Overview of Eyelid Tumors

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Overview and Epidemiology

Eyelid is a common place for skin cancer to occur and constitute 5–10% of all skin cancers. Eyelid neoplasms comprise a variety of benign and malignant growths (Table 1.1). Significant majority of these growths are benign in nature and constitute 82–98% of all neoplasms (Table 1.2). There is wide, racial, and probable geographical variation reported in the incidence of the various eyelid tumors. Eyelid malignancies vary in distribution and presentation. The most common malignant eyelid tumor in western literature is basal cell carcinoma (BCC) comprising 86–91% incidence among the Caucasians [7, 12]. However, in one of the largest series from China and India this incidence is much lower, consequently sebaceous gland carcinoma (SGC) constitutes 32% of all eyelid tumors [6, 13]. In studies from Asian countries [2, 14, 15] it is the sebaceous gland carcinoma which constitutes the majority (67–77%). The mean age for benign tumor is lower than that of malignant tumors. Epithelial tumor and dermoid cysts are the most common eyelid tumor in children [16]. Malignant eyelid tumor in children is extremely rare. When it presents, is usually a part of a systemic process, genetic defects or following radiation treatment [17, 18]. Merkel cell carcinomas (MCC) of the eyelid are rare neuroendocrine tumor constituting 5–20% of the head and neck tumor, predominantly in Caucasians [19].

Classification of Eyelid Tumors

Eyelid tumors can arise from various histological layers eyelid is composed of. Eyelid tumors are classified as benign or malignant or according to the tissue or cell of origin (Tables 1.2, 1.3 and 1.4). They can be subdivided into non-melanocytic and melanocytic tumors. Benign epithelial proliferations such as squamous papilloma, pseudoepitheliomatous hyperplasia, seborrheic keratosis, keratoacanthoma cysts and nevi are common. Among the malignant, BCC (Figs. 1.1 and 1.2) is the most common in Caucasians and SGC among the Asians (Fig. 1.3), followed by squamous cell carcinoma (SCC) and malignant melanoma (MM) (Figs. 1.4, 1.5, 1.6, 1.7 and 1.8). The large majority of BCC (93%) was seen in 71% of females [2] SGC has predilection for the upper lid [20]. Merkel cell cancer has higher prevalence in men. Primary malignant melanomas of the eyelid skin are rare and account for 0.2–13% of all reported

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Table 1.1 Regional incidence of benign and malignant eyelid neoplasms in the epidemiological studies

	Author, country, year	Study period	Total number of patients	Biopsy proven tumors	Benign (%)	Mean age years (gender preponderance)	Premalignant %	Malignant (%)	Mean age years (gender preponderance)
	Asian studies								
1.	Huang, Taiwan, 2015 [1]	1995–2015	4521	4521	4294 (95.0)	55.4		227 (5)	72.5
2.	Chang CH, Taiwan, 2003 [2]	1994–1998	144	129	126 (87.5)	-(f)	nil	12.5%	61 (f)
3.	Toshida H, Japan, 2012 [3]	1993–2007	118	118	106 (89.8)	47.8 (f)	ns	12 (10.2%)	53.1 (f)
4.	Sihota, India 1996 [4]	1982–1992	313	313	135 (43.1)	ns	ns	178 (56.8)	ns
5.	Rathod A, India, 2015 [5]	2007–2009	100	100	61 (61)	37.02	1	39 (39)	58.59
6.	Ni Z, China, 1996 [6]	1953–1992	3510	3510	2413 (68.7)	na		1097 (31.2)	na
	Western literature								
7.	Deprez, Switzerland, 2009 [7]	1989–2007	4981	4981	4087 (82)	–		894 (18)	–
8.	Paul S, USA, 2011 [8]	2004–2007	855	855	649 (75.9)	<60 (nil)	18	206 (24.1)	>60 (m)
9.	Mclean, AFIP, USA, 1994 [9]	1984–1989	846	456	456 (54)	–	–	390 (46)	
10.	Font, USA, 2006 [10]	1980–1982	1474	NOS	880 (60)		–	594 (40)	–
	Middle East								
11.	Bagheri, Tehran, Iran, 2013 [11]	2000–2010	182	182	82	46.4 (f)		100	63.9 (m)
12.	Gundogan FC, Turkey, 2015 [12]	2008–2012	1502 (1541)	1541	1424 (92.4)	50.08 (f)	6%	22 (1.5%)	68.6 (m)

