Clinical presentation, causes and management of ocular tumors by Azuamah, Y. C. is licensed under a Creative Commons Attribution-NonCommercial-NoDerivatives 4.0 International License.
ABSTRACT
Ocular tumors describe abnormal growth of cells of tissues of the eye that can be pigmented or non-pigmented. Non-pigmented ocular tumors include conjunctival papilloma, conjunctival intraepithelial neoplasia, invasive squamous cell carcinoma, lymphoid tumors and Kaposi’s sarcoma. Pigmented tumors include malignant melanomas of the conjunctival, eyelid, iris and choroid, epithelial melanosis, primary acquired melanosis and pyogenic granuloma. Benign lesions such as the conjunctival naevus does not require any excision while malignant ocular tumors can be treated with topical chemotherapy as an adjunct to surgical procedures like excision, cryotherapy, radiation therapy, argon laser photoablation, enucleation and exenteration. While some causes remain unknown, common risk factors include long-term exposure to ultraviolet radiation, exposure to toxic chemicals and radiation, human papilloma virus, AIDS and inherited genes. Adequate funding and research is advocated toward the proper understanding of the causes of ocular tumors and the development of medical diagnostic centers for excision biopsy and proper treatment of the tumors.

INTRODUCTION
TUMORS
A tumor is a term that describes any swelling but which is generally used to refer to an abnormal mass of tissue that forms when cells in a specific area reproduce at an increased rate (The British Medical Association, 2002). The term is also equated with neoplasms. In modern medicine, the term tumor means a neoplasm that has formed a lump. Some neoplasms do not form a lump (Willis, 2002). Prior to abnormal growth, cells often undergo an abnormal pattern of growth, such as metaplasia and dysplasia. However, metaplasia and dysplasia do not always progress to neoplasia. Neoplasms may be benign, premalignant or malignant. Cancer is the common term for all malignant tumors (Hanahan and Weinberg, 2000).

All tumors, benign or malignant have two basic components: proliferating neoplastic cells that constitute their parenchyma and a supportive stroma made up of connective tissue and blood vessels (Kumar, et al., 2008). Although parenchymal cells determine the
behavior and pathologic consequence of neoplasms, the growth and evolution of neoplasms are critically dependent on their stroma. An adequate blood supply is requisite, and the stromal connective tissue provides the framework for the parenchyma. In some tumors, the stromal support is scant and so the neoplasm is soft and fleshy. Some tumors, for example, some cancers of the female breast are hard or scirrhou (Kumar, et al., 2008).

**Benign Tumors**

In general, benign tumors are designated by attaching the suffix –oma to the cell of origin. Tumors of mesenchymal cells generally follow this rule. For example, a benign tumor arising from fibroblastic cells is called a fibroma, a cartilaginous tumor is a chondroma, and a tumor of the osteoblasts is an osteoma. Adenoma is the term applied to a benign epithelial neoplasm that forms glandular patterns as well as to tumors derived from glands but not necessarily producing glandular patterns. Benign epithelial neoplasms producing microscopically or macroscopically visible finger-like projections from epithelial surfaces are referred to as papillomas. Those that form large cystic masses, as in the ovary, are referred to as cystadenomas (Coussens and Werb, 2002). Some tumors produce papillary patterns that protrude into the cystic spaces and are called papillary cystadenomas. When a neoplasm, benign or malignant, produces a macroscopically visible projection above a mucosal surface and projects, for example, into the gastric or colonic lumen, it is termed a polyp. The term polyp is preferably restricted to benign tumors. Malignant polyps are better designated polypoid cancers (Kumar, et al., 2008).

**Malignant Tumors**

The nomenclature of malignant tumors essentially follows the same schema used for benign neoplasms, with certain additions. Malignant tumors rising from mesenchymal tissue are usually called sarcomas. Malignant neoplasms of epithelial cell origin, derived from the three germ layers are called carcinomas. Thus, cancer arising in the epidermis of ectodermal origin is a carcinoma, as is a cancer arising in the mesodermally derived cells of the renal tubules and the endodermally derived cells of the linings of the
gastrointestinal tract. Carcinomas may be further qualified. One with a glandular growth pattern microscopically is termed an adenocarcinoma, and one producing recognizable squamous cells arising in any epithelium of the body is termed squamous cell carcinoma. It is common practice to specify, when possible, the organ of origin (e.g. a renal cell adenocarcinoma). Not infrequently, however, a cancer is composed of undifferentiated cells of unknown tissue origin, and must be designated merely as a poorly differentiated or undifferentiated malignant tumor (Kumar, et al., 2008). In benign and in differentiated malignant neoplasms, the parenchymal cells bear a close resemblance to each other, as though all were derived from a single cell, as is the case with cancers. Infrequently, divergent differentiation of a single line of parenchymal cells into another tissue creates what are called mixed tumors. The best example of this is the mixed tumor of salivary gland origin. These tumors contain epithelial components scattered within a myxoidstroma that sometimes contains islands of apparent cartilage or even bone. All these elements arise from epithelial and myoepithelial cells of salivary gland origin; thus the preferred designation of these neoplasms is pleomorphic adenoma. The great majority of neoplasms, even mixed tumors are composed of cells representative of a single germ layer. Teratomas, in contrast, are made up of a variety of parenchymal cell types representative of more than one germ layer, usually three (Houghton and Polsky, 2002).

**Pathogenesis of Tumor Growth**

The natural history of most malignant tumors can be divided into four phases (Kumar, et al., 2008):

i) Malignant change in the target cell, referred to as transformation

ii) Growth of the transformed cells

iii) Local invasion

iv) Distant metastases

In the great majority of instances, a benign tumor may be distinguished from a malignant tumor with considerable confidence based on morphology. Sometimes, however a neoplasm defies categorization. Ultimately, morphologic diagnosis cannot predict the biologic behavior or clinical course of a neoplasm with absolute certainty. Occasionally,
this prediction is confounded by a mixed discrepancy between the morphologic appearance of a tumor and its behavior.

**Differentiation and Anaplasia**

Differentiation refers to the extent to which neoplastic cells resemble comparable normal cells, both morphologically and functionally. Lack of differentiation is called anaplasia (Raghavan, 2005). Well differentiated tumors are composed of cells resembling the mature normal cells of the tissue of origin of the neoplasm. Poorly differentiated or undifferentiated tumors have primitive-appearing, unspecialized cells. In general, benign tumors are well differentiated. The neoplastic cell in a benign smooth muscle tissue, a leiomyoma, so closely resembles the normal cell that it may be impossible to recognize it as a tumor by microscopic examination of individual cells. Malignant neoplasms, in contrast, range from well differentiated to undifferentiated. Malignant neoplasms composed of undifferentiated cells are said to be anaplastic. Lack of differentiation or anaplasia, is considered a hallmark of malignant transformation. Anaplasia literally means “to form backward”, implying a reversion from a high level of differentiation to a lower level. Most cancers do not represent “reverse differentiation” of mature normal cells, but in fact arise from stem cells that are present in all specialized tissues (Coussens and Werb, 2002). The well differentiated cancer evolves from maturation or specialization of undifferentiated cells as they proliferate, whereas the undifferentiated malignant tumor derives from proliferation without complete maturation of the transformed cells. Lack of differentiation or anaplasia is marked by a number of morphological changes (Willis, 2002).

