

Clinical Approach to Orbital Disease

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- I. History
 - a. Progression:
 - i. Acute (Hours to days)-hemorrhage or infarction of pre-existing tumor (e.g. lymphangioma), acute orbital cellulites, IOI
 - ii. Sub-acute (days to weeks)- IOI, thrombophlebitis, TRO, metastatic disease, some malignancies (rhabdomyosarcoma, neuroblastoma, granulocytic sarcoma)
 - iii. Chronic (months to years)- most solid tumors (dermoids. neurogenic tumors, cavernous hemangioma, etc
 - b. Pain:
 - i. sign of infection, infarction, inflammation, orbital hemorrhage
 - ii. seen in perineural invasion associated with malignant lacrimal gland tumors
 - c. Worsening of symptoms with URI- characteristic of lymphangioma
- II. Exam
 - a. Periorbital changes
 - i. Salmon colored subconjunctival mass-lymphoproliferative disease
 - ii. Eyelid retraction, EOM insertion injection/edema-TRO
 - iii. Vascular discoloration-lymphangioma, varix(blue), capillary hemangioma (red)
 - iv. S-shaped lid-neurofibroma
 - v. Ecchymosis-neuroblastoma, leukemia
 - vi. Prominence of temple-sphenoid wing meningioma
 - vii. Black encrusted lesions-mucormycosis
 - viii. Corkscrew conjunctival vessel- A-V fistula
 - ix. Frozen globe-metastasis, epithelial malignancies of lacrimal gland
 - b. Proptosis
 - i. Hertel exophthalmometry-measures globe protrusion from latera orbital rim to corneal apex
 - ii. Unilateral-TRO, lymphoproliferative, IOI, intraconal lesions
 - iii. Bilateral-TRO, IOI, lymphoproliferative
 - iv. Axial proptosis- indicative of intraconal lesions or lesions which enlarge the EOMs or diffusely involve the orbital soft tissues
 - v. Displacements-globe pushed away from tumor site
 - 1. downward-rhabdomyosarcoma, leukemia
 - 2. downward/medial-lacrimal gland tumors, dermoids
 - 3. downward/lateral-mass in superolateral quadrant inc. dermoid, encephalocele
 - 4. superior-inc. maxillary sinus tumor, lacrimal sac disease
 - vi. Pseudoproptosis

1. Increased axial length of globe
 2. EOM weakness
 3. Contralateral enophthalmos
 4. Orbital bony asymmetry
 5. Palpebral fissure asymmetry
- c. Palpation
- i. If gaze is directed to examiner's finger, septum is relaxed allowing deeper palpation of orbital structures
 - ii. Detection of resistance to retrodisplacement
- d. Pulsation
- i. Vascular lesions with high flow
 - ii. Transmission of CSF pressure via orbital bony defect
- e. Bruits
- i. In high flow lesions
 - ii. Found in orbital CC fistulas, dural-sinus fistulas
- III. Orbital cellulites
- a. Preseptal cellulites
- i. Confined to eyelids anterior to septum
 - ii. Related to trauma or external lesion such as hordeolum
- b. Orbital cellulites
- i. Most common cause of proptosis in childhood
 - ii. Usually due to extension of paranasal sinusitis
 - iii. Clinical findings-pain, fever, proptosis, adnexal swelling, restricted motility, decreased vision
- c. Subperiosteal or orbital abscess formation is an indication for surgical drainage
- d. Complications
- i. Cavernous sinus thrombosis- severe complication with bilateral proptosis, ophthalmoplegia, cranial neuropathy, papillary abnormalities, intracranial signs
 - ii. Brain abscess, meningitis, osteomyelitis of skull
- e. Mucormycosis
- i. Potentially fatal fungal infection
 - ii. Typical organism of sinuses and nasal cavity causing infection in setting of severe systemic acidosis and immunosuppression (as in DKA)
 - iii. Commonly presents as orbital apex syndrome
 - iv. Pathology features vascular invasion by large nonseptate branching hyphae causing thrombosis
 - v. Treated by correction of metabolic abnormality, local excision of infected tissues, amphotericin B
- IV. Idiopathic Orbital Inflammation
- a. May be acute, subacute, or chronic and is subject to frequent relapses
 - b. Can be unilateral or bilateral, can involve any orbital tissue including fat, EOM, lacrimal gland, tenon's, optic nerve sheath

