Orbital Tumors: General Information

Tumors and inflammatory processes can occur behind the eye. They often push the eye forward causing a bulging of the eye called proptosis. The most common causes of proptosis are thyroid eye disease and lymphoid tumors (lymphoma and atypical lymphoid hyperplasia).

Other tumors include vascular tumors (e.g. hemangiomas, lymphangioma), lacrimal gland tumors (e.g. dacrystoadenitis, benign mixed tumor, sarcoidosis and adenoid cystic carcinoma), and growths that extend from the sinuses into the orbit (e.g. squamous carcinoma, mucocele). Metastatic cancer can also form an orbital tumor. Lastly, an orbit tumor can also be caused by inflammation (e.g. pseudotumor, sarcoidosis) or infection (abscess).

Most patients with orbital tumors notice a bulging of the eyeball. Infections, inflammations and certain orbital cancers can also cause pain. Less commonly, orbital tumors may be an incidental finding on CT or MRI of the head, sinuses and orbit.

Though CT, MRI’s and ultrasound can help in determining the probable diagnosis, most orbital tumors are diagnosed by a surgical biopsy called an orbitotomy (anterior or lateral). A specimen is sent to an ophthalmic pathologist who helps determine the exact diagnosis.

Treatments
When possible, orbital tumors are totally removed. If they cannot be removed or if removal will cause too much damage to other important structures around the eye, a piece of tumor may be removed and sent for evaluation by an eye-pathologist. Occasionally an orbital tumor is too big or involves the sinuses and requires more extensive surgery with bone-flaps.
If tumors cannot be removed during surgery, most orbital tumors can be treated with external beam radiation therapy. Certain rare orbital tumors require removal of the eye and orbital contents. In certain cases orbital radiotherapy may be used to treat any residual tumor (in an effort to spare vision and the eye).

Some important types of orbital tumors are:

- Lacrimal gland tumors
- Adenoid Cystic Carcinoma of the Lacrimal Gland

- Cavernous Hemangioma of the Orbit
- Lymphangioma of the Orbit
- Orbital Mucocoele
- Orbital Rhabdomyosarcoma
- Optic Nerve Sheath Meningioma
- Lymphoid Tumors
- Orbital Meningiomas and Schwannomas
- Metastatic Tumors
- Cystic Lesions (Dermoid cysts, Teratoma)
- Optic Nerve Gliomas
- Metastatic Tumors: Neuroblastoma

**Lacrimal Gland Tumors**

*Computed tomography shows an flattened round mass located between the lateral bony orbital wall*

Mass lesions of the lacrimal gland can be classified broadly into inflammatory and neoplastic subtypes. Inflammatory etiologies, while not uncommon, include dacryoadenitis, sarcoidosis, and orbital inflammatory pseudotumor. For the purposes of this discussion, the focus will be on
neoplastic lesions of the lacrimal gland. Most of the neoplastic lesions in the lacrimal gland are epithelial in origin, with approximately 50% classified as benign and 50% as malignant.

Benign lesions include pleomorphic adenomas (benign mixed cell tumors), benign reactive lymphoid hyperplasia, and oncocytomas. These lesions are slowly growing masses more commonly found in adults in their forth to fifth decades of life. Malignant tumors of the lacrimal gland include adenoid cystic carcinoma, adenocarcinoma, squamous cell carcinoma, mucoepidermoid carcinoma, and malignant lymphomas.

Adenoid cystic carcinoma is the most common malignant lacrimal gland tumor, comprising 50% of malignant tumors of lacrimal gland and 25% of all lacrimal gland tumors. Most cases are seen in the third decade of life with a second bimodal peak in the teenage years.

**Adenoid Cystic Carcinoma of the Lacrimal Gland**

Adenoid cystic carcinoma is a type of cancer that affects glandular structures of the lacrimal gland. Orbital adenoid cystic carcinoma usually occurs in patients 20-50 years old.

