

CONTENTS

1. Tumors of the Conjunctiva and Caruncle	1
Anatomy and Histology	1
Lymphatic Drainage	2
Classification and Frequency	2
Epithelial Neoplasms	3
Papilloma	3
Keratoacanthoma	4
Hereditary Benign Intraepithelial Dyskeratosis	5
Other Benign Epithelial Lesions	6
Epithelial Cysts	6
Nonspecific Keratotic (Leukoplakic) Lesions	6
Pseudoepitheliomatous (Pseudocarcinomatous) Hyperplasia	6
Leukoplakic Lesions (Ultraviolet Light Related)	6
Conjunctival Intraepithelial Neoplasia	7
Squamous Cell Carcinoma	10
Variants of Squamous Cell Carcinoma	12
Mucoepidermoid Carcinoma	12
Spindle Cell Carcinoma	12
Adenoid Squamous Cell Carcinoma	15
Other Carcinomas of the Conjunctiva	15
Basal Cell Carcinoma	15
Sebaceous Carcinoma	15
Lymphoepithelioma-Like Carcinoma	16
Melanocytic Tumors	16
Nevi	16
Nevus Variants	19
Other Benign Melanocytic Lesions	19
Racial Pigmentation and Benign Epithelial Melanosis	19
Ephelides	19
Congenital Pigmentation (Ocular Melanocytosis, Oculodermal Melanocytosis)	20
Primary Acquired Melanosis	20
Melanoma	23
Soft Tissue Tumors	26
Vascular Tumors	27
Myxoma	27
Fibrous Histiocytoma	28

Embryonal Rhabdomyosarcoma, Botryoid Subtype	28
Kaposi's Sarcoma	29
Lymphoid Tumors	30
Plasmacytoma.	31
Metastatic Tumors of the Conjunctiva	31
Tumor-Like Congenital Lesions	32
Dermoid	32
Dermolipoma	32
Ectopic Lacrimal Gland	33
Complex Choristoma.	33
Smooth Muscle Hamartoma	33
Osseous Choristoma.	33
Tumors of the Caruncle	33
2. Tumors of the Uveal Tract	41
Anatomy and Histology	41
The Iris	41
The Ciliary Body	42
The Choroid	42
Vascularization	43
Innervation	43
General Considerations	43
Melanocytic Tumors of the Uvea	43
Nevi	43
Melanocytoma (Magnocellular Nevus)	47
Malignant Melanoma.	48
Diffuse Uveal Melanocytic Proliferation	62
Nonmelanocytic Tumors of the Uvea	63
Mesenchymal Tumors	63
Benign Peripheral Nerve Tumors	66
Osteoma	67
Xanthomatous Lesions	68
Reactive Lymphoid Hyperplasia and Primary Non-Hodgkin's B-Cell Lymphoma of the Uvea.	68
Secondary and Metastatic Tumors	70
Leukemia	70
Lymphoma	70
Plasmacytoma	71
Metastatic Tumors	71

Cysts	72
Staging and Grading of Uveal Melanoma	74
Clinical Staging	74
Pathologic Staging	74
3. Tumors of the Retina	85
Anatomy and Histology	85
Classification and Frequency	86
Retinoblastoma	86
Retinocytoma	103
Glial Tumors and Tumor-Like Conditions	104
Astrocytoma	104
Astrocytic Hamartoma	104
Massive Gliosis of the Retina	105
Vascular Tumors	105
Angiomatosis Retinae	105
Cavernous Hemangioma	106
Melanogenic Neuroectodermal Tumor of the Retina	106
Lymphoid Malignancies	106
Primary Intraocular Lymphoma	106
Leukemia	112
Tumors of the Retinal Pigment Epithelium	112
Metastatic Neoplasms	114
Neuroepithelial Tumors	115
Congenital Tumors of the Ciliary Epithelium	115
Benign Acquired Tumors of the Ciliary Epithelium	121
4. Tumors of the Optic Nerve and Optic Nerve Head	133
Anatomy and Histology	133
Classification and Frequency	134
Meningioma	135
Juvenile Pilocytic Astrocytoma	140
Melanocytoma	145
Astrocytic Hamartoma and Giant Calcified Drusen	147
Hemangioma and Hemangiopericytoma	147
Medulloepithelioma	148
Juvenile Xanthogranuloma	149
Juxtapapillary Hamartoma	150
Malignant Astrocytoma	151
Primary Malignant Melanoma	151
Secondary and Metastatic Tumors	152

5. Tumors of the Eyelids	155
Anatomy and Histology	155
Tumors	158
Benign Epithelial Tumors	159
Squamous Cell Papilloma	160
Seborrheic Keratosis	160
Inverted Follicular Keratosis	160
Benign Lichenoid Keratosis	160
Large Cell Acanthoma	161
Pseudocarcinomatous Hyperplasia	161
Keratoacanthoma	161
Precancerous Epithelial Lesions	162
Actinic Keratosis	162
Bowen's Disease	162
Radiation Dermatitis	163
Xeroderma Pigmentosum	163
Malignant Epithelial Tumors	164
Basal Cell Carcinoma	164
Nevoid Basal Cell Carcinoma Syndrome	168
Squamous Cell Carcinoma	169
Spindle Cell Squamous Carcinoma	170
Adenoid Squamous Cell Carcinoma	170
Melanocytic Tumors	170
Nevocellular Nevus	171
Giant Congenital Melanocytic Nevus	173
Blue Nevus, Cellular Blue Nevus, and Nevus of Ota	174
Freckle	176
Lentigo Simplex	176
Solar Lentigo	176
Malignant Melanoma	176
Lentigo Maligna and Lentigo Maligna Melanoma	177
Superficial Spreading Melanoma	177
Nodular Melanoma	178
Acral Lentiginous Melanoma	178
Melanoma Arising from a Nevus	179
Malignant Melanoma	180
Dysplastic Nevus Syndrome	181
Benign Sebaceous Gland Tumors	181