Table 1.2 Eyelid tumors originating from epidermis

Subtypes	Benign	Premalignant	Malignant
Non-melanocytic	Squamous cell papilloma	Actinic(solar) keratosis	Basal cell carcinoma
	Seborrheic keratosis	Intraepithelial neoplasia	Squamous cell carcinoma
	Inverted follicular keratosis	Sebaceous nevus (of Jadassohn)	Mucoepidermoid carcinoma
	Reactive hyperplasia (pseudoepitheliomatous hyperplasia)	Xeroderma pigmentosa	Keratoacanthoma
		Keratoacanthoma	
Melanocytic	Ephelis or freckles	Congenital dysplastic nevus	
	Lentigo simplex	Lentigo maligna (melanotic freckle of Hutchinson)	Melanoma arising from nevi
	Solar Lentigo		Melanoma arising in lentigo maligna
	Junctional nevus		Melanoma arising de novo
	Intradermal nevus		
	Compound nevus		
	Spitz nevus		
	Balloon cell nevus		
	Blue nevus		
	Cellular blue nevus		
	Oculodermal nevus of Ota		
Eyelid tumors arising from adnexal and cystic lesions			
Sebaceous gland tumors	Sebaceous gland hyperplasia	–	Sebaceous gland carcinoma
	Sebaceous gland adenoma		
Sweat gland and lacrimal gland tumors	Syringoma		Sweat gland (eccrine) adenocarcinoma
	Papillary syringadenoma		Mucinous sweat gland adenocarcinoma
	Eccrine spiradenoma		Apocrine gland adenocarcinoma
	Eccrine acrospiroma		Adenoid cystic carcinoma
			Porocarcinoma
Eyelid tumors arising from hair follicle			
	Trichoepithelioma		Carcinoma of hair follicles
	Trichofolliculoma/trichoadenoma		
	Trichilemmoma		
	Pilomatrixoma (calcifying epithelioma of Malherbe)		
Other cystic lesions			
	Epidermal inclusion cyst		
	Sebaceous cyst		
	Retention cyst		
	Eccrine hidrocystoma		
	Apocrine hidrocystoma		
	Trichilemmal cyst		
	Other benign cystic lesion		

Table 1.3 Fibrous, fibrohistiocystic, and muscular eyelid tumors

	Benign	Intermediate	Malignant
Fibrous	Fibroma		Fibrosarcoma
	Keloid		Congenital fibrosarcoma
	Nodular fasciitis		
	Proliferative fasciitis		
	Fibromatosis		
Fibrous histiocytic	Xanthelasma		Malignant fibrous histiocytoma
	Xanthoma	Atypical fibroxanthoma	Malignant giant cell fibrous histiocytoma
	Dermatofibroma	Dermatofibrosarcoma protuberans	Malignant fibroxanthoma
	Xanthogranuloma	Angiomatoid fibrous histiocytoma	
	Fibrous histiocytoma		
	Juvenile xanthogranuloma		
	Necrotic xanthogranuloma		
	Reticulohistiocytoma		

	Benign	Malignant
Smooth muscle	Leiomyoma	Leiomyosarcoma
	Angiomyoma	
Skeletal muscle	Rhabdomyoma	Rhabdomyosarcoma

Table 1.4 Eyelid tumors arising from vascular, perivascular, neural, lipomatous, cartilage, bone lymphoid tumors, hamartomas, and choristomas

