Competencies in Oculoplastics

By Don O. Kikkawa, MD

As part of the “Road to Recertification” article series in New Retina MD, Don O. Kikkawa, MD, provides an overview of some of the topics that retina specialists may want to review in more detail for Maintenance of Certification. As with every article in this series, Dr. Kikkawa’s overview is not meant to take the place of a comprehensive review course; rather, its purpose is to highlight some key areas within the oculoplastics subspecialty and to encourage a more thorough review prior to taking the Demonstration of Ophthalmic Cognitive Knowledge exam.

- Diana V. Do, MD

ORBIT

Orbital anatomy

The orbit consists of 7 bones: ethmoid, sphenoid, maxilla, palatine, zygoma, lacrimal, and frontal. The sphenoid contributes to all of the orbital walls except the floor. The roof consists of the frontal and the sphenoid; the lateral consists of the zygomatic and the sphenoid; the floor consists of the maxilla, the zygoma, and the palatine; and the medial wall consists of the maxilla, the lacrimal, the ethmoid, and the sphenoid.

The volume of the orbit is 30 ccs. The orbital width is greatest approximately 1 cm behind its anterior most surface. The distance from orbital rim to the orbital apex is approximately 40 mm. The medial orbital walls are parallel and if both lateral walls were to extend posteriorly, they would intersect at a 90º angle.

The majority of the orbital volume consists of orbital fat. Orbital fat is derived from neural crest origin. Deeper orbital fat has a more whitish color while the more anterior pre-aponeurotic fat is more yellow. Clinically, the upper eyelid has 2 fat pads, divided by the trochlea, while the lower lid has 3 clinically apparent fat pads. The medial fat pad in the lower lid is separated from the central fat pad by the inferior oblique muscle and the central fat pad is separated from the lateral fat pad by the arcuate expansion of the inferior oblique muscle.

Most of the neurovascular structures enter the orbit within the superior and inferior orbital fissures, except the optic nerve, which enters through the optic canal. The extracranal portion of the superior orbital fissure transmits the lacrimal nerve (sensory), the frontal nerve (sensory), the trochlear nerve (CN IV), while the intracranal portion transmits the superior and inferior divisions of the oculomotor nerve (CN III), the nasociliary nerve (sensory), and the abducens nerve (CN VI). The inferior orbital fissure transmits the infraorbital and zygomatic nerves (sensory).

Evaluation of Orbital Disorders

Orbital signs and symptoms are best evaluated by the 6 P’s: pain, progression, proptosis, periorbital changes, palpation, and pulsation. Although pain is suggestive of inflammatory disorders or infection, perineural invasion of adenoid cystic carcinoma can also cause pain. Rapid progression also suggests infection or inflammation while tumors grow more slowly. Proptosis is measured with an exophthalmometer and is clinically significant if more than 2 of relative difference between eyes. Gender and ethnic variations exist. Periorbital changes can give clues to the underlying diagnosis. Skin discoloration, such as the strawberry nevus of capillary hemangioma are helpful. Palpation can also aid in orbital diagnosis, for example the “bag-of-worms” feel of a plexiform neurofibroma. Pulsation can be seen in arteriovenous fistulas and also in neurofibromatosis with absence of the greater wing of the sphenoid.

Imaging is recommended in the diagnosis of orbital disease. CT imaging is useful as an initial study. It also gives good examination of the orbital bones and sinuses. MRI is preferred for soft tissue detail and imaging at the orbital apex and intracranial region just posterior to the orbit. Ultrasound requires an experienced operator, but can be useful to dynamically examine the posterior globe and extraocular muscles widths. Doppler studies aid in examination of blood flow velocity and course.

