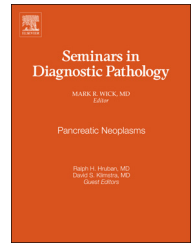


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Epithelial, non-melanocytic and melanocytic proliferations of the ocular surface



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ABSTRACT

Ocular surface tumors are commonly encountered by ophthalmologists and ophthalmic pathologists. These tumors have varied clinical manifestations. In this article, we discuss the most commonly encountered non-melanocytic and melanocytic ocular surface tumors, with emphasis on their common clinical features, morphologic patterns, and prognostic factors.

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The ocular surface includes the conjunctiva and cornea. Primary tumors at that site may be acquired or congenital and they include a variety of epithelial, stromal, hematolymphoid, and secondary lesions (Table 1). These lesions can cause significant morbidity and also have the potential to be life-threatening. In this brief review, we consider the epidemiological, clinical, and histopathological features of the non-melanocytic and melanocytic tumors of the ocular surface.

Histology

Considering the functional, embryological and anatomical aspects of this region, the “ocular surface system” encompasses the surface and glandular corneal and conjunctival epithelium, accessory lacrimal glands, Meibomian glands, the apical and basal matrices, the lacrimal glands with the lacrimal drainage system, the eyelashes, and the glands of Zeis and Moll.

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Table 1 – Epithelial tumors of the ocular surface.

Non-melanocytic	
Benign	
	Squamous papilloma
	Keratoacanthoma
	Pseudoepitheliomatous hyperplasia
	Oncocytoma
	Dacryoadenoma
Pre-malignant	
	Conjunctival corneal intraepithelial neoplasia (CCIN)
Malignant	
	Squamous cell carcinoma
Melanocytic	
Benign	
	Complexion-associated melanosis (racial melanosis)
	Secondary melanosis
	Ocular melanocytosis
Nevi	
	Primary acquired melanosis
Pre-malignant	
	Melanoma in situ
Malignant	
	Malignant melanoma

Conjunctiva

The conjunctiva functions to produce the mucous component of the tear film and helps to attach the globe to the eyelids. Anatomically, the conjunctiva is divided into 3 zones—the palpebral conjunctiva (lining the inner surface of the eyelids), the forniceal conjunctiva (lining the superior and inferior fornices) and the bulbar conjunctiva (covering the sclera). Histologically, it consists of a non-keratinizing, stratified squamous or columnar goblet cell-containing epithelium, separated from the underlying substantia propria by a basement membrane (Fig. 1). The epithelium is 2–3 layers thick with the basal layer containing a variable number of melanocytes. In addition to a rich vascular and lymphatic network, the substantia propria contains conjunctiva-associated lymphoid tissue (CALT), constituting a mucosal immune system.

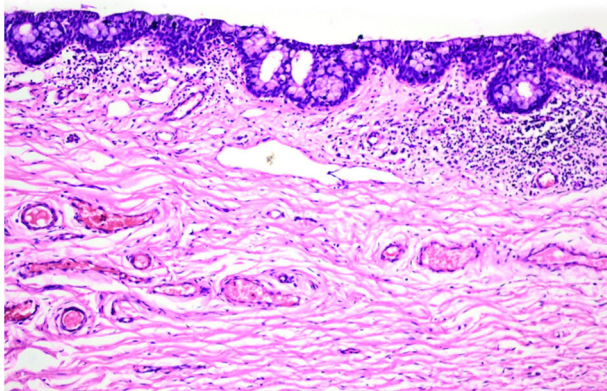


Fig. 1 – Conjunctiva with a non-keratinizing, stratified squamous or columnar goblet cell-containing epithelium and underlying substantia propria.

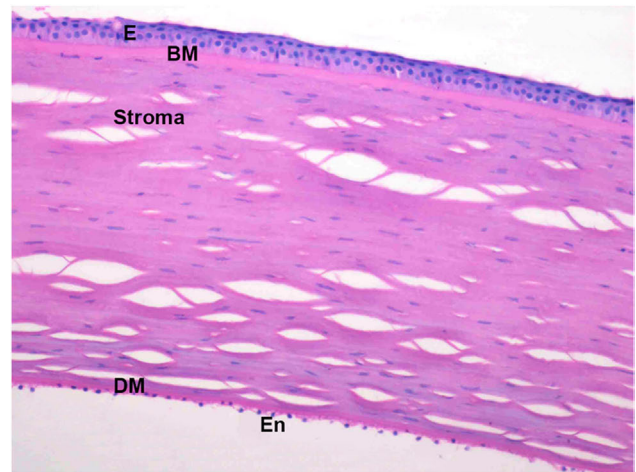


Fig. 2 – Layers of the cornea—(top to bottom) epithelium (E), Bowman's membrane (BM), stroma, Decemet's membrane (DM), and endothelium (En).

Cornea

The cornea is a transparent and avascular tissue that acts as an optical medium for light to enter the eye (Fig. 2). The stratified squamous, non-keratinized corneal epithelium is 3–6 layers thick. Posterior to it is the Bowman's membrane (anterior limiting membrane), an acellular band of collagen. The corneal stroma accounts for 90% of its thickness and comprises a lattice-like arrangement of collagen fibrils with sparsely distributed keratocytes. Posterior to the stroma is Descemet's membrane, a modified basement membrane of the corneal endothelium and the posterior-most corneal layer.

The corneal endothelium is a squamous or cuboidal monolayer responsible for exchange of fluid between the aqueous humor and the corneal stroma. Unlike the corneal epithelium, the corneal endothelium lacks a capacity for regeneration. Recently, Dua et al.¹ have identified a novel, acellular pre-Descemet's layer (Dua's layer); it plays a role in formation of such posterior corneal lesions as acute hydrops, descematomyces, and pre-descemet's dystrophies.

Epithelial tumors

Epithelial ocular surface tumors may be melanocytic or non-melanocytic (Table 1).

Non-melanocytic epithelial tumors

Squamous papilloma

Squamous papillomas are common benign tumors that are seen in both children and adults. They present most commonly in the inferior fornix as solitary or multiple, sessile or pedunculated, pink to red fleshy lesions with finger-like projections (Fig. 3).² Testing with the polymerase chain reaction (PCR) has shown an association with human papilloma virus (HPV) in 44–92% of these lesions, most commonly

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