**Pleomorphism.** Both the cells and the nuclei characteristically display pleomorphism, a variation in size and shape. Cells may be found that are many times larger than their neighbors, and other cells may be extremely small and primitive appearing.

**Abnormal nuclear morphology.** Characteristically, the nuclei contain an abundance of DNA and are extremely dark staining (hyperchromatic). The nuclei are disproportionately large for the cell, and the nucleus-to-cytoplasm ratio may approach 1:1 instead of the
normal 1:4 or 1:6. The nuclear shape is very variable, and the chromatin is often coarsely clumped and distributed along the nuclear membrane (Kumar, et al., 2008).

**Mitosis.** As compared with benign tumors and some well differentiated malignant neoplasms, undifferentiated tumors usually possess large numbers of mitoses, reflecting the higher proliferative activity of the parenchymal cells. The presence of mitoses, however, does not necessarily indicate that a tumor is malignant or that the tissue is neoplastic. Many normal tissues exhibiting rapid turnover, such as bone marrow, have numerous mitoses, and non-neoplastic proliferations such as hyperplasias contain many cells in mitosis. More important as a morphologic feature of malignant neoplasia are atypical, bizarre mitotic figures, sometimes producing tripolar, quadripolar, or multipolar spindles (Houghton and Polsky, 2002).

**Loss of Polarity.** In addition to the cytologic abnormalities, the orientation of anaplastic cells is markedly disturbed (i.e. they lose normal polarity). Sheets or large masses of tumor cells grow in an anarchic, disorganized fashion.

**Other Changes.** Another feature of anaplasia is the formation of tumor giant cells, some possessing only a single huge polymorphic nucleus and others having two or more nucleus. In the cancer giant cell, the nuclei are hyperchromatic and large in relation to the cell. Although growing tumors obviously require a blood supply, often the vascular stroma is scant, and in many aplastic tumors, large central areas undergo ischemic necrosis (Raghavan, 2005).

**Rates of Growth**

The rate of growth of a tumor is determined by three main factors: the doubling time of tumor cells, the fraction of tumor cells that are in the replicating pool, and the rate at which cells are shed and lost in the growing lesion. The proportion of cells within the tumor population that are in the proliferative pool is referred to as the growth fraction. As tumors continue to grow, cells leave the proliferative pool in ever-increasing numbers owing to shedding, lack of nutrients or apoptosis (Kumar, et al., 2008; Willis, 2002). Ultimately the progressive growth of tumors and the rate at which they grow are
determined by an excess of cell production over cell loss. In some tumors, especially those with a relatively high growth fraction, the imbalance is large resulting in more rapid growth than in those in which cell production exceeds cell loss by only a small margin. Some leukemias and lymphomas and certain lung cancers have a relatively high growth fraction, and their clinical course is rapid. By comparison, many common tumors such as cancers of the colon and breast have low growth fractions, and cell production exceeds cell loss by only 10%; they tend to grow at a much slower pace (Hanahan and Weinberg, 2000).

Fast-growing tumors may have a high cell turnover, implying that the rates of both proliferation and apoptosis are high. Obviously, for the tumor to grow, the rate of proliferation should exceed that of apoptosis. An original transformed cell (approximately 10µm in diameter) must undergo at least 30 population doublings to produce 10⁹ cells (weighing approximately 1 gram), which is the smallest clinically detectable mass. If every one of the daughter cells remained in the cell cycle and no cells were shed or lost, it would take about 90 days for one transformed cell to produce a clinically detectable tumor containing 10⁹ cells. In reality, the latent period before which a tumor becomes clinically detectable is unpredictable but typically much longer than 90 days; up to many years for most solid tumors, emphasizing that human cancers are diagnosed only after they are fairly advanced in their life cycle (Kumar, et al., 2008). The total cell-cycle time for most tumors is equal to or longer than that of corresponding normal cells. In general, the growth of tumors correlates with their level of differentiation, and thus most malignant tumors grow more rapidly than benign tumors.

**Local Invasion**

Nearly all benign tumors grow as cohesive expansile masses that remain localized to their site of origin and do not have the capacity to infiltrate, invade or metastasize to distant sites as do malignant tumors. Because they grow and expand slowly, they usually develop a rim of compressed connective tissue, sometimes called a fibrous capsule, which separates them from the host tissue. The growth of cancers is accompanied by progressive infiltration, invasion, and destruction of the surrounding tissue. In general,
malignant tumors are poorly demarcated from the surrounding normal tissue, and a well-defined cleavage plane is lacking. Most malignant tumors are obviously invasive and can be expected to penetrate the wall of the colon or uterus, for example, or fungate through the surface of the skin. Such invasiveness makes their surgical resection difficult. Next to the development of metastases, invasiveness is the most reliable feature that differentiates malignant from benign tumors (Coussens and Werb, 2002).

Metastasis

Metastases are tumor implants discontinuous with the primary tumor. Metastasis unequivocally marks a tumor as malignant because benign neoplasms do not metastasize. The invasiveness of cancers permits them to penetrate the blood vessels, lymphatics, and body cavities, providing the opportunity for spread. With few exceptions, all cancers can metastasize. The major exceptions are most malignant neoplasms of the glial cells in the central nervous system, called gliomas, and basal cell carcinomas of the skin. Both are locally invasive form of neoplasia but they rarely metastasize. In general, the more aggressive, the more rapidly growing; and the larger the primary neoplasm, the greater the likelihood that it will metastasize or already has metastasized (Kumar, et al., 2008).

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Benign</th>
<th>Malignant</th>
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<tbody>
<tr>
<td>Differentiation/</td>
<td>Well differentiated; structure may be typical of tissue of origin</td>
<td>Some lack of differentiation with aplasia; structure is often atypical</td>
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<tr>
<td>anaplasia</td>
<td></td>
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<tr>
<td>Rate of growth</td>
<td>Usually progressive and slow; may come to a standstill and regress; mitotic figures are rare and normal</td>
<td>Erratic and may be slow to rapid; mitotic figures may be numerous and abnormal</td>
</tr>
<tr>
<td>Local invasion</td>
<td>Usually cohesive and expansile well-demarcated masses that do not invade or infiltrate surrounding normal tissues</td>
<td>Locally invasive, infiltrating the surrounding normal tissues; sometimes may be seemingly cohesive and expansile</td>
</tr>
<tr>
<td>Metastasis</td>
<td>Absent</td>
<td>Frequently present; the larger and more undifferentiated the primary, the more likely are metastases</td>
</tr>
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OCULAR TUMORS

Eyelid Tumors

Benign eyelid lesions

**Hordeolum:** An external hordeolum (stye) is a small abscess caused by acute staphylococcal infection of a lash follicle and its associated gland of zeis or moll. The gland of zeis are modified sebaceous glands associated with the lash follicle. Gland of moll are modified sweat glands whose ducts open into the lash follicle. Examination shows a tender inflamed swelling of the lid margin which points anteriorly through the skin. It resolves spontaneously or discharges anteriorly. Resolution is enhanced by warm compress. Use of antibiotics is also helpful (Khurana, 2005).