Laboratory testing is crucial in cases of suspected thyroid related orbitopathy (T3, T4, and TSH), vasculitis, and Wegener’s granulomatosis (c-ANCA). Angiotensin converting enzyme (ACE) and lysozyme can help detect sarcoidosis.
Infections/Inflammatory Disorders

Preseptal cellulitis causes swelling and erythema in the eyelids with sparing of the orbit and globe. Oral antibiotic therapy is typically effective. Orbital cellulitis is the most common cause of proptosis in children. Imaging is required for diagnosis. It occurs primarily as spillover from infection in the sinuses. Proptosis, limitation in extraocular motility, ptosis and erythema of the eyelids may occur. In children, it is most often caused by a gram-positive organism but adults can have mixed bacterial causes. Parenteral antibiotics are indicated. Clinical course and imaging should be followed. If abscess formation is noted in adults, urgent drainage is indicated. In children, the literature supports careful observation of medial and inferior abscesses in children younger than 9 years of age.

Other types of infection in the orbit include fungal and parasitic causes. Invasive fungal disease typically occurs in immunocompromised patients or poorly controlled diabetics (mucor). Aspergillus can invade the orbit in healthy adults, but a more common presentation is allergic fungal sinusitis with secondary orbital involvement. Treatment usually consists of wide debridement in invasive disease with adjunct antifungal therapy, while allergic disease is treated with corticosteroids. Parasitic infections are rare but include cysticercosis and Echinococcus granulosus. Cysticercosis typically involves the extraocular muscles and Echinococcus presents as a hydatid cyst.

Thyroid eye disease (TED) is the most common cause of proptosis in adults. It is caused by a poorly understood autoimmune process. Deposition of extracellular matrix in the orbital tissues causes swelling with some patients also developing fat hypertrophy. Orbital women have a 6:1 predilection for the disease. Smoking is not causal but is associated with worsening of disease. Eye findings include: proptosis, eyelid retraction, restrictive strabismus, chemosis, conjunctival injection, eyelid edema and erythema, and optic neuropathy in severe cases. Approximately 90% of patients with eye findings are hyperthyroid with the remaining 10% being either euthyroid or hypothyroid. CT imaging can reveal enlarged extraocular muscles and/or enlarged fat compartments in the orbit. The most commonly enlarged extraocular muscles are the inferior followed by the medial rectus. The tendon is typically spared. Treatment is based on symptoms and severity of disease. Patients with optic neuropathy require urgent treatment with steroids followed by orbital decompression if resistant. In patients requiring elective surgical intervention, a staged approach is recommended (with not all stages being required) in order: orbital decompression, strabismus surgery, and lid surgery.

Nonspecific orbital inflammation, formerly known as pseudotumor, is an autoimmune benign inflammatory process. It can have varying presentation but is likely to be acute and includes swelling, pain, proptosis and erythema. Imaging may show soft tissue swelling of the lacrimal gland, extraocular muscles, anterior orbit, or orbital apex. If a myositic pattern is present, extraocular muscle involvement typically involves the tendon. In contrast to TED when the extraocular muscles are involved, it is usually in the field of gaze of the muscle, showing paresis and not restriction in the opposite field of gaze (as seen in TED). After ruling out other causes, steroids are helpful to reduce symptoms but recurrences are common. Biopsy is recommended to rule out other causes such as lymphoma.

Periorbital Cysts and Orbital Neoplasms

Rhabdomyosarcoma most commonly affects children between the ages of 8 to 10. It is the most common malignant orbital tumor in children. There is a predilection for the superior orbit with bony involvement. Urgent biopsy is recommended in cases with suspected rhabdomyosarcoma. Pathological subtypes include embryonal (most common), alveolar (most malignant), pleomorphic (least common), and botryoid. Multimodality treatments with radiation and chemotherapy have improved survival rates.

Dermoid cysts most commonly present in the superior lateral orbit in children. They arise at the frontozygomatic suture and are palpable as discrete lesions. Imaging can occasionally reveal a dumbbell-shaped dermoid with the deeper portion of the mass extending through the lateral wall of the orbit. Excision is performed through a lid crease incision. Lateral orbitotomy may be required if deeper extension is seen.

