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RESIDENT PHYSICIAN: Michael Underbrink, MD

FACULTY PHYSICIAN: Shawn Newlands, MD

SERIES EDITORS: Francis B. Quinn, Jr., MD and Matthew W. Ryan, MD

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INTRODUCTION

Although the orbital area is generally considered within to fall within the domain of the ophthalmologist, many orbital related disease processes require the involvement of other subspecialties. Some related orbital disease processes, such as paranasal sinus disease, may demand the involvement of the otolaryngologist. In some cases, patients with primary orbital lesions may present initially to the ENT clinic. It is therefore, imperative that we possess a basic understanding of the complex anatomy and potential pathology of the orbit.

Anatomy

The orbit is a pyramidal or cone-shaped space comprised of seven bones (frontal, greater and lesser wings of the sphenoid, zygoma, maxilla, lacrimal, palatine, and ethmoid). The inferior wall, or orbital floor is composed mainly of the orbital plate of the maxilla, the zygomatic bone anterolaterally, and the orbital plate of the palatine bone posteriorly. The inferior orbital fissure separates the floor from the lateral wall, providing passage for the maxillary division of CNV, the infraorbital artery, branches from the sphenopalatine ganglion, and branches of the inferior ophthalmic vein to the pterygoid plexus. The lateral wall is composed of the frontal process of the zygoma and the greater wing of the sphenoid lateral to the optic foramen. Along the lateral wall, we find Whitnall's tubercle, a bony prominence just deep to the rim and just above the midpoint of the lateral wall, where the lateral canthal ligament attaches. The superior orbital fissure demarcates the lateral wall from the orbital roof. The major orbital nerves and vessels pass through here (except the optic nerve and ophthalmic artery which pass through the optic foramen). Posteriorly, the superior orbital fissure communicates with the cavernous sinus and middle cranial fossa. The superior orbital wall, or orbital roof, is comprised of the orbital plate of the frontal bone with a small contribution, posteriorly, from the lesser wing of the sphenoid. Anterolaterally, the fossa of the lacrimal gland creates a shallow depression, and medially, there is a spine or depression for the trochlea about 5mm deep to the rim. The medial wall is composed of the frontal process of the maxilla, the lacrimal bone, the lamina papyracea of the ethmoid, and a portion of the lesser wing of the sphenoid. Posteriorly, due to its somewhat eccentric placement (medial and superior to the geometric apex), the optic foramen is related to the posterior ethmoid air cells. The foramina for the anterior and

posterior ethmoid arteries lie within the frontoethmoid suture line. Anteriorly, the lacrimal fossa can be located between the anterior and posterior lacrimal crests.

The orbit is compartmentalized by several interconnecting fasciae. The periorbital, or periosteum lining the orbital walls, is continuous with the dura mater at the optic foramen and superior orbital fissure. Several septae pass from the periorbital to divide the orbital fat into lobules. There is a potential space between the periosteum and the bony walls called the subperiosteal space. Continuous with the periorbital at the orbital rim is a fibrous sheet that extends across the entrance of the orbit called the orbital septum. It fuses with the levator aponeurosis in the upper lid and the sheath of the inferior rectus in the lower lid. A firm fibrous sheath, known as Tenon's capsule (bulbar fascia), surrounds the entire globe except for the cornea and separates the eye from other orbital contents. The episcleral space is a potential surgical plane between the bulbar fascia and the globe. Lastly, there is a dense muscular fascia created by the fusion of the fibrous sheaths of the extraocular muscles. Thickening of this fascia from the medial and lateral recti form the medial and lateral canthal tendons. Along with contributions from the fascia of the inferior rectus these fascia form a fibrous sling which supports the globe-the suspensory ligament of Lockwood. Within the muscular cone formed by this fibrous sheath is the central space. Outside the cone lies the peripheral space.

