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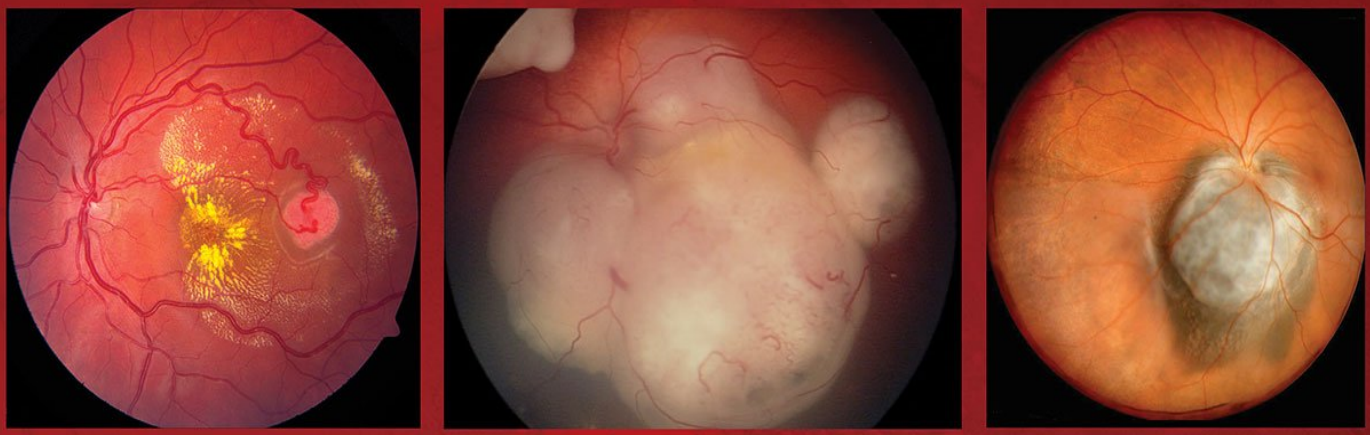
Intraocular Tumors

AN ATLAS AND TEXTBOOK

THIRD EDITION



JERRY A. SHIELDS

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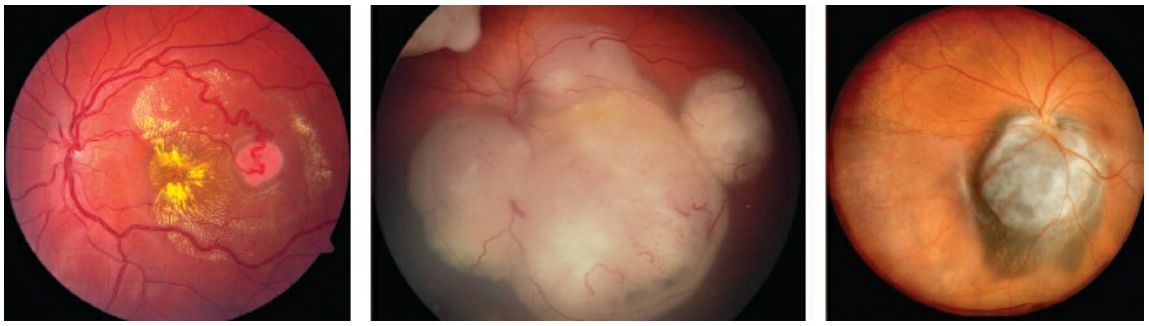

THIRD EDITION

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This book is dedicated to our seven children,
Jerry, Patrick, Bill, Maggie Mae, John, Nellie, and Mary Rose.
They were toddlers and young children during the first two editions of this atlas
and now they are grown into young adults, developing their own careers.
We wish them satisfaction and success in their work
and unending happiness in life.

FOREWORD 1

For more than four decades, Jerry and Carol Shields have dedicated a significant amount of their professional lives to the study of ocular oncology. Guided and inspired by several mentors in this field, but particularly on the medical side by J. Donald M. Gass and on the histopathological side by W. Richard Green, they have worked diligently and productively on the Oncology Service at Wills Eye Hospital in Philadelphia, indisputably the world's leading institution in this field. Nowhere has their primary work been assimilated and enhanced more than on this legendary service which has emerged as a citadel for this division of ophthalmology. Their original, innovative, and lasting work has not been limited to the malignant tumors alone such as choroidal melanoma, retinoblastoma, and metastatic chorioretinal disease, but it has also included important discoveries in degenerative, inflammatory, infectious, and other rare chorioretinal diseases. The results have been hundreds of clinical/scientific papers, virtually all in peer-review journals, numerous books as authors and editors, and perhaps above all, the training of countless residents and retina/ocular oncology fellows worldwide.

Several years ago, they realized the importance of intraocular pathology throughout the world, specifically in terms of the prevalence and severe impact on visual disabilities. This led to an attempt to meet the needs of the eye care community with a comprehensive clinical atlas. That comprehensive atlas became the standard reference in the field of intraocular tumors, read and referenced by the medical community, residents and fellows in ophthalmology, and the rest of the related eye care community. Prior to their original atlas, there was no standard for the study of intraocular tumors. That stand-alone atlas is still the standard in our field. So, why is there a need for a new edition? The answer is simple. Medical knowledge is simply a moment in time. Advances in all phases of medical-scientific disciplines have occurred since their first edition, particularly multimodal imaging. This new edition has addressed intraocular tumor findings with optical coherence tomography and its many sub-technological devices, the intravenous angiographies and beyond. Through these advances in medical sciences, particularly technological developments with advanced imaging, Jerry and Carol now have a new atlas which describes new entities, new manifestations of old disorders, better explanations for presumed mechanisms for the pathogenesis of these diseases, and a myriad of suggested treatment modalities and approaches for their management. This new atlas will clearly meet the expectations of its readers, students, distinguished colleagues, and friends alike. It will provide a treasure of knowledge and experience in the diagnosis and treatment of intraocular tumors. For sure, Jerry and Carol's combined efforts will be rewarded by the gratitude of clinicians, scientists, ophthalmologists, retinal specialists, students, and patients, and by the incalculable pleasure that will result on the part of the casual as well as the discerning reader of this masterpiece in intraocular tumors.

Yours very truly,

Lawrence A. Yannuzzi, MD

Vitreous Retina Macula Consultants
of New York, New York, USA
LuEsther T. Mertz Retinal Research Center,
Manhattan Eye, Ear and Throat Hospital,
New York, USA

FOREWORD 2

In Greek mythology, Atlas (Ancient Greek: Ατλας) was the primordial Titan who held up the celestial spheres.

In the sphere of ocular tumors, ophthalmologists, pathologists, and oncologists have depended historically on our **Shields Atlas** for support. And this new edition gives unprecedented service!

Intraocular Tumors: An Atlas and Textbook is a work of Olympian proportions: this third edition includes an expanded and updated text; over 2,000 photographs illustrate the spectrum of ocular neoplastic disease, including both common and rare intraocular tumors; spectral domain enhanced depth imaging optical coherence tomography examples abound, as well as a panoply of new ultrasound, fundus autofluorescence, fluorescein angiographic, indocyanine green, magnetic resonance imaging, and computed tomographic tumor images.

Also new to this edition of the atlas are a phalanx of tables marshalled to combat ocular tumors, including those listing updated classifications, risk factors, clinical features, differential diagnoses, and therapeutic options.

The atlas serves as well as a substantive reference work, with the bibliographic material organized into major reviews, and small case series, and subdivided into imaging, genetics/pathology, therapy, and case reports.

Chapters are color coded for fast reference, and there is an expanded and updated emphasis on surgical illustrations and photographs. In short, this third atlas edition features major upgrades and additions: the new *ne plus ultra* for state of the art, comprehensive information on ocular tumors.

Jerry and Carol Shields, who rule the rarified heights of the 14th floor here at 840 Walnut Street in Philadelphia – and whose reach of course extends around the world – are specially honored this year as we celebrate the 40th anniversary of the Ocular Oncology Service at Wills Eye Hospital. A fitting time indeed to publish this titanic *magnum opus*, crowning as it does four decades of globally and historically unparalleled clinical experience with ocular oncology. This is a work of medicine and of art, reflecting a heroic partnership with generations of patients and their families, and with trainees from all over the world: a new standard for the field. The broad shoulders of the Shields team support and epitomize in the finest way the words of the Wills motto emblazoned on our seal: Skill with compassion. This Atlas is both metaphorically and in actuality the fruition of that noble and herculean labor.

Julia A. Haller, MD
Ophthalmologist-in-Chief, Wills Eye Hospital
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Professor and Chair of Ophthalmology
Thomas Jefferson University
Philadelphia, Pennsylvania, USA

PREFACE

Forty years in the management of intraocular tumors

Forty years is a long time. Forty years is half a lifetime and beyond the time of most careers. Forty years represents the number of years that we have devoted our medical and surgical practice to the study of intraocular tumors. Unlike nearly every other ophthalmologist worldwide, we focus every minute of our career on the topic of ocular tumors, benign or malignant, and the numerous simulating lesions. Every working day, from early morning before the sunrise, we traveled to work, met and examined patients, reviewed imaging and testing to establish the diagnosis, provided treatment for a spectrum of intraocular tumors, finished a day's work, and traveled home in the early evening. Forty years represents the time that we used to discover, think, design, critique, and perform hours-on-end research that culminated in numerous ocular oncology projects with information pushing the field forward. Slow and steady progress, but looking back, we participated in giant leaps of knowledge.

And why choose such a rigorous vocation? For a singular goal—to improve outcomes for patients with intraocular tumors.

Forty years ago, intraocular tumors were dismissed as rare, often requiring enucleation. There was little clinical interest in this field. The subspecialty of ocular oncology did not exist. But we saw the need for improvement in patient care and our Ocular Oncology Service at Wills Eye Hospital was established. Over this 40-year period, we participated in and witnessed the evolution of treatments such as plaque radiotherapy for melanoma and retinoblastoma, targeted chemotherapy into the ophthalmic artery for retinoblastoma, and photodynamic therapy for hemangioma and other lesions.