Neural tumors include neurofibromas, glioma, meningioma, and schwannoma. Plexiform neurofibroma is commonly seen in NF 1 and can affect the eyelids and orbit. Typically, this produces an S-shaped lid and is palpable as a “bag of worms.” Other signs of NF 1 included pulsatile proptosis due to greater wing of the sphenoid dysplasia. Optic nerve glioma is most commonly seen in children and also is associated with neurofibromatosis. Proptosis and vision loss are common. Imaging reveals fusiform enlargement of the optic nerve. Treatment is mainly observation, but surgical excision can be considered with rapid growth and threatening of the optic chiasm. Meningioma is seen more commonly in adults. They can arise in either the optic nerve sheath or the dura. Lesions in the sphenoid ridge cause proptosis and temporal fossa swelling. CT imaging of optic nerve meningioma can show calcification and the typical railroad track sign, where meningioma of the sphenoid ridge shows hyperostosis with contrast enhancement of the involved soft tissue component. Treatment for optic nerve meningioma is observation if vision is good but fractionated stereotactic radiation can be considered if growth and vision loss occur. Craniotomy is indicated in sphenoid ridge meningioma if vision loss and significant proptosis are present. Schwannomas are proliferations of Schwann cells. Pathologically they can have two patterns Antoni A (solid) and Antoni B (myxoid). They are typically...
well circumscribed and can be removed if symptomatic with decreased vision or significant proptosis.

The most common vascular lesions in the orbit include hemangioma, lymphangioma, hemangiopericytoma, arteriovenous lesions and varices. Capillary hemangiomas typically present in the first few months of life. They typically exhibit rapid growth during the first year of life then slowly involute. Associated syndromes include the Kasabach Merritt syndrome with visceral lesions and thrombocytopenia. Vision threatening and disfiguring lesions can be treated medically with propranolol. Steroid injections and surgery are second line treatments. Cavernous hemangioma is the most common benign tumor in adults. They typically present in women in middle age. CT imaging typically shows a well-circumscribed lesion. Complete excision is recommended if functional deficits occur. Lymphangiomass are hemodynamically isolated lesions that do not enlarge with Valsalva. They can enlarge during upper respiratory illness. They can have spontaneous hemorrhage with associated sudden onset proptosis. Imaging can show infiltrative lesions with cystic spaces. Treatment should be conservative, unless vision threatening hemorrhage occurs. Preventive treatment with intracystic sclerosing agents shows promise. Hemangiopericytomas are also well circumscribed on CT imaging. Complete excision is often difficult because lesions are friable. Malignant transformation can occur. Arteriovenous fistulas can be either high flow (carotid cavernous) or low flow (dural cavernous). Signs and symptoms include proptosis, dilated conjunctival vessels, and elevated intraocular pressure. Imaging can show a dilated superior ophthalmic vein and enlarged extraocular muscles. Treatment involves closure of the fistula with interventional radiology techniques. Varices are low flow lesions that massively enlarge with Valsalva. Treatment is symptom specific. Observation is typical unless vision is threatened or pain and disfigurement is severe.

Lymphoid lesions commonly present with an anterior mass in the orbit. There may be an associated salmon patch lesion of the conjunctiva. CT imaging may reveal an orbital mass that molds around orbital structures. Biopsy should be performed with immunohistochemical staining and flow cytometry. Systemic work up is mandatory. Chemotherapy and/or radiation are often helpful.

Secondary tumors can affect the orbit. They can be either metastatic or contiguous from adjacent orbital structures. Common metastases to the orbit in adults are: breast (causing enophthalmos or muscle enlargement), prostate (orbital bony involvement), and lung. Metastatic neuroblastoma in children often causes ecchymosis to the inferior orbit. Tumors adjacent to the orbit can arise from the sinuses (squamous cell carcinoma and sinonasal undifferentiated carcinoma), the nasopharynx (nasopharyngeal carcinoma), eyelids (basal and squamous cell carcinomas), and globe.