![Figure 1: External hordeolum](image)

Internal hordeolum is a small abscess caused by acute staphylococcal infection of the meibomian glands. Examination shows a tender inflamed swelling within the tarsal plate which is usually more painful than a stye. The lesion discharges either anteriorly through the skin or posteriorly through the conjunctiva.

![Figure 2: External hordeolum](image)
**Chalazion (Meibomian cyst):** A chronic inflammatory lesion caused by blockage of meibomian gland orifices and stagnation of sebaceous secretions. The meibomian glands are sebaceous glands located in the tarsal plates that secrete outer lipid layer of the precorneal tear film. Examination shows a painless, roundish, firm lesion in the tarsal plate. Treatment is by surgical incision and use of systemic antibiotics (Kanski, 1998).

![Figure 3: Chalazion](image)

**Molluscumcontagiosum:** An infection caused by one of the pox viruses. Examination shows a pale, waxy nodule. Treatment include shave excision, cauterization and cryotherapy (Khurana, 2005).

![Figure 4: Molluscumcontagiosum](image)

**Strawberry naevus:** Presents within the first 6 months of birth. Examination shows a raised red lesion. The tumor usually grows until the age of 12 months and then starts to involute spontaneously. Complete resolution occurs in 75% of patients by the age of 3 years (Kanski, 1998).
Figure 5: Strawberry naevus

**Cyst of Moll:** Small, round, translucent fluid-filled lesion on the anterior lid margin.

Figure 6: Cyst of Moll

**Cyst of Zeis:** Contains oily secretions and less translucent.

Figure 7: Cyst of Zeis

**Malignant eyelid tumors**

**Kaposi’s sarcoma:** A vascular tumor tumor which is frequently associated with AIDS. Examination shows a pink, red-violet to brown lesion. Treatment is with low dose radiotherapy (Spalton, et al., 2006).
**Eye lid melanoma:** Malignant melanoma of the eyelid is rare but potentially lethal.

**CONJUNCTIVAL TUMORS**

**Conjunctival papilloma**

Papillomas of the conjunctiva can be pedunculated or sessile. Pedunculated papillomas most commonly affect children and young adults and are thought to be caused by an infection with the papillomavirus (Kanski, 1998; Kanski and Bowling, 2011). They may be bilateral and multiple, and associated with verruca vulgaris of the eyelid or elsewhere on the skin. They are usually located on the palpebral conjunctiva, fornix or caruncle.
Figure 10: Conjunctival papilloma - (a) Sessile (b) Pedunculated (c) Juxtalimbal and forniscal (d) Confluent

Treatment of small lesions may be unnecessary because of high rate of spontaneous resolution over a period of 2 to 3 years. Sessile papillomas usually affect older adults and are not infectious. They are invariably single and unilateral, and located either on the bulbar conjunctiva or at the limbus.

**Conjunctival intraepithelial neoplasia**

Conjunctival intraepithelial neoplasia is a term now used for Bowen’s disease, carcinoma in situ, conjunctival dysplasia and intraepithelial epithelioma (Khurana, 2005). The tumor usually begins near the limbus and spreads to involve the fornices and cornea. Examination shows a slightly elevated fleshy mass with tufted blood vessels, although occasionally it may appear as a gelatinous avascular tumor. As the tumor is superficial to the basement membrane, the conjunctiva moves freely over the underlying epithelial tissue.
Invasive squamous cell carcinoma

Invasive carcinoma is characterized by deep invasion of the stroma with fixation to underlying tissues. If untreated, the tumor may penetrate to reach the inside of the eye where it may grow rapidly. Examination shows a white, roughened, raised lesion at the limbus in the interpalpebral fissure. Occasionally, a papillary or sessile growth pattern is seen. As a result of its rarity, the tumor may be misdiagnosed as chronic conjunctivitis or atypical pterygium (Kanski, 1998).

Kaposi’s sarcoma

Kaposi’s sarcoma is a vascular tumor which eventually affects 25% of patients with AIDS. Examination shows a bright red mass, most often in the lower fornix. A very early
lesion may be mistaken for a chronic subconjunctival hemorrhage, a foreign body granuloma or a cavernous hemangioma.

Figure 13: Kaposi’s sarcoma

**Choristoma**

Choristomas are congenital overgrowths of normal tissue in abnormal locations. They are the most common type of epibulbar tumor in children. The two main types are dermoids and lipodermoids.

**Dermoids** consists of collagenous connective tissue covered by epidermoid epithelium. They appear as solid white masses most frequently located at the limbus. They are smooth, soft, yellowish, subconjunctival masses most frequently located at the inferotemporallimbus and showing protruding hair. Occasionally the lesions are very large and may virtually encircle the limbus (complex dermoid).

Figure 14: Simple limbaldermoid
**Lipodermoids**(*Dermolipoma*) consists of adipose tissue with surrounding dermis-like connective tissue. They appear as soft, yellow, movable soft subconjunctival masses located most commonly at the limbus or outer canthus. The surface is usually keratinized and may have hairs, reflecting its origin from ectopic skin. Occasionally the lesion may extend into the orbit or anteriorly toward the limbus. Treatment should be avoided because surgery may be complicated by scarring, ptosis, dry eye and ocular motility problems. However, if they are particularly unsightly, then debulking the anterior may improve cosmesis without compromising ocular motility.

![Figure 15: Dermolipoma](image)

**Lymphoid tumors**

The conjunctiva may be involved in a great variety of benign and malignant lymphoid lesions. Hodgkin’s disease, however, only rarely involves the conjunctiva and when it does there is invariably associated widespread systemic disease. Examination shows a salmon-colored subconjunctival infiltrate which must be differentiated by biopsy from a malignant lymphoma(Jereb, et al., 2004).

**Lymphoproliferative tumors**

Most conjunctivally lymphoproliferative lesions are reactively lymphoid hyperplasia, a proliferation of B and T cells with germinal follicle formation. Conjunctival lymphoma may arise in three clinical settings (a) de novo (b) extension from orbital lymphoma and
(c) occasionally associated with systemic involvement (Catania, 1998). Sometimes reactive lymphoid hyperplasia undergoes transformation to lymphoma. Most conjunctival lymphomas are B cell lymphomas and arise from mucosa-associated lymphoid tissue (MALT). It presents in late adult life with irritation or painless swelling which may be bilateral. The signs are a slowly-growing, mobile, salmon-pink or flesh-colored infiltrate on the epibulbar surface or in the fornices; rarely a diffuse lesion may mimic chronic conjunctivitis (Spalton, et al., 2006).