The blood supply for the majority of the orbit is via the ophthalmic artery, which branches off the internal carotid artery and traverses through the optic foramen with the optic nerve. It gives off numerous branches, including the anterior and posterior ethmoid arteries which exit at their respective foramina into the anterior cranial fossa and eventually enter the nose. Parts of the inferior orbit are supplied by the infraorbital artery, which is a branch off of the internal maxillary artery. Parallel to the arteries run numerous veins which eventually unite to form a superior and inferior ophthalmic vein. These vessels pass through the superior orbital fissure and empty into the cavernous sinus. A branch off the inferior portion passes through the inferior orbital fissure to communicate with the pterygoid plexus. There are no lymphatics or lymph nodes within the orbit.

Another important aspect of the orbit is the lacrimal system, which has a secretory and an excretory system. The secretory system is composed of the lacrimal gland (reflex secretor) and various basic secretors, which include conjunctival goblet cells, accessory subconjunctival lacrimal glands and oil secreting tarsal meibomian glands. The excretory system removes tears via the muscular contraction of the eyelids which drive the lacrimal secretions medially. There is a punctum medially in each lid, which empties into a canaliculus. These canaliculi empty into the lacrimal sac, located in the lacrimal fossa, which empties almost vertically into the nasolacrimal duct.

Evaluation

The approach to a patient with an orbital tumor must be centered on ophthalmic evaluation. A good history and thorough physical examination is essential to the development of a diagnosis. The history should include information about the onset, duration, and progression of the orbital disease process. The most important clinical manifestation of orbital disease is proptosis, however, this may not be the chief complaint. Often, the patient will first report changes in visual acuity, diplopia, eyelid droop/fullness, discharge, and/or pain. A history of allergies, sinus infections, epistaxis, nasal discharge or obstruction and tearing should be elicited to rule out a sinonasal origin for the disease process. It is also important to ask about other medical illnesses, such as thyroid disorders, granulomatous diseases, and autoimmune disorders.

The physical examination should pay careful attention to visual acuity, visual fields, pupillary responses, ocular motility and inspection of the external surface of the globes, eyelids and surrounding

structures. Exophthalmos can be most easily discerned by visualizing the eyes from above or below. Exophthalmos, or proptosis, is the most common sign of orbital disease, occurring with about 90 percent of primary orbital tumors. Protrusion of an eye more than 21 mm beyond the orbital rim or forward displacement of one globe 2mm greater than the other is clinically significant and should raise suspicion for an orbital disease process. Also, the direction of displacement of the proptotic eye may clue the clinician to a possible etiology. Displacement of the eye downward and laterally may indicate frontal or ethmoid sinus disease. Tumors of the maxilla will displace the globe upward. A purely axial displacement may indicate pathology within the muscle cone, such as an optic nerve glioma or Grave's ophthalmopathy. Medial displacement of the eye may indicate a lacrimal gland tumor or temporal fossa mass. Palpation of the orbital contents is important and may help identify anterior pathology. A complete head and neck examination is essential with focus on the nose, paranasal sinuses, nasopharynx and neck. Evaluation of cranial nerve function should also be performed.

Laboratory evaluation with a complete blood count, sedimentation rate and thyroid function tests should be obtained for evidence of infection, inflammation or thyroid disease. Defining the extent and localization of orbital tumors is most dependent on imaging techniques. Ultrasonography is a relatively inexpensive and safe method of evaluating the orbit. However, its use has been largely supplanted by computerized tomography and magnetic resonance imaging. It is still valuable as an office technique and as an adjunct to discriminate cystic, solid, angiomatic, and infiltrative tissues. CT scanning is currently the best available technique for detection and localization of the extent of orbital lesions. Coronal and axial views of the orbit should be obtained, which help to define the relationship of an orbital tumor to the optic nerve, extraocular muscles, blood vessels, globe, sinuses and brain. CT scans provide more information regarding bony landmarks, and are therefore, indispensable in surgical planning. Magnetic resonance imaging studies are particularly useful for evaluating possible intracranial extension of orbital tumors. MR imaging is also better for delineating vascular orbital lesions with the exception of orbital varices, which are easier to detect with coronal CT scan. Arteriography is useful for differentiating hemangiopericytoma from cavernous hamangioma (encapsulated venous malformations) and preoperative embolization of vascular tumors.