Currently, ocular oncology has emerged as a vitally important subspecialty, bursting with innovative treatments and remarkable success. We have achieved ultimate goals of saving the patient's life and the eye, and we are now focused on visual outcomes with minimal local or systemic toxicities. Daily electronic communication and online publication allow rapid spread of information to remote regions of the world, at a button's click. Organized local, national, and international societies for ocular oncology educate physicians and patients. The playing field for ocular cancer worldwide is gradually equalizing.

Retinoblastoma management has reached unsurpassed heights with novel methods of intravenous, intra-arterial, and intravitreal chemotherapy with astounding high rates of globe salvage, patient safety, and preservation of visual acuity. Remarkably, some children are cured with two or three doses of single-agent chemotherapy delivered into the ophthalmic artery. Vitreous retinoblastoma seeds, a previously doomed finding, are now reversed with intravitreal chemotherapy, a technique that was previously avoided for fear of tumor seeding outside the eye. Lack of complications and reliable success has fueled this avenue of therapy.

Uveal melanoma management has taken strong forward steps regarding early tumor detection and unraveling genomic status to allow a more accurate estimation

of ultimate metastatic risk. A thin needle is slipped into the tumor to aspirate only 10 or 20 cells for DNA or RNA analysis, leading to a genetic profile of the tumor, predicting high or low risk for metastasis. Uveal melanoma can now be detected at an incredibly small size, under 1 or 2 mm in thickness, using optical coherence tomography (OCT) combined with established risk factors. Imagine, micro-millimeter melanoma detection—a promise for better survival.

So looking back over these 40 years and particularly over the past 10 years since the second edition of our atlases, we are proud of the enormous advancements in the field of ocular oncology. A young girl with retinoblastoma that would have lost her eye in the 1970s now will have the globe retained without cosmetic deformity and likely with decent visual acuity. The uncle that might have lost his eye to a large melanoma in the past will now possibly have detection of the tumor at a 2.0-mm stage for early treatment and favorable prognosis. This remarkable progress in ocular oncology has been achieved through collaboration worldwide.

This third edition of our book, *Intraocular Tumors: An Atlas and Textbook*, displays the unparalleled headway in the field of ocular oncology. We have organized this volume based on anatomic tumor origin with extensive documentation of clinical and imaging features of nearly every intraocular tumor, benign or malignant. This book is a treasure box for you to read and enjoy, and to use as a guide for patient management.

Jerry A. Shields, MD
Carol L. Shields, MD

ACKNOWLEDGMENTS

This atlas is our masterpiece, our magnum opus, representing our careers in ocular oncology. This atlas represents not only our work, but it embodies the work of our team, a collaborative team working together each day in patient care and research to achieve a magnificent, singular goal of excellence in ocular oncology. This team includes physicians, nurses, technicians, photographers, administrators, secretaries, and many others.

We are grateful to our professors for teaching us to the basic concepts of the various benign and malignant tumors of the eye. This knowledge provided a solid foundation for our understanding and further exploration of intraocular tumors. We are thankful to our patients for granting us the honor to provide their medical care and assist them in making medical decisions. Each patient story, with its ups and downs, has added to our understanding of intraocular tumors.

From the Ocular Oncology Service at Wills Eye Hospital, we would like to thank our outstanding team of ophthalmic photographers, Tika Siburt, Tessa Tintle, Jacqueline Hanable, and Sandor Ferenczy for their masterful talents in capturing the features of ophthalmic tumors. Each photograph is a resplendent display of tumor characteristics. We thank Linda Warren for the illustrative surgical drawings. Importantly, we would like to commend the entire staff on the Ocular Oncology Service at Wills Eye Hospital under the direction of David Lashinsky for their devotion and service to our patients. We particularly acknowledge the work of Sandra Dailey in helping with day-to-day matters related to this book. Our staff is the quintessential example of teamwork and dedication, with genuine care and respect for each patient.

We are grateful to the medical staff at Wills Eye Hospital of Thomas Jefferson University, including Julia Haller MD, the Ophthalmologist-in-Chief, and the members of the Pathology, Retina, Uveitis, Cornea, Oculoplastics, Pediatric Ophthalmology, Glaucoma, Neuro-Ophthalmology, and other services for assisting with our patients and sharing ideas.

It has always been our practice to seek the best care for our patients from among the several medical institutions in the Philadelphia region. Special thanks to the entire staff at the Department of Medical Oncology at Thomas Jefferson University, particularly Takami Sato, MD, who has devoted his career to better understanding and management of systemic uveal melanoma metastasis. In addition, we recognize the world class, remarkably precise catheterization skills of Pascal Jabbour, MD, in the Department of Neurosurgery, Endovascular Division. His provision of intra-arterial chemotherapy to hundreds of babies worldwide has truly changed the lives of those patients.

In addition, we would like to credit our pediatric oncology colleagues at The Children's Hospital of Philadelphia, especially the brilliantly accomplished Anne Leahey, MD who designs and delivers systemic chemotherapy to children with intraocular cancers. For her leadership regarding our patients and hundreds worldwide, we are grateful. We credit the excellent work of Emi Caywood, MD at Dupont Nemours Children's Hospital of Thomas Jefferson University, who monitors

all children receiving intra-arterial chemotherapy. We also recognize our team of radiation oncologists at Drexel University College of Medicine, Hahnemann Hospital, under the direction of Lydia Komarnicky, MD. For the past 40 years, they have provided cutting edge radiation therapy for our patients, including custom-designed plaque radiotherapy and various methods of stereotactic teleradiotherapy. We also recognize the leadership work of our genetics team at the University of Pennsylvania under the direction of Arupa Ganguly, PhD, who have skillfully and authoritatively pioneered both retinoblastoma and melanoma genetic assessment.

Importantly, we would like to credit our ocular oncology associates who have shared in the medical and surgical care of our patients. These include Arman Mashayekhi, MD, an expert par excellence in intraocular tumors and laser treatments, Sara Lally, MD, a maestro clinician with marvelous skills in patient medical and surgical care, and Emil Say, MD, an outstanding specialist with tremendous research abilities. In addition, there are hundreds of fellows and visitors to the Oncology Service who should be recognized and commended for their dedication to the field of ocular oncology.

There is one individual who deserves special recognition, our long-time friend and ophthalmic pathologist, Ralph C. Eagle Jr, MD. Over the many years that we have worked together, he has provided magnificent histopathology consultation on ocular tumors, most of which were particularly challenging. We are indebted to him for his dedication and consummate diagnostic acumen. Dr. Eagle has provided numerous high-quality photographs of gross and microscopic specimens of many conditions in this book, a testament to his bountiful pathology experience and excellent photography skills. We would also like to thank Hormoz Eyha, MD, an outstanding cytopathologist from Fox Chase Cancer Center, able to establish a tumor diagnosis based on a few floating cells.

Finally, we would like to thank our seven children for allowing us the time and support to complete this third edition of the atlases. When we wrote the first edition, they were babies and toddlers, and with the second edition, they were young children and teenagers. Now with this third edition, they are unfolding their own careers as adults.

So, we have said enough. Now it is time for you to sit back and peruse the charm of the 25 chapters in this textbook and atlas. You will note many new items in this third edition including new illustrations, updated references and text, instructive tables and classifications, and cutting edge imaging with current techniques including autofluorescence and spectral domain optical coherence tomography. In the end, our hope is that you will appreciate this work and find it useful for your clinical practice.

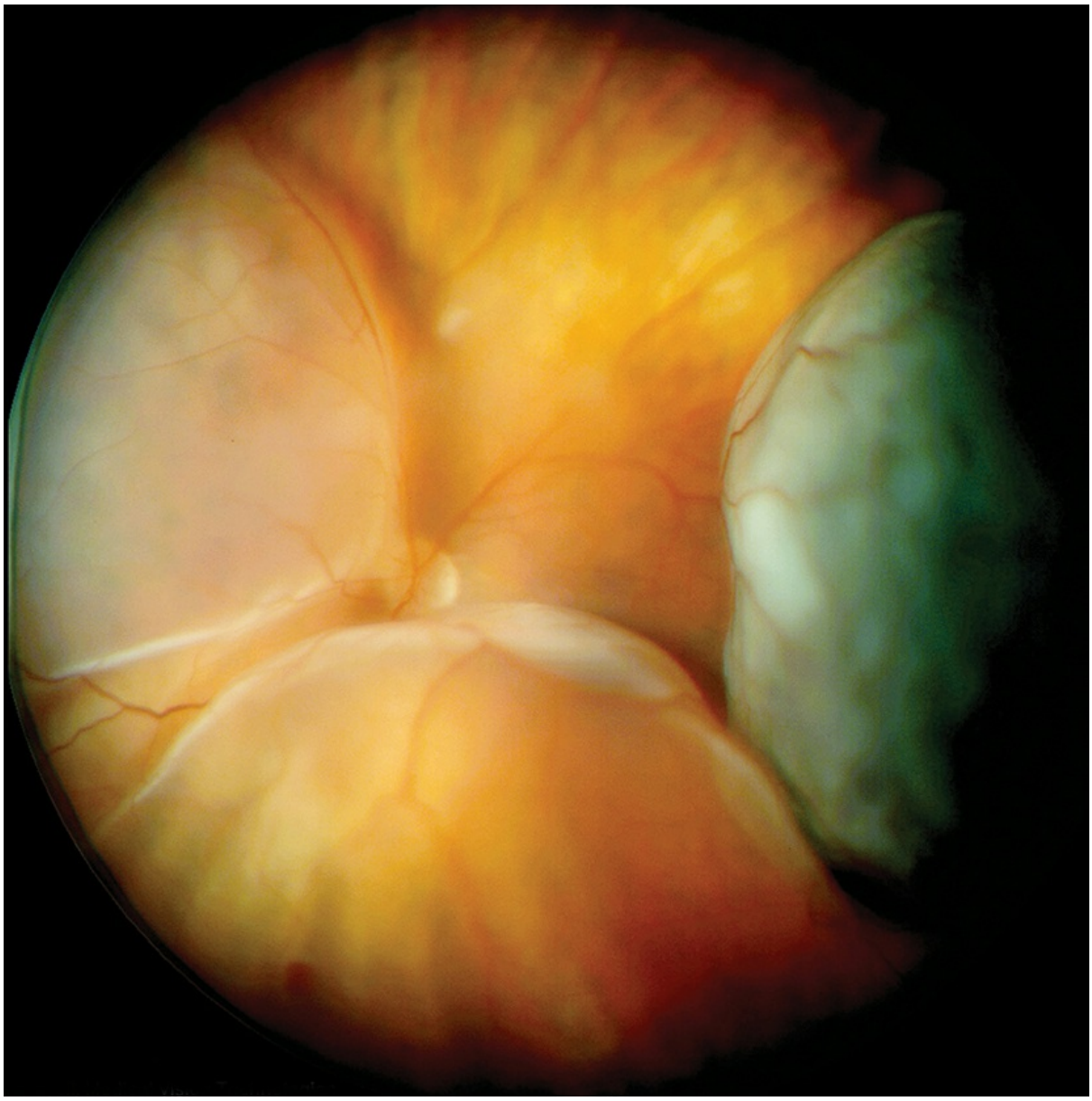
Jerry A. Shields, MD
Carol L. Shields, MD