Orbital Trauma

The orbit can be affected by trauma in many ways. Both bony and soft tissue injury can occur. Clinical presentation and treatment vary based on cause and findings.

Most common bony injuries are fractures. Several types of fractures involve the orbit. The most common are blow out fractures. Other fractures include tripod or zygomatic fractures, Le Fort fractures, and nasoethmoidal fractures. Orbital apex fractures involving the optic canal can cause vision loss from direct optic neuropathy.

Blow out fractures occur when an object strikes the orbital entrance. Two theories have been proposed: 1) the indirect hydraulic theory with posterior pressure in the orbit causing a blow out and 2) the direct buckling theory causing the possibility of a blow out as well as a blow in fracture. Clinical symptoms include diplopia due to inferior rectus muscle entrapment, enophthalmos, infraorbital nerve hypesthesia, and hypoglobus. CT imaging with axial and coronal thin cuts provide the best views for diagnosis and treatment decision making for this condition.

Indications and timing of repair are controversial but urgent indications include: the white-eyed blow fracture in children where the fracture site incarcerates the rectus muscle causing possible ischemia and in those cases where the oculocardiac reflex is caused by muscle entrapment. Other indications for repair within 2 weeks are: diplopia within 30° of primary associated with a positive forcedduction test, clinically significant enophthalmos greater than 2 mm, and a fracture size greater than 50% of the orbital floor. Repair typically is via a transconjunctival approach with elevation of orbital contents from the fracture site and placement of an alloplastic implant.

Tripod or zygomatic fractures affect the orbit by also causing orbital floor fractures but with a different mechanism. The zygoma becomes fractured along the frontozygomatic, zygomatic-maxillary, and zygomatic-temporal suture lines. Additionally there is involvement of the zygomatic maxillary buttress. Malar eminence flattening and lateral canthal dystopia can occur in displaced fractures. Le Fort II and III fractures also involve the orbit, with Le Fort III fractures having a higher incidence of optic neuropathy. Nasoethmoidal fractures cause flattening of the nasal bridge and telecanthus with possible lacrimal system involvement. Early intervention is warranted in patients with significantly displaced fractures, as deformity can be severe if untreated.

Indirect optic neuropathy can occur after blunt trauma to the orbit or forehead. It is thought that deformation of the optic canal occurs with external force that leads with shearing and swelling of the intracanalicular portion of the optic nerve. Vision is reduced as well as presence of an afferent pupillary defect. Controversy exists as to the best treatment. Both high dose steroids and optic canal decompression have been advocated, but a multicenter trial did not show either to be beneficial.
Soft tissue injuries of the orbit can lead to contusion and hemorrhage both within the orbit and eyelids. Most contusion and hemorrhage can be observed as long as vision and intraocular pressure (IOP) are stable. In cases of retrobulbar hemorrhage, an orbital compartment syndrome can occur leading to vision loss. Urgent intervention with canthotomy and cantholysis can be useful at the bedside with operative intervention necessary if vision and IOP do not normalize. Orbital foreign bodies can be organic or nonorganic. All organic foreign bodies should be removed, as there is a high potential for infection and inflammatory foreign body reaction. Metallic foreign bodies can be tolerated if not causing functional problems.

**Eyelids**

**Surface Anatomy and Dimensions**

Normal eyelid contour has the highest peak of the upper lid just nasal to the pupil and the lower point of the lower lid just lateral to the pupil. The lateral canthus normally rests 1-2 mm higher than the medial canthus. This is referred to as a mongoloid slant. If the lateral canthus is lower than the medial canthus this is referred to as an antimongoloid slant. The normal horizontal palpebral width is approximately 30 mm with the intercanthal width being approximately 30 mm as well. Widening of the intercanthal distance is referred to as telecanthus.

Vertical lid position is measured by the margin-to-reflex distance (MRD), using a light source with the reflex being centered in the pupil and the distance of the lid margin being measured in mm from the reflex. Normal levator function or lid excursion is 15 mm.

