Idiopathic Orbital Inflammatory Syndrome in a Pediatric Patient, a Diagnosis of Exclusion.

A 15 year old male with prior diagnosis of orbital cellulitis and worsening symptoms despite treatment presents to the clinic with findings consistent with Orbital Inflammatory Syndrome.

I. Case History
• 15 yo black male
• Presents to clinic for severe eye pain and worsening orbital swelling that has been progressing over the past 2 days. Patient was recently seen in the ER and diagnosed with Preseptal Cellulitis and treated with Keflex and Bactrim since being discharged by the Emergency Room. Patient is afebrile but reports his eye feels “tighter” and more painful. Patient reports being unable to keep down his oral antibiotics, prescribed to him before he was discharged.
• Pt. had previously been seen by an Ophthalmologist who diagnosed him with orbital cellulitis and sent him directly to the ER for orbital imaging, IV antibiotics, and hospital admission. The Emergency Room preformed requested imaging and then chose to discharged patient with a prescription for Bactrim BID and Cefalexin 500mg TID.
• Past Medical History: Asthma, eczema, severe allergies, and acute otitis media.
• Past Surgical History: Sinus surgery and adenoidectomy.
• Medications: Cetirizine, Fluticasone Propionate, Proventil Inhaler, Bactrim, Cefalexin
• No recent URI, trauma, current fever, or malaise.

II. Pertinent findings
• Clinical:
  - Best Corrected Snellen Visual Acuity : 20/25 OD, 20/20 OS
  - Elevation deficit of the right eye with diplopia reported on left gazes. Pt reports diplopia when eye is manually opened and pain in right and left gazes.
  - Color Vision was normal OD, OS
  - Pupils were equal, round, and reactive to light. No afferent pupillary defect.
  - Patient is afebrile and denies feeling ill
  - Hertel Exophthalmometer readings were 24mm OD/ 19mm OS with a base of 112
• Physical:
  - External Evaluation: RUL is swollen shut. The patient’s skin is not tense, but is extremely erythematous.
  - Globes are resistance to retropulsion.
  - Severe pain on lacrimal gland palpation.
- Biomicroscopy exam revealed 4+ temporal chemosis with 2+ injection of the right eye, all other anterior segment findings were normal. All posterior pole findings were normal.

- Laboratory studies:
  - Pt had an abnormal complete blood count with differential showing elevated white blood count, and the patient also had and elevated C-Reactive Protein and Erythrocyte Sedimentation Rate

- Radiology studies:
  - A primary CT of the orbits without contrast was taken in the Emergency Room showing preorbital soft tissue swelling over the right eye with no fluid collection or retro-orbital extension. These finding were consistent with preseptal cellulitis.
  - A second CT of the orbits with contrast was preformed before hospital admission two days later with similar, but slightly worsened findings.
  - An MRI of the orbit, face, neck with and without contrast was performed before hospital discharge to rule out an orbital mass. The scan showed an enlarged right lacrimal gland compared to the left side with preseptal swelling, without any orbital masses or neoplasms

III. Differential diagnosis
- Primary diagnosis: Idiopathic Orbital Inflammatory Syndrome
- Others:
  - Orbital Cellulitis
  - Pre-septal Cellulitis
  - Thyroid Eye Disease
  - Wegner Granulomatosis
  - Sarcoidosis
  - Primary Orbital Malignancy
  - Lymphoproliferative disease
  - Tolosa-Hunt Syndrome
  - Carotid Cavernous Fistula

IV. Diagnosis and discussion
- Idiopathic Orbital Inflammatory Syndrome is a benign, non-infective syndrome of unknown etiology characterized by inflammation of the orbit and surrounding structures. It is the third most common orbital malignancy.
- Patients typically present with symptoms of deep, boring headache with orbital pain, chemosis, redness, proptosis, uveitis, disc edema, and eosinophilia. Symptoms usually progress very quickly.
- This condition is rare in pediatric populations, and pediatric patients often present with more severe symptoms than in adult populations.
• Pediatric patients are more likely to present with bilateral periorbital edema, however, when adult patients present with this finding, an systemic evaluation for systemic vasculitis or lymphoproliferative disease
• While the patient had previously been diagnosed with Preseptal Cellulitis, the presence of proptosis, pain on eye movement, and dense conjunctival injection are symptoms more associated with Orbital Cellulitis. However, as Idiopathic Orbital Inflammatory Syndrome is a diagnosis of exclusion, it was necessary to first treat the patient as if he actually had orbital cellulitis.
• The fact that the patient did not improve as expected with IV antibiotics, was afebrile, and had abnormal complete blood count with differential, elevated C-Reactive Protein, and Erythrocyte Sedimentation Rate all supported the diagnosis of Idiopathic Orbital Inflammatory Syndrome.

V. Treatment, management
• Idiopathic Orbital Inflammatory Syndrome’s most common and accepted treatment is corticosteroid therapy, but antimetabolites, alkylating agents, cytotoxic agents, and other immunosuppressive agents have recently been considered for alternative therapy. Antibiotics are often used in conjunction with corticosteroid therapy. Low-dose radiation may be considered if patient does not respond to oral corticosteroid therapy or if disease recurs during taper period, but should only be used after an orbital biopsy has ruled out all other etiologies
• Orbital decompression surgery is rarely needed, and is only necessary when there is evidence of compressive optic neuropathy
• Once the examination was complete, Pediatric Service was contacted and agreed to meet patient in the Emergency Room for evaluation and admission. Patient was treated with 600mg IntraVenous Clindamycin Q8H. Patient’s symptoms showed minor improvement over the next 24 hours, so patient was given 1g of IV solumedrol overnight. Patient then had significant improvement and was discharged a few days later with prescription for oral prednisone 60mg daily, which he will continue for 60 days and oral antibiotics for 7-10 days. Patient returned to clinic on 2 weeks later where all symptoms had resolved and only minor conjunctival injection remained.
• The patient will be monitored by his pediatrician and thoroughly evaluated by rheumatology to rule out any systemic inflammatory disorders or lymphoma.

• Bibliography:


VI. Conclusion

- Idiopathic Orbital Inflammatory Syndrome is a diagnosis of exclusion. It is necessary to rule out an infectious etiology before progressing with corticosteroid treatment.
- While this disease usually responds well to treatment, it is necessary to warn patients that recurrence is common.
- It is essential to properly diagnose Idiopathic Orbital Inflammatory Syndrome and rule out all differential diagnoses, especially those that may be life threatening, like systemic vasculitis, orbital malignancies, or lymphoma.
- Proper consults are medically necessary to properly diagnosis this condition and manage it properly. Ophthalmology, pediatric, and oculoplastics consults were all critical parts of this patient’s care. A rheumatology consult is also necessary to rule out any systemic inflammatory disorders or lymphoma.