	Benign	Malignant
Vascular	Nevus flammeus (port wine stain)	Angiosarcoma
	Papillary endothelial hyperplasia	Lymphangiosarcoma
	Capillary hemangioma	Kaposi's sarcoma
	Cavernous hemangioma	
	Venous hemangioma	
	Epithelioid hemangioma (angiolymphoid hyperplasia)	
	Arteriovenous malformation	
	Lymphangioma	
Perivascular	Hemangiopericytoma	Malignant hemangiopericytoma
	Glomus tumor	Malignant glomus tumor
Neural	Traumatic neuroma	Malignant peripheral nerve sheath tumor
	Neurofibroma	Merkel cell tumor
	Plexiform neurofibroma	
	Schwannoma (neurilemoma)	
	Neuroglial choristoma	
Lipomatous	Lipoma	Liposarcoma
	Hibernoma	
Cartilage	Chondroma	Chondrosarcoma
	Osteoma	Mesenchymal chondrosarcoma
		Osteosarcoma
Lymphoid	Benign lymphoid hyperplasia	Lymphoma
	Plasmacytoma	Leukemic infiltration
Hamartomas and choristomas	Dermoid cyst	
	Phakomatous choristoma	
	Ectopic lacrimal gland	
Others	Myxoma	

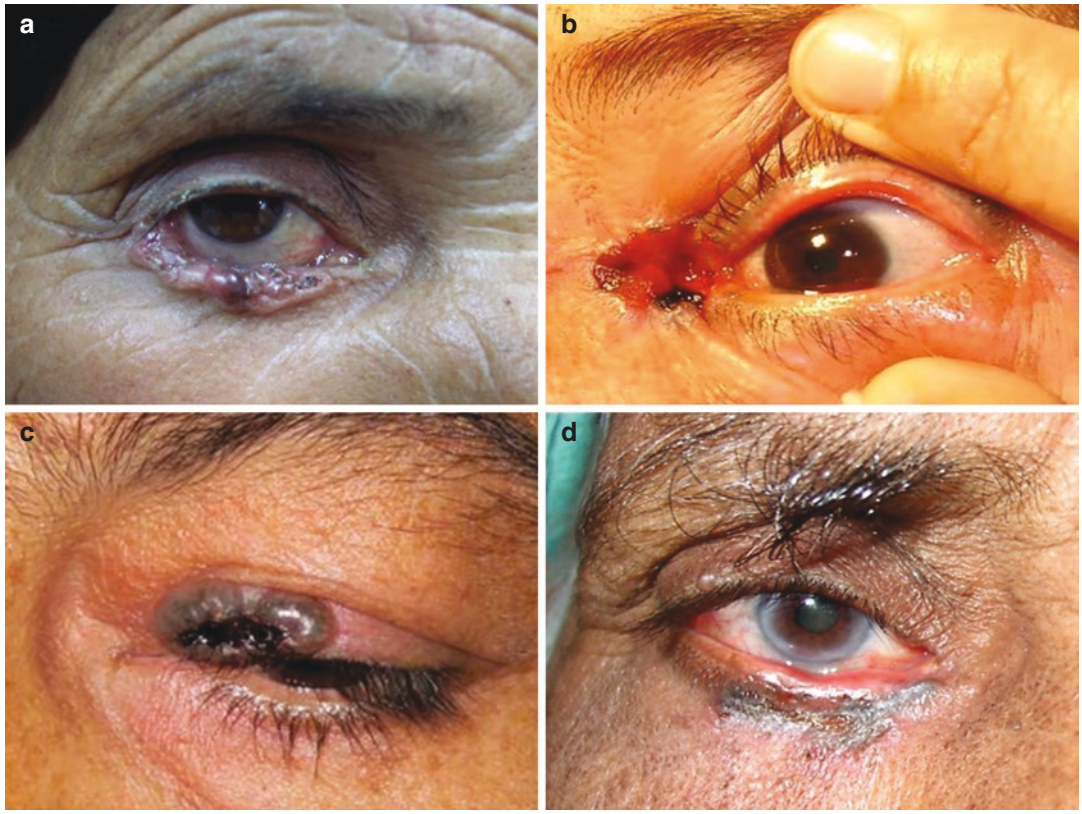

Fig. 1.1 (**a**) Basal cell carcinoma involving the lower lid. (**b**) Ulcerative basal cell carcinoma involving the medial canthus. (**c**) Basal cell carcinoma involving the upper lid with central necrotic area. (**d**) Morpheaform type of basal cell carcinoma involving the lower lid