**Eyelid Disorders**

*Ptosis.* By definition, ptosis exists when the MRD is less than +2. Several categories of ptosis exits: myogenic, aponeurotic, synkinetic, neurogenic and mechanical. Congenital ptosis is likely myogenic in nature due to dysgenesis of the levator palpebrae superioris muscle. Acquired ptosis is likely caused by aponeurotic dehiscence or thinning. Marcus Gunn jaw winking ptosis is a type of synkinetic ptosis caused by aberrant innervation of CN V to the lateral pterygoid and branches of CN III to the levator. CN III palsy causes neurogenic ptosis likely with poor supra, infra and adduction. Capillary hemangiomas and plexiform neurofibromas cause mechanical ptosis by a mass effect and interference with levator excursion. Myasthenia gravis (MG) can also cause ptosis and should be ruled out. Variability, Cogan’s lid twitch, and fatigue are suggestive of MG.

Levator function should be measured in every ptosis patient. Levator function is divided into 3 categories: 0-5 poor; 5-10 fair; and 10-15 good. Repair of ptosis is dependent upon several factors: age of patient, symptoms, degree of ptosis and levator function. Symptoms include blockage of the superior visual field and interference with the performance of daily activities. Congenital ptosis should ideally be repaired in the preschool years unless amblyopia develops. Visual field testing both with and without taping of the lid should be documented. Protective mechanisms should also be tested, including tear production, orbicularis function, and presence of Bell’s phenomenon.

Type of repair is predicated upon levator function. In cases of good function, levator advancement is typically indicated. For moderate function, levator resection is indicated. For poor function, a maximal levator resection or frontalis suspension is performed. For mild amounts of ptosis, posterior approach conjunctival Muellerectomy can be effective.

*Entropion and ectropion.* Entropion is an inward turning of the eyelid margin. It can be either involutional or cicatricial in nature. Symptoms include foreign body sensation, tearing and ocular irritation.

Involuitional entropion is caused by 3 factors: horizontal lid laxity, disinsertion of the lower lid retractors, and override of the preseptal portion over the pretarsal portion of the orbicularis muscle. Cicatricial causes include: trachoma, ocular cicatricial pemphigoid, alkali burns, ocular surface neoplasia, and chronic use of topical ocular medications. Treatment depends on the underlying cause. Involuotional cases are repaired with lid shortening and reinsertion of the lid retractors. Cicatricial cases are typically treated by margin rotation or posterior lamellar grafting after malignancy has been ruled out.

Ectropion is an outward turning of the lid. It also has involutional and cicatricial causes. Symptoms mainly include tearing and ocular irritation. Involuotional ectropion is typically caused by poor orbicularis tone, lid laxity and retractor disinsertion. Cicatricial ectropion is typically caused by anterior lamellar forces causing the lid to rotate outward and downward. This could be caused by trauma-induced scarring, actinic skin damage, or prior eyelid surgery from skin cancer or blepharoplasty. Treatment includes lid tightening with or without retractor reinsertion for involutional cases and scar release with or without skin grafting in cicatricial cases.

*Lid retraction.* When the lid retracts above the limbus in the upper lid or below the limbus in the lower lid, retraction is present. Another term for this is *scleral show.* Lagophthalmos (incomplete lid closure) may also result from lid retraction. The most common cause of lid retraction is thyroid related orbitopathy. Other causes include dorsal midbrain syndrome (Collier’s sign), uncompensated contralateral ptosis, and previous blepharoplasty.

*Lid lesions.* Most lid lesions are benign. The most common lid lesions are chalazion, external hordeolum, papilloma, seborrheic keratosis, and actinic keratosis. Chalazion is a granulomatous inflammation within the Meibomian glands and is typically treated initially with warm compresses, with incision and curettage being reserved for chronic lesions. External hordeolum typically arise from the gland of Zeis and respond also to warm compresses. Papillomas are...
pedunculated proliferations of squamous epithelium while seborrheic keratosis arises from epithelial proliferation with horn cysts. Actinic keratosis is considered a precancerous lesion that can give rise to squamous cell carcinoma.