CONTENTS

PART 1 TUMORS OF THE UVEAL TRACT

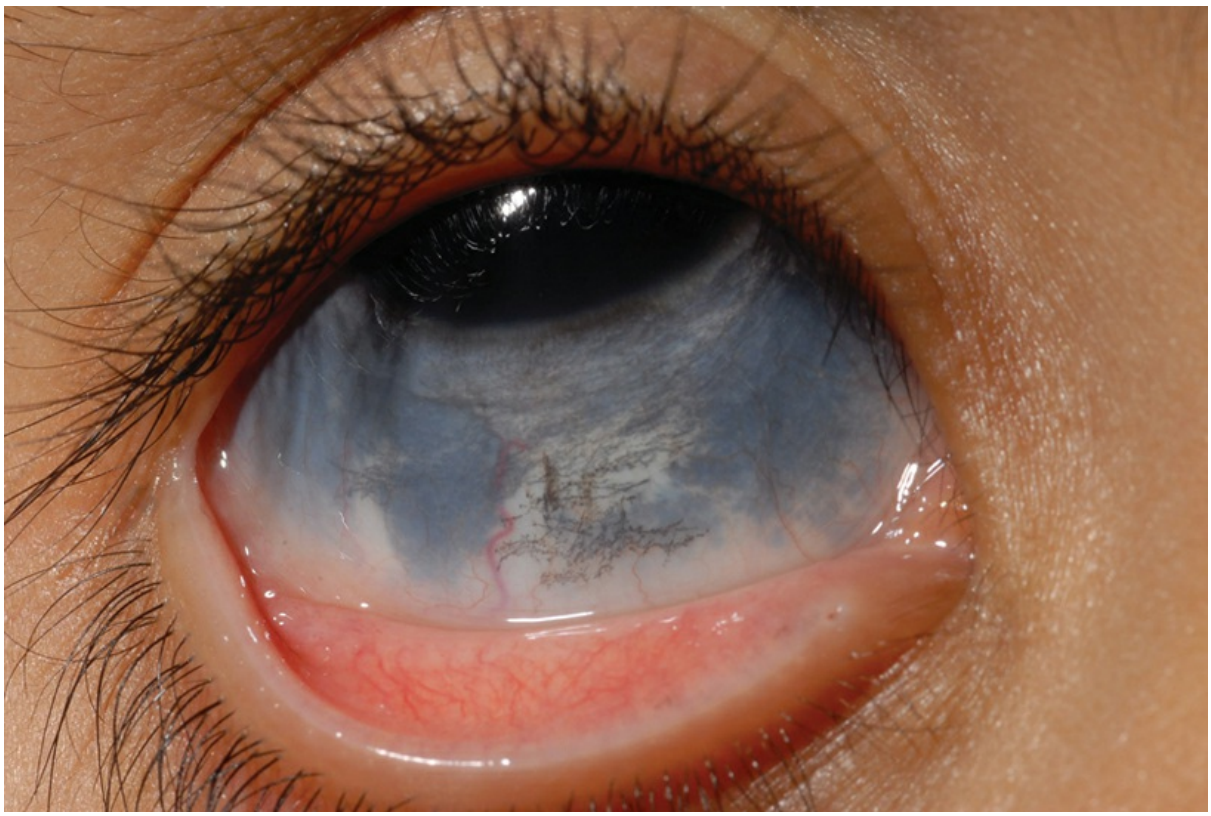
PART 1

TUMORS OF THE UVEAL TRACT



CHAPTER 1

CONGENITAL UVEAL LESIONS



INTRAOCULAR LACRIMAL GLAND CHORISTOMA

General Considerations

Several congenital abnormalities can affect the uvea, such as coloboma and aniridia, but most of them are not directly related to the differential diagnosis of intraocular tumors. The relationship between sporadic aniridia and nephroblastoma is well known, but it is not usually associated with intraocular neoplasms. Other than the systemic hamartomas and retinoblastoma, which are discussed in subsequent chapters, there are only a few congenital abnormalities of the uvea that have importance in the field of ocular oncology. Congenital iris cysts, that can simulate iris tumors, are more appropriately discussed in Chapter 4, which covers both congenital and acquired cysts. The two that are discussed here are intraocular lacrimal gland choristoma and congenital ocular melanocytosis.

Ectopic lacrimal gland tissue can occur in the orbit, conjunctiva, or eye (1). Intraocular lacrimal gland choristomas are rare and most have occurred in the iris and with a few involving both the iris and ciliary body. Several theories on the pathogenesis of intraocular lacrimal gland choristoma have been published (2).

Clinical Features

Clinically, intraocular lacrimal gland choristoma usually is recognized in early infancy as a fleshy reddish pink mass of the iris and/or ciliary body (1–11). It has a slightly lobulated surface that appears almost identical to the normal lacrimal gland as visualized at the time of orbital surgery. Clear cysts can sometimes appear within the lesion early in the clinical course. These have been likened to lacrimal gland cysts (dacryops). With regard to the natural course of intraocular lacrimal gland choristoma, the main mass does not tend to grow substantially, but the cysts within the lesion can progressively enlarge and cause iris atrophy, cataract, secondary glaucoma, and hyphema. The differential diagnosis includes iris nevus, melanoma, juvenile xanthogranuloma, and other iris granulomas in young patients. Although our experience has been limited mostly to larger lesions, it is quite possible that lacrimal gland choristoma can occur as a small, asymptomatic lesion that may remain clinically insignificant. Fine needle aspiration biopsy (FNAB) has been used to make the diagnosis in such cases (8).

Pathology

Histopathologically, intraocular lacrimal gland choristoma is a mass composed of normal-appearing lacrimal gland tissue. On occasion, the ducts or acini within the lesion become dilated due to accumulation of clear fluid, probably consistent with tears, in the lumen. This explains the “cysts” that are sometimes seen clinically.

Management

Because iris lacrimal gland choristoma is usually diagnosed in young children and is often stationary or slow growing, periodic observation is often an acceptable initial management. As mentioned earlier, FNAB has been employed to make the diagnosis based on recognition of typical benign epithelial cells consistent with lacrimal gland. In such cases, observation may be initially employed because many cases remain fairly stable. However, we believe that the appearance of a progressively enlarging cyst within the lesion should prompt early surgical removal of the mass to prevent glaucoma and visual impairment (9). If surgical excision of the lesion is necessary, iridocyclectomy may be required to remove the entire lesion (2,9). It is feasible that aspiration of the cyst could be a temporizing procedure, although we have not yet performed cyst aspiration in this rare condition.

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INTRAOCULAR LACRIMAL GLAND CHORISTOMA

Intraocular lacrimal gland choristoma has characteristic clinical features and is generally managed by local resection, which can be difficult in some cases due to the size and extent of the lesion. Two clinicopathologic correlations are shown.

1. Shields JA, Eagle RC Jr, Shields CL, et al. Natural course and histopathologic findings of lacrimal gland choristoma of the iris and ciliary body. *Am J Ophthalmol* 1995;119:219–224.
2. Shields JA, Hogan RN, Shields CL, et al. Intraocular lacrimal gland choristoma involving iris and ciliary body. *Am J Ophthalmol* 2000;129:673–675.

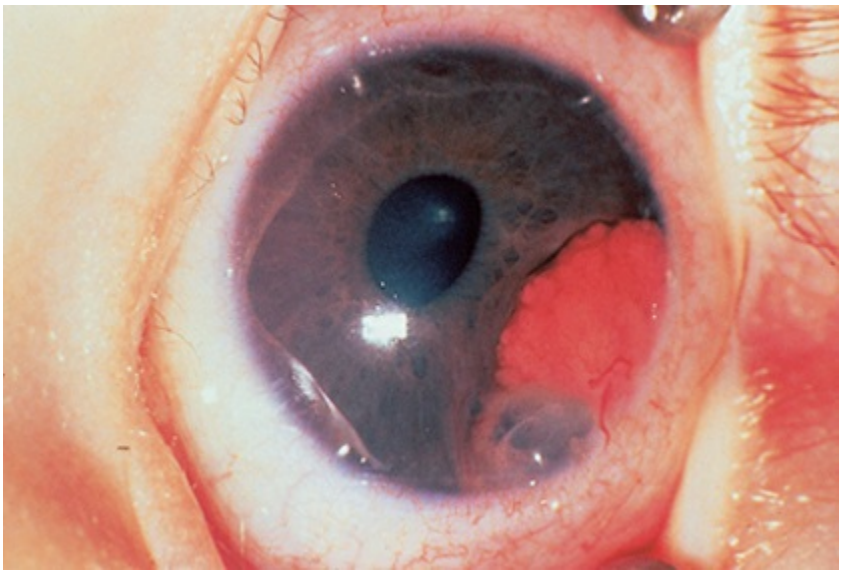

Figure 1.1. Appearance of the lacrimal gland choristomas of the left iris in a 7-week-old baby girl. Note the pink color of the lesion, with the clear cyst in the inferior part of the mass. The lesion was initially followed without treatment.