Pediatric Orbital Tumors

A child presenting with proptosis is usually very disconcerting to all parties involved. The pediatric patient with an orbital tumor differs substantially from the adult patient with a much greater incidence of congenital lesions, higher frequency of infection, and unique benign and malignant tumors involving the orbit. The most common orbital masses in children are cystic lesions of the orbit, mainly dermoids. Vasculogenic lesions are the second most common and include capillary hemangiomas, lymphangiomas, or cavernous hemangiomas. The remaining number of cases in any particular category is small, although a differential diagnosis should include inflammatory lesions, adipose-containing lesions, lacrimal gland masses, lymphoid tumors and leukemia, optic nerve and meningeal tumors, osseous and fibro-osseous masses, rhabdomyosarcoma, and metastatic lesions (most frequently neuroblastoma). The most common malignant processes include rhabdomyosarcoma, metastatic disease, lymphomas and leukemia. Most orbital tumors in children are benign and familiarity with the more common orbital lesions and their presentations will help the clinician make a timely and accurate diagnosis.

Cystic Lesions (Dermoid cysts, Teratoma)

Dermoid cysts likely arise from trapped embryonic ectoderm within the suture lines between the orbital bones. The sequestered tissue forms a cyst lined with keratinized epithelium and dermal elements. Cysts consisting of squamous epithelium without adnexal structures are called epidermoids. The orbital

dermoids can be classified into juxtasutural, sutural, and soft-tissue types. The most common type is the juxtasutural appearing in the superotemporal and superonasal quadrants. Clinically, this type of cyst presents in the preschool child as a painless mass in the superotemporal area at the lateral portion of the eyebrow. It is usually unattached to overlying skin, mobile, smooth and nontender. Soft tissue or sutural dermoids usually grow more slowly over a long period of time and often present with proptosis. Most patients have no visual symptoms. CT scan reveals a well circumscribed lesion with a low density lumen and sometimes bony remodeling. Deeper orbital lesions may show complete bony defects with calcium deposition. Occasionally, episodes of inflammation can be seen with small ruptures of the cyst wall. Management is surgical with complete excision of the cyst lining taking care to avoid rupture. Surgery is delayed until the risk of accidental rupture from trauma outweighs the general anesthesia risks, usually around 1 year of age. Smaller cysts can be followed for signs of growth, visual disturbances, or cosmetic deformity.

Teratomas are rare congenital germ-cell tumors which arise from primordial germ cells. These tumors are characterized by the presence of ectodermal, mesodermal and endodermal components. These tumors typically present at birth, and although benign with no bone invasion, often cause orbital enlargement with significant morbidity. With large intraconal masses, massive proptosis accompanied by conjunctival keratinization, exposure keratopathy and corneal ulceration may be seen. Massive teratomas traditionally are treated by orbital exenteration, however salvage of the globe is possible with smaller lesions (typically without subsequent normal visual acuity).

Vasculogenic Lesions (Capillary hemangioma, lymphangioma)

Capillary hemangiomas are common orbital masses in the pediatric population. One third are diagnosed at birth, and over 90% are visible by 6 months of age. The most common presentation is superficial involvement appearing as tumor and telangiectatic vessels in the skin that with time develops the typical strawberry-like appearance. Deeper lesions may appear as raised, soft, purplish nodules. Deep orbital involvement may present solely with proptosis and no skin changes. Orbital hemangiomas frequently produce proptosis and globe displacement and enlarge with Valsalva maneuvers or crying. The typical course of these lesions is a normal appearance at birth, with the lesion first noticed at one month and enlarging until the patient is 1 to 2 years of age. This is followed by a period of stabilization and then spontaneous involution by age 4 to 8 years of age. Long term cosmetic sequelae are minimal following involution though visual complications such as amblyopia and astigmatism from distortion of the globe are common. Major complications include superinfection, ulceration, and necrosis with possible hemorrhage. Rare but serious complications include Kasabach-Merritt syndrome and high-output cardiac failure (large lesions). Orbital involvement is best evaluated with CT or MRI which show a diffusely infiltrating non-encapsulated mass, conforming to the surrounding orbital structures. Bony erosion is not seen, although expansion of the orbital walls is possible. Ultrasonography is also a valuable noninvasive test. Lesions which do not affect visual or orbital development can be observed. Indications of treatment include any of the complications discussed previously. Medical therapy involves the use of intralesional steroid injection, systemic steroids, or interferon. Radiation therapy can be very effective in the control of these lesions, however concerns regarding radiation-induced malignancies in children has led to a reduction in its use. Surgical resection is reserved for well-circumscribed lesions or those causing severe sequelae unresponsive to medical therapy. Complete surgical removal is often not possible.