- c. Symptoms include pain, edema/chemosis, proptosis, motility limitation, vision loss
 - d. In children often bilateral (1/3), associated with papillitis, iritis, pachymeningitis
 - e. CT scan- infiltration of involved tissues
 - i. EOM involvement does not spare tendon of insertion as in TRO
 - ii. Ring sign seen in Tenonitis/posterior scleritis
 - f. Pathology-variable infiltration of lymphocytes, plasma cells, eosinophils with a background of fibrosis
 - g. Treatment- biopsy to confirm diagnosis, steroids (systemic), radiation or orbital decompression in progressive resistant cases
- V. Developmental pediatric tumors
- a. Terminology
 - i. Hamartoma-growth consisting of disorganized tissues normally found at the involved site
 - ii. Choristoma-growth consisting of tissues not normally found at the involved site
 - b. Cysts
 - i. Dermoids-
 - 1. lined by mature skin with dermal appendages
 - 2. often found near bony sutures, due to pinching off of ectoderm during fetal skull formation
 - 3. Treated with complete surgical excision
 - 4. May caused inflammation if they rupture subcutaneously
 - c. Lipodermoids
 - i. Solid tumors which are usually sub-conjunctival
 - ii. Tend to be superotemporal with deep posterior extension
 - iii. Resection should be limited to exposed anterior portion to limit postoperative incidence of strabismus and ptosis
 - d. Teratomas
 - i. Consist of tissues derived from multiple germinal layers
 - ii. Often cause dramatic exophthalmos at birth
 - iii. Typically cystic, can sometimes be removed with preservation of globe
 - iv. CT- orbital enlargement, ZMF suture separation
- VI. Vascular tumors
- a. Capillary hemangioma
 - i. Appear in first weeks of life, grow over 1-5 years, then begun to involute
 - ii. Red, dimpled, elevated surface (strawberry nevus), deeper lesions are bluer
 - iii. May cause amblyopia due to ptosis, strabismus, anisometropia
 - iv. Pathology-vascular channels with complete endothelial lining
 - v. Treatment indicated when vision affected, injection of intralesional steroids
 - b. Lymphangioma

- i. Occurs in 1st decade, does not spontaneously involute
 - ii. Can suddenly enlarge in cases of URI or intralesional hemorrhage
 - iii. Anatomically involves eyelids, conjunctiva and deeper orbital structures, have superonasal predilection
 - iv. Lesions can also occur in the sinuses and oropharynx
 - v. Pathology-large serum filled spaces with incomplete endothelial lining and scattered lymphoid tissue
 - vi. Difficult to treat due to infiltrative nature and high likelihood of hemorrhage
 - c. Cavernous hemangioma
 - i. Most common benign orbital tumor of adults, more common in women
 - ii. Slow progressive proptosis, may cause optic nerve compression
 - iii. CT-encapsulated intraconal mass
 - iv. High internal reflectivity on Bscan
 - v. Pathology-large cavernous blood filled spaces with complete endothelial lining
 - vi. Surgically excised via orbitotomy when indicated
 - d. Hemangiopericytoma
 - i. Uncommon, M>F, middle age presentation
 - ii. Similar course to cavernous hemangioma but more rapid progression
 - iii. CT-well circumscribed encapsulated mass with more contrast enhancement than cavernous hemangioma
 - iv. Pathology- plump pericytes in vascular network, +staghorn vessels, +reticulin stain
 - v. Must be completely excised to prevent recurrence, malignant transformation, metastasis
 - e. AV malformation
 - i. Developmental anomalies consisting of anastomosing arteries and veins with no intervening capillary network
 - ii. Often dilated corkscrew episcleral vessels
 - f. AV fistula
 - i. Abnormal acquired arteriovenous communications caused by trauma (basal skull fractures) or degeneration
 - ii. Signs include surface vascular tortuosity, pulsatile proptosis, bruits, elevated IOP
 - g. Varices
 - i. Occur secondary to intracranial AVM
 - ii. May increase size with Valsalva causing variable proptosis
 - iii. CT-size is position dependant, Ca++ phleboliths
 - iv. Pathology-venous channels, thrombosis
 - v. Observe, drainage of lesions which are anterior
- VII. Neural tumors
 - a. Optic nerve glioma
 - i. 1st decade (2-6 yrs of age)

- ii. associated with NF1 (20-50%), pathognomonic for NF1 when bilateral
 - iii. gradual painless axial proptosis with vision loss, APD, papilledema progressing to atrophy
 - iv. pathology-juvenile pilocytic astrocytoma with glial hypercellularity, areas of myxomatous degeneration, eosinophilic Rosenthal fibers, surrounded by arachnoid hyperplasia (mimics meningioma)
 - v. Treatment
 - 1. Observe if stable Va, minimal deformity
 - 2. Orbital excision to confirm path, or to relieve deformity or pain from globe exposure
 - 3. intracranial surgery-prior to chiasmal involvement
 - 4. radiation-for unresectable tumors
 - vi. CT/MRI-fusiform optic nerve enlargement, look for intracranial extension
- b. Meningioma
- i. Originate in arachnoid villi
 - ii. May involve optic nerve sheath or secondarily involve orbit when cranial lesion grow in sphenoid wing
 - iii. Symptoms
 - 1. sphenoid wing-temporal mass, proptosis, lid edema
 - 2. field defects, papilledema or optic atrophy
 - iv. Radiology-tubular enlargement of optic nerve sheath or hyperostosis, bone destruction and intralésional Ca⁺⁺ in cranial lesions
 - v. Treatment
 - 1. Observe if Va good
 - 2. Resect if Va decreasing or growth is progressive (esp. younger patients)
 - 3. Combined neurosurgical approach for intracranial lesions
 - vi. Pathology-benign appearing meningothelial cells, Psammoma bodies (Ca⁺⁺)