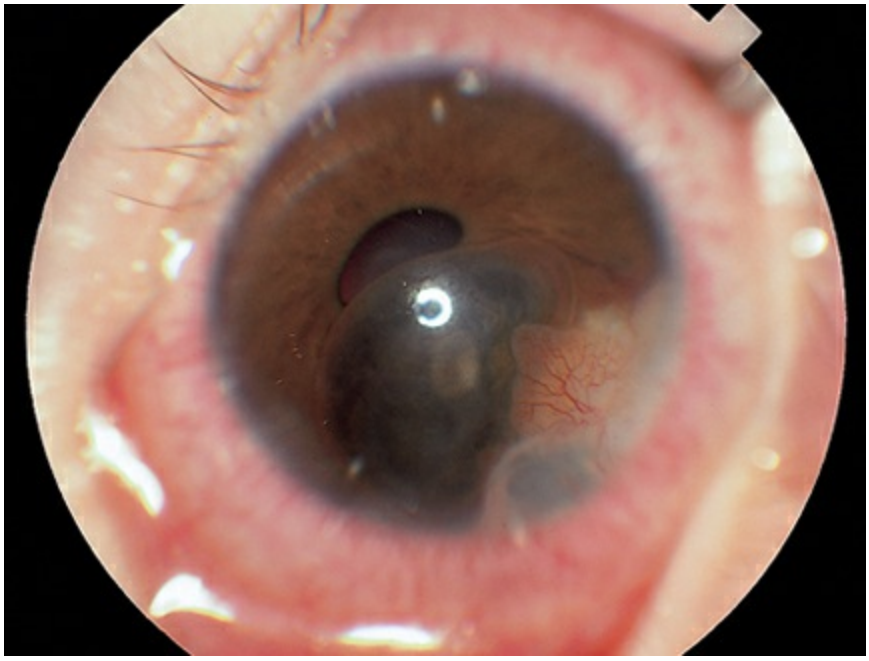

Figure 1.2. Clinical appearance of the mass when the infant was 19 months old. Avascularized corneal pannus is now present over the peripheral aspect of the mass. The inferior cyst is unchanged, but a new cyst is emanating from the mass and filling almost half of the anterior chamber inferotemporally and obscuring part of the pupil. The lesion was removed by a sector iridectomy. About 1 year later, retinal detachment ensued and surgical repair was attempted elsewhere. According to unconfirmed history, enucleation was eventually performed elsewhere.

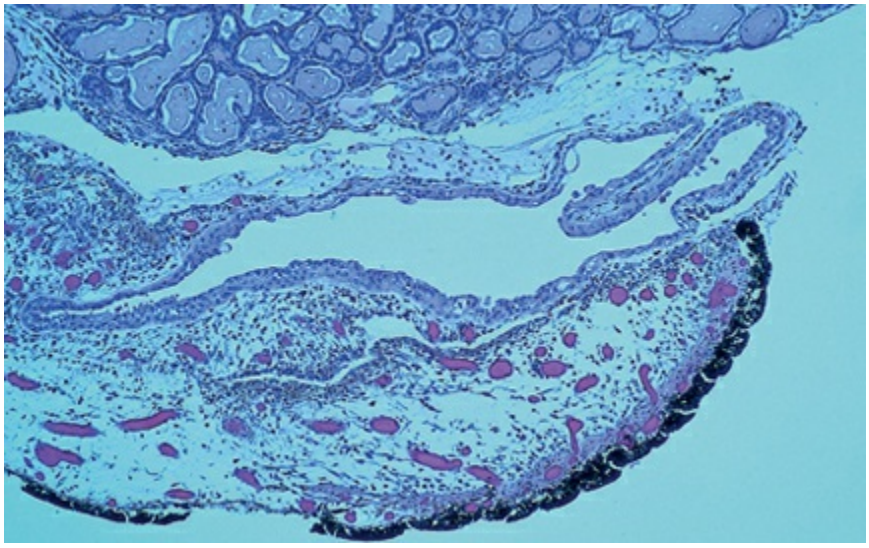

Figure 1.3. Low-magnification photomicrograph showing glandular mass (above) and irregular, partially collapsed cyst (below). (Hematoxylin–eosin ×10.)

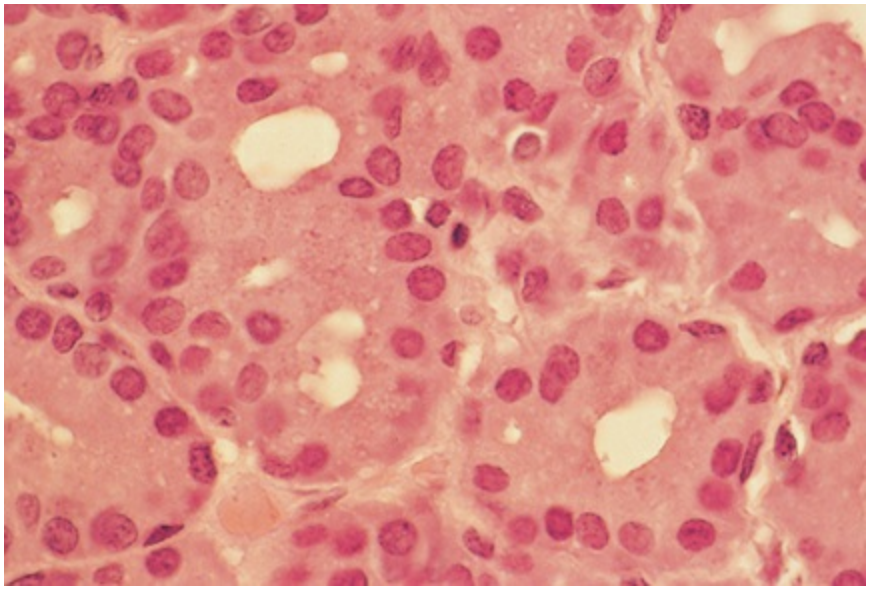

Figure 1.4. Photomicrograph of the solid portion of the lesion, showing glandular tissue identical to normal lacrimal gland. (Hematoxylin–eosin ×200.)

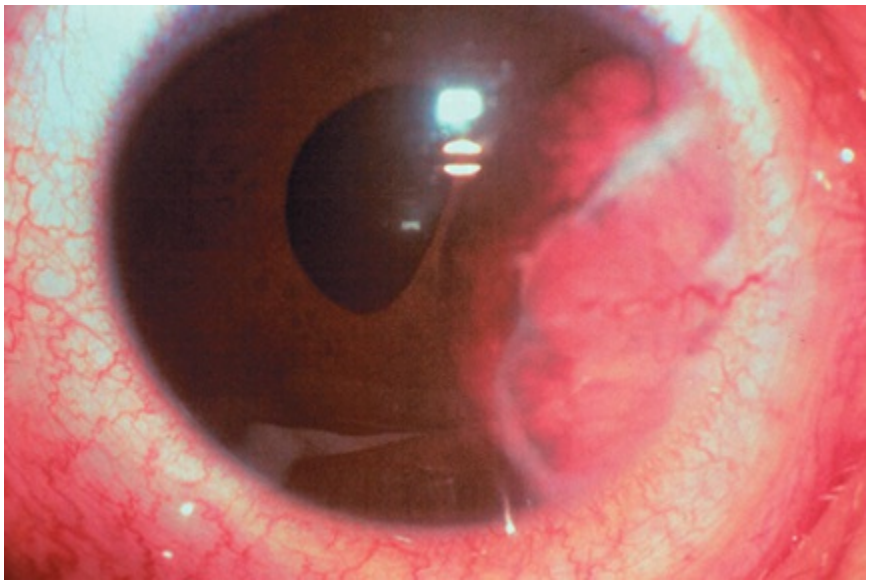

Figure 1.5. Clinical appearances of another lacrimal gland choristoma of the iris and ciliary body in a 12-month-old boy. The lesion had been present since birth. Note the remarkable similarity to the prior case.

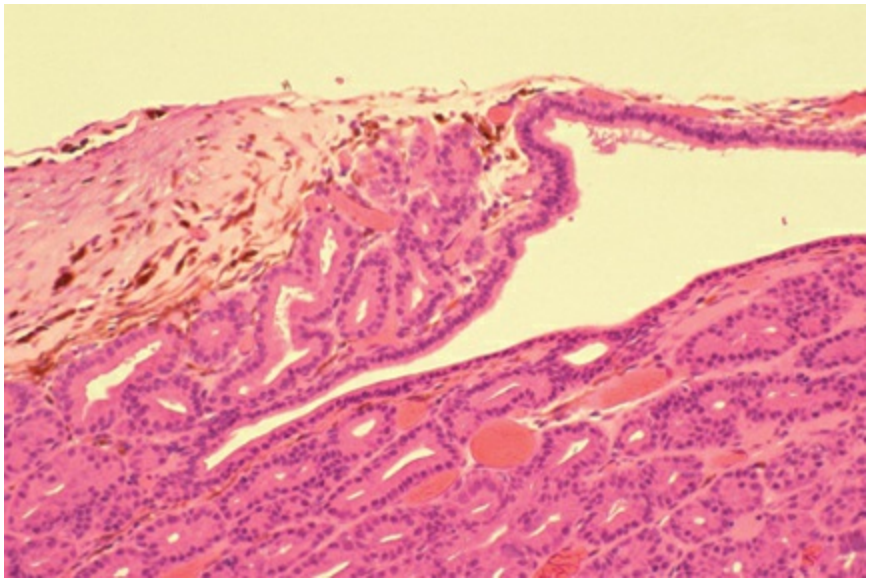

Figure 1.6. Histopathology of lesion seen in Figure 1.5, showing dense fibrous tissue (above) and normal lacrimal gland tissue (below). (Hematoxylin–eosin ×40.)