Figure 16: Conjunctival lymphoma

**Conjunctivalnaevus**

The conjunctivalnaevus is the most common melanocytic conjunctival tumor. The overall risk of malignant transformation is 1% (Kanski and Bowling, 2011). It usually presents during puberty or early adult life. Its classification is similar to that of the skin, with only minor modifications. Most are compound or subepithelial. Examination shows a solitary, sharply demarcated, flat or slightly elevated lesion. The single most common site is the juxtalimbal area, followed by other epibulbar sites, the plica, caruncle and eyelid margin. Naevi may be focal other diffuse but they are never multifocal. Cystic spaces are frequently seen within the substance of a naevus and the amount of pigment is variable. Pigmented naevi virtually always contains some shade of brown, ranging from dark tan to deep chocolate. About 30% of naevi are almost non-pigmented, although careful examination may uncover a fine stippling of pigment (Agarwal, et al. 2002). At puberty, the amount of pigmentation and size of the lesion may increase. As naevi on the palpebral and fornical conjunctiva are extremely rare, any pigmented lesion at these sites should be suspected of being a melanoma precursor or a frank melanoma and should be
biopsied. As juxtalimbalnaevi do not extend unto the peripheral cornea, a pigmented lesion which straddles the cornea should be considered clinically a malignant melanoma. Occasionally, a non-pigmented naevus can become inflamed and vascularized, and may be mistaken for an angiomatous tumor.

Figure 17: Conjunctivalnaevus

Conjunctival epithelial melanosis

Conjunctival epithelial (racial) melanosis is frequently seen in blacks and other individuals with dark complexions. It develops during the first few years of life and becomes static by early adulthood (Kanski, 1998). The pigmentation is bilateral, although its distribution and intensity may be asymmetrical. Examination shows areas of flat, patchy, brownish pigmentation scattered throughout the conjunctiva. It is most prominent in the interpalpebral fissure and typically fades in intensity toward the fornices. The pigmentation may be particularly marked at the limbus and around the perforating branches of the anterior ciliary nerves as they enter the sclera (Agarwal, et al., 2002). Juxtalimbal pigmentation may extend on to the periphery of the cornea. With the slit lamp, the pigment is seen to be within the epithelium, and the conjunctiva moves over the sclera.
Figure 18: Conjunctival Epithelial Melanosis

**Oculodermal melanosis**

Oculodermal melanosis consists of hyperpigmentation of facial skin and mucous membranes in the distribution of the ophthalmic, maxillary, and occasionally, the mandibular division of the trigeminal nerve. There are three possible patterns (Khurana, 2005).

*Dermal melanosis* in which only the skin is involved, occurs in about one-third of cases.

*Ocular melanosis* in which only the episclera is involved, is rare. When seen on the slitlamp, subepithelial melanosis consists of multifocal slate-grey pigmentation which lies beneath the epithelium. Because of its deep location, the lesion does not move over the globe.

*Oculodermal melanosis* is the complete syndrome in which both the skin and eye are affected. It occurs in about two-thirds of patients. The term “congenital ocular melanocytosis” includes patients with either ocular involvement alone or both ocular and dermal involvement.

The melanocytosis frequently affects the ipsilateral uvea; rarely, it may also involve the orbital tissues and, in some cases, it may extend beyond the orbit to the meninges and brain. Other ocular findings include glaucoma associated with hyperpigmentation of the trabeculum, melanocytosis of the cornea and lens, uveitis, cataract and hyperpigmentation of the optic nerve head.
Primary acquired melanosis

Primary acquired melanosis (PAM) typically develops in middle-aged or elderly white patients. It is extremely rare in blacks and, in contrast to racial melanosis, it is almost always unilateral. It usually presents during the sixth or seventh decade of life (Spalton, et al., 2006). Examination shows unifocal or multifocal, flat, indistinct areas of brown (ranging from golden brown to chocolate brown) conjunctival pigmentation. As any part of the conjunctiva may be involved, it is necessary to examine both the inferior and superior fornices by everting the eyelids. The lesions can be moved freely over the globe. In contrast to conjunctivalnaevi, cystic spaces are absent. PAM may shrink or remain stable for long periods of time, lighten or darken focally or enlarge centrifugally (radial growth phase). Malignant transformation to melanoma should be suspected by the sudden appearance of one or more nodules in otherwise flat lesions.

Figure 19: Primary Acquired Melanosis

Malignant conjunctival melanoma

Conjunctival melanoma accounts for approximately 2% of all eye malignancies. It is therefore much less common than melanoma of the choroid. The three clinical and pathological forms are (Kanski and Bowling, 2011):

Melanoma with PAM which occurs where the tumor has evolved indirectly with a variably protracted course of PAM. The most dramatic evidence of malignancy in a
patient followed up for PAM is the sudden appearance of one or more nodules in otherwise flat lesions. The adjacent skin is also occasionally involved.

*Primary melanoma*, a melanoma without PAM.

*Melanoma derived from a pre-existing benign naevus* is very rare.

The primary melanoma presents during the early fifties. The tumor is extremely rare in blacks and during the first two decades of life. Examination shows a solitary, pigmented, or non-pigmented nodule. Melanoma arising from a pre-existing naevus appears as a black or grey vascularized nodule that may be fixed to the episclera. Non-pigmented tumors have a characteristic, smooth, vascularized, or fish-flesh appearance. A common site is the limbal area although the tumor may develop anywhere in the conjunctiva.

![Malignant conjunctival melanoma](image)

**Figure 20: Malignant conjunctival melanoma**

**Pyogenic granuloma**

A pyogenic granuloma is a fibrovascular proliferation in response to a tissue insult involving the conjunctiva such as surgery, trauma and less frequently, inflammation. Spontaneous lesions are rare. Histology shows granulation tissue, chronic inflammation and proliferation of small blood vessels, similar to a cutaneous pyogenic granuloma (Agarwal, et al., 2002). Presentation is a few weeks after surgery for chalazion, strabismus or enucleation. Examination shows a fast-growing pink, fleshy, vascularized conjunctival mass near the conjunctival wound that bleeds easily. Treatment with topical steroids is usually successful; resistant cases require excision.
Ocular surface squamous neoplasia (OSSN)

OSSN describes a spectrum of benign, pre-malignant and malignant unilateral slowly-progressive epithelial lesions of the conjunctiva and cornea. Risk factors include ultraviolet light exposure, human papilloma virus (type 16) infection, AIDS, xerodermapigmentosum and stem cell therapy (Khurana, 2005). It presents usually late in adult life with ocular irritation or a mass. Signs are variable and clinical differentiation between the three histological types is unreliable. Most tend to occur within the interpalpebral fissure mostly at the limbus, although they may involve any part of the conjunctiva or cornea (Catania, 1998). Examination shows a gelatinous mass with superficial vessels; a white leukoplakic plaque that covers the lesion; papillomatous lesion with corkscrew-like surface blood vessels; squamous cell carcinoma is a fleshy, pink, papillomatous mass with feeder vessels or occasionally it may exhibit diffuse growth and masquerade as chronic conjunctivitis. Corneal involvement may occur but intraocular extension is uncommon.
Figure 22: Squamous neoplasia

Miscellaneous conjunctival tumors (Spalton, et al., 2006)

Episcleral hemangioma or telangiectasia may be associated with Sturge-Weber syndrome.