Lymphangiomas are benign congenital vascular malformations which may affect the conjunctiva, eyelids or deep orbit. Classically, they are viewed as separate from the vascular system, although some overlap has been noted. Typically, the tumor is identified within the first two decades of life. The course may present as slow enlargement with increasing proptosis over many years, or one of sudden proptosis from intralesional hemorrhage (chocolate cyst). These tumors have been known to show expansion during an acute upper respiratory infection. Superficial lesions are more common and have a better

prognosis for vision than deeper lesions. There is no enlargement of the tumor with Valsalva maneuvers. Imaging studies include CT and MRI, which both show the multi-compartmental nature of the venous-lymphatic malformations. MR imaging is preferred over CT because it delineates the internal structure of the various cysts of the lesion. The management of orbital lymphangioma is difficult due to the infiltrative nature of the tumor. Surgical debulking and cyst drainage is the treatment of choice for significant proptosis, corneal exposure or optic nerve compression. Complete surgical excision is often not possible.

Rhabdomyosarcoma

This is the most common orbital malignant tumor found in children. It presents early in the first decade with rapid unilateral proptosis and displacement of the globe. The eyelid may be erythematous with conjunctival chemosis. Occasionally ptosis, tearing, headache, and nosebleed are complaints. A palpable mass is present in about 25% and with large intraconal lesions papilledema and retinal vascular congestion may be seen. Sinusitis and epistaxis may occur with extension of the mass into the paranasal sinuses. CT scan shows an irregular tumor with moderately well-defined margins, soft tissue attenuation, and often evidence of bony destruction (50%). MR imaging demonstrates a signal similar to muscle on T1 and higher than muscle on T2-weighted images. A complete blood count is helpful to distinguish from orbital cellulitis and leukemia, and biopsy of a suspected rhabdomyosarcoma should be performed as soon as possible for a definitive diagnosis. During biopsy, as much tumor as can be removed without violating critical structures should be excised. Histologically, it can be divided into four main types: embryonal, alveolar, pleomorphic, and botryoid. The embryonal type is the most common, occurring in 2/3 of cases. The alveolar is second most common and is the most malignant with a high frequency of metastases. The pleomorphic type is the most differentiated type with the best prognosis. Once a tissue diagnosis is obtained, staging of the disease with bone marrow biopsy, CXR, LFTs, CBC and lumbar puncture is required. With the recent advances in treatment using chemotherapy and radiation, this once fatal disease now carries a much better prognosis. Localized disease (Group I & II) carries a 90% 5-year survival. However, disseminated or gross residual disease (Group II & IV) has a 35% 5-year survival rate.