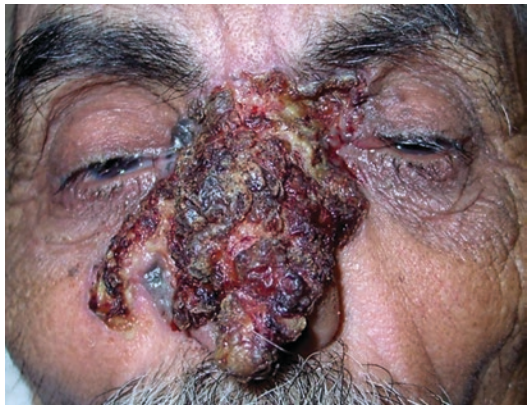

Fig. 1.2 Extensive basal cell carcinoma involving both medial canthi, nose and cheek

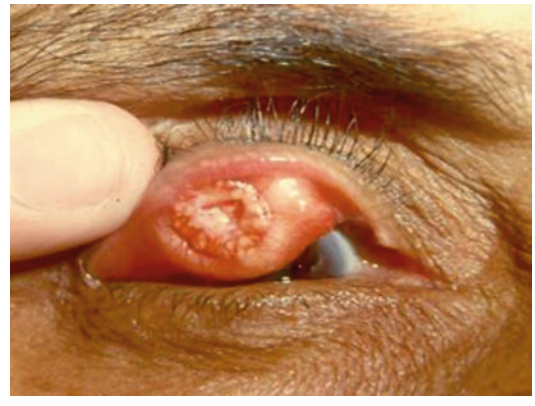

Fig. 1.3 Sebaceous gland carcinoma misdiagnosed as chalazion and surgically intervened

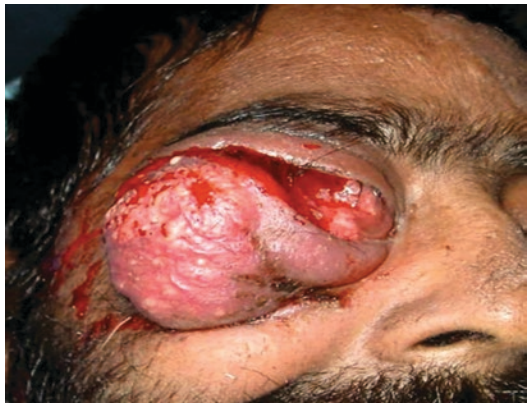

Fig. 1.4 Malignant melanoma involving lower lid and conjunctiva

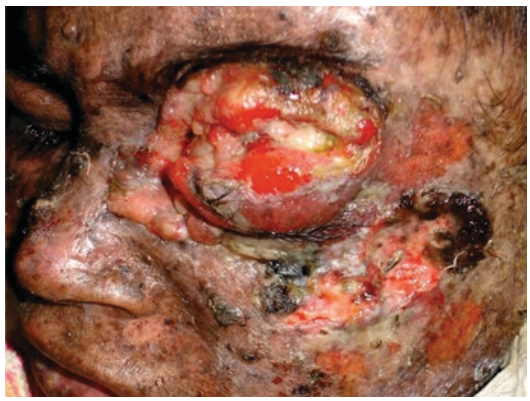

Fig. 1.5 Extensive malignant melanoma involving both the eyelids in a patient with xeroderma pigmentosa

cases [2, 7]. They occur 20 years later than other non-melanoma tumor and have 2.6 times predilection for the lower lid. Eyelids can also be involved by secondary and metastatic lesions.