*Computed Radiographic Tomography (CT) demonstrates and Adenoid Cystic Carcinoma of the Lacrimal Gland with Orbital Extension (arrow)*
When an adenoid cystic carcinoma of the lacrimal gland grows, it typically pushes the eye down, towards the nose and forward. It can cause bulging of the eye (called proptosis). Another characteristic of adenoid cystic carcinoma is that it also invades local nerves causing pain. Therefore, pain and proptosis are the most common symptoms of adenoid cystic carcinoma of the lacrimal gland.

A complete eye examination with a clinical history and ophthalmic examination are crucial to the diagnosis of adenoid cystic carcinoma of the lacrimal gland and orbit, or any kind of lacrimal gland tumor.

CT scans, MRI’s and ultrasounds are also helpful in determining the diagnosis of adenoid cystic carcinoma of the lacrimal gland. When the eye cancer specialist sees a well-defined tumor in the superior-temporal (upper - outer) part of the orbit, that may have eroded into adjacent bone and/or extend into the orbital apex, he or she should suspect the tumor might be an adenoid cystic carcinoma. Other tumors to consider include: benign mixed tumor, adenocarcinoma, dacryoadenitis, or other processes affecting the lacrimal gland. All of these kinds of tumors have a similar clinical presentation.

When possible, an adenoid cystic carcinoma should be totally removed. This usually requires a surgery called a lateral orbitotomy. Unfortunately, total removal is often impossible due to the tumor’s size, shape, and presence of invasion. Should the adenoid cystic carcinoma be found to have a capsule, and should your doctor be able to remove it within its “capsule,” surgery offers the best prognosis.

In many cases, the adenoid cystic carcinoma extends beyond the capsule. In these cases, removal of the entire adenoid cystic carcinoma may require removal of the orbital contents, bones and adjacent structures. Due to the poor (local control) results from this type of extensive surgery; combinations of surgical removal and subsequent high dose local radiation therapy are offered as an alternative. There are no comparative studies to prove if extensive surgery or surgery with subsequent radiation therapy is better for survival.

**Cavernous Hemangioma of the Orbit**
Hemangioma is a benign tumor that is found to grow within the orbit. Most commonly found right behind the eye (in the muscle cone), it can push the eye forward causing a bulging
doctors call proptosis.

Computed tomography (CT) shows a flattened round mass located between the lateral and medial rectus muscles behind the globe. The mass itself has well-defined borders, which suggest a non-infiltrative tumor.

Cavernous hemangioma of the orbit is most commonly seen in middle-aged women. Most are found within the muscle cone, but can be found anywhere in the orbit.

If large, they can indent the back of the eye causing choroidal folds, or push on the optic nerve causing atrophy, and subsequent visual loss.

Rarely, the eye can become pushed out (proptotic) so far that there are corneal exposure problems, dry eye, corneal erosions and ulcers.

Cavernous hemangioma of the orbit is usually a slow-growing tumor. Tumor growth occur, will be measured by eye examinations including (but not limited to) visual acuity, color vision assessment, Hertel exophthalmometry (a measure for eye-bulging), as well as an evaluation for double vision (strabismus), corneal exposure, retinopathy, vascular damage, and optic...
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neuropathy (optic nerve damage).

Surgical treatment of orbital hemangioma is indicated when tumor growth causes optic nerve compression, and corneal exposure, or evidence of vision loss. Some more subtle indications regard tumor location within the orbit, and its anatomic relations with important orbital structures.

The goal of local resection for choroidal hemangioma should be complete removal of the tumor. This usually involves careful dissection of the tumor to protect the tumor's capsule (as possible). Connecting vascular feeder vessels should be identified and cauterized. A lateral orbitotomy is usually required to keep the large tumor intact.

**Lymphangioma of the Orbit**

Lymphangioma is rare (less than 7% of childhood orbital tumors). Patients can present with acute proptosis (bulging eye) after minor head trauma, as a gradual proptosis, or after an upper respiratory infection.

*Computed Tomography (CT) shows a large lymphangioma, pushing the eye out of the orbit and compression on the optic nerve (left image).*
Lymphangioma tends to start in the superior and nasal quadrants of the orbit. More than half the times affect anterior (conjunctival and adnexal) structures. Typically, the lymphangioma bleeds into itself causing cysts of blood (blood-fluid bubbles) within the tumor. If the cyst forms behind the eye, it pushes the eye forward. If the tumor forms in the eyelid or structures around the eye "adnexa", blood filled lymphatic channels called "lymphangiectasias" can be seen beneath the conjunctiva.