Optic Nerve Gliomas

Optic nerve glioma is the fifth most common primary intraorbital tumor (1.5% to 3.5% of all orbital tumors) and the third most common orbital tumor in children. These tumors may occur randomly, but are often associated with Neurofibromatosis type I (18 to 50% of cases), and are often bilateral when occurring with this disease. The mean age of presentation is about 8 years. The typical presentation is proptosis and visual loss or visual field changes. Intracranial extension may produce headaches and pain. Upon eye examination, optic disk swelling or atrophy may be noted as well as decreased motility. Diagnosis can usually be made based on clinical exam and radiography due to the characteristic appearance on CT. Radiographically, they appear as fusiform enlargement of the optic nerve which is isodense to brain. Intracranial extension into the optic canal and chiasm is best evaluated with MRI. Despite their apparently benign and slow growth pattern, gliomas are associated with significant morbidity and mortality, especially when the tumor spreads to the optic chiasm, hypothalamus and brain. Because of this, these tumors must be excised while still confined to the optic nerve. Once the tumor has extended to the chiasm, surgery is not indicated. Radiotherapy at this point does not seem to improve prognosis and chemotherapy is unproven. Gliomas are indolent enough to warrant conservative management with serial radiographic studies when vision is intact. However, once the tumor extends to the optic canal or the eye becomes blind and/or proptotic, surgical excision is necessary.

Fibrous Dysplasia

This lesion is the most frequently seen fibro-osseous tumor and develops almost exclusively in children during the first 2 decades of life. This lesion is characterized by the replacement of normal bone with abnormal tissue composed of collagen, fibroblasts, osteoid and giant cells (immature woven bone). There are two types of fibrous dysplasia: polyostotic (Albright's syndrome) and monostotic. Polyostotic fibrous dysplasia involves multiple bones, but not generally the orbit, and is characterized by abnormal skin pigmentation and precocious puberty. Monostotic fibrous dysplasia occurs most often in the bones of the face and is the most common type in the orbit. The orbital roof is the most common site of orbital involvement. Although the onset of disease is within the first couple of years of life, the usual presentation is the adolescent child with proptosis, globe and orbit displacement and facial asymmetry. The CT will show thickened abnormal bone with sclerotic lesions with a "ground-glass" appearance. Biopsy is usually necessary to confirm the diagnosis and to rule out more aggressive lesions. Conservative management with close observation is the hallmark of treatment, since these lesions usually stabilize after puberty. Indications for surgical treatment are significant cosmetic deformity and vision loss. Complete resection followed by immediate craniofacial reconstruction is recommended, usually with the neurosurgeon. If total resection is not possible, removal of as much of the lesion as necessary to protect sight and correct the cosmetic deformity should be performed.

Metastatic Tumors: Neuroblastoma

Neuroblastoma is the most frequent metastatic orbital disease in children. Other encountered metastatic diseases include Ewing's sarcoma, leukemia, and lymphoma. Neuroblastoma is common in children and accounts for about 10% of all malignancies. The majority of cases occur before age 5 (median 22 months). Bilateral disease is common with eyelid ecchymoses and proptosis being the most common presenting signs. Additional symptoms may include abdominal fullness/pain, edema and hypertension owing to the primary lesion, most commonly in the adrenal gland. Urinalysis is positive for catecholamines in 90 to 95%. Incisional biopsy is necessary to confirm the diagnosis. Children presenting with orbital disease are stage IV, and have a survival rate of less than 15%. Therapy includes surgery, chemotherapy, radiation therapy and bone marrow transplantation.

Adult Orbital Tumors

In the adult population, the more common types of orbital tumors vary significantly from children. The most common tumor groups in this population include carcinomas (paranasal sinus, secondary & metastatic), inflammatory masses (pseudotumor), lacrimal gland tumors, cysts, lymphomas, meningiomas, and vascular tumors (cavernous hemangiomas). Secondary tumors commonly invade the orbit and include mucoceles, squamous cell carcinoma, meningioma, vascular malformations and basal cell carcinoma.

Paranasal Sinus Masses

A mass developing in the paranasal sinuses has the potential to extend into the orbit and is a common cause of an orbital mass lesion. The most common mass lesion of the orbit originating in the sinus is the mucocele. Mucoceles often result from obstruction of a sinus ostium leading to an enlarging fluid filled sinus which eventually may erode through the orbital bony wall. The median age of presentation is around 50 years, although they may present at any age. Most arise from the ethmoid and frontal sinus. Patients will present with unilateral proptosis with globe displacement away from the mass, lid swelling and sometimes a palpable mass. CT scan reveals a well-defined homogeneous mass extending into the orbit through a bony defect associated with an opacified sinus cavity. Treatment involves drainage of the obstructed sinus and often obliteration of the sinus.