CONGENITAL OCULAR MELANOCYTOSIS

General Considerations

Congenital ocular melanocytosis is a well-known condition that has been the subject of many reports, mainly because of its relationship to uveal melanoma (1–31). It can be divided into ocular melanocytosis and oculodermal melanocytosis, or nevus of Ota (1–5,10–12). They have identical episcleral and uveal pigmentation, but the latter has periocular cutaneous pigmentation as well. Both predispose to uveal melanoma, as well as melanoma of the ipsilateral skin, orbit, meninges, and central nervous system, in areas where there are excess melanocytes as a part of this condition. It has been estimated that about 1 in 400 Caucasian individuals with ocular or oculodermal melanocytosis will develop uveal melanoma, sometimes in childhood (6,9,11,12). Conversely, about 3% of patients with uveal melanoma will have ocular melanocytosis (11). In extremely rare instances, the scleral pigmentation can give rise to melanoma.

Several patients with bilateral ocular melanocytosis have developed bilateral uveal melanoma (11). It has recently been found that uveal melanoma associated with ocular melanocytosis has a more aggressive clinical course and an increased chance of developing metastasis (11,12). About 10% of patients with ocular melanocytosis have ipsilateral elevated intraocular pressure, seemingly related to melanocytic hyperpigmentation of the anterior chamber angle (5).

Clinical Features

The most evident finding is unilateral (occasionally bilateral) hyperpigmentation of the sclera and uveal tract. The scleral pigmentation is characterized by flat, gray-to-brown patches of pigmentation that is quite different from the more localized nodule of extraocular extension of uveal melanoma (11,12). Both the uveal and the scleral pigment can have either a diffuse or sector distribution. Heterochromia is often a predominant feature, with part or all of the affected iris being darker than the iris of the fellow eye. The choroidal pigmentation is greater than in the opposite eye. This condition most often involves the entire choroid but it can occur in a partial or sector distribution in the choroid (10). With time, there develops overlying degeneration of the retinal pigment epithelium and numerous drusen in the area of choroidal involvement. A higher incidence of melanocytoma of the optic nerve in affected eyes has been recognized (16). Multiple uveal melanoma can be seen in patients with ocular melanocytosis (24,29). The presence of foci of orange pigment over choroidal melanocytosis is a suggestion that the lesion is becoming thicker and potentially evolving into choroidal melanoma.

Another interesting variation of ocular melanocytosis is iris mammillations (22,25,28). These are numerous, villiform, closely compact dark-brown nodules that occupy the anterior aspect of the iris. They can be appreciated to some degree in most patients with ocular melanocytosis. In some instances the patient has only iris mammillations, which is considered to be an incomplete expression (forme fruste) of ocular melanocytosis (25). They are also seen in some patients with neurofibromatosis and are different from the discrete iris Lisch nodules seen with that condition. A relationship of ocular melanocytosis to phacomatosis

pigmentovascularis and nevus flammeus has recently been recognized and affected patients also have an increased risk for uveal melanoma (14).

Diagnostic Approaches

The diagnosis of ocular and oculodermal melanocytosis is best made by recognition of its typical clinical features described above. The recent use of optical coherence tomography has confirmed increased choroidal thickness in the area of pigmentation and such areas must be checked periodically to detect very early melanoma (15).

Pathology

Histologically, ocular melanocytosis is characterized by dense, heavily pigmented melanocytes in the affected uveal tract (16,17). The melanoma that can occur with ocular melanocytosis usually arises in the choroid and/or ciliary body in patients of any age (9–12). Iris melanoma in patients with ocular melanocytosis is rare. However, we have observed a nodular iris melanoma arising from sector iris melanosis in a child with ocular melanocytosis (30).

Management

Because of the increased incidence of melanoma, patients with ocular melanocytosis should be carefully examined periodically for their entire life, looking for evidence of uveal, orbital, or brain melanoma.

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CONGENITAL OCULAR MELANOCYTOSIS: EXTERNAL FEATURES

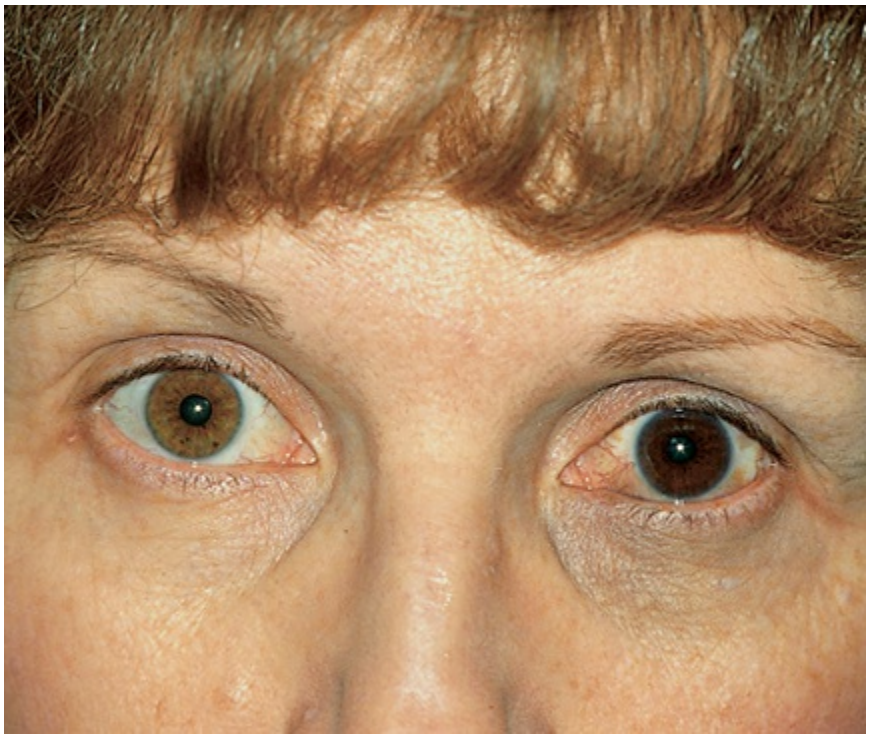

Figure 1.7. Iris heterochromia secondary to ocular melanocytosis in the left eye of a 48-year-old woman. Note that the left iris is darker. Note also the subtle skin hyperpigmentation on the sclera and left lower eyelid. These findings are typical of oculodermal melanocytosis (nevus of Ota).

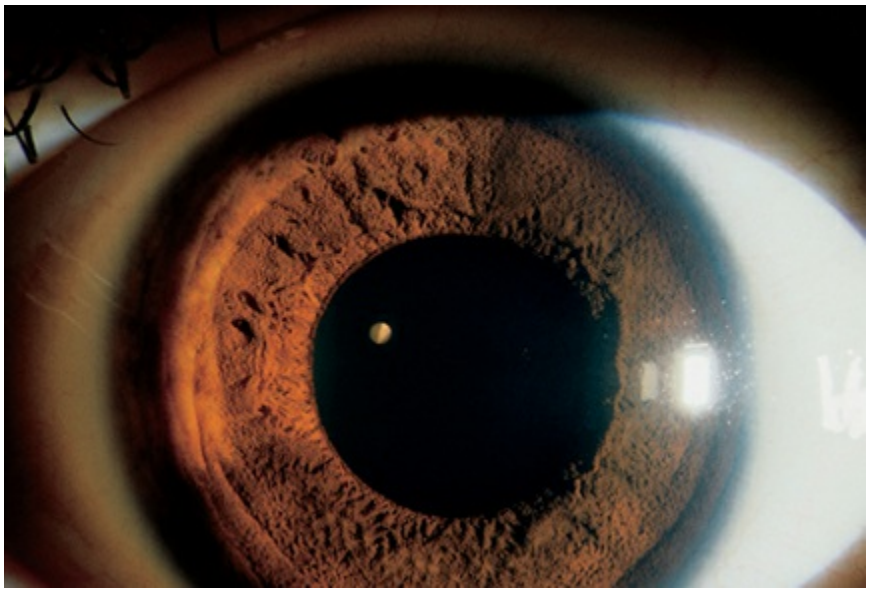

Figure 1.8. Closer view of affected iris in congenital ocular melanocytosis. Note that most of the iris is dark brown and has numerous small nodules called mammillations, best seen inferiorly.

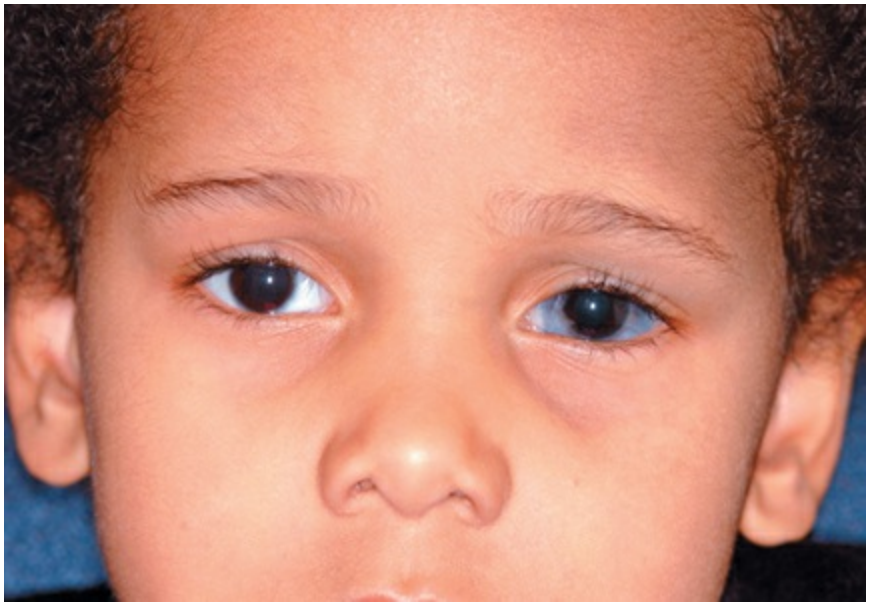

Figure 1.9. Marked scleral involvement with melanocytosis in a child. Note that this child also has very subtle pigmentation of the left lower eyelid, meeting the criteria for oculodermal melanocytosis.

