#### ORBITAL & PERIOCULAR TUMORS

### Orbit:

**Orbital lymphoma:** This is the most common type of cancer of the orbit in adults. It is usually a form of B-cell non-Hodgkin's lymphoma.

There are several types of B-cell lymphomas that occur in the orbit. These include MALT, follicular lymphoma, large B-cell lymphoma and mantle cell lymphoma. Less-common types include natural killer T-cell lymphoma, mycosis fungoidis and Hodgkin's lymphoma.

Diagnosing orbital lymphoma often starts with a CT scan followed by a surgical biopsy. Treatment may include radiation, chemotherapy, targeted therapy, or a combination of these, depending on type of lymphoma and the stage of the cancer.

**Orbital sarcoma:** Sarcomas arise from muscles or fatty tissue. The most common sarcoma in the orbit is rhabdomyosarcoma, which is found most frequently in children.

The most common type of orbital rhabdomyosarcoma can be treated with a combination of chemotherapyy and radiation. This combination can prevent the need for removal of the eye and orbital content in the majority of patients.

**Orbital and optic nerve meningiomas:** These tumors are benign, meaning they are not cancerous and do not spread. They can be associated with the optic nerve. They can also form in the meninges (membranes that line the skull and enclose the brain and top of the spinal cord) in the base of the skull and extend into the orbit.

These tumors are usually diagnosed with an imaging exam, such as an MRI of the orbit. Treatment usually includes radiation.

**Metastatic orbital tumors:** Almost all types of cancer can metastasize (spread) to the orbit (the space behind and around the eye). The most common are breast, lung and prostate cancer; melanoma; and carcinoid tumors. Metastatic tumors of the orbit are treated with chemotherapy, radiation and occasionally surgery.

### **Lacrimal Gland:**

**Lymphoma:** Lymphomas in the lacrimal gland usually form as lesions in the upper outer part of the eye. Diagnosis starts with a surgical biopsy. Treatment may involve radiation to the orbit, chemotherapy or a combination of these treatments.

**Adenoid cystic carcinoma:** This rare cancer has the potential to spread and become lifethreatening. For smaller or less aggressive tumors, eye-preserving surgery followed radiation and chemotherapy. We are also treating these patients with intra-arterial chemotherapy to shrink the size of these tumors so that complete excision is possible. This is done through a multi-disciplinary approach with oncology, interventional radiology and radiation oncology.

**Pleomorphic adenoma:** This is the most common benign (non-cancerous) tumor of the lacrimal gland. It is diagnosis based on clinical signs and imaging studies. Treatment includes total excision of the tumor.

**Other lacrimal gland carcinomas:** Other forms of carcinoma such as adenocarcinoma or squamous carcinoma or mucoepidermoid carcinoma of lacrimal gland are managed similarly to adenoid cystic carcinoma.

## **Lacrimal Sac and Nasolacrimal Duct:**

**Squamous cell carcinoma:** Diagnosis is made with CT scan. Surgery is the main treatment. Radiation therapy, is sometimes used after surgery to decrease the likelihood of recurrence.

**Transitional cell carcinoma:** Diagnosis is made clinically and with CT scan. Treatment for transitional cell carcinoma of the lacrimal sac includes surgery, and often radiation therapy.

**Lacrimal sac lymphoma:** Treatment of this disease begins with a surgical biopsy to establish exact type of lymphoma. Treatment of lacrimal sac lymphoma is similar to other forms of orbital lymphoma. Depending on the type and stage of lymphoma, very low-dose radiation therapy or chemotherapy can be used.

# Conjunctiva & Ocular Surface:

**Squamous cell carcinoma:** This is the most common type of conjunctival eye cancer. It can affect the area around the cornea on the eye surface or the inner conjunctival layer of the eyelids. It has a very low risk of spreading to the lymph nodes.

This eye cancer is most often treated with surgery and freezing treatments. Sometimes topical chemotherapy in the form of eye drops is used after surgery.

If this cancer returns in an aggressive form, the eye and eye socket may have to be removed.

**Melanoma:** Melanoma can occur on the conjunctiva on the surface of the eyeball (bulbar conjunctiva) or on the conjunctival covering of the inside of the eyelid (palpebral conjunctiva). Conjunctival melanomas can spread to the lymph nodes and other parts of the body. This risk is higher for thicker melanomas.

Conjunctival melanoma is treated with surgery combined with cryotherapy along the surgery's edges. We may consult ENT for a sentinel lymph node biopsy to look for signs of early metastasis. Sometimes doctors use chemotherapy eye drops after surgery to reduce the chance of recurrence.

Mucosa-associated lymphoid tissue lymphoma (MALT): This form of lymphoma occurs on the surface covering of the eye and the inside layer of eyelid (conjunctiva).

Treatments for this eye cancer may include chemotherapy, targeted therapy or radiation therapy.

# **Eyelid:**

**Basal cell carcinoma**: More than 90% of eyelid cancers are basal cell carcinomas, a type of skin cancer. This cancer is usually not aggressive and does not spread to lymph nodes or distant organs.

The disease is treated with surgery to remove the tumor. The eyelid tissue will also be reconstructed to preserve the patient's vision, maintain comfort and restore the eye's appearance. Recently, we have been using a medication, vismodegib, to treat advanced cases involving the orbit. In some cases, targeted therapy and adjuvant radiation therapy may be appropriate.

**Squamous cell carcinoma:** This skin cancer occurs less often on the eyelid than basal cell carcinoma, but it is more aggressive. It can spread to nearby lymph nodes and other parts of the body.

The main treatment for this type of eye cancer is surgical removal. Radiation therapy or other treatments may be used in addition to surgery if a large area is affected or if the tumor cannot be fully removed.

**Melanoma:** This type of skin cancer accounts for about 1% of eyelid cancers. It is potentially life-threatening. It can affect the eyelid skin or the conjunctiva.

Melanomas of the eyelid are surgically removed. We work with dermatology and ENT to treat these patients. ENT may perform a sentinel lymph node biopsy can find microscopic signs of metastasis, which can help doctors diagnose and treat metastatic melanomas in the early stages.

**Sebaceous carcinoma (meibomian gland carcinoma):** This rare type of eyelid carcinoma is also known as sebaceous gland or sebaceous cell carcinoma. If the orbit is involved then orbital imaging (CT or MRI) is necessary.

Sebaceous cancer in the eyelid is surgically removed, and the eyelid is reconstructed. We sometimes also use topical chemotherapy in the form of eye drops or cryotherapy to treat disease. Sebaceous carcinoma can spread to the regional lymph nodes. For larger sebaceous carcinomas, we may consult ENT to perform a sentinel lymph node biopsy to look for microscopic signs of the disease's spread.

**Merkel cell carcinoma:** This is a rare but aggressive cancer that starts in the eyelid's touch receptors. It usually shows up as a fast-growing purplish or flesh-colored mass.

Treatment for this cancer typically starts with surgery to remove the mass and reconstruct the eyelid. Adjuvant radiation therapy can also be used in some cases. ENT is typically involved to perform a sentinel lymph node biopsy to determine if the cancer has spread to nearby lymph nodes.

In about 30% to 50% of patients, the cancer has spread to the lymph nodes. In these cases, the nodes also need to be treated surgically or with radiation therapy. For tumors larger than 20 mm (about an inch), adjuvant chemotherapy may be used to prevent the cancer's return.