62 WM sinusitis, migraines, c/o transient blur/headaches. BVA 20/20, normal exam except residual corneal shrapnel. Right orbit CT demonstrates unexpected 1.7x1.16cm mass. DDx Cavernous Hemangioma/Hemangiopericytoma, Fibrous Histiocytoma, Orbital Pseudotumor, Lymphoma, Schwannoma will be discussed.

CASE REPORT

I. Case History

A. Demographics and Chief Complaint:
   a. 62 y.o. white male presents for optometry consult at Jesse Brown VAMC with complaints of transient blurry vision
   b. Secondary complaint: Headaches occurring all over his head, 2-3 headaches a month, and some that awaken him from sound sleep

B. Ocular History:
   a. Trauma c surgery OS (1970): shrapnel in corner eye removed as well as back of head
   b. Denies history of glaucoma and other ocular conditions
   c. LEE: 5-6 years ago, not at V.A.

C. Medical History:
   a. Hypertension, controlled
   b. Hypercholesteremia
   c. COPD
   d. Migraines (longstanding, since service) vs. Cluster headaches
   e. Shrapnel back head
   f. Chronic Left Knee Pain
   g. h/o left leg muscle injury

D. Allergies:
   1. Lisinopril

II. Pertinent Findings

A. Clinical:
   a. Distance entering aided acuities: 20/25-3 OD PH 20/20, 20/40- OS PH 20/20-
   b. Distance corrected acuities with refraction: 20/20 OD, 20/20 OS
   c. Preliminary Testing:
      i. Pupils: Normal OD, OS (-) APD
      ii. EOMS: Full range of Motion OD, OS
      iii. Confrontation Fields: Full to finger counting OD, OS
      iv. Color: Normal OD, OS
v. Exophthalmometry: 18/18 base@96
vi. Retropulsion: Symmetrical OU
vii. Red Cap Desaturation: Equal in both eyes
viii. Humphrey Visual Field testing: Essentially full OD, OS

d. Slit Lamp (abnormal findings only):
   i. Lids/Lashes:
      1. OD: 1-2mm of dark subepithelial deposit, soft, mobile, ?retained debri from shrapnel injury
   ii. Cornea:
      1. OS: small corneal scar superior temporal to pupil
   iii. All other slit lamp findings non-contributory

e. Intraocular Pressures:
   i. OD: 16mmHg, OS: 16mmHg

f. Dilated Fundus Exam:
   i. C/D: OD 0.25/0.3, OS 0.25/0.25
   ii. ONH: flat, sharp, good color, OU
   iii. Macula: mild RPE changes OU
   iv. Vessels, Vitreous, Periphery: Unremarkable

g. Assessment:
   i. Leading diagnosis is sinusitis causing headaches

h. Plan:
   i. Order CT of orbit and sinuses

B. Radiology Studies
   i. X-ray of orbit revealed:
      1. Clouding of left maxillary antrum due to either sinusitis, large cyst or polyp
      2. All other paranasal sinuses normal
      3. No lytic or sclerotic bony lesions
      4. Small metallic foreign bodies seen in left maxilla
   ii. CT scan w/o contrast of maxillofacial region revealed:
      1. Polypoid lesions in right and left maxillary sinuses
      2. Mild mucosal thickening of right and left maxillary, ethmoid, frontal, sphenoid sinuses consistent with paranasal sinusitis
      3. UNEXPECTED FINDING: soft tissue mass lesion, 1.7cm x 1.16cm in size, in right retrobulbar region.
      4. Extraconal muscles normal
      5. No evidence of abnormal increased density at fatty tissue posterior to the right globe
   iii. CT scan with contrast of orbits:
      1. Minimal enhancement of lesion, minimal central vascularity
2. Lesion not affecting optic nerve, located in inferior anterior orbital region
3. Lesion not affecting extraocular muscles
   iv. Unable to MRI due to foreign bodies of maxilla

C. Differential Diagnosis for orbital mass lesion that will be discussed including treatment and management:

a. Cavernous Hemangioma
   i. Affects 40-60 y.o. range and is benign, slowly progressive tumor that in its later stages shows signs and symptoms of decreased vision, restricted motility, proptosis, and choroidal folds
   ii. CT scan shows well-defined, oval-round, encapsulated lesion with minimal enhancement

b. Fibrous Histiocytoma
   i. Affects 40-60 y.o. with signs and symptoms of decreased vision, restricted motility, proptosis, diplopia, ptosis and epiphora
   ii. Benign or malignant mesenchymal tumor
   iii. CT scan shows well-defined round mass, more commonly found in nasal quadrant

c. Orbital Hemangiopericytoma
   i. Rare, vascular tumors caused by the over-proliferation of pericytes. Affect all ages and are painless masses that do not present with any associated signs and symptoms
   ii. CT scan will show well defined mass
   iii. Biopsy is best method for confirming diagnosis

 d. Lymphoma
   i. Affects 50-70 y.o. with signs and symptoms proptosis, diplopia, ptosis, and lid edema
   ii. Broad spectrum of variations being benign to malignant, can progress from benign to malignant years after
   iii. CT scan shows mass that molds or encompasses adjacent structure
   iv. Important to check for retroperitoneal involvement:
      1. Chest radiographs, serum immunoprotein electrophoresis, thoraco-abdominal CT, and depending on the case bone marrow aspiration