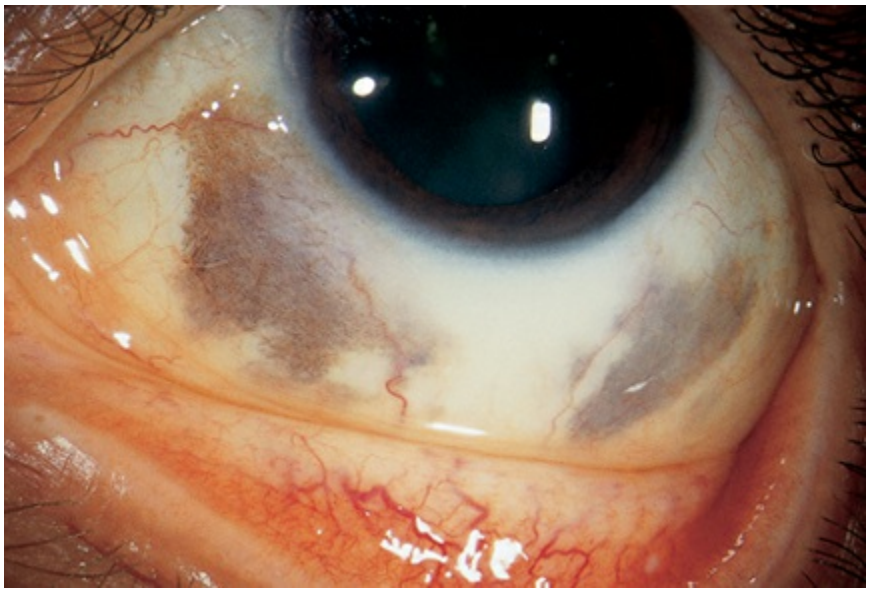

Figure 1.10. Inferior scleral melanocytosis in a 56-year-old woman.

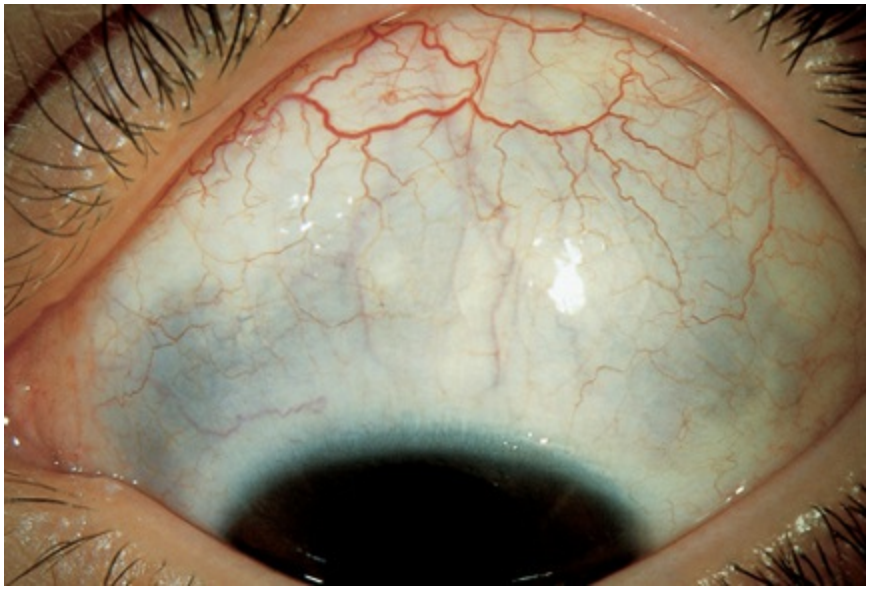

Figure 1.11. Superior scleral melanocytosis in a 40-year-old man.

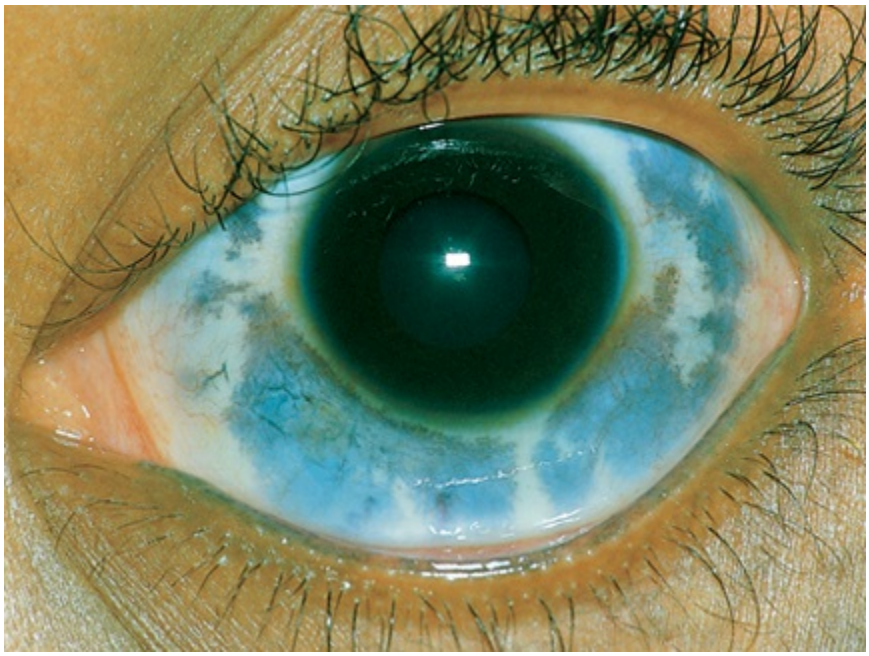

Figure 1.12. More severe melanocytosis in the left eye of a 30-year-old woman. Note that the scleral pigment has a blue-gray color in this case.

CONGENITAL OCULAR MELANOCYTOSIS: FUNDUS FEATURES

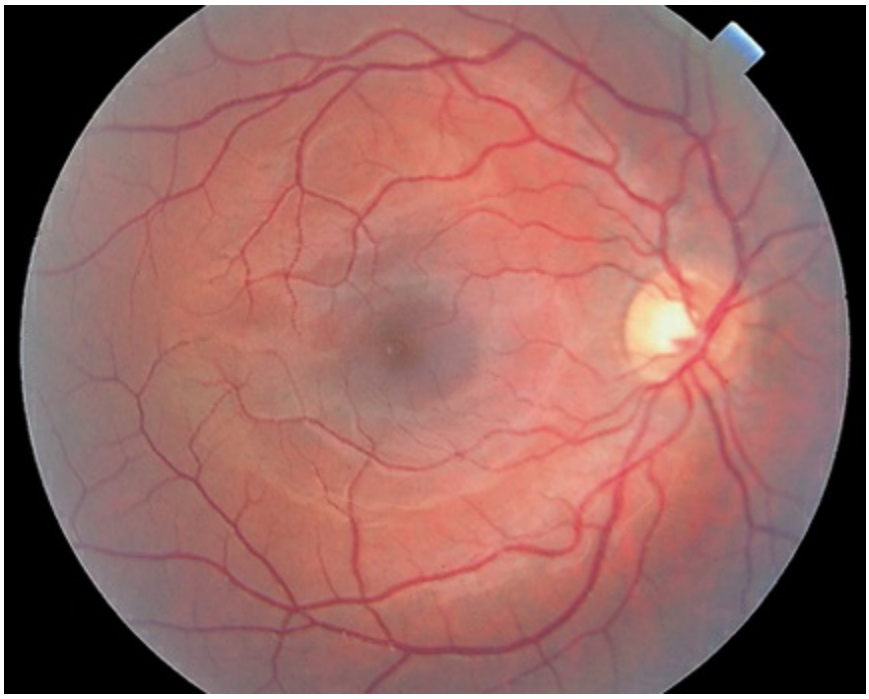

Figure 1.13. Fundus photograph of the unaffected right eye of the patient with contralateral ocular melanocytosis. The background fundus color is normal.

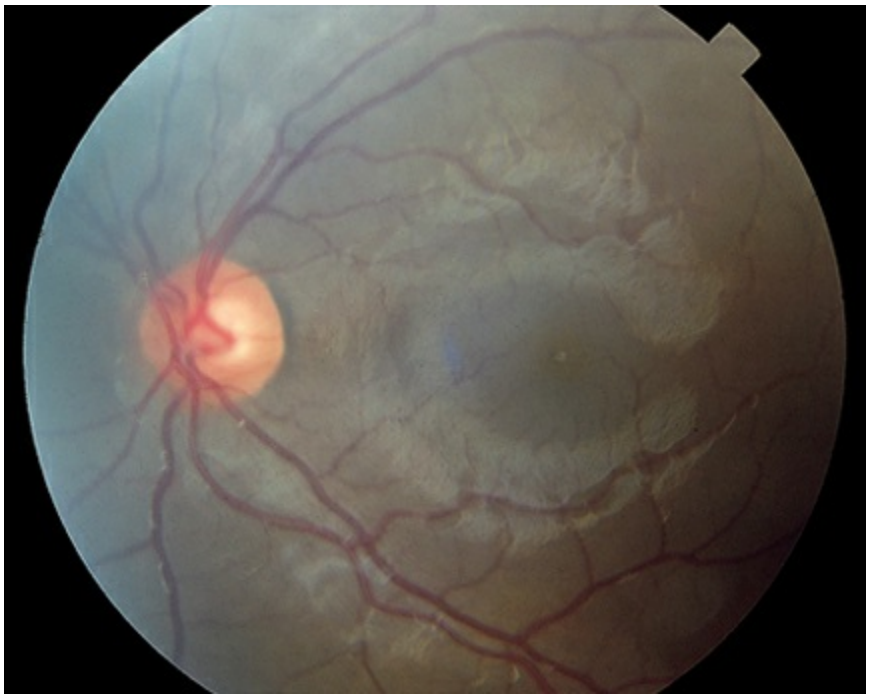

Figure 1.14. Fundus photograph of the affected contralateral left eye of the patient shown in Figure 1.13. The background fundus color is darker than the fellow right eye.