Reactive pseudoepitheliomatous hyperplasia is a rapidly-growing, white, hyperkeratotic, juxtalimbal nodule which develops secondary to irritation.

Benign hereditary intraepithelial dyskeratosis is a rare bilateral juxtalimbal hyperplastic translucent lesion with dilated vessels.

Melanocytoma is a rare, congenital, black, slowly-growing lesion which cannot be moved freely over the globe.

Myxoma is a benign soft tissue tumor that can be mistaken for amelanotic naevus.

Leiomyoma is a noncancerous tumor that arises from the smooth muscle of the blood vessels or embryonic tissue present in the medial canthus.

Figure 23: (a) hemangioma (b) pseudoepitheliomatous hyperplasia (c) intraepithelial dyskeratosis (d) Melanocytoma
TUMORS OF THE UVEA

Iris naevus: Iris naevi are common benign tumors composed of small spindle and dendritic naevus cells. A naevus consists of a pigmented, flat or slightly elevated lesion in the superficial layers of the iris. When situated near the pupil, they may cause distortion, ectropionuveae or localized lens opacities (Agarwal, 2002).

Figure 24: Iris naevus

Iris melanoma: Slow growing and composed of spindle cells. It presents usually during the fifth and sixth decade of life. Examination shows a solitary pigmented or non-pigmented nodule, most often located in the lower half of the iris.

Figure 25: Iris melanoma

Iris cysts: They arise from the iris pigment epithelium or rarely, the stroma. Those from pigment epithelium are globular, dark brown structures which transilluminate. Those from the stoma are larger, solitary with clear anterior wall.
Iris cyst: Iris cysts can be divided into two categories: primary and secondary.

**Primary Iris cyst:** These cysts are usually small and benign. They may be caused by trauma, inflammation, or surgery.

**Secondary Iris cyst:** These cysts develop secondarily to trauma, inflammation, or tumor growth.

**Iris leiomyoma:** Iris leiomyomas are rare tumors that arise from smooth muscle in the iris. They are typically small and asymptomatic.

**Ciliary body melanoma:** Ciliary body melanomas are more common than iris melanomas. They cannot usually be visualized unless the pupil is widely dilated.

**Choroidal melanoma:** Choroidal melanomas are more common than ciliary body melanomas and usually affect the retina. They can present as a pigmented, oval-shaped mass with a brown color. However, they can also be mottled with dark-brown or black pigment or virtually amelanotic (Spalton, 2006). In some patients, choroidal melanomas do not cause any symptoms and are detected during a routine examination.

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**Figure 26:** Iris cyst – (a) Primary Iris cyst (b) Large secondary iris cyst

**Figure 27:** Iris leiomyoma

**Figure 28:** Extraocular extension of ciliary body melanoma
examination. Decreased visual acuity or defective visual field depends on size, location and the presence or absence of secondary exudative retinal detachment.

![Choroidal melanoma](image)

**Figure 29: Choroidal melanoma**

**Choroidalnaevus:** An asymptomatic, flat or minimally elevated, oval or circular, slate-grey lesion less than 5mm in diameter (Khurana, 2005). A typical naevus should be observed every 18-24 months and a suspicious naevus, every 6 months.

![Choroidalnaevus](image)

**Figure 30: Choroidalnaevus**

**Choroidalhaemangioma:** The diffuse choroidal thickening caused by this tumor gives the fundus a deep red color which can easily be missed unless a comparison is made with the fellow eye (Kanski, 1998).

**RETINAL TUMORS**

**Retinoblastoma:** Retinoblastoma is the most common primary, malignant, intraocular tumor of childhood and the second most common of all age groups. The average age at diagnosis is 18 months and vast majority is apparent before 3 years. Children with
bilateral tumors present earlier than those with unilateral involvement. A white pupillary reflex is the most common mode of presentation.

Figure 31: Retinoblastoma – (a) fundus background (b) White pupil

There is a third, very rare type of retinoblastoma called trilateral retinoblastoma. In trilateral retinoblastoma, a tumor also develops on the pineal gland in addition to both eyes. It is also called pinealoblastoma (Aitken, et al., 2013).

Figure 32: Trilateral retinoblastoma

**Retinal capillary hemangioma:** Retinal capillary hemangiomas are associated with systemic lesions in about 25% of cases (Kanski, 1998). Examination initially shows a tiny lesion, no larger than a microaneurysm, located within the capillary bed between an arteriole and a venule. With the passage of time, the tumor grows into a small red nodule, and then into a larger round orange-red tumor.
TUMORS OF THE ORBIT

Rhabdomyosarcoma: The most common primary malignant orbital tumor in children. It presents usually in the first decade of life with a rapidly progressive proptosis which may initially be mistaken for an inflammatory process. Examination shows a mass in the upper part of the orbit, although the tumor can involve any part of the orbit (Khurana, 2005). The diagnosis is confirmed by biopsy. Treatment is with high dose local radiotherapy followed by chemotherapy. Exenteration is reserved for the rare recurring or radiotherapy-resistant tumor. In cases unresponsive to treatment, death usually occurs within 18 months.

Figure 33: Rhabdomyosarcoma

Dermoid cyst: Dermoids are lined by keratinized stratified squamous epithelium, have a fibrous wall and contain dermal appendages such as sweat glands, sebaceous glands and hair follicles. It presents typically in infancy with an asymptomatic, firm, round, localized lesion in the upper temporal aspect of the orbit. There are no associated bony defects and the globe is neither displaced nor proptotic. In patients with a ruptured spinal dermoid cyst, fatty droplets can disseminate in the cerebrospinal fluid or in a dilated central canal of the spinal cord (Agarwal, 2002). Treatment is based on cosmetic indications.
Neuroblastoma: A malignant tumor that arises from primitive neuroblasts of the sympathetic chain, most commonly in the abdomen, followed by the thorax and pelvis. Eventual metastasis to the orbit occurs in about 40% of cases (Sofi, et al., 2012).