Neoplasms of the paranasal sinuses are uncommon, but frequently extend to involve the orbit when they do occur. Both epithelial and mesenchymal neoplasms may develop here. Benign tumors tend to push the periorbita aside, while malignant lesions tend to invade the periosteum. By far the most common malignancy is squamous cell carcinoma. This disease is usually advanced at presentation with orbital invasion in almost two thirds of the patients. Presentation with orbital extension imparts a poor prognosis, especially with extension above an imaginary line from the medial canthus through the angle of the mandible (Ohngren's line). Glandular malignancies may arise from either minor salivary glands or from the basal lamina of the respiratory epithelium. Adenocarcinomas arising from the ethmoid sinuses are frequently associated with wood workers. Salivary tumors are usually adenoidcystic or mucoepidermoid and are more common in the antrum. Adenoid cystic carcinomas have the propensity for perineural spread via the infraorbital nerve and have been known to cause bilateral orbital invasion with blindness. Locally invasive neoplasms may arise from the olfactory neuroepithelium-esthesioneuroblastoma. Due to the proximity to the ethmoid, early involvement of this sinus is common. Orbital involvement is a late finding, due to its indolent growth. A benign paranasal neoplasm, not uncommonly seen, is the inverted papilloma. This is a benign growth arising from the lateral nasal wall in the region of the middle meatus, or from the sinus mucosa. The tumor usually involves the ethmoid and maxillary sinuses with gradual enlargement through the bony septa and sinus walls. Most patients present after nasal obstruction or epistaxis have occurred. True violation into the orbital fat is rare, however, compression of the orbital contents can cause visual loss. In 5 to 10% of cases, inverted papillomas are associated with SCCA so aggressive surgical resection is mandatory.

Evaluation of the paranasal sinus mass is best done radiographically with CT scan, because of the ability to detect early lesions and note bony destruction with either orbital or intracranial extension. MRI scans are useful in detecting intracranial extension and distinguishing certain neoplastic diseases from one another. After radiographic evaluation, tissue should be obtained with biopsy prior to definitive evaluation. This can optimally be done in the operating room, especially when exploration of the involved sinus is required. Biopsy should not be performed in the case of juvenile angiofibroma, tumors with direct cranial cavity communication, cerebrospinal fluid leak, or masses with synchronous pulsations with the heart beat (i.e. encephalocele). Malignant tumors originating from the paranasal sinuses will require radical resection with wide margins. Penetration through the periorbita is an indication for sacrifice of the orbital contents. Postoperative radiation therapy often accompanies surgical resection. Invasion of the orbit by a malignancy of the paranasal sinuses usually carries a grave prognosis, and often palliation is the primary treatment goal.

Orbital Pseudotumor (Idiopathic Orbital Inflammation)

This descriptive term was first used in 1905 by Birch-Hirschfeld to describe an inflammatory condition of the orbit of unknown etiology. It is now applied to a large spectrum of non-specific idiopathic inflammations of the orbit and ocular adnexa, excluding systemic diseases (sarcoidosis, Wegener's, thyroid orbitopathy, etc.). It is now restricted to lesions which show histological appearance of polymorphous inflammatory infiltration of T&B cells, plasma cells, eosinophils and neutrophils, with a variable degree of fibrous tissue proliferation. This disease is a common cause of proptosis presenting anywhere from the 2nd to 7th decade of life. Multifocal involvement is common and any orbital structure may be involved. The onset of symptoms is typically a few days, however, subacute or chronic forms have been described. The typical symptoms are dull orbital pain which is worse with eye movement. Proptosis is the most common finding although eyelid swelling, chemosis, and diplopia commonly occur. Visual loss may occur with scleral, uveal, or optic nerve inflammation. CT findings show hazy enlargement of affected structures with enhancement after intravenous contrast injection. MR T1-weighted images show lesions with similar signal to muscle that enhance with contrast. T2-weighted images have increased signal similar or greater than fat. A range of therapeutic options have been employed including surgery, steroids, immunosuppressive agents, and radiation therapy. Symptom relief

