Tumours - eyelid, lacrimal and orbital

Mr David H Verity, MD MA FRCOphth
Consultant Ophthalmic Surgeon

Qu: **What is a tumour?**

A tumour refers to a new growth of tissue, and can be benign (implying limited growth with no distant spread), pre-malignant (with potential to become malignant) or malignant (implying an ability to grow, to invade and replace adjacent structures, and to spread to other parts of the body – or 'metastasise').

The character of malignant tumours can vary from those which take many years to grow (and with little metastatic potential), to those which grow rapidly (and which can even metastasise before the primary tumour is discovered).

Qu: **What are the causes of tumours?**

In most cases the exact cause of a tumour is not known, but factors which are thought to play a role are:

(i) The environment: the best example is exposure to sunlight which increases the risk of eyelid skin tumours, particularly in fair-skinned individuals.
(ii) The immune system (which also protects against infections), which maintains constant vigilance against abnormally proliferating cells; if there is a breakdown of this 'surveillance', a tumour can develop. For this reason, patients on strong drugs which suppress the immune system are at greater risk of developing tumours.
(iii) Inheritance: most have a genetic component, although only exceptionally rarely are tumours inherited on a 'one to one' basis from a parent.

Qu: **What are the symptoms?**

A very wide variety of benign, premalignant, and malignant tumours can occur around the eyelids and the structures adjacent to the eye. The symptoms vary according to location and nature of the tumour. Tumours of the eyelids are generally visible to the patient, and can sometimes cause local swelling, loss of the eyelashes, and/or a red or uncomfortable eye, but those on the rear surface of the eyelids, or within the eye socket, may not be so apparent. The latter can present in many different ways, with symptoms including pain, swelling, blurred vision, double vision, or displacement of the globe (either forward and/or to one side).

Qu: **What eyelid tumours can occur?**

Neoplastic diseases of the lid include basal cell carcinoma (BCC, a common and *relatively* indolent growth with virtually no metastatic potential), squamous cell carcinoma (SCC), sebaceous cell carcinoma, melanoma, lymphoma, Merkel cell tumour, and Kaposi's sarcoma (these last three being very rare).

To be certain of a diagnosis, a biopsy is necessary, although in many cases dermatologists are able to treat typical BCCs without performing a biopsy first. When a tumour is close to the eyelids, in view of the complex anatomical nature of the eyelids, a biopsy is usually preferred before beginning treatment.

BCCs, SCCs, and melanomas account for 90 %, 5%, and <1% of all neoplastic eyelid lesions respectively. SCC can develop from pre-existing 'pre-malignant' disease (see below). SCC is more aggressive than BCC, with ~ 5% of patients developing local periocular recurrence despite complete excision on histological examination, and with a mortality rate from SCC related death of about 2 %.

Sebaceous gland carcinoma (SGC) is a rare and aggressive tumour of the sebaceous (meibomian) glands in the eyelids which can spread both locally and by metastasis. A history of *presumed* recurrent chalazion, or chronic unilateral blepharoconjunctivitis (red eyelid margins) always raises suspicion.

Melanomas are often irregularly pigmented, although some may not be, and can present with inflammation and bleeding. Clinical forms include lentigo maligna, superficial spreading, and nodular melanoma.

Qu: **What are the 'premalignant' tumours of the eyelids?**

Premalignant disease is quite common, particularly in fair-skinned individuals. These lesions include

(i) Actinic keratosis (sun damage of the superficial layers of the skin), which can progress to squamous cell carcinoma,

(ii) Bowens disease (malignancy in the basal layers of the skin - 'intraepithelial'. About 4 % of these lesions progress to squamous cell carcinoma.

(iii) Lentigo maligna – slowly spreading superficial skin pigmentation which becomes invasive in about a third of all patients.

Qu: What is the treatment of eyelid tumours?

The management of eyelid tumours depends on the many factors such the age and general health of the patient, the nature of the tumour, and whether there could be local or peripheral spread. Frequently, a general medical and/or oncology opinion are necessary even if a tumour appears to be localised to the eyelid or the orbit.

Eyelid tumours are generally managed with *complete* excision (this can include Mohs' micrographic surgery, performed by a dermatologist) and subsequent reconstruction.

Reconstruction methods include allowing the defect to heal naturally – often with excellent results, directly closing the defect, or by using a combination of local tissue flaps, skin grafts and /or other grafts (such as ear cartilage or hard palate). Some tumours can be managed with local freezing (cryotherapy), irradiation, or medical treatments which stimulate the immune system to destroy abnormal tumour cells.

Patients with certain tumours (SCC, Sebaceous cell carcinoma, and melanoma) also require a general oncological review to exclude disease elsewhere, and this may include surgery to investigate the presence of lymph node involvement in the neck.

Qu: **What tumours can affect the lacrimal system?**

Tumours which affect the lacrimal system are unusual, these including lymphoma around the lacrimal sac, and intrinsic tumours within the sac itself. A biopsy is required, and the treatment depends on the nature of the tumour, and can include radiotherapy, chemotherapy and wide local excision with reconstruction.
N.B. The vast majority of swellings at the inner corner of the eyelids (over the lacrimal sac) are due to a benign mucocoele (build up of mucus) – tumours in this position are very rare indeed.

Qu: **Can tumours pose a risk to the eye and vision?**

Yes – either as a result of infiltration and distortion of the eyelid with reduced corneal protection (BCC, SCC, SGC), direct involvement of the corneal surface (SGC), and/or spread into the orbit (any tumour). In addition, if left untreated for a long time, complete excision can result in a larger defect (and even *loss of the eye* in rare cases), this necessitating more extensive reconstructive surgery with greater risk to overall protection of the eye.

Qu: **What tumours can affect the orbit?**

A very wide range of tumours can occur in the orbit, reflecting the different types of tissue in this area. Generally, dedicated CT imaging and a biopsy (of part or of all) of the irregular tissue growth is required before a definitive diagnosis can be made, and treatment started. Certain 'masses' require complete intact excision (based on the history and CT findings), while in others a biopsy is necessary first – this frequently identifying inflammation without any sinister changes. The treatment depends on the severity and extent of the disease, the propensity for spread outside the orbit, and whether there is already disease elsewhere. Lymphoma, for example, always requires a general oncology / haematology review and investigations first, followed by

treatment which ranges from observation alone to chemotherapy and adjunctive radiotherapy.

Qu: **What is the follow up period after surgery to treat a tumour?**

Because of the risk of recurrence, patients are reviewed regularly (at increasing intervals) for up to five years after treatment.

Qu: **Is there a risk of a tumour spreading to the other eye?**

This is a commonly asked question; although some tumours such as BCC and SCC can occur *de novo* (or recur) on any of the eyelids, they do not 'spread' to the other side of the face (unless left untreated for a very long period). Some orbital 'swellings' which are subsequently proved to be due to *inflammations* (such as sarcoidosis), can occur simultaneously or consecutively in each orbit, but do not represent a 'spread' from one side to the other.