Malignant neoplasms do occur on the lid with increasing frequency with the aging population. Basal cell carcinoma is the most common, followed by squamous cell carcinoma, sebaceous cell carcinoma and malignant melanoma. More rare types of malignancies include Merkel Cell carcinoma, the violaceous appearing tumor of neuroendocrine origin.

Basal cell carcinoma, the pearly tumor caused by proliferation in the basal cell layer of the epidermis can clinically be divided into nodular, morpheaform, basosquamous and pigmented. Certain syndromes, such as basal cell nevus (Gorlin-Goltz) syndrome and xeroderma pigmentosum, have a high incidence of this tumor. Metastasis is rare.

Squamous cell carcinoma is cause by uncontrolled cell growth in the stratum corneum. It is more common on the upper lid and can metastasize via lymphatic spread. It typically has a keratinized appearance on its surface.

Sebaceous cell carcinoma can arise from the meibomian glands, the gland of glands, or sebaceous glands located in the caruncle. Clinical presentation can vary from a chalazion appearing lesion to chronic unilateral blepharitis. Histopathology with oil staining is recommended for diagnosis. Metastasis can occur.

Malignant melanoma is a typically pigmented tumor arising from melanocytes. Worrisome clinical findings include lesions of increasing size irregular pigmentation and irregular borders. Staging is based on depth of lesion invasion. Sentinel lymph node dissection can be useful for staging and prognosis. Metastasis and death can occur.

Treatment of lid lesions varies. Benign lesions can be observed unless symptomatic or affecting lid function. Treatment of malignant lesions typically involves surgical excision with either frozen or permanent sections. Moh’s micrographic technique can be useful for basal and smaller squamous cell carcinomas. Sebaceous cell carcinoma and melanoma typically require larger margins and permanent sections for accurate margin control.

Eyelid Reconstruction

Eyelid reconstruction is often required in cases after tumor removal and trauma. It is dependent on the size of the defect, the depth of involvement (either anterior lamella, posterior lamella or both), the involvement of the lacrimal system, and whether either canthus is involved. There is certainly practitioner preference and bias involved with lid reconstruction, and there may be more than 1 way to rebuild the eyelid. Certain common principles, however, do apply.

Common principles are:
1) Either the posterior or anterior lamella must provide the vascular supply. You cannot place a free graft on the bed of a free graft.
2) If possible, anterior lamella is best supplied by adjacent tissue transfer. If inadequate, free full thickness skin grafts and be used from following: upper lid, retroauricular, supraclavicular, and inner upper arm.
3) Posterior lamellar grafts can be used from the upper tarsus, ear cartilage, hard palate or nasal chondromucosa.
4) Lid-sharing procedures should be avoided in children in the amblyogenic age.

Defects less than 25% of either the upper lid or lower lid margin can be closed primarily. A lateral canthotomy and cantholysis can add additional length if needed. For defects between 25-50% percent of the lid, a Tenzel semicircular advancement flap is my preferred method for both upper and lower defects.

For defects greater than 50% of the lid margin of the lower lid, a Hughes tarsocconjunctival flap with full-thickness skin graft is preferred. The flap can be opened as early as 2-3 weeks. A Mustarde cheek rotational could also be used with a posterior lamellar graft, such as a free tarsal graft. For defects greater than 50% of the upper lid, a Cutler-Beard pedicle or Leone flap (tarsocconjunctival flap from the lower lid with full thickness skin graft) are preferred.

Lacrimal System

Anatomy

The lacrimal system is broadly divided into secretory and excretory systems. The lacrimal secretory system consists of the lacrimal and accessory lacrimal glands.

The lacrimal excretory system can be divided into the upper and lower systems. The upper system consists of the puncta and canaliculi. The lower system consists of the lacrimal sac and nasolacrimal duct.