should be the goal. Surgery may be effective for localized lesions, but is generally reserved for biopsy only as it may exacerbate the inflammatory process. Steroid therapy (oral prednisone 60-100 mg/day) is a mainstay of treatment and usually provides symptomatic relief, however, relapse rates are high when discontinued. Some cases require immunosuppressive medications, such as cyclophosphamide or cyclosporine. Radiation therapy is indicated for lymphoid infiltrates and is effective in nonfibrotic lesions with marked cellularity or prominent inflammatory features, and is an alternative when steroid therapy is contraindicated or has adverse effects.

Lacrimal Gland Tumors

Masses of the lacrimal fossa are caused by numerous conditions. Enlargement of this area with swelling and erythema often indicates infection or inflammation of the gland. However, inferior and medial displacement of the globe with no inflammatory signs or symptoms should raise suspicion of a neoplastic process. About half of the lacrimal tumors are epithelial neoplasms, while the other half are lymphoproliferative disorders. Lymphoid lesions include benign lymphoid hyperplasia, malignant lymphoma and leukemias. Lymphoid lesions rarely cause bony changes or destruction and appear as smooth enlargement of the gland on CT scans. Epithelial neoplasms appear more irregular on CT and include pleomorphic adenomas (benign), adenoid cystic carcinoma, adenocarcinoma, mucoepidermoid carcinomas, and undifferentiated carcinomas.

Primary epithelial neoplasms of the lacrimal gland are rare, and by far the most common of these is the pleomorphic adenoma (benign mixed tumor). It occurs in adults primarily between the ages of 20 and 50 years. The presenting symptoms are painless proptosis with inferior and medial displacement of the globe that has been present for many months or years. These patients should undergo complete excisional biopsy of the gland for diagnosis confirmation. There is a 32% rate of recurrence with a chance for malignant degeneration over the patient's lifetime if the capsule is incised without complete removal.

The most common malignant epithelial neoplasm of the lacrimal gland is adenoid cystic carcinoma. In comparison to the benign mixed tumor, malignant epithelial neoplasms will present with a more progressive onset of proptosis and globe displacement. Pain and numbness are also more common in malignant tumors. CT scans will often show bony destruction and infiltration of the lacrimal mass. An incisional biopsy is indicated for these findings before proceeding with treatment. The mortality rate is significant (>50%), and therefore aggressive surgical resection, many times with exenteration and removal of soft tissue and bone, followed by radiotherapy.

Lymphoid Tumors

Lymphoid tumors are one of the more common orbital tumors despite the orbit not containing lymph nodes or a well defined lymphatic vasculature. The incidence is between 4 and 13% of all orbital tumors. Orbital lymphomas may be primary or associated with systemic disease. Although most orbital lymphomas are localized to the orbit at diagnosis, many patients will develop systemic lymphoma over time. Approximately 20% of patients with lymphoid tumors of the conjunctiva, 35% of patients with orbital tumors and 67% with eyelid disease will eventually develop systemic lymphoma. Orbital lymphoma is an adult disease process usually presenting between the age of 50 and 70 years. The course is usually one of an anterior mass which enlarges slowly causing progressive painless proptosis over several weeks to months. The classic lesion is a smooth, pink-orange mass ("salmon patch") under an intact conjunctiva. CT scan shows a homogeneous mass with well defined borders that does not destroy surrounding structures or bone. Most lesions are extraconal and in the superior orbit. The lacrimal gland may be affected but enlarges with a normal shape unlike primary lacrimal gland tumors. To make a definitive diagnosis a generous biopsy is needed and must be sent for immunohistochemical studies

(fresh) as well as permanent section (formalin). Once a diagnosis of lymphoma is made, patients should be worked up for systemic disease including a complete physical exam, a complete blood count, bone marrow biopsy and CT scans of the thorax, abdomen and pelvis. Patients with localized lymphoma to the orbit may be treated with primary XRT, and patients with systemic disease may respond to a combination of orbital radiotherapy and systemic chemotherapy. Consultation with an oncologist should be obtained.

