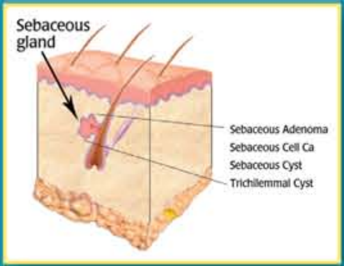


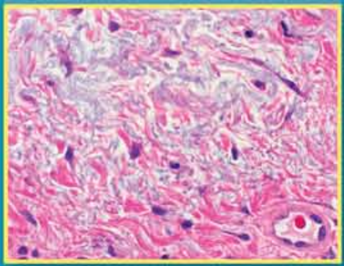

Diagnostic Atlas of Common Eyelid Diseases

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Preface

For any clinician dealing with ophthalmic diseases, individual lesions of the eyelid and conjunctiva can be extremely confusing. From a practical perspective such lesions are either benign or malignant, and can be cystic or solid, melanotic or amelanotic. Certainly the most important diagnostic question is whether the lesion represents a malignant tumor that requires biopsy and more definitive treatment. Often, following biopsy, the histopathologic diagnosis is difficult to interpret since most are histologically based on specific tissue cells of origin. The question arises, of course, as to the clinical relevance of the diagnosis. For the majority of benign lesions the treatment will be the same; that is, observation or, if of cosmetic or functional significance, surgical excision. Some lesions may be amenable to ancillary therapy such as steroid injection, cryotherapy, laser ablation, or radiotherapy.

Malignant tumors of the eyelid present a special category of concern. Some, like the basal cell carcinoma, rarely metastasize, but can be locally aggressive; when small they are less of an immediate threat. Others, such as sebaceous cell carcinoma and malignant melanoma, have a metastatic potential that requires more immediate and aggressive intervention. A high index of suspicion and a low threshold for biopsy will lead to a correct diagnosis much of the time.

In the pages that follow we present the current state of our knowledge on a number of eyelid diseases with which all ophthalmic clinicians should be familiar. Several introductory chapters discuss eyelid anatomy, examination, evaluation and decision making, and biopsy and reconstructive techniques. The main body of the atlas is divided into two sections, eyelid malpositions and eyelid lesions. In Chapter 7: Eyelid Malpositions, we discuss congenital and acquired dystopias of the eyelids, such as ptosis, ectropion, epicanthus, and lagophthalmos. In Chapter 8: Eyelid Lesions, we present conditions such as seborrheic keratosis, basal cell carcinoma, and hemangioma.

The concept of this book grew out of the need for a quick and easy-to-use reference to specific clinical and histopathologic information on common eyelid malpositions and diseases. For each condition we give an introduction, clinical presentation, and treatment, with appropriate illustrations. For eyelid lesions, we also include histopathology and differential diagnosis. Available information has been condensed into minimal text without cited references. Within each section, diseases are arranged alphabetically to make it easier to find specific entries. The reader will find the same information, such as clinical presentation or histopathology, in the same sequence for every disease. At the end of each disease entry we include selected references with no attempt at presenting a comprehensive literature review.

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Alan D. Proia

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Peter Rubin, M.D.	Pilomatrixoma
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Contents

Chapter 1

Anatomy of the Eyelids

INTRODUCTION

The eyelids serve several valuable functions. Most importantly they provide mechanical protection to the globe. They also provide vital chemical elements to the precorneal tear film, and help distribute these layers evenly over the surface of the eye. During the blink phase the eyelids propel tears to the medial canthus where they enter the puncta of the lacrimal drainage system. The eyelashes along the lid margins sweep air-borne particles from in front of the eye, and the constant voluntary and reflex movements of the eyelids protect the cornea from injury and glare.

In the young adult the interpalpebral fissure measures 10 to 11 mm in vertical height. In middle age this is reduced to only about 8 to 10 mm (1) and in old age the fissure may be only 6–8 mm or less. The horizontal length of the fissure is 30 to 31 mm. The upper and lower eyelids meet at an angle of approximately 60 degrees medially and laterally. In primary position of gaze the upper eyelid margin lies at the superior corneal limbus in children and 1.5 to 2.0 mm below it in the adult. The lower eyelid margin usually rests at the inferior corneal limbus or just slightly above it.