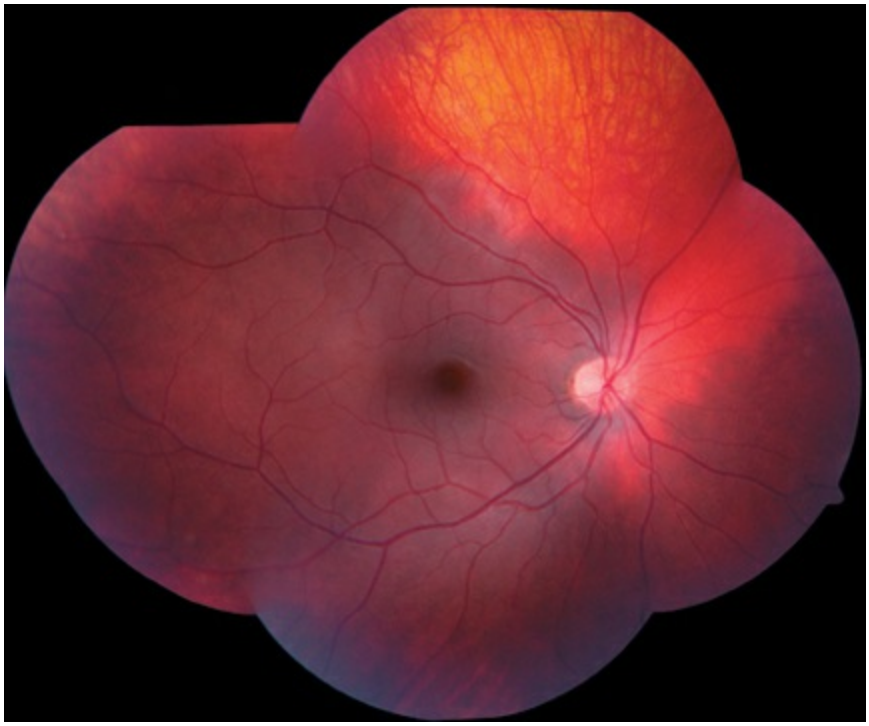

Figure 1.15. Montage fundus photograph of sector choroidal melanocytosis in a young man. In this case the choroidal hyperpigmentation is nasal, inferior, and temporal, but it spares the superior quadrant.

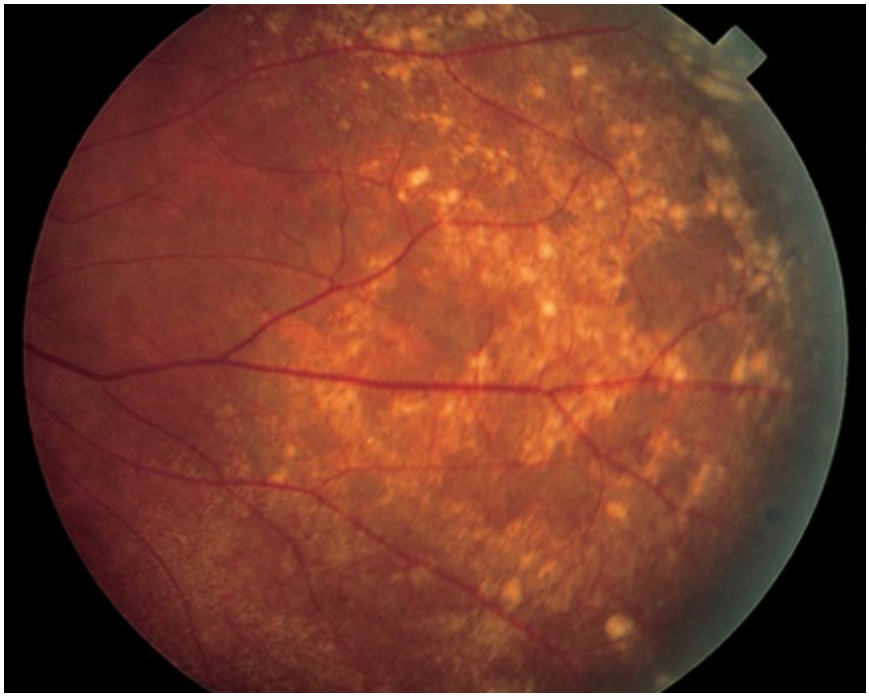

Figure 1.16. Typical peripheral retinal pigment epithelial alterations and drusen in a 48-year-old person with ocular melanocytosis. The extent and severity of these pigment epithelial changes increase with age.

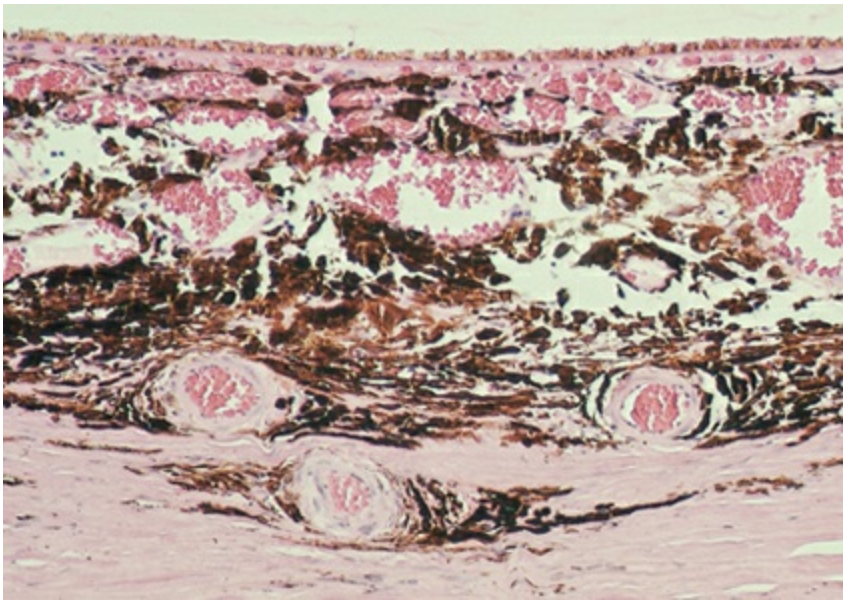

Figure 1.17. Histopathology of the choroid in a patient with ocular melanocytosis. There is increased choroidal thickness and hyperpigmentation secondary to increased number of choroidal melanocytes. (Hematoxylin–eosin ×40.)

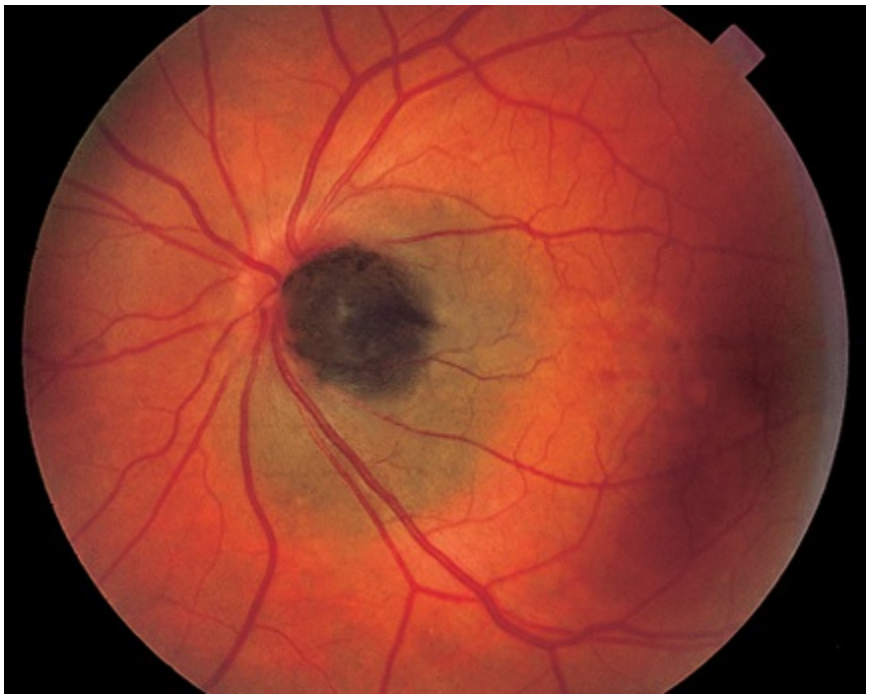

Figure 1.18. Melanocytoma of the optic disc in a patient with scleral melanocytosis in the opposite eye. There appears to be a slight increased incidence of melanocytoma of the optic disc in patients with ocular melanocytosis (personal observations). Note the juxtapapillary choroidal component of the lesion.

CONGENITAL OCULODERMAL MELANOCYTOSIS (NEVUS OF OTA) IN NON-CAUCASIANS

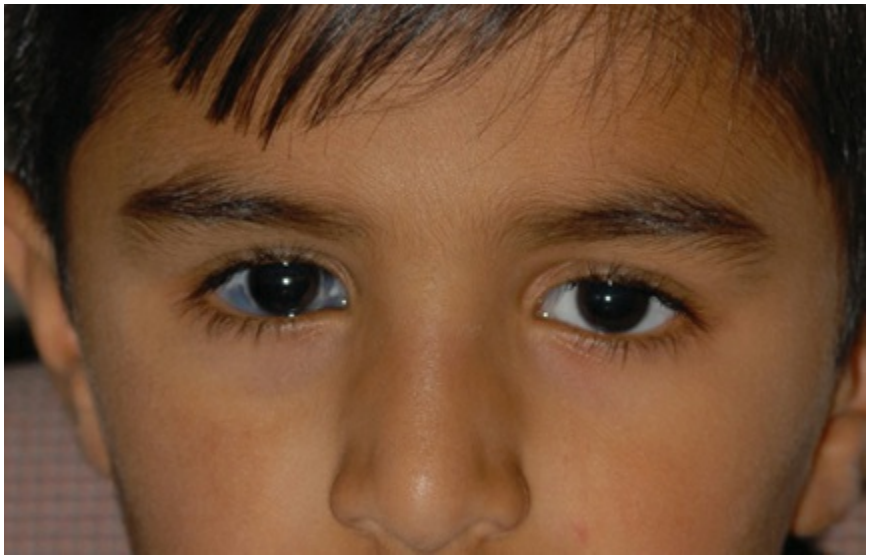

Figure 1.19. Oculodermal melanocytosis of right eye in an 8-year-old Indian boy.

