**Title:** Sudden Sunken Eye: A case of sudden onset unilateral enophthalmos in the absence of trauma

**Abstract:**
Enophthalmos in the absence of trauma is rarely encountered in the clinical setting. This case report examines the evaluation, management and treatment of the differential diagnoses of unilateral enophthalmos.

I. Case History
   o Patient Demographics
     ▪ 52 year old black male
   o Chief Complaint
     ▪ Referral from the emergency department: Occasional left eye pain that feels like something is “pulling his eye in” starting 3-4 days ago. Pulling sensation increases when right eye is covered. Left eye appears more sunken in than it has in the past. Accompanied by feeling of fluid draining down from the inside of nose on the left side.
   o Ocular, Medical History
     ▪ Ocular History
       • LEE: Unknown
       • Denies trauma, diplopia, surgeries
       • History of “lazy eye” when he was younger, was not treated
     ▪ Medical History
       • (-) DM, HTN
       • h/o of appendectomy
       • Smoker: ~10 cigarettes/day
       • NKDA

II. Pertinent Findings
   o DVAsc:
     ▪ OD: 20/20-2
     ▪ OS: 20/30-2 PH: 20/20-2
   o PERRL-APD
   o EOM: Mild elevation deficit on ductions and versions OS: otherwise smooth, accurate and extensive OU
   o Confrontational Fields: Full to finger counting OU
   o Cover Test sc: ~15^ Constant alternating exotropia at near, ~15^ intermittent alternating exotropia at distance
   o Hertel Exophthalmometry
     ▪ OD: 16mm
     ▪ OS: 12mm
- **Base:** 102

- **Slit Lamp Exam**
  - **Lids/Lashes:** 1+ stenosed glands OU
  - **Conjunctiva:** Clear OU
  - **Cornea:**
    - OD: Arcus 360
    - OS: Small, circular, non inflamed phlyctenule inferior nasal limbus, (+) Staining; arcus 360
  - **Iris:** Flat and clear OU
  - **Anterior Chamber:** Deep and quiet OU
  - **Angles:** Open OU

- **Intraocular Pressures**
  - OD: 11mmHg @ 9:49am GAT
  - OS: Unable to obtain pressures with GAT or tonopen (due to enophthalmos and patient cooperation)
  - **Tactile:** Tender and equal to palpation
  - Attempted IOP measurement post dilation OS: 10mmHg @ 11:06am Tonopen

- **Dilated Fundus Exam (1gtt 1% Tropicamide and 1gtt 2.5% Phenylephrine hydrochloride @ 10:03am)**
  - **Lens:** Clear OU
  - **Vitreous:** Clear OU
  - **C/D:**
    - OD: 0.50 round, pink and distinct margins
    - OS: 0.50 round, pink and distinct margins
  - **Macula:** Flat and clear OU
  - **Vessels:** 2/3 OU
  - **Periphery:** Flat and intact, no holes, tears, breaks or detachments 360 OU

- **Radiological Imaging: CT Scan of Orbits with Contrast**
  - Marked volume loss of the left intraorbital fat and marked crowding of the left intraorbital soft tissue structures
  - Mild enlargement of the left medial rectus with small amount of “dirty fat” infiltrating the surrounding fat planes and around the anterior portion of the left superior rectus
  - Mildly more prominent left optic nerve sheath complex with mild uniform peripheral enhancement and mild enlargement of the left superior ophthalmic vein
  - Multiple small roundish calcifications at left inferolateral extraconal space and a tiny round calcification in the left superomedial extraconal space
  - Minimal inflammatory mucoperiosteal thickening in the maxillary sinuses and focally in some anterior ethmoidal air cells
- Orbital walls and orbital floor including the maxillary floor intact
- Incidental findings of marked periodontal disease

III. Differential Diagnosis
   a. Primary/Leading
      i. Orbital varix
      ii. Inflammatory orbital pseudotumor
      iii. Silent sinus syndrome
      iv. Scirrhous metastatic cancer
   b. Others
      i. Pseudoenophthalmos
      ii. Age related fat atrophy
      iii. Injection related tissue atrophy