The margin of each eyelid is about 2 mm thick. Posteriorly the marginal tarsal surface is covered with conjunctival epithelium, interrupted by the meibomian gland orifices (Fig. 1). Anteriorly the margin is covered with cutaneous epidermis from which emerge the eyelashes. The gray line is a faint linear zone separating these two regions. Between the skin and conjunctiva at a level 5 mm above the tarsus are, layered from front to back, the orbicularis muscle, the orbital septum, the preaponeurotic fat pockets, the levator aponeurosis, and Müller's supratarsal muscle.

EYELID SKIN

The skin covers the external surface of the body and provides significant protection against trauma, solar radiation, temperature extremes, and desiccation. It also allows for major interaction with the environment. The skin of the eyelid is the thinnest in the body owing to only a scant development of the dermis and subcutaneous fat.

The epidermis is the outer layer of the skin averaging about 0.05 mm in thickness on the eyelids, compared to the palms and soles where it can attain a thickness of 1.5 mm. It contains no blood vessels and is dependent upon the underlying dermis for its nutrients (Fig. 2). There are four layers to the epidermis consisting of keratinocytes layered from deepest to most superficial in progressive stages of differentiation. These keratinocyte cells proliferate and push more formed cells upward into successively higher layers. As they move upward the keratinocytes produce a fibrous protein, called keratin. Bundles of tonofilaments help distribute stress enabling the epidermis to withstand a fair amount of surface abuse. The layers of the epidermis are from top to bottom:

- Stratum corneum
- Stratum granulosum
- Stratum spinosum
- Stratum basale

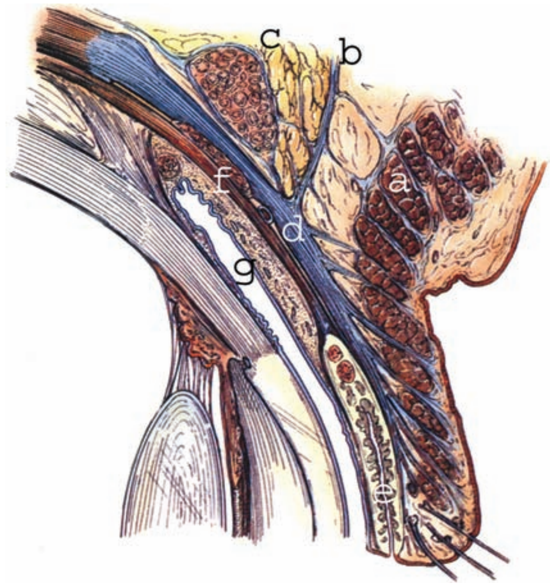

Figure 1 Sagittal cross-section of the upper eyelid. **a.** Orbicularis muscle; **b.** orbital septum; **c.** preaponeurotic fat pad; **d.** levator aponeurosis; **e.** tarsal plate; **f.** Muller's supratarsal muscle; **g**. conjunctiva.

At the base of the sequence is the stratum basale (= basalis) containing cuboidal or columnar cells arranged in a single row and containing large nuclei (Fig. 3). The basal layer of columnar cells with mitotic figures serves for keratinocytic proliferation and was formerly referred to as the stratum germinativum. Together, the strata basale and germinativum are called the basal cells. The stratum spinosum is the next higher layer and is composed of polygonal cells which form spines when they shrink during histologic preparation because of the presence of desmosomes between adjacent cells. The cytoplasm is filled with tonofilaments and phospholipids. The stratum granulosum is 3 to 5 cells thick; these cells contain keratohyalin granules rich in histidine and precursors of the protein filaggrin that promotes keratin filament aggregation. Toward the top of this layer the cells lose their nucleus and become more flattened. Changes in enzyme function cause cell death as the cells enter the upper-most layer, the stratum corneum. This layer is made of dead, flattened keratinocytes that shed about every few weeks in a process known as desquamation. This layer provides a waterproof covering that also resists minor abrasions. In thicker skin such as the palms and soles a layer called the stratum lucidum is found between the granular and cornified strata; this appears as a clear homogeneous amorphous zone.