The lengths of the lacrimal system components are as follows: puncta, 2 mm; canaliculi, 8 mm; lacrimal, sac 10 mm; and nasolacrimal duct, 12 mm. The nasolacrimal duct exits beneath the inferior turbinate. Classicly the valve of Rosenmuller exists at the level of the common canaliculus and the valve of Hasner at the opening of the nasolacrimal duct.

The main lacrimal gland resides in the superotemporal quadrant of the orbit just behind the superior orbital rim. The lateral horn of the levator divides the gland into orbital and palpebral lobes. The ductules of the lacrimal gland pass closely on the surface or within the palpebral lobe to drain into the superolateral fornix. The accessory gland of Wolfring and Krause are located at the superior tarsal border and upper fornix respectively.

Lacrimal System Disorders

Upper system abnormalities involve the punctum and canaliculus. Punctal abnormalities can be divided into congenital and acquired problems. Congenital abnormalities of the puncta are punctal atresia and an imperforate puncta.
In punctal atresia, there is no formed opening or mound where the puncta is normally formed. Typically, there is canalicular atresia as well. Imperforate puncta is a condition where there is a thin membrane covering the opening of the puncta with a fully formed upper system. Opening of the membrane is typically curative.

Canalicular obstruction can have many causes. Infection, topical medications, trauma and idiopathic causes are the majority. Canalicitis is an infection within the canalculus most often caused by Actinomyces. Curettage is generally effective in relieving the obstruction. In cases of both canalicular obstruction and canalicular atresia, treatment consists of conjunctival dacryocystorhinostomy with Jones tube (DCR).

Lacrimal sac disorders include infections and neoplasms. Disorders of the lacrimal sac usually cause swelling. The classic teaching is that if the swelling is below the medial canthal ligament, this is indicative of infection. If swelling is above the ligament, neoplasm is the most likely cause.

Dacryocystitis is infection within the lacrimal sac. Acute dacryocystitis causes swelling, erythema and tenderness, is most often caused by gram-positive bacteria. Chronic dacryocystitis can cause mucoid discharge and swelling, but there is typically no erythema or tenderness. The underlying cause is nasolacrimal duct obstruction. In the case of acute dacryocystitis, the valve of Rosenmuller allows inflow but not outflow into the lacrimal sac. Chronic dacryocystitis, there is some incompetence of the valve with reflux being present with external pressure. Acute dacryocystitis requires systemic antibiotics to treat the infection followed by dacryocystorhinostomy (DCR) to relieve the obstruction. Patients with chronic dacryocystitis benefit directly from DCR.

Nasolacrimal duct obstruction (NLDO) can either be congenital or acquired. Congenital NLDO is reported to occur in up to 20% of newborns. Symptoms include epiphora, mucoid discharge, and conjunctivitis. Conservative management is external massage and short-term topical antibiotics. NLDO probing should be performed if symptoms do not resolve before one year. If probing fails, the next treatment is typically silicon stent placement and if still unsuccessful, followed by DCR.

Acquired NLDO is typically idiopathic in nature and consists of chronic inflammation in the duct, followed by eventual fibrosis. In the absence of dacryocystitis, symptoms are likely to be epiphora only. In early stages, partial or functional NLDO can be treated with silicone stent placement and eyelid tightening but in late stages only likely to be improved by DCR. It is important to note that NLDO can be caused by intranasal and sinus pathology and these causes must be ruled out before proceeding with DCR.

Lacrimal gland disorders can also have multiple causes. Acute dacryoadenitis causes tenderness, swelling, and fullness of the lateral upper eyelid and orbit. There may be expressible purulence from the lacrimal gland. Imaging is recommended for all suspected lacrimal gland disorders. Systemic antibiotics are generally curative with infective dacryoadenitis.