Orbital Meningiomas and Schwannomas

These are most common in adults during the fourth to seventh decade of life and are rarely seen in children. Although primary orbital meningiomas may arise from the optic nerve, 70% invade the orbit from the cranium. The most common presenting symptom is proptosis, yet visual disturbances are usually the chief complaint. Other symptoms may include headache and diplopia. Imaging studies usually make the diagnosis. CT scan will reveal a fusiform enlargement of the optic nerve, which is homogeneous, infiltrative, and enhancing. The classic "railroad track" describes calcifications of the tumor along the optic nerve in the subarachnoid space. MRI is used to evaluate intracranial extension, showing a hyperintense tumor after contrast administration. In the older patient, these tumors may be followed if no evidence of intracranial extension exists. In the younger patient (<40), these lesions are more aggressive and should be excised at an early stage. Unfortunately, optic nerve sheath meningiomas often cause severe visual deficits after removal.

A schwannoma, or neurilemoma, is a benign, noninvasive, peripheral nerve tumor that may arise from any nerve within the orbit. They are relatively rare and usually occur in adults from age 20 to 70 years. These patients present with painless proptosis and diplopia. Rarely, progressive visual loss or a relative afferent pupillary defect may occur. Diagnosis is made with CT and/or MR imaging. CT shows a well-circumscribed, homogeneous, elongated ovoid mass displacing surrounding structures. The tumor is hypointense on T1-weighted images of MRI, and hyperintense on T2-weighted images. The tumor may be extraconal when associated with the trochlear nerve (IV), but is more commonly intraconal. The definitive therapy is complete surgical excision and the prognosis is excellent with rare recurrences.

Cavernous Hemangioma (Encapsulated Venous Malformation)

An encapsulated venous malformation, commonly referred to as the cavernous hemangioma, is a relatively common vascular lesion in the orbit. It is the most common vascular lesion of adults and is the most common primary intraconal orbital lesion in adults. The peak incidence is during middle age with the average age of onset around 40 years. It is generally more common in women (70%) than men (30%) and is generally unilateral. These lesions present with a slowly progressive painless proptosis over a period of several years. Interestingly, these malformations are isolated from the orbital vascular system and therefore do not enlarge with Valsalva maneuvers. It is thought that these lesions are formed from pre-existing vascular malformations, the vessels of which are initially collapsed outside of the fibrous capsule. Once these vessels enlarge due to changes in the systemic circulation pressures, they become enlarged and eventually incorporated by a newly formed reactive fibrous capsule.

Imaging with either CT or MRI reveals a well-defined mass with an oval shape. Most are intraconal, but occasionally can be found outside the muscular cone. On CT they are homogeneous with increased density. With MRI they appear homogeneous and isointense to muscle on T1-weighted images and hyperintense on T2-weighted images. Following contrast addition, the lesions enhance inhomogeneously. The treatment of choice is complete surgical excision of the tumor and capsule with an excellent prognosis. Recurrences are very rare even with incomplete resection.

Metastatic Tumors

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 mets). On CT, the most common finding is a well-defined, contrast enhancing, intraconal mass. The orbital bony walls are also a common site for metastasis, especially with prostate cancers. 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.

Conclusion

The orbit has a very complex anatomy in close association with the paranasal sinuses and cranial vault. A wide variety of lesions may develop in the orbit with significant variation according to the age of the patient. It is not uncommon for these patients to present to the otolaryngologist either primarily or via consultation from another subspecialty. It is therefore important for the otolaryngologist to possess a basic understanding of the anatomy of the orbit and the more common disease processes which may affect it. Since vision is of primary concern with disease processes of the orbit, it is always advisable to involve the ophthalmologist in the management of orbital tumors. In some cases it may be necessary for several specialties to manage these diseases together. A combined approach with the help of neurosurgeons, oncologists, radiation therapists, otolaryngologists and ophthalmologists will often give the best outcome to the patient afflicted with an orbital tumor.

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