Neurofibromatosis: Also called Reckinghausen’s disease, neurofibromatosis is characterized by numerous neurofibromas (soft, fibrous swellings, varying significantly in size), which grow from nerves, and by café au lait spots (pale, coffee-colored patches) on the skin (Aitken, et al., 2013). The eyes are affected and blindness can result.
CAUSES OF OCULAR TUMORS

While the cause of most ocular tumors remains unknown, some reported causes and risk factors include:

- **Ultraviolet radiation:** Ultraviolet (UV) light is found in sunlight and is emitted by electric arcs and specialized lights such as mercury lamps and black lights. Ultraviolet radiation can be classified into UVA, UVB, and UVC with wavelengths of 400-315nm, 315-280nm and 280-100nm respectively. High intensities of UVB light are hazardous to the eyes and exposure can cause cataracts, pterygium, pinguecula and conjunctival tumor formations. UV light is absorbed by molecules known as chromophores, which are present in the eye cells and tumors. Chromophores absorb light energy from the various wavelengths at different rates. If too much UV light is absorbed, eye structures can be damaged (Finger, 2005).
- Blue, grey or green eyes
- Unusual brown spots on your eyes
- Lots of unusually shaped or large moles (atypical mole syndrome)
- A weakened immune system – people who have HIV/AIDS or who are taking medicines that suppress their immune system (Lital, et al., 2012)
• Inherited genes
• Stem cell therapy
• Human papilloma virus (type 16) infection
• Chemical injury
  
  (Ogun, et al., 2009)

Other risk factors (Mandal, 2012)

Race or ethnicity – Caucasians or whites are at a higher risk of melanomas than African Americans or Asian Americans. Whites are also at a higher risk of skin melanomas. Eye melanomas follow a similar pattern in terms of risk association.

Age – Primary intraocular melanoma commonly occurs after the age of 50. It is rare in children and those who are over 70 years of age.

Sex – Eye melanoma affects both men and women with same frequency.

Colors of the eyes or iris – People with light colored eyes have raised risk of intraocular melanoma. Among these, those with blue, grey or green or any light eyed individuals are at higher risk than brown eyed individuals. The actual reason is unknown but it is speculated that the risk assessment is similar to whites being more prone to skin and eye melanomas.

Occupational hazards – Workers like farmers, fishermen, welders, or chemical and laundry workers have a greater risk of getting eye melanomas.

TREATMENT/ MANAGEMENT PROCEDURES

EXCISION AND CRYOTHERAPY

Excision is the cutting out of a disease tissue from the healthy tissue (The British Medical Association, 2002). Excision is indicated in malignant tumors of the conjunctiva such as conjunctival melanoma and squamous carcinoma. However, excision alone has been found to be an inadequate treatment for both conjunctival melanoma and carcinoma (Yousef and Finger, 2012; Finger, 2005). Primary excision alone has been associated with high recurrent rates. This finding is because the tumor edges typically extend
beyond its visible margins. Conjunctival melanomas are known to include edges that are without pigment. This is why cryotherapy is included to extend the margins an additional 3mm. This is the use of temperatures below freezing to destroy the tumor cells. Cryosurgery destroys cells in several ways:

- First, the rapid creation of intracellular ice (within cancer cells) is lethal.
- Second, as ice forms outside a cell, the water inside is drawn out. This shrinks the cell and collapses cellular membranes resulting in a release of proteins and chemicals that kills cancer cells.
- Third, as ice (that surrounds shrunken cells) begins to thaw, large amounts of free water (produced by the thawing ice) rush back inside the cancer cells making them burst.

Modern cryosurgery is performed in a manner to produce predictable tissue response in the target cancer. Factors that influence the efficacy of cryo destruction include the cooling rate, tissue temperature, the freeze-thaw cycle, and the number of repetitions. Special techniques must be used to prevent or limit intraocular freezing that might affect vision.

Figure 37: Excision of eyelid melanoma- (a) Large melanoma of eyelid (b) Lesion is excised (c) Reconstructed eyelid
TOPICAL CHEMOTHERAPY

Chemotherapy is the treatment of diseases by use of drugs that have a specific toxic effect upon the cancer cells. Finger (1993) published the first case where mitomycin chemotherapy eye drops were used to treat malignant melanoma of the conjunctiva and cornea. More recently, Interferon alpha eye drops are also being used for the treatment of conjunctival melanoma. Some tumors are multiple (multifocal) while others can be amelanotic (invisible to clinical examination). If large or multifocal, standard surgery and cryotherapy may be inadequate to completely destroy the tumor. When they are amelanotic or difficult to see, they may also be missed. This is why a topical eye drop can be used to treat all of the affected conjunctival surfaces.

RADIATION THERAPY

While the primary treatment of ocular tumors is frequently surgical, several forms of ophthalmic radiation have also been used to treat these malignancies. Radiation therapy uses high energy radiation from x-rays and other sources to kill cancer cells and shrink tumors. The goal of radiation therapy is to eradicate tumor burden in a manner that maintains visual function and preserves surrounding sensitive ocular tissues. Ophthalmic radiation may be used as a curative therapy, as adjuvant treatment following surgical excision, or as a palliative therapy for advanced cases of tumors. The two main types of radiation therapy are the external beam radiotherapy and plaque brachytherapy. External
beam radiation therapy employs radiation that comes from a machine outside the body while plaque brachytherapy employs radiation that is administered by placing a radiation plaque over or very near the tumor (Jereb, et al., 2004).

Figure 39: Plaque brachytherapy

Proton therapy

A cyclotron is a particular type of radiotherapy machine specifically used to treat eye tumors. It directs a proton radiation beam precisely at the affected area, causing as little radiation exposure as possible to the surrounding healthy eye tissue. Before the treatment, a minor operation to attach small metal tags to various parts of the eye. The tags act as markers for the radiation beam (Raghavan, 2005).

ENUCLEATION

Enucleation is the surgical removal of the eye, leaving eye muscles and the contents of the eye socket intact. The eyelids, eyelashes, brow and surrounding skin all remain. This procedure is done when there is no other way to remove the cancer completely from the eye. Unfortunately, loss of vision for the eye removed is permanent because an eye cannot be transplanted. The eye is removed and a spherical implant made of coral or hydroxyapatite is placed into the orbit. This allows the blood vessels to grow into the porous coral material. Occasionally, porous polyethylene implants are used. The muscles that help give movement to the eye are then sutured to the implant, which will allow for some movement of the prosthesis. After enucleation, there is reduced visual field on the side of the body when looking straight ahead and there is loss of depth perception. Many
of the skills of depth perception can be relearned and with time, almost all patients are able to do all the things they used to do before losing their eye (Shields, et al., 2011).