IV. Diagnosis and Discussion
   a. Enophthalmos is a posterior displacement of the globe. Unilateral enophthalmos is normally characterized by a difference of >2mm on Hertel exophthalmometry. Trauma is the leading cause of cases seen in a clinical setting (4).
   b. Orbital varix
      i. Rare congenital disorder affecting men and women equally (3).
      ii. Low pressure and low flow vascular hamartoma with distendable vessels (6).
      iii. May be undetectable on CT scan, but becomes more evident when the CT scan is done while performing a Valsalva maneuver (8). Thrombosis in the vessels can be visualized as phleboliths or tiny round calcifications (2).
      iv. Pathogenesis: Enophthalmos may be caused by pressure induce orbital fat atrophy (1,2).
   c. Inflammatory Orbital Pseudotumor
      i. Usually presents with unilateral proptosis and painful ophthalmoplegia and may be associated with systemic autoimmune disease such as Wegener’s Granulomatosis
      ii. CT imaging may show “dirty fat” and involve a single extraocular muscle (9).
      iii. Pathogenesis: Thought to occur post-inflammation. Fibrosis replacing areas of inflammation causes contracture of the orbital socket (14).
   d. Silent sinus syndrome
      i. A rare condition in which patients present with painless unilateral enophthalmos. Patients may be asymptomatic and present due to cosmetic appearance.
      ii. Pathogenesis: atelectasis of the maxillary sinus in the absence of trauma. Occlusion of the osteomeatal complex causes mucous accumulation, eventually causing a low grade
inflammation leading to osteolysis of the sinus walls and orbital floor. A negative pressure is created towards the imploding sinus pulling in the contents of the orbit along with it (14).

e. Scirrhous metastatic cancer
   i. In women, enophthalmos may be an indicator of metastatic breast cancer. Breast cancer is more likely to metastasize to the orbit than any other cancer. It may also be indicative of lung, prostate, and stomach cancers (4).
   ii. Pathogenesis: Soft tissue dysplasia can cause contracture from fibrosis and muscle invasion (1).

f. Pseudoenophthalmos
   i. May be a result of altered lid position, microphthalmos, phthysis bulbi, facial asymmetry, contralateral exophthalmos, or structural lesions (4).

g. Age related atrophy
   i. Natural cause that is more likely to cause an appearance of bilateral enophthalmos (4).

h. Injection related fat atrophy
   i. Repeated injections of corticosteroids or recreational drugs into the same area have been shown to produce soft tissues atrophy.

i. Leading differential diagnosis based on current findings and testing:
   Orbital varix or inflammatory pseudotumor
   i. On initial presentation, the leading differential diagnosis was silent sinus syndrome, however the CT scan showed intact orbital bones and an intact maxillary sinus.
   ii. It is less likely to be metastatic cancer due to patient demographics.
   iii. Based on the CT scans, findings of “dirty fat” surrounding the medial rectus points to an etiology more inflammatory in nature. Calcifications found on the CT may also be indicative of a possible orbital varix that was not visible on the CT scan that was taken. Further blood work up would be necessary to single out a diagnosis.

V. Treatment, Management
a. Following ocular examination, the patient was referred for a CT scan as described above. Further imaging with MRI and laboratory testing for ESR, C-reactive protein, cANCA, ANA, and anti single stranded DNA antibodies to rule out any inflammatory or infectious etiologies
b. Treatment of the underlying cause should be initiated. Treatment of orbital varices is normally conservative in nature. If the cause is inflammatory in nature, a course of systemic steroids can be started. Periorbital fillers can be used to improve cosmesis. Referral to
oculoplastics for reconstruction of the orbit would be necessary if the etiology was due to structural changes.

VI. Conclusion
a. Patients presenting with unilateral enophthalmos in the absence of trauma should be carefully examined in order to determine an underlying cause. Symptoms may be absent hence it is up to the clinician to perform a detailed case history and examination of externals. Hertel exophthalmometry should be utilized for evaluation to assess the difference in the two eyes. Imaging of the orbit can provide more information regarding structural changes. A blood workup should also be performed to rule out inflammatory or infectious causes enophthalmos.

References:


