Acute Idiopathic Orbital Inflammatory Syndrome
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Abstract
Orbital inflammatory pseudotumor is a diagnosis of exclusion. Laboratory testing and imaging are essential to rule out an infection or systemic autoimmune disease. Oral corticosteroid therapy typically gives a swift recovery of clinical symptomatology.

- Case History: 72 year old Caucasian male
  - CC: red left eye with pain on eye movement and horizontal binocular diplopia when looking to the left worsening over past 1.5 weeks
  - Ocular History: Unremarkable
  - Medical History: Hypertension, bipolar disorder, recent toothache
  - Medications: Divalporex sodium 500mg nightly, hydrochlorothiazide 25mg ½ tab daily, aspirin 81mg daily

- Pertinent Findings
  - Clinical
    - Entrance testing:
      - VA: 20/20 OD, OS
      - Pupils: mild physiological anisocoria, round and reactive to light; no APD
      - EOM: Abduction deficit OS
      - CTsc:
        - 2°pd exophoria @ 6m (primary gaze)
        - 2°pd exophoria @ 6m (right gaze)
        - 10°pd constant left esotropia @ 6m (left gaze)
    - Externals:
      - Hertel exophthalmolmetry: 18 OD, 20 OS, 113 base
      - Mild resistance to retropulsion OS
      - Forced duction testing: attempted with ambiguous results
    - SLE:
      - Lids: 4mm left ptosis, mild left upper lid edema
      - Conjunctiva: mild-mod chemosis and bulbar injection OS
    - DFE:
      - Optic nerve head: 0.30 round with distinct margins OD, OS
  - Visual field testing: SITA fast 24-2
    - Unremarkable OD, OS
  - Laboratory Studies
    - CBC, ESR, ACE, ANA, cANCA, thyroid panel, fasting glucose: Non-contributory except for mildly elevated WBC and neutrophils
  - Radiology studies
    - MRI of head with and without contrast: Asymmetric thickening of the muscle belly of the left medial rectus muscle with surrounding inflammatory changes and diffuse infiltration of the intraconal fat.
• Differential Diagnosis
  o Primary / leading: Orbital pseudotumor/mass
  o Others:
    ▪ Orbital cellulitis
    ▪ Orbital lymphoma
    ▪ Graves disease
    ▪ Wegener’s granulomatosis
    ▪ Carotid cavernous fistula

• Diagnosis and Discussion
  o Orbital pseudotumor, also known as idiopathic orbital inflammatory syndrome, is an acute benign inflammatory condition that can affect any soft tissue of the eye. It is unrelated to any systemic autoimmune disease, and there is no sex, age, or race predilection. It may involve the entire orbit or present in a localized fashion involving the anterior orbit, posterior orbit, lacrimal gland, or extraocular muscles. Lymphocytes, plasma cells, macrophages, and polymorphonuclear cells infiltrate the tissue causing inflammation that then leads to fibrosis. It is a diagnosis of exclusion. Corticosteroids are the treatment of choice and a reduction in symptoms can be seen in a few days; therefore, a hallmark sign of orbital pseudotumor is a prompt response to steroid treatment. Oral prednisone starting at a dose of 80-100 mg daily is used for 2-3 weeks followed by a taper.
  o Initially, the leading diagnosis for this patient was an orbital mass or inflammation causing lid ptosis, globe proptosis, and an abduction deficit. However, with a presentation of lid edema and pain, an infectious etiology such as orbital cellulitis must be considered. In this case, the patient was afebrile, had only a slightly elevated WBC count, and his symptoms evolved slowly over one week making acute orbital cellulitis unlikely. Additionally, thyroid eye disease as well as systemic autoimmune disease was ruled out with laboratory studies such as thyroid function tests (TSH, free T4), ESR, ANA, ACE, and cANCA being non-contributory. Anti-nuclear antibody testing was ordered to rule out systemic lupus erythematosus; angiotensin converting enzyme testing was ordered to rule out sarcoid; anti-neutrophil cytoplasmic antibody testing was ordered to rule out Wegnener’s granulomatosis.
  o In cases suspicious for an orbital mass, imaging is indicated, and computed tomography is the typically the most common form. Magnetic resonance imaging, however, offers advantages in distinguishing inflammation from neoplasia and hemorrhage due to it is superior soft tissue imaging properties. This patient underwent MRI of the head which showed asymmetric thickening of the muscle belly of the left medial rectus with surrounding inflammatory changes; therefore, this patient’s presentation was most consistent with myositic orbital pseudotumor.
  o Symptoms of myositic orbital pseudotumor include the sudden onset of unilateral pain, swelling, and double vision. Because the belly of the left medial rectus was enlarged the patient demonstrated an abduction deficit OS. This case serves to highlight the distinction that must be made between a muscle restriction and a true cranial nerve palsy. Even though the patient demonstrated an incomitant esotropia on left gaze, his diplopia was not due to a neurological problem (ie left
abducens palsy) but rather could be attributed to an infiltration of the medial rectus muscle itself.

- **Treatment and Management**
  - The patient was co-managed with an oculoplastics specialist and prescribed prednisone 80mg daily for 2 weeks. At the four day follow up, the patient had subjective improvement in symptoms. There was minimal pain with left gaze and mild diplopia on far left and upgaze. The ptosis had resolved. At the one week follow up, the orbital pain, lid swelling, erythema and diplopia had all improved significantly. The patient was to continue 80mg prednisone for another 1-2 weeks and then begin a slow taper.

- **Conclusion**
  - Individuals with acute idiopathic orbital inflammatory syndrome tend to present with symptoms of sudden onset of eye pain, restricted ocular motilities, diplopia, eyelid edema, erythema and proptosis. Orbital pseudotumor may present as localized or diffuse inflammation, and depending on the tissue affected, it can result in permanent vision loss. Laboratory testing and imaging is necessary for a proper diagnosis. Treatment involves high dose oral prednisone with gradual tapering. Although a rapid improvement in symptoms is expected, the inflammation can recur and if not treated properly, may become chronic.

- **Bibliography**