The epidermis also contains several specialized types of cells. Melanocytes of neural crest origin are scattered among the keratinocytes in the stratum basale and the deeper layers of the stratum spinosum. They are pale cells which produce long processes that extend between the keratinocytes, and produce melanin which is stored in granules called melanosomes. These

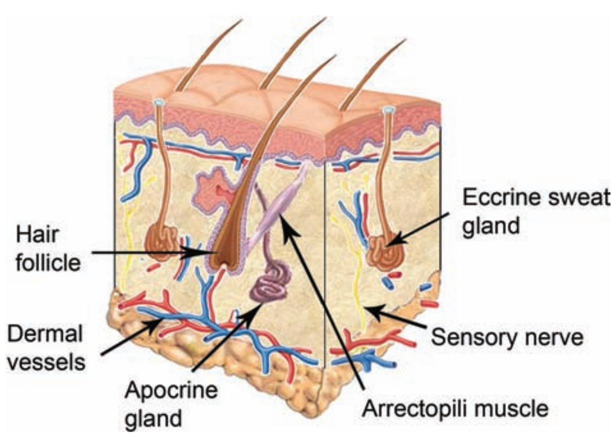

Figure 2 Anatomic section of eyelid skin showing major dermal adnexal structures.

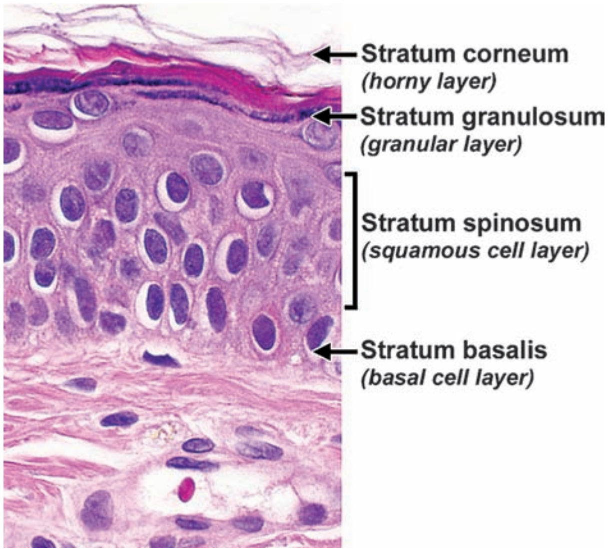

Figure 3 Histologic section of eyelid skin showing the four layers of the epidermis, and the dermis.

are ultimately transferred to the keratinocytes where they accumulate on the sun-exposed side of the nucleus to protect it from UV radiation. Langerhans cells, or epidermal dendritic cells, are macrophages that originate in the bone marrow and migrate to the epidermis. They are present in all epidermal layers, but are concentrated in the stratum spinosum. They serve as antigen-presenting cells that ingest and process foreign antigens and present them to lymphocytes for activation of the immune system. The function of the Merkel cell is not completely understood. They are derived from neural crest cells and are attached to keratinocytes by desmosomes. They are found throughout the epidermis but are particularly abundant in the basal layer. Merkel cells are associated with nonmyelinated nerve endings with which they form the Merkel disc which may act as a mechanosensory receptor for light touch.

Beneath the epidermis is the basement membrane and below that the dermis. The stratum basale is connected to the basement membrane by protein fibers. On the dermal side anchoring fibers of collagen tether the dermis to the basement membrane. The basal membrane is irregular and epidermal extensions project into the dermis to form a system of rete ridges. These are more prominent in areas where the skin undergoes shearing stress.

The dermis lies beneath the basement membrane and on the eyelids is about 0.3 mm in thickness compared to other parts of the body where it may be up to 3 mm thick. It contains three types of tissues that are not layered: collagen, elastic tissue, and reticular fibers. The upper papillary dermal layer contains a thin arrangement of collagen fibers. The lower reticular dermal layer is thicker and contains thick collagen fibers arranged parallel to the surface. The reticular layer also contains fibroblasts, mast cells, nerve endings, lymphatics, and epidermal appendages surrounded by a ground substance of mucopolysaccharides, chondroitin sulfates, and glycoproteins.

The fibroblast is the major cell type in the dermis. These secrete elastin and a procollagen that is then cleaved by proteolytic enzymes into collagen which becomes cross-linked. Collagen makes up nearly 70% of the dermis by weight.

A number of epidermal appendages lined with epithelium lie within the dermis. These include hair follicles associated with an arrectopili muscle attached to the dermal-epidermal junction. Apocrine sweat glands of Moll are coiled glands in the deep dermis that empty via a long ductule into the uppermost portion of the hair follicle. Apocrine glands secrete by cellular decapitation with the apical portion of the secretory cell mixed with sialomucin producing a more viscous secretion with cellular debris. They are concentrated along the eyelid margins.