Sebaceous Gland Carcinoma	182
Tumors of the Eccrine and Apocrine Glands of the Eyelid	191
Syringoma	191
Pleomorphic Adenoma	192
Eccrine Acrospiroma	193
Mucinous Sweat Gland Adenocarcinoma	193
Primary Cutaneous Adenoid Cystic Carcinoma	195
Adenocarcinoma of Eccrine Sweat Glands	195
Adenoma and Adenocarcinoma of the Glands of Moll.	195
Tumors of the Pilar Structures of the Eyelid	196
Trichoepithelioma	196
Trichofolliculoma.	197
Trichilemmoma	198
Pilomatrixoma	199
Adnexal Carcinoma	200
Vascular Tumors	200
Capillary Hemangioma and Nevus Flammeus	200
Cavernous Hemangioma	202
Lymphangioma	202
Glomus Tumor	203
Cutaneous Angiosarcoma	203
Xanthomatous Lesions.	203
Xanthelasma	204
Fibrous Histiocytoma	204
Tuberous Xanthoma.	204
Juvenile Xanthogranuloma	205
Other Xanthogranulomatous Lesions	205
Lipoid Proteinosis.	205
Miscellaneous Lesions	207
Mycosis Fungoides	207
Lymphomatoid Granulomatosis	208
Nodular Fasciitis.	209
Juvenile Fibromatosis.	210
Phakomatous Choristoma	211
Granular Cell Tumor	211
Merkel Cell Tumor	212
Intravascular Papillary Endothelial Hyperplasia	212
Erdheim-Chester Disease	213

Necrobiotic Xanthogranuloma with Paraproteinemia	214
Carney's Complex	215
Metastatic Tumors	216
6. Tumors of the Lacrimal Gland	223
Anatomy and Histology	223
Classification and Frequency	223
Benign Epithelial Tumors	225
Pleomorphic Adenoma (Benign Mixed Tumor)	225
Oncocytoma (Oncocytic Adenoma, Oxyphilic Adenoma)	228
Warthin's Tumor (Adenolymphoma)	228
Myoepithelioma	228
Malignant Epithelial Tumors	229
Adenoid Cystic Carcinoma	229
Malignant Mixed Tumor	234
Primary Adenocarcinoma	236
Mucoepidermoid Carcinoma	237
Miscellaneous Malignant Epithelial Tumors	238
Primary Ductal Adenocarcinoma	238
Acinic Cell Carcinoma	239
Sebaceous Adenocarcinoma	239
Lymphoid Tumors	239
Mesenchymal Tumors	240
Secondary and Metastatic Tumors	240
Tumor-Like Lesions	240
Chronic Dacryoadenitis	240
Benign Lymphoepithelial Lesion	240
Lacrimal Gland Cysts	241
Staging and Grading of Lacrimal Gland Tumors	242
7. Tumors of the Lacrimal Drainage System	247
Anatomy and Histology	247
Classification and Frequency	247
Epithelial Tumors (Papillomas and Carcinomas of the Lacrimal Sac)	250
Benign Epithelial Tumors	250
Malignant Epithelial Tumors	253
Nonepithelial Tumors	260
8. Tumors of the Orbit	265
Anatomy and Histology	265
Classification and Frequency	266

Fibrous Tumors	267
Fibromatosis	267
Nodular Fasciitis	267
Fibroma	268
Fibrous Histiocytoma	268
Fibrosarcoma	270
Lipomatous Tumors	271
Lipoma	271
Liposarcoma	271
Myogenic Tumors	271
Smooth Muscle Tumors	271
Rhabdomyosarcoma	272
Miscellaneous Skeletal Muscle Tumors	276
Vascular Tumors	276
Capillary Hemangioma	276
Cavernous Hemangioma	278
Lymphangioma	279
Hemangiopericytoma	280
Solitary Fibrous Tumor	281
Epithelioid Hemangioma	283
Arteriovenous and Venous Anomalies	284
Intravascular Papillary Endothelial Hyperplasia	284
Angiosarcoma	285
Peripheral Nerve Sheath Tumors	285
Neurofibroma	285
Schwannoma (Neurilemoma)	287
Granular Cell Tumor	289
Malignant Peripheral Nerve Sheath Tumor	289
Bone Tumors	290
Fibrous Dysplasia	291
Ossifying Fibroma	291
Osteosarcoma	292
Mesenchymal Chondrosarcoma	293
Lymphoid Tumors	294
Reactive Lymphoid Hyperplasia	295
Lymphoma	295
Orbital Plasmacytoma	297

Leukemic Disorders	297
Granulocytic Sarcoma	297
Histiocytosis	298
Langerhans Cell Histiocytosis	298
Histiocytic Disorders of Mononuclear Phagocytes	299
Miscellaneous Orbital Tumors	301
Epithelial Cystic Choristoma	301
Teratoma	303
Endodermal Sinus Tumor	303
Pigmented Retinal Choristoma	304
Ewing's Family of Tumors	304
Primary Orbital Carcinoid and Paraganglioma	304
Primary Melanoma	304
Alveolar Soft Part Sarcoma	305
Secondary Tumors	305
Metastatic Tumors	306
Staging and Grading of Sarcomas of the Orbit	307
9. Pathologic Examination of Ocular Specimens	321
The Globe	321
Enucleation	321
Excision	322
Biopsy	322
Fine-Needle Aspiration Biopsy	323
Cytologic Samples	324
The Eyelids	324
The Conjunctiva and Caruncle	324
The Orbit and Optic Nerve	325
The Lacrimal Gland and Sac	325
Index	329