VIII. Other Neural Tumors

- a. Neurofibromatosis 1-autosomal dominant with irregular penetrance
 - i. Multiple hamartomas of skin, eye, CNS
 - 1. Café au lait spots
 - 2. axillary freckling
 - 3. plexiform neurofibromas-S-shaped upper lid
 - 4. sphenoid wing hypoplasia with secondary encephalocele
 - 5. pigmented iris Lisch nodules
 - 6. congenital glaucoma
 - 7. optic nerve glioma
- b. Neurofibromas
 - i. Types
 - 1. Plexiform-pathognomonic of NF1

- 2. Diffuse
 - 3. Isolated
 - 4. All are infiltrative vascular tumors causing disorganization of tissue and tend to bleed during resection
 - ii. Pathology-nonencapsulated proliferation of axons, schwann cells, and well developed vascular supply
 - iii. Treatment
 - 1. Plexiform-debulking
 - 2. Isolated-excision (malignant forms do exist)
 - c. Schwannoma (neurilemmoma)
 - i. Well-encapsulated peripheral nerve sheath tumors
 - ii. May be painful
 - iii. Are encapsulated by the perineurium of the nerve of origin
 - iv. Pathology-
 - 1. Antoni A areas-pallisaded spindle cells
 - 2. Antoni B areas-stellate cells in loose myxoid substance
- IX. Lacrimal glands tumors
- a. Overview
 - i. 75% of lesions inflammatory or lymphoproliferative
 - ii. 25% are epithelial (50% benign)
 - b. Pleiomorphic adenoma (benign mixed tumor)
 - i. Most common epithelial of lacrimal gland
 - ii. Occur in 4th to 5th decade, develops slowly
 - iii. Cause a firm, painless mass in the supertemporal quadrant pushing the globe downward and medially
 - iv. CT globular gland enlargement with bone molding
 - v. Treatment-complete excision without violation of capsule or biopsy (opening of capsule may cause recurrence and possible malignant degeneration)
 - c. Adenoid cystic carcinoma
 - i. Most common malignancy of lacrimal gland
 - ii. Infiltrative, rapidly progressive tumor associated with pain due to perineural invasion
 - iii. CT-nonencapsulated tumor with evidence of bone destruction
 - iv. Pathology-benign appearing epithelial cells in nests, tubules, and in cribriform (Swiss-cheese) pattern
 - v. Treatment-biopsy followed by exenteration with removal of involved bone
 - d. Nonepithelial (infiltrative) lesions
 - i. Inflammatory- includes IOI, sarcoid
 - ii. Lymphoproliferative-benign or malignant
 - iii. Mikulicz syndrome (lymphoepithelial lesions)-loss of glandular parenchyma, lymphocyte infiltration, myoepithelial islands (called Sjogren's disease if associated with rheumatoid arthritis)
- X. Lymphoproliferative lesions
- a. Spectrum

- i. Benign (reactive) lymphoid hyperplasia
 - ii. Atypical lymphoid hyperplasia
 - iii. Malignant lymphoma
 - b. Features may include progressive proptosis, motility loss, enlargement of lacrimal gland, or conjunctival salmon patch lesions depending on tissue involved at presentation
 - c. Systemic evaluation
 - i. Physical with attention to adenopathy
 - ii. CBC/diff
 - iii. Bone marrow bx
 - iv. CT of chest, abdomen, pelvis
 - v. Bone scan
 - vi. Serum protein electrophoresis
 - d. Biopsy-requires fresh tissue for marker studies and cytogenetic analysis
 - e. Lymphoid hyperplasia
 - i. Benign-mature lymphocytes with germinal centers
 - ii. Polyclonal by definition
 - iii. Atypical lesions- less well organized may have small detectable monoclonal populations
 - f. Malignant lymphoma
 - i. Diffuse collection of atypical immature lymphocytes
 - ii. Vast majority are B-cell tumors
 - iii. Monoclonal
 - g. Treatment-
 - i. Radiotherapy for localized orbital disease
 - ii. Chemotherapy for systemic disease
- XI. Mesenchymal tumors
- a. Rhabdomyosarcoma
 - i. Most common primary orbital malignancy of childhood
 - ii. Rapid growth with lid edema, proptosis, more common in superior orbit
 - iii. CT-solid well-defined mass with bone destruction
 - iv. Pathology
 - 1. embryonal-most common
 - 2. alveolar-most malignant
 - 3. pleomorphic-most well differentiated
 - v. Treatment-
 - 1. Immediate biopsy
 - 2. workup with chest/abdomen CT, LP, bone marrow bx
 - 3. high-dose local RT with systemic chemo
- XII. Metastatic pediatric tumors
- a. Neuroblastoma
 - i. Presents with ecchymotic proptosis and bone destruction
 - ii. Metastases occur late in course-primary tumor in neck, abdomen, mediastinum