Sebaceous glands contain epithelium that is an outgrowth of the external root sheath of the hair follicle. These are holocrine glands that shed the entire epithelial cell along with secretory products of complex oils, fatty acids, wax, and cholesterol esters called sebum. A large sebaceous gland is associated with each hair follicle and empties its secretions directly into

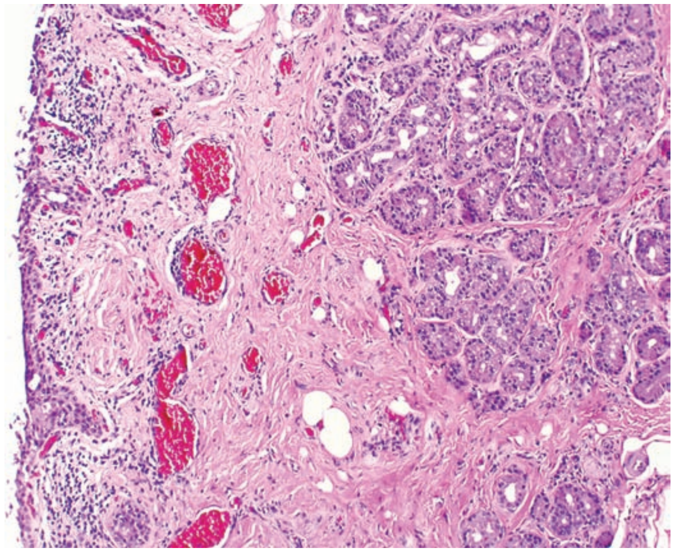

Figure 4 Histologic section of the conjunctiva with numerous glands of Wolfring.

the follicle. The hair follicle along with the sebaceous and Moll glands form the pilosebaceous unit. Additional small sebaceous glands called glands of Zeis are present between follicles and discharge their contents directly onto the skin surface. Eccrine sweat glands are also present in the dermis but they are not associated with the hair follicle. They open directly onto the epidermal surface via a long straight ductule. Eccrine glands secrete a clear fluid composed of water, salts, glycogen, and sialomucin.

Blood vessels and nerve endings course throughout the dermis where they derive from similar structures in the sub-dermis and deep fascia. Specialized sensory structures called Meissner's and Vater-Pacini corpuscles within the dermis transmit sensations for touch and pressure.

Beneath the dermis is a subcutaneous layer of fat and connective tissue. Subcutaneous fat is very sparse beneath the preseptal portion of the eyelid skin, and absent from the more distal pretarsal portions. Beneath the skin within the eyelid are also found other structures that can be the focus for disease processes. On the subconjunctival side of the eyelid these accessory structures include the accessory lacrimal glands of Krause and Wolfring beneath the conjunctiva and are concentrated on the lateral side (Fig. 4), and the meibomian glands which are modified sebaceous glands within the tarsal plates (Fig. 1).

THE ORBICULARIS MUSCLE

The orbicularis oculi is a complex striated muscle that lies just below the skin. It is divided anatomically into three contiguous parts—the orbital, preseptal, and pretarsal portions (Fig. 5). The orbital portion overlies the bony orbital rims. It arises from insertions on the frontal process of the maxillary bone, the orbital process of the frontal bone, and from the common medial canthal tendon. Its fibers pass around the orbital rim to form a continuous ellipse without interruption at the lateral palpebral commissure, and insert just below their points of origin.

The palpebral portion of the orbicularis muscle overlies the mobile eyelid from the orbital rims to the eyelid margins. The muscle fibers sweep circumferentially around each lid as a half ellipse, fixed medially and laterally at the canthal tendons. Although this portion forms a single anatomic unit in each eyelid, it is customarily further divided topographically into two parts, the preseptal and pretarsal orbicularis.

The preseptal portion of the muscle is positioned over the orbital septum in both upper and lower eyelids, and its fibers originate perpendicularly along the upper and lower borders of the medial canthal tendon. Fibers arc around the eyelids and insert along the lateral horizontal raphé. The pretarsal portion of the muscle overlies the tarsal plates. Its fibers originate from the medial canthal tendon via separate superficial and deep heads, arch around the lids and