The differential diagnosis for chronic swelling of the lacrimal gland is larger. It includes inflammatory conditions, lymphoid disorders and neoplasia. Imaging provides some clues as to the diagnosis. The bony lacrimal gland fossa should be closely examined for destruction. Malignant tumors, such as adenoid cystic carcinoma and pleomorphic adenocarcinoma, cause bone destruction with invasion of cortical bone. Long-standing slow enlargement of the gland can cause bone erosion and remodeling but in general, not destruction. A globular or round enlargement of the gland is suggestive of a pleomorphic adenoma, whereas molding of the gland around the globe and orbit is more likely to be caused by lymphoma. Inflammatory disorders, such as nonspecific orbital inflammation, are likely to show involved surrounding soft tissue involvement.

Treatment varies on the underlying cause. Lacrimal gland lesions should be biopsied prior to instituting treatment. Anterior orbitotomy with incision biopsy is useful for most lesions, but complete excisional biopsy should be considered if pleomorphic adenoma is suspected. Nonspecific orbital inflammation is best treated with oral corticosteroids with radiation being reserved in cases of treatment resistance. Patients with lymphoid lesions, including lymphoma, should have a medical workup looking for systemic involvement, which occurs in roughly 50% of cases. Most cases of orbital lymphoma are B-cell lesions that are CD 20 positive. With isolated orbital involvement, radiation can be considered, but if systemic involvement, chemotherapy or targeted B-cell therapy with rituximab is required.

Malignant tumors typically required complete tumor excision. Bony removal orbital exenteration may be required. Adjunct external radiotherapy will likely be required in addition to surgical removal.

SOCKET AND FORNICES

Globe removal should be reserved for cases of severe trauma, malignancy, and pain control in a non-seeing eye. Improved cosmesis can also be an indication but must be considered with the desires of the patient and concern to salvage the globe. Options for globe removal include evisceration and enucleation. Exenteration is an operation typically reserved as a life-saving procedure in cases of malignancy. Evisceration involves removal of the globe contents with preservation of the sclera. The cornea can be either spared or removed. Indications include trauma and pain relief, but this operation should not be performed in cases of suspected malignancy. After removal of all intraocular contents, an orbital implant is placed within the scleral remnant and closure of the sclera. Tendon’s capsule, and conjunctiva is performed in multiple layers. Prosthetic fitting is performed after several weeks. With evisceration, there is a miniscule risk of sympathetic ophthalmia with several case reports in the literature.

Enucleation is considered the procedure of choice in cases of suspected intraocular malignancy and severe globe trauma.
After conjunctival peritomy, the muscles are detached from the globe and the optic nerve is severed. An orbital implant is then placed within the muscle cone. Orbital implants vary in composition and size. Nonporous implants include silicone and polymethyl methacrylate. Porous implants include porous polyethylene, hydroxyapatite and bovine demineralized bone. Porous implants have the advantage of fibrovascular ingrowth and improved implant motility. Integrated peg placement into the implant with prosthetic coupling has been described but does risk implant exposure and infection.

Orbital exenteration involves removal of all orbital contents, including the globe, extraocular muscles and orbital fat. This is a disfiguring operation and should be reserved for life threatening malignancy or infection. Portions of the bony orbit may be removed if involved with the disease process. Reconstruction depends on the disease and time frame after surgery. Disease surveillance may be hindered if free tissue flaps are placed into the orbit but provide a viable option if the underlying disease is under control. Split thickness skin grafting allows for faster healing but secondary intention should be considered in cases of infection (invasive fungal infection). Orbital exenteration prostheses can be fit into the bony orbital cavity with limitations of static eyelids and lack of prosthetic movement.

Socket contraction with shortening of the fornices can occur with any socket procedure. The end result is inability to retain an ocular prosthesis. Examination of the contracted socket should be focused on 2 aspects, volume and surface area. If volume is deficient, volume replacement can be performed with additional alloplastic implants with either implant exchange or subperiosteal augmentation. If surface area is deficient, additional surface area can be added with mucosal membrane grafts or hard palate mucosa. If both are deficient, dermis fat grafting may be useful. Severe socket contraction typically results from scarring from trauma or multiple operations. Success of reconstruction and ultimate prosthetic retention may be limited in these cases.


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