- iii. Involvement of cervical ganglia-causes Horner's with heterochromia
 - b. Leukemia
 - i. Orbital disease most common in ALL
 - ii. AML-causes granulocytic sarcoma (chloroma) presenting before blood or marrow involvement
- XIII. Thyroid orbitopathy (Graves disease)
 - a. Most common cause of unilateral and bilateral proptosis in adults
 - b. Women affected 8X more than men, onset 20-45 years
 - c. Systemic disease
 - i. Most cases hyperthyroid (can occur in hypothyroid or euthyroid cases)
 - ii. Infiltrative orbitopathy
 - iii. Infiltrative dermopathy
 - d. Clinical features
 - i. Exophthalmos
 - ii. Upper/lower eyelid retraction
 - iii. Lid lag and lagophthalmos
 - iv. Restrictive myopathy and diplopia
 - v. Eyelid edema
 - vi. Conjunctival chemosis and injection
 - vii. Compressive optic neuropathy
 - e. CT-EOM enlargement (esp. IR, MR) with sparing of tendons
 - f. Pathology-lymphocytic/plasmocytic infiltration with fibroblast activation and mucopolysaccharide production
 - g. Treatment-
 - i. Lubrication
 - ii. Possible systemic prednisone
 - iii. Surgery
 - 1. orbital decompression
 - 2. EOM surgery
 - 3. lid retraction repair
 - iv. Radiation for refractory cases
- XIV. Vasculitis
 - a. Giant cell arteritis
 - i. May involve orbital vessels-esp. posterior ciliary arteries
 - ii. Pathology-granuloma of ILM
 - iii. Treatment-high dose steroids after ESR with prompt biopsy
 - b. Wegener's granulomatosis
 - i. Features include erosive sinus disease, tracheobronchial necrotic lesions, cavitory lung lesions, glomerulonephritis
 - ii. Orbital inflammation due to extension of sinus lesions
 - iii. Pathology-necrotizing vasculitis with granulomas
 - iv. Treatment-cytoxan
 - c. Lethal midline granuloma
 - i. Massive progressive necrosis of nose, sinuses, palate

- ii. Response to high dose RT
- XV. Sarcoidosis
 - a. Eye findings include uveitis, lacrimal gland involvement, enlarged EOM
 - b. Facial palsy-Heerfordt's syndrome
 - c. Pathology-non-caseating granulomas with Langhan's giant cells
 - d. Treatment-corticosteroids, possible surgical debulking
- XVI. Secondary orbital tumors
 - a. Sinus tumors
 - i. Mucocele
 - 1. Expansile cystic lesions arising from paranasal sinuses causing erosion of orbital walls due to obstruction of sinus excretory ducts
 - 2. CT-well-defined margins with sinus opacification
 - 3. Pathology-cyst lined by pseudostratified ciliated columnar epithelium
 - 4. Treatment-external removal of sinus mucosa and obliteration of sinus cavity
 - ii. Carcinomas
 - 1. Squamous cell carcinoma is most common tumor secondarily invading the orbit
 - 2. early signs include sinusitis and epistaxis
 - 3. late symptoms-nasal obstruction, epiphora
 - b. Neglected eyelid tumors
 - c. Intraocular tumors with orbital extension
- XVII. Metastatic tumors
 - a. Rare-1/10th as common as intraocular metastases
 - b. Sources-breast, lung, GI, prostate
 - i. Breast-most common orbital met in women
 - 1. may be years after initial diagnosis
 - 2. fibrosing forms may cause enophthalmos
 - ii. Bronchogenic-most common orbital met in men
 - iii. Prostatic-pseudotumor-like picture
 - c. Findings-pain, proptosis, bone destruction, ophthalmoplegia
 - d. Treatment
 - i. Biopsy/debulking
 - ii. Radiation or chemotherapy depending on tumor type and workup