All primary carcinomas of the eyelid can be classified based on their clinical and histological presentation using the TNM [tumor, nodes (lymph), metastasis] by AJCC (8th Ed) classification system [21]. TNM staging describes the size of tumor, number and location of regional lymph nodes which have malignant cells in them and whether the malignant cells have spread or metastasized to another part of the body. The TNM classification of eyelid carcinomas reflects both morbidity and mortality risks in order to provide useful guidelines for patient management.

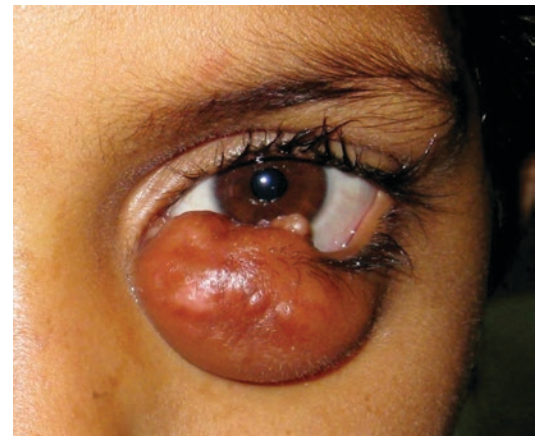

Fig. 1.6 Rapidly growing squamous cell carcinoma of the eyelid and extending to the orbit

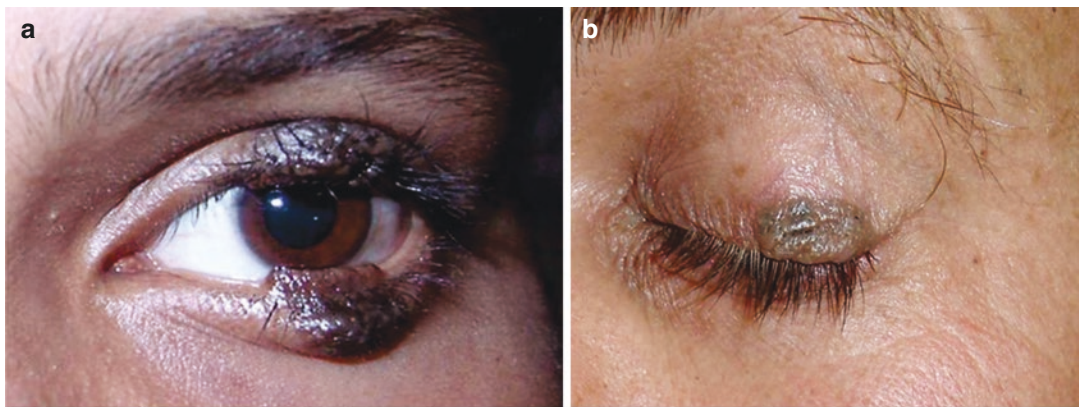

Fig. 1.7 (**a**) Kissing nevus in a young adolescent girl. (**b**) Nevi involving the upper lid in a young adult with a history of recent growth. (**c**) Nevus involving the lid margin in a young adult. (**d**) Keratoacanthoma