Figure 40: Enucleation, before and after insertion of prosthetic eye

**EXENTERATION**

Exenteration is the surgical removal of the eye and soft tissues surrounding it. Both the eye and the total contents of the eye orbit are removed including the eyelids. It is performed in bulky tumors that have arisen from the palpebral conjunctiva and spread to the eye lids and anterior orbit (Kanski, 1998). Most patients prefer to wear a patch after exenteration, rather than a prosthesis, especially with the larger reconstructions. Attempts have been made to preserve more of the orbital tissues including palpebral and bulbar conjunctiva to achieve a reconstruction of the socket, which allows retention of a standard ocular prosthesis using mucosal grafts in addition if needed (Tylers, 2006; Goldberg, et al., 2003). This is feasible in only a small minority of exenterated patients.
PHOTODYNAMIC THERAPY

Photodynamic therapy (PDT) destroys cancer cells with a fixed-frequency laser light in combination with a photosensitizing agent that is injected into the bloodstream. The photosensitizing agent alone is harmless and has no effect on either healthy or abnormal tissue. However, when laser is directed onto tissue containing the drug, the drug becomes activated and the tissue is rapidly destroyed. The laser light used in PDT is directed through a fiber-optic placed close to the tumor to deliver the proper amount of light and selectively target only the abnormal tissue. An advantage of PDT is that it causes minimal damage to the healthy tissue. However, PDT makes the skin and eyes sensitive to light for about 6 weeks after treatment. Patients are advised to avoid direct sunlight for at least 6 weeks after PDT treatment (Murray, 2012).

Barbazetto, et al. (2004) noted tumor regression after one month of treatment of patients with conjunctival squamous cell carcinoma with photodynamic therapy. The patients received one to three treatments of verteporfin (6 mg/m² body surface area intravenously). The light dose was calculated as 50 J/cm². All tumors were irradiated 1 minute after injection.

ARGON LASER PHOTOABLATION

A laser is a device that transfers light of various frequencies into an extremely intense, small, and nearly non-divergent beam of monochromatic radiation in the visible region, with all the waves in phase; capable of mobilizing intense heat and power when focused at close range. Argon laser is a laser with ionized argon as the active medium, whose
beam is in the blue and green visible light spectrum, used for photocoagulation or photoablation. Shin, et al. (2013) reported a complete removal of conjunctival naevi in over 230 patients with no significant complications such as infections, corneal damage, or permanent injection. Argon laser photoablation is a safe and effective treatment for benign superficial conjunctival pigmentation.

Figure 42: (a) Conjunctival naevus before photoablation
(b) After photoablation

CASE PRESENTATIONS

CASE 1: CONJUNCTIVAL PAPILLOMA

A 58-year-old otherwise healthy man presented with an asymptomatic, slowly growing papule near his right medial canthus of 6 years’ duration (Figure 43a). Upon retraction of the lower eyelid, several large exophytic pedunculated tumors in the right inferior fornix and palpebral regions were discovered (Figure 43b). He had a history of a similar lesion in the same location removed 15 years prior. The lesions were grossly removed via shave biopsy technique with cauterization of each base. All of the tissue was submitted for paraffin-embedded sections. Microscopic examination of the specimen demonstrated a typical frond-like pattern with prominent fibrovascular cores covered by an irregular proliferation of non-keratinized stratified squamous epithelium containing goblet cells. This histology is characteristic of a conjunctival papilloma. Four months after removal of the patient’s already recurrent lesions, a small re-recurrence of a 2-mm papule was noticed and the patient was referred to ophthalmology (Litak, et al., 2012).
CASE 2: SQUAMOUS CELL CARCINOMA

A 94-year-old woman had noticed that the conjunctiva of her right eye was hyperemic 10 months earlier. She was suspected of having a conjunctival tumor at a private clinic 5 months later but she refused to undergo further examinations because of her age. After a rapid increase in the size of the mass, she was referred to Chiba University Hospital, Japan. At first examination, her visual acuities were Hand motion at 30 cm OD and 0.8 OS. Slit-lamp examination showed a large, irregular-surface mass on the nasal conjunctiva. The mass was so large that it covered the pupil of the eye. The ocular movements were full in both eyes. She had neither history nor symptoms or signs of systemic cancers. Cytopathological examination from a scraping biopsy showed a class IV, well-differentiated squamous cell carcinoma. Because the size of the tumor was large, a systemic CT examination was carried out. The CT examination showed a large tumor surrounding the appendix, multiple legions in the liver and lungs. No other orbital lesions were found on CT examination. The tumor was excised and the patient given 0.04% Mitomycin C eyedrops 2 times/day. The tumor was almost completely excised (figure44c) and her family were very happy with the results of the operation. She declined treatment for the primary systemic cancer because of her age. The patient died of the primary systemic cancer one month after the surgery (Mitamura, et al., 2011).
CASE 3: EYE TUMOR/CANCER

In January 2011, a father noticed that the left eye of his 6-year-old son had a shifted position, which made him take the boy to an optical service hospital in Orlu. He disclosed that his son underwent treatment there for some time with the cause not being diagnosed. He went further to reveal how they were referred to one Egyptian who is based in Aba Abia state, who also offered his own services to no avail. In search of solution and cure for their only son’s ailment their ship anchored at the Federal Medical Centre (FMC) and General Hospital Umuguma all in Owerri Imo state, with several weeks of visitations and treatment, yet neither the cause nor any solution was proffered. The boy was referred to an Eye Foundation Hospital in Ikeja Lagos, from where they were referred to Lagos University Teaching Hospital (LUTH).

It was at LUTH, that the ailment was diagnosed to be tumor/cancer of the eye before he was referred to University of Nigeria Teaching Hospital (UNTH) Enugu for an affordable accessibility since the parents live in Imo state. The treatments and chemotherapy, received in UNTH showed a sign of improvement for two weeks but what followed after the two weeks was a shocking rapid growth of the tumor which resulted in the removal of the boy’s damaged/infected left eye in a surgical operation. This operation again seems a respite of hope, only to escalating again after two weeks to what is shown in the figure 45. Medical experts in the University Teaching Hospital (UNTH) Enugu has advised that the only option to save the boy’s life is to fly him to India for a surgical operation which is estimated to gulp not less than three million Naira (N3m) and so far not up to 10% of the said amount has been realized for the trip although the international passports was done and delivered by the Nigeria Immigration Services (NIS) within a record time of 24hrs in order to contribute their quota in saving the life of a poor innocent boy whose
sight will send pity and compassion into the spines of even the conscienceless and the heartless. He later died in his home town Amaifeke, Orlu L.G.A., Imo state (Anonymous, 2014).