e. Orbital Pseudotumour
   i. Affects younger population (less than 50 y.o.)
   ii. Diagnosis of exclusion
   iii. Pain is key symptom along with associated proptosis and EOM restriction
   iv. Can present as optic neuropathy with visual field loss, abnormal color vision, ophthalmoplegia, and afferent pupillary defect
v. Laboratory studies that should be ordered include CBC with differential, ESR ANA, and ACE
vi. CT scan shows poorly defined margins, enhancement due to infiltration
vii. MRI is useful tool to differentiate inflammation from hemorrhage and neoplasm
viii. If the diagnosis is uncertain, then an orbital biopsy is necessary

f. Neurilemoma/Schwannoma
   i. Affects all ages, however, rarely children
   ii. Benign, slowly progressive tumors arising from schwann cells
   iii. Occur as a single cystic lesion in orbit and can mimic intraconal lesions like cavernous hemangiomas
      1. MRI is useful differentiation between cavernous hemangioma vs. Schwannoma
   iv. Located typically around sensory nerves of orbit: supratrochlear, lacrimal, and supraorbital nerve
   v. Diagnosis can be confirmed with biopsy showing encapsulated, well-defined spindle cell lesions

D. Diagnosis and Discussion
a. Diagnosis:
   i. Cavernous Hemangioma

b. Features of Condition:
   i. Most common adult orbital tumor, affects 40-60 y.o.
   ii. Patients present with painless, decreased visual acuity or diplopia
   iii. If pressing on globe, may see choroidal folds, induced hyperopia, increased IOP, and strabismus
   iv. Rare cases patient may complaint of gaze-evoked transient blurring or headaches
   v. Benign, slow-growing with symptoms that occur in later stages
   vi. During pregnancy it may enlarge
   vii. Bilateral cases are rare
   viii. Orbital CT scan should be ordered with contrast:
      1. Well-defined intraconal or extraconal mass
      2. No bony erosion
      3. Adjacent structures are not destroyed or invaded, however, there may be some displacement of structures
   ix. If MRI not contraindicated will show:
      1. Hypointense on T₁-weighted, Hyperintense on T₂-weighted
      2. Significantly homogenous on T-2 weighted is key feature
         a. There is “progressive enhancement”, meaning there is an initial small area of enhancement followed by more enhancement sustained at an “equilibrium” into later phases

c. Incidence:
   i. Accounts for 80% of orbital tumors
ii. 9.5% to 18% of tumors that expand in size
iii. Most common presenting symptom is painless proptosis

d. Treatment:
   i. Severity of the condition determines the management plan. Severity depends on whether or not the mass is affecting vision and causing symptoms
      1. Normal findings with exam and ancillary testing rules out compressive optic neuropathy secondary to mass
      2. Monitor the condition, patient will return to clinic 1 year for repeat visual field testing, color vision, exophthalmometry, and dilated fundus examination
         a. Depending on severity of symptoms and visual field, patients should be monitored more frequently with semi-annual visits
      3. Consider orbital excisional biopsy in future if necessary. Not necessary for an invasive procedure at this time into the orbit when there no threat to vision
   ii. In this particular case, observation is the best option. Patient did not present with proptosis, EOM restriction, decreased vision, diplopia, and other ocular signs and symptoms
   iii. Most cases surgical intervention is not required
   iv. In cases where symptoms and signs are present:
      1. Orbital excisional biopsy should be performed to confirm diagnosis
      2. Most cases these lesions are encapsulated and easy to remove
      3. Surgical method is based on location of the lesion, in most cases a lateral or anterior orbitotomy is performed
      4. Cryoprobe can be used, but disadvantage is the potential for freezing adjacent structures when using with deeper lesions

e. Recent Literature:
   i. Indicates excisional surgery should be withheld for asymptomatic lesions until threaten to the vision or undesirable cosmetic appearance

f. Surgical Risks
   i. Capsular rupture resulting in bleeding during excision can lead to mortality
   ii. Risks associated with general anesthesia
   iii. Further pitfalls discussed

g. Conclusion
   i. The first diagnostic tool to perform is a CT scan. It is important for differentiating between a cystic, non-infiltrative, and infiltrative lesions
   ii. The clinician to narrow the differentials, must determine whether it is encapsulated versus non-encapsulated, furthermore, whether or not the lesion is inflammatory or infectious in nature
      1. Infectious and inflammatory lesions will show moderate to maximum enhancement
iii. Location of the lesion can assist the clinician in ruling out certain types
   1. Cavernous Hemangiomas typically do not invade adjacent structures
   2. Lymphoma typically molds to structures and encompass in area
   3. Fibrous Histiocytoma tend to be attached to surrounding bone
   4. Orbital Pseudotumor vary in location
   5. Schwannoma typically intraconal lesions

iv. Differentiating between schwannoma and cavernous hemangioma:
   1. MRI shows distinct features
      a. Orbital cavernous haemangiomas, significantly homogenous
         is key feature, hyperintense with T-2, more defined-ovoid
         intraconal
         i. MRI shows more “progressive enhancement”
      b. Schwannomas are heterogeneously, isointense or mildly
         hyperintense, located either intraconal or extraconal region,
         and not as defined in shape
   2. Location of schwannomas are typically near sensory nerves of the
      orbit, resulting in being located outside of orbital muscle cone versus
   3. Schwannomas affect all ages, while hemangiomas affect older
      population
BIBLIOGRAPHY