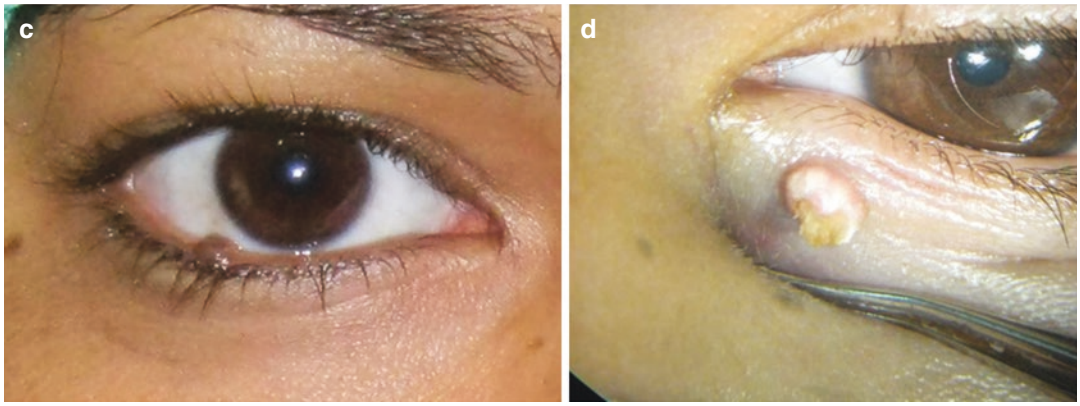

Fig. 1.7 (continued)

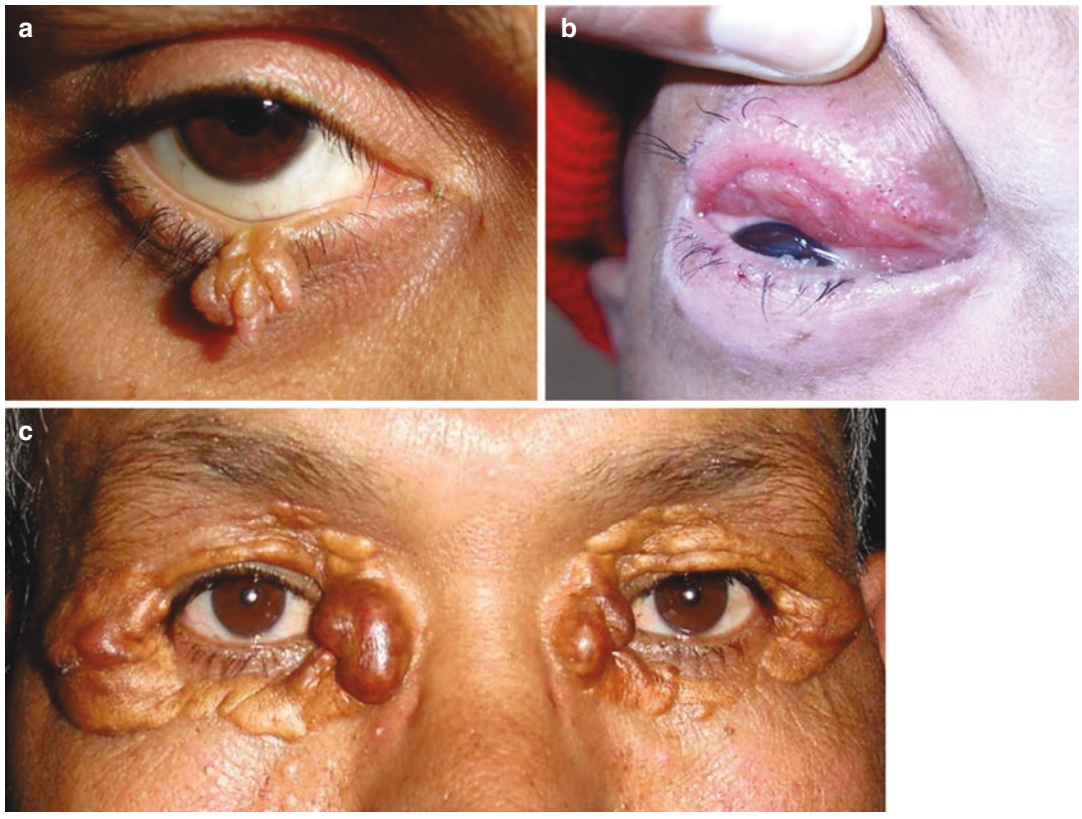

Fig. 1.8 (**a**) Squamous papilloma of the lower eyelid. (**b**) Lymphangioma diffusely involving the lids and orbit. (**c**) Extensive Xanthelesma involving all four lids

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