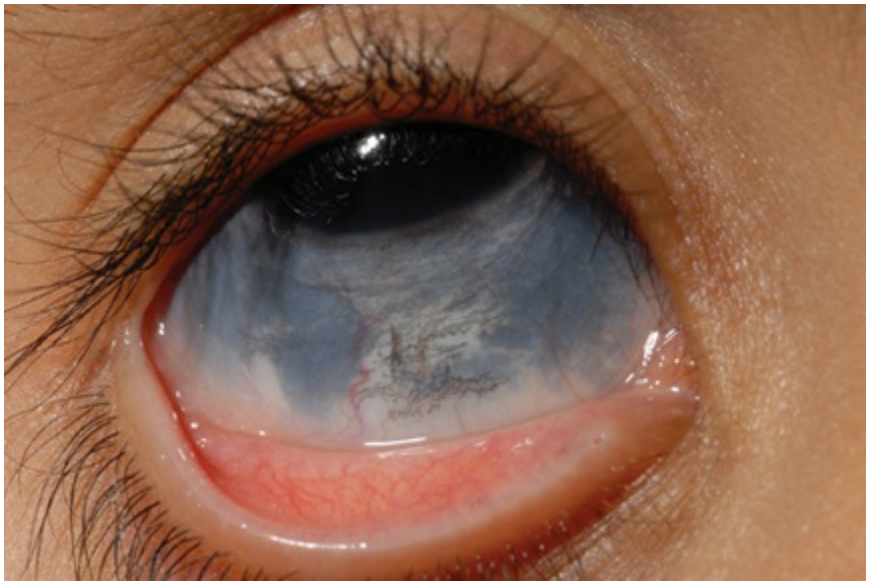

Figure 1.20. Heavy scleral pigmentation is noted in patient in Figure 1.19.

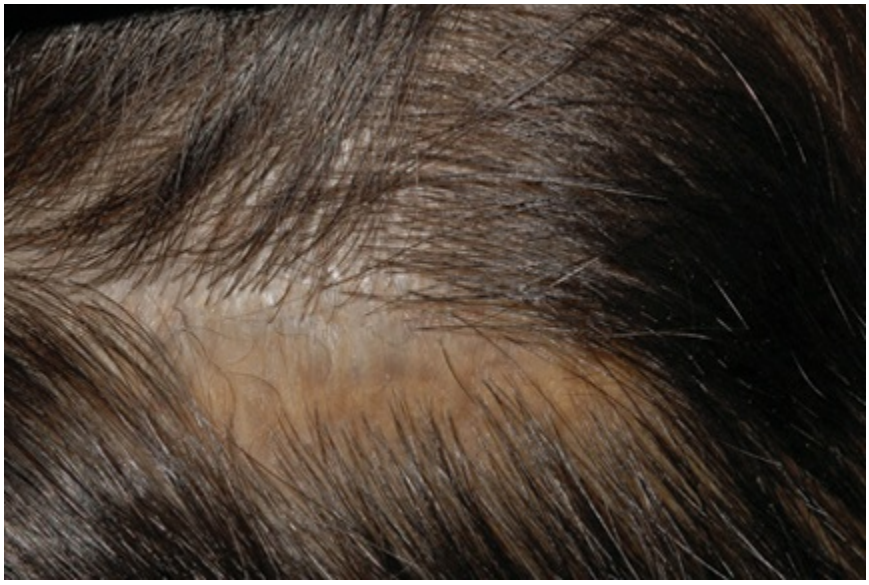

Figure 1.21. Ipsilateral blue scalp melanocytosis is found in patient in Figure 1.19.

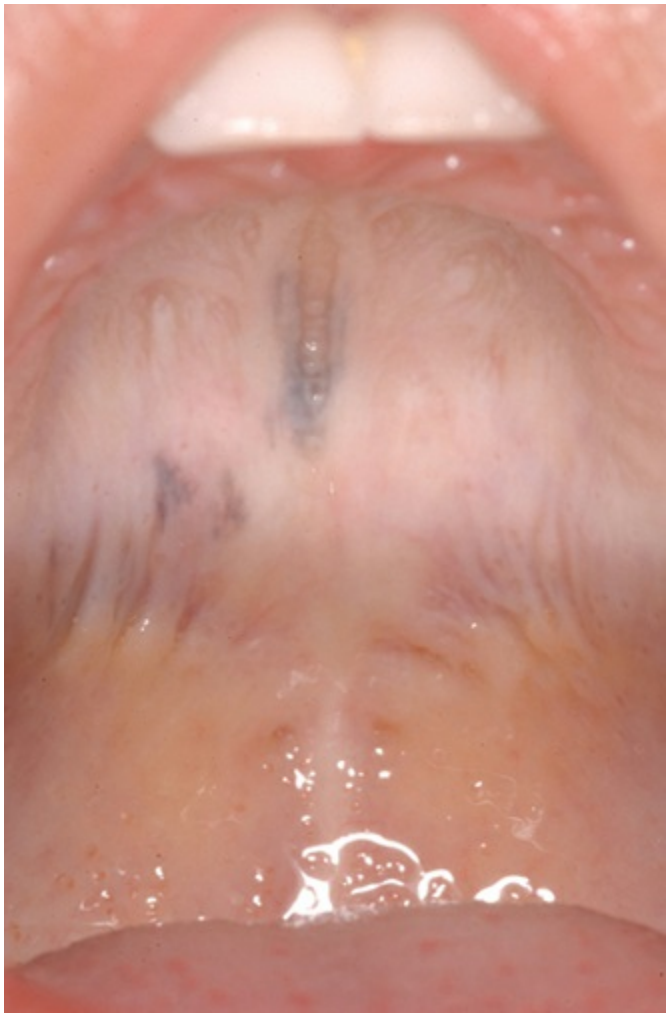

Figure 1.22. Ipsilateral subtle, pigmentation of the palate of patient in Figure 1.19. This subtle pigmentation can be overlooked in patients with oculodermal melanocytosis.

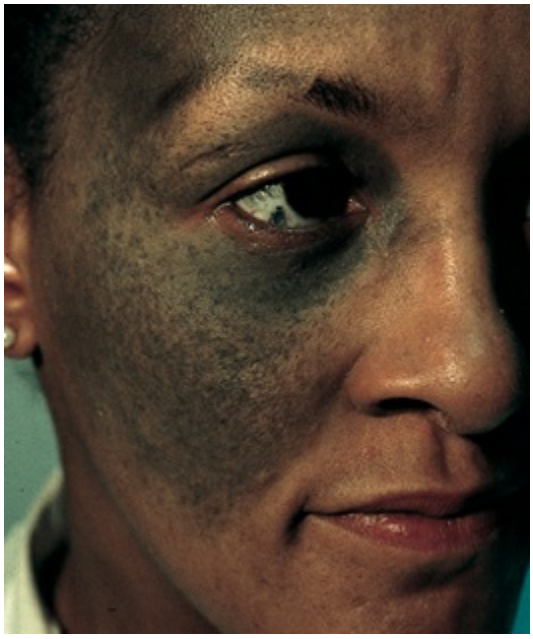

Figure 1.23. Oculodermal melanocytosis on the right side in a 36-year-old African-American patient. Oculodermal melanocytosis may be more difficult to diagnosis with dark-skinned individuals. However, as in white patients, affected black patients also have a higher incidence of melanoma in the pigmented areas.

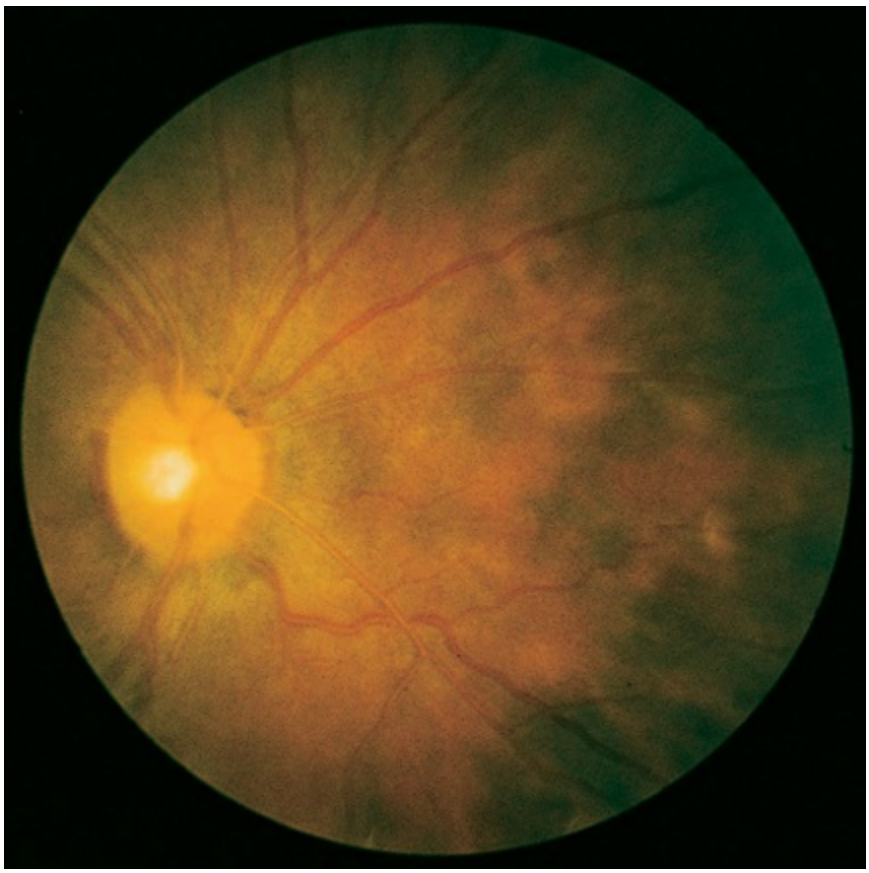

Figure 1.24. Fundus of the right eye in patient shown in Figure 1.23. Note the marked pigment epithelial alterations secondary to the thickened, hyperpigmented choroid.

Ophthalmology

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