Figure 45: Eye tumor/cancer

**CASE 4: PRIMARY ORBITAL NEUROBLASTOMA**

A 5-year-old boy was brought to the Outpatient Department of a hospital with complaints of painless, rapidly progressing protrusion of the left eye since five months which was associated with loss of vision. The mass between the lids was red and firm in appearance and impeded any view of his left globe (figure 46). The patient was physically normal and had no signs of congenital malformation. There were gross restriction in movement of proptosed mass in all positions of gaze. Symblepharon was present. No, other details could be seen. The lids were edematous and ecchymosed. The mass was non-tender and globular. It was immobile and non-pulsatile. It was not expansible on coughing or crying. Right eye appeared normal. Complete hemogram, including general blood picture, ESR, urine and stool examination were found to be within normal limits. Computed tomography (CT) scan showed soft tissue density enhancing mass in the left retro-orbital region measuring 3x2.7 cm pushing the left eye ball anteriorly. The optic nerve could not be separated out and lesion seemed to be adherent to the eye ball. There was no evidence
of mass in the sinuses, nasal cavities or intra-cranial cavities. Ultrasomography (USG) scan of the left eye ball was performed using high frequency and showed a large hypoechoic mass. Fine needle aspiration cytology (FNAC) of the tissue showed features suggestive of Neuroblastoma.

Subsequent medical evaluation including chest X-ray, USG of the abdomen, whole body CT scan and bone scintigraphy showed no evidence of systemic involvement or metastasis. The tumor was diagnosed as primary orbital neuroblastoma. The whole orbital exenteration operation was done under general anesthesia and the patient further received 11 cycles of chemotherapy and 36 Gy local EBRT. Adequate treatment was given and patient was further referred to pediatric oncology department for further management (Sofi, et al 2012).

Figure 46: Primary orbital neuroblastoma

CASE 5: RHABDOMYOSARCOMA

A 15year old boy experienced what he thought was his first migraine headache. What he and his family didn’t know was that he had a deadly, malignant monster rapidly expanding inside and that would soon be bursting out of his skull, ravaging his right eye and disfiguring his face. CAT scan revealed the tumor was pressing against his brain. Haiti’s overwhelmed health-care system could offer little assistance. The doctors affiliated with another Haitian relief organization, Project Medishare, suggested that he be taken to the Dominican Republic for palliative, not curative treatment. As everyone would eventually learn, the patient was suffering from a rare form of cancer called
rhabdomyosarcoma. Although the tumor tends to grow (and potentially metastasize) very quickly, it also usually responds quickly to aggressive treatment provided that the treatment (in this patient’s case, chemotherapy, radiation and, ultimately, surgery) is administered soon enough. He’ll require, at minimum, three months of potent bi-weekly chemotherapy, followed by six weeks of daily radiation (Dorfman, 2014). The hope is that the tumor will shrink enough to be surgically excised.

Figure 47: Rhabdomyosarcoma

**CASE 6: MALIGNANT EYE TUMOR**

The Haitian toddler was hospitalized in Port-au-Prince, Haiti with a fever. A tumor was noticed growing in her left eye. It was diagnosed as a malignant eye tumor by a hospital in Haiti. As authorities were trying to secure passports and emergency visas for her and her mother to leave the country for surgery, she suddenly died in the hospital and the exact cause of her death could not be ascertained (Anonymous, 2011).
CASE 7: NEUROFIBROMATOSIS

A 51-year-old man from Portugal has one of the most extreme facial tumours ever seen in medical history. Unemployed, the patient is blind in one eye as a result of the giant weeping growth that has consumed his features. For years he has struggled to eat properly or carry out everyday tasks. The tumor has taken over his mouth and tongue, ballooning his lips, twisting his gums and breaking his teeth off. The deep, 15cm long growth which started as a birth mark when he was just 11 was sparked by abnormalities in his capillaries and veins. It started to get much bigger when he hit 16 and has been expanding ever since. It also bleeds frequently and most mornings his pillow is covered with blood from the night before.

"Children see me and start crying," he mumbles sadly from behind the monstrous 12lb growth that is eating his face. "My face is ugly but my heart is not," he pleads. "I just want people to understand who I am. I don't like to speak very much because I dribble all the time." He lives in Lisbon and spends most of his days sitting in one of the city's main squares watching the world go by. Many take photos on camera phones of the man known locally by the hurtful nickname 'StainedSkin.' And his looks have also made him an unlikely 'celebrity' in the area. After his mother died several years ago, his younger sister who has a 9-year-old daughter of her own became his primary carer. She balances life as a hairdresser with looking after him, ordering his food and being on call 24/7. Recently the patient flew to the UK to discuss the possibility of surgery to remove part of the tumor. But because his mother was a Jehovah's Witness, he refused any surgery.
because his religion means he can't have blood transfusions. This made his sister fume and her brother's stubbornness led to her storming out of a surgeon's surgery. Doctors in London came up with a plan to remove parts of the growth bit by bit without transfusion. And after days of uncertainty, he finally agreed to go under the knife after returning to Portugal to think it over. He is expected to come back to the UK soon once a date is set for the operation which will transform his face - and his life – forever (Anonymous, 2010).

![Figure 49](image)

Figure 49: (a) Neurofibromatosis patient (b) & (c) with his sister

**CASE 8: NEUROFIBROMATOSIS**

A Chinese male, with a giant facial tumor used to be recognized as a tumor man. He is believed to have the world's worst case of neurofibromatosis, a disorder that affects tumor growth. At a very young age of four, he suffered from a rare genetic disease called Neurofibromatosis, obscuring his external appearance and warping his backbone. His face and head swelled when he was a child and, according to a neighbor, his mother said he should be a government official when he grew up because his head was so big. Tumor growths on his face caused him to have a trouble to hear, to speak and even to eat. It grew larger and rapidly as he aged, knocking out his teeth, crushing his left ear to hang down his shoulder level, blocking his left eye and deforming his bone as well. The tumors cause him constant physical pain and have stunted his growth. The patient, from Hunan province, China has gone through several operations in the past to remove the tumors on his face that have at one point weighed as much as 15 kg (Anonymous, 2010).
CONCLUSION

Ocular tumors may be benign or malignant; melanocytic or non-melanocytic. Benign conjunctival tumors such as the conjunctival naevus is the most common melanocytic conjunctival tumor. It usually presents at puberty or early adult life. Malignant ocular tumors can progress and lead to death. Long term exposure to ultraviolet radiation from the sun exposure to radiation and toxic chemicals and inherited genes are the most common causes of ocular tumors. Early detection and regular check-ups to monitor the progression of a benign tumor is important to prevent its transformation to a malignant case. Surgical treatment procedures such as radiation therapy may not completely destroy the tumor cells and recurrences may occur after excision. Chemotherapy with topical eyedrops are helpful in treating all affected areas.

RECOMMENDATIONS

Patients with benign tumors are advised to visit the eye clinic regularly to monitor any possible progression of the tumor. Diagnostic centers with fully equipped laboratory facilities for excision biopsy should be set up across the country for appropriate diagnosis on the type of tumor present. Proper funding by governmental and non-governmental agencies is required toward adequate diagnosis and treatment of conjunctival
tumors. Further research toward adequate treatment procedures and proper understanding of the causes should be encouraged. Public education and enlightenment programs should be frequently organized especially in the rural areas to properly educate people on the dangers of ocular tumors.

REFERENCES