A careful history may reveal sudden painful proptosis (bulging of the eye), after facial trauma or upper respiratory infection.

Physical examination may reveal bluish discoloration of or blood vessels within the eyelid skin. Should the vessels extend under the conjunctiva, they are called lymphangiectasias. Severe cases can be associated with corneal exposure, ulceration and optic nerve damage.

Though lymphangioma patients can present with a history of sudden proptosis (due to bleeding within the tumor), orbital lymphangioma is typically a slow growing tumor. Therefore, most lymphangiomas are followed by observation for growth (clinical and radiographic studies) prior to considering intervention.

Treatment of lymphangioma is typically indicated when it is associated with growth, optic nerve compression, corneal exposure problems (keratitis sicca), glaucoma or evidence of vision loss.

When treatment of lymphangioma is considered, the goal is rarely complete removal. This is because the edges of most orbital lymphangiomas are poorly defined. Most patients undergo several debulking surgeries to relieve acute optic nerve compression or corneal exposure. In rare cases, orbital lymphangioma patients may require exenteration of the orbit, or radiation therapy for relief of pain.

**Orbital Mucocele**

Orbital mucocele can occur when sinus mucoceles cannot naturally drain through the nose. Instead, they grow and slowly invade adjacent orbital tissues.
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A CT scan image shows an orbital mucocele as a gray mass incompletely surrounded by bony borders (white thin lines), that makes contact with the optic nerve (ON). LM: lateral rectus muscle MR: medial rectus muscle.

Generally arising from the ethmoid or frontal sinuses, orbital mucoceles are mucous or fluid-filled cysts which can displace the eye. Frontal sinus mucoceles can force the eye down, ethmoid tumors will push the eye out and maxillary lesions can elevate or push the eye in. Lastly, orbital mucocele originating in the sphenoid sinus can compress the optic nerve resulting in loss of vision.

Patients with mucocele of the orbit, typically have a history of facial trauma or chronic sinus disease. They tend to be in their mid 40's or older. They can have painless proptosis, or complain of headaches, double vision, or loss of vision.

Though magnetic resonance imaging (MRI) can be consistent with a mucous or serous fluid-filled tumor, a drainage procedure is typically required and found to be diagnostic. The mucoid or serous fluid (which is found to make up the mucocele) should be sent for culture and sensitivity as well as cytologic examination. Mucoceles can be infected. In those cases, the choice of antibiotics can depend on cultures taken during surgery.

The treatment of mucocele of the orbit is surgical. Your orbit surgeon is the best suited doctor to perform adequate resection and reconstruction.

Treatment involves removal of as much of the cyst and its lining as possible. This usually requires an orbitotomy and sinusectomy. It is most important to re-establish or create a new drainage pathway for the mucous to exit the nose.

Orbital Rhabdomyosarcoma

Rhabdomyosarcoma is the most common primary malignancy of the orbit in children. It can also occur in adults. The average age of patients affected by rhabdomyosarcoma is 7 - 8 years. Most parents first notice a droopy eyelid (called ptosis), and/or that the eye is more prominent (called proptosis), or that their child has a tumor under the conjunctiva.

Computed axial tomography (CT-scan) and magnetic resonance imaging (MRI) typically show a mass adjacent to or attached to one of the ocular or orbital muscles. CT is particularly helpful because it shows if the orbital bones have been invaded by the rhabdomyosarcoma.

Rhabdomyosarcoma is usually found in the superonasal orbit (that is under the upper lid near the nose). Though location can vary, as in this case.

Rhabdomyosarcoma can grow rapidly and if the tumor grows into the brain or spreads to the lung, survival is poor. Prompt biopsy of a rhabdomyosarcoma followed by a combination of chemotherapy and irradiation offers the best chance of survival. In fact, recent reports suggest that current treatments offer greater than 90% survival from rhabdomyosarcoma.

