemia	177
Abusive Head Trauma	189
Age-Related Macular Degeneration	191
Polypoidal Choroidal Vasculopathy	195
Macular Dystrophies	196
Diffuse Photoreceptor Dystrophies	198
Neoplasia	202
Retinoblastoma	202
Retinocytoma	207
Medulloepithelioma	209
Fuchs Adenoma	210
Combined Hamartoma of the Retina and RPE	210
Adenomas and Adenocarcinomas of the RPE	211
12 Uveal Tract	213
Highlights	213
Topography	213
Iris	213
Ciliary Body	215
Choroid	215
Developmental Anomalies	216
Aniridia	216
Coloboma	217
Inflammation (Uveitis)	218
Infectious Uveitis	218
Noninfectious Uveitis	219
Degenerations	222
Rubeosis Iridis	222
Hyalinization of the Ciliary Body	222
Choroidal Neovascularization.	222
Neoplasia	223
Iris	223
Choroid and Ciliary Body	225
Metastatic Tumors	232
Other Uveal Tumors.	233
13 Eyelids	237
Highlights	237
Topography	237
Developmental Anomalies	239
Distichiasis.	239

Phakomatous Choristoma	239
Dermoid Cyst	240
Inflammation	240
Infectious Inflammatory Disorders	240
Noninfectious Inflammatory Disorders	241
Degenerations and Deposits	243
Xanthelasma	243
Amyloidosis	243
Cysts	245
Epidermoid Cysts	245
Ductal Cysts	245
Neoplasia	245
Epidermal Neoplasms	245
Dermal Neoplasms	252
Neoplasms and Proliferations of the Dermal Appendages	252
Merkel Cell Carcinoma	256
Melanocytic Neoplasms	257
14 Orbit and Lacrimal Drainage System	261
Highlights	261
Topography	261
Bony Orbit and Soft Tissues	261
Developmental Anomalies	262
Cysts	262
Inflammation	263
Noninfectious Inflammation	263
Infectious Inflammation	266
Degenerations	269
Amyloid	269
Neoplasia	269
Lacrimal Gland Neoplasia	270
Lymphoproliferative Lesions	272
Soft-Tissue Tumors	274
Vascular Tumors	274
Tumors With Fibrous Differentiation	275
Tumors With Muscle Differentiation	278
Peripheral Nerve Sheath and Central Nervous System Tumors	280
Adipose Tumors	282
Bony Lesions of the Orbit	282
Secondary Tumors	282
Lacrimal Sac Neoplasia	283
15 Optic Nerve	285
Highlights	285
Topography	285
Developmental Anomalies	287
Colobomas	287
Optic Pits	287

Inflammation	287
Infectious Optic Neuritis	288
Noninfectious Optic Neuritis	289
Degenerations	291
Optic Atrophy	291
Optic Nerve Head Drusen	292
Neoplasia	294
Melanocytoma	294
Glioma	295
Meningioma	296

PART II Intraocular Tumors: Clinical Aspects 299

16 Introduction to Part II 301

17 Melanocytic Tumors 303

Highlights	303
Introduction	303
Iris Nevus	304
Iris Melanoma	304
Ciliary Body and Choroidal Nevi	305
Melanocytoma of the Iris, Ciliary Body, and Choroid	309
Melanoma of the Choroid and Ciliary Body	312
Clinical Characteristics	313
Diagnostic Evaluation	314
Differential Diagnosis	318
Classification	323
Metastatic Evaluation	324
Treatment	327
Prognosis and Prognostic Factors	330
Epithelial Tumors of the Uveal Tract and Retina.	331
Adenoma and Adenocarcinoma	331
Acquired Hyperplasia	332
Simple Hamartoma	333
Combined Hamartoma	333
Fine-Needle Aspiration Biopsy	334

18 Vascular Tumors 335

Highlights	335
Introduction	335
Choroidal Vascular Tumors	335
Choroidal Hemangiomas.	335
Retinal Vascular Tumors.	339
Prenatal Retinal Vascular Tumors: Non-Leaking Lesions	339
Postnatal Retinal Vascular Tumors: Leaking Lesions	341

19	Retinoblastoma	347
	Highlights	347
	Introduction	347
	Diagnostic Evaluation	348
	Clinical Examination	348
	Ancillary Imaging	350
	Differential Diagnosis	354
	Retinoblastoma Classification	357
	Treatment	358
	Enucleation	360
	Chemotherapy	361
	Local Consolidation Therapy	363
	Laser Photocoagulation	363
	Cryotherapy	363
	Plaque Radiotherapy	363
	Intravitreal Chemotherapy	364
	External Beam Radiotherapy	364
	Spontaneous Regression	365
	Genetic Counseling	365
	Associated Conditions	367
	Retinocytoma	367
	Primitive Neuroectodermal Tumor	367
	Prognosis	368
20	Ocular Involvement in Systemic Malignancies	371
	Highlights	371
	Secondary Tumors of the Eye	371
	Metastatic Carcinoma	371
	Direct Intraocular Extension	380
	Lymphoid Tumors	380
	Primary Intraocular Lymphoma	380
	Primary Uveal Lymphoma	383
	Secondary Involvement of Systemic Lymphoma	386
	Ocular Manifestations of Leukemia	386
	Additional Materials and Resources	389
	Requesting Continuing Medical Education Credit	391
	Study Questions	393
	Answers	401
	Index	409