Patients will develop problems typically seen after chemotherapy and irradiation of the eye, but if there is no recurrence after 3 years, it is likely that the rhabdomyosarcoma has been controlled.

Optic Nerve Sheath Meningioma

Orbital and optic nerve meningioma can extend from the brain into the orbit (behind the eye) and push the eye forward causing a bulging of the eye called proptosis.

Patient with orbital meningioma typically have proptosis (bulging eye). Optic nerve compression can cause optociliary shunt vessels to form and loss of vision. Depending on the location, size and degree of optic nerve involvement; patients can develop monocular and/or junctional visual field defects.

In making this diagnosis, one should look at the optic nerve blood vessels, and for dilated retinal veins. The nerve head can appear raised. Enlarged blood vessels are called "optociliary shunt vessels" and indicate that the meningioma has disrupted the natural circulation through the optic nerve to the retina and choroid. Ultrasonography can be used to measure the optic nerve sheath diameter.
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The picture is taken through the pupil and is a standard way to examine and document the retina's and optic nerve's appearance. Dilated varicose veins are seen around the edges of the optic nerve disc, these are optociliary shunt vessels.

Computed tomography (CT) shows that the eye is pushed forward by the meningioma. Notice that the tumor looks like a bright spot that wraps around the optic nerve (which measures 7.4mm), this effect is what causes the nerve to atrophy and die away.

MRI image shows an optic nerve stealth meningioma. Meningioma wraps around the nerve. The classic rail-trac appearance (white lines) surround the optic nerve core (dark gray).

Orbital meningioma is typically a slow-growing tumor. Once diagnosed, meningioma can be observed for growth prior to considering intervention. Treatment is indicated when there is a risk of spread to the central nervous system (in primary optic nerve sheath meningioma), documented progressive vision loss, or for rapid growth.

Though microsurgical resections have been tried (in an effort to spare the optic nerve), most eventually fail. The goal of local resection should be complete removal of the meningioma. This usually involves removal of the involved optic nerve. Whenever complete surgical removal is not possible, radiation therapy is commonly employed.

Metastatic Tumors

Metastasis is the medical term used to refer to tumors (mostly cancerous) that have spread to the rest of the body. Metastasis is an important cause of orbital disease in the adult, representing approximately 8% of all orbital tumors. Orbital metastases may signal either reactivation of treated disease or new systemic malignancy. Breast carcinoma is the most common metastatic tumor found in women followed by lung carcinoma. In men the most common are lung and prostate. The common presenting symptoms are proptosis, diplopia, pain and vision loss. Physical examination may reveal upper eyelid ptosis and a palpable mass. The average age at presentation is in the 7th decade, most being female (due to the higher incidence of breast metastasis - 50% of all metastasis). On CT, the most common finding is a well-defined, contrast enhancing, intraconal mass.

The orbital metastasis can locate at the bony walls themselves. Prostate cancers especially. Biopsy may be necessary for diagnosis and the prognosis with orbital metastasis of systemic cancer is very poor (avg. survival - 10 months). Radiation therapy is the usual modality of treatment for orbital metastasis with chemotherapy and hormonal therapy occasionally used.
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CT scan of the orbit shows an ovoid mass deeply located within the orbit. The mass seems to be located with the medial rectus muscle. Biopsy proved to be a metastasis breast tumor.

Complex Cranio-Orbital Tumors

All too often orbital tumors invade back into the cranial cavity into the brain. On the contrary, skull brain tumors can also invade the orbit. A wide variety of tumors can produce skull invasion and vice versa. Complex microsurgical skull base approaches are necessary to safely remove such tumors. Neurosurgical skills and training are desirable when dealing with this kind of tumors. If needed, the orbit surgeon may seek neurosurgical consultation.

A middle-aged woman presents with a blind eye and a droopy eyelid.

Orbit CT scan showed a rather large orbit-cranial tumor. Skull base invasion affected temporal cranial fossa, lateral cavernous sinus, and disrupted into the orbit, destroying its bony walls.

Black-empty area remains after total resection of tumor.