Abstract
A case of recurrent orbital myositis presenting with progressive diplopia and strabismus is discussed. The pertinent clinical findings, investigations, treatment and management are presented, with emphasis on the clinical response to corticotherapy.

Case History
Demographics and Chief Complaint: 27 yo White Female was referred for neuro-ophthalmology consultation for progressive double vision for 3 months. She also reported recent history of redness and pain in the left eye for the past 10 years.

Ocular History
Orbital Myositis for 10 years.
Recurrent episodes of orbital inflammation were treated with varying strengths of oral steroids. Patient is now steroid dependent.

Medical History
Migraine
Pseudo tumor removal from the sinuses.

Medications
Prednisolone 25mg qd and Methotrexate 1ml subcutaneous weekly for orbital myositis, Ortho Tri-cyclen qd, Multi-vitamins and Folic acid supplements.

Clinical ocular findings
The Best Corrected Visual Acuities (BCVA) was 20/20 with a correction of plano-1.00x100 OD and plano-1.75x76 OS. Cover test revealed 20-prism diopters (pd) esotropia in the left eye, worsening in right gaze (35pd) and 10 pd hypertropia in the right eye in primary gaze. Sensory exam with Worth-Four dot test showed diplopia at distance and near. Stereoscopic acuity was 800 arc seconds at 40cms. Extra-ocular movements were full in the right eye. Left eye showed restricted elevation and abduction. Color vision and visual fields (Humphrey 30-2) were unremarkable. Intraocular pressures were 20 in each eye. Dilated fundus exam was unremarkable in both eyes. Review of MRI from the past revealed thickening of medial rectus muscle in the left eye involving the tendon. Investigations for potential autoimmune disorders including TSH, ACE, ANA, ESR, RPR and CBC were negative. After observation for one month, the patient showed worsening of esotropia in primary gaze (50 pd esotropia in the left eye with 20 pd hypertropia in the right eye at distance). Surgical intervention was opted at this time. Forced Duction Test showed restriction in abduction and elevation in the left eye, consistent with the MRI findings noted before. The left medial rectus was recessed by 10 mm and the left inferior rectus was recessed by 6mm. During follow-up, three months after the surgery, patient had developed a consecutive exotropia in the left eye, measuring greater than 50 pd with 30pd left hypertropia at distance. Non-comitancy was noted in all the secondary gazes. To reduce the diplopia and for cosmesis, a lateral rectus recession (8mm) was performed in the left eye, with a post-operative alignment of 30pd left exotropia and 30pd left
hypertropia at distance and an intermittent exotropia (8pd) with 12pd left hypertropia at near. To minimize diplopia in primary gaze and down daze, a 10pd base down Fresnel prism was dispensed in the left eye. At one-month follow-up, the exotropia was intermittent at distance and near with persistent left hyper deviation (16 pd at near and 30 pd at distance). The patient reported significant reduction in diplopia and was happy with the surgical outcome. She will be followed in 3 months. Future course of treatment to reduce the vertical diplopia would be to perform an anterior transposition of the left inferior oblique muscle.

**Diagnosis**
Orbital Pseudo tumor causing restrictive orbital myopathy.

**Differential Diagnosis**
Thyroid Eye Disease; Tolosa-Hunt Syndrome.

**Discussion**
Orbital pseudo tumor is a benign inflammatory condition involving the tissues in the orbit and is often classified based on the anatomical structure affected within the orbit. When single or multiple extra ocular muscles are involved the condition is termed orbital myositis. The pathophysiology is poorly understood. The disease is commonly seen in middle-aged adults with 2:1 female preponderance. Common symptoms of this disease include orbital pain exacerbated on eye movements and diplopia. Other less common symptoms include conjunctival chemosis, conjunctival injection and periorbital edema. Orbital myositis can commonly be confused with Thyroid eye disease. However, the latter is characterized by acute and severe onset of pain and rapid response to systemic steroid therapy. About 90-95% of myositis is unilateral. In a retrospective study of 100 patients with orbital myositis, fifty one (68%) had single muscle involvement, with the lateral and medial recti being affected most frequently. Positive response to steroid therapy was noted in fifty one (68%) of patients. Systemic involvement in orbital pseudo tumor includes Crohn’s disease, scleritis, systemic lupus erythematositis and rheumatoid arthritis. Imaging studies like computerized tomography (CT), Magnetic Resonance Imaging (MRI) and Ultrasound have often been helpful to identify the exact location of inflammation. Histology of the pseudo tumor consists of nonspecific polymorphic, lymphocytic infiltrates with macrophages, polymorphonuclear leukocytes and eosinophils.
**Treatment and management**
Systemic corticosteroid therapy has been the treatment of choice for orbital pseudo tumor with dramatic improvement noted within 24-48 hours of treatment. Common starting dosage includes 1.0-2.0mg/Kg/day and is tapered when improvement is noted. Other treatment options include cytotoxic agents like methotrexate, cyclophosphamide and infleximab. Radiation therapy has been considered in cases resistant to corticosteroid therapy. In a retrospective study\(^5\) of 133 patients with orbital inflammations, radiation therapy was found to be successful in nine out of fourteen patients found resistant to corticosteroids. Recurrences have been noted in bilateral cases.

In this case, diplopia management was an important component to treatment to improve the patient’s quality of life. The strabismus observed in these patients is usually non-comitant, due to the restrictive myopathy. Myositis usually responds well to oral corticosteroid therapy. In large angle non-comitant deviations, surgery has been indicated. One study\(^6\) reported persistent diplopia in a patient with orbital myositis, despite multiple surgeries to correct a large exotropia. The same is being observed in our patient discussed here. Fresnel prisms are valuable in minimizing the residual diplopia, which surgeries failed to correct. They are also valuable in cases where the diplopia is transient.

**Conclusion**
Orbital pseudo tumor is a benign disease commonly affecting females and accounts for 5-10% of inflammatory conditions in the orbit. A hallmark and a unique identifier of this disease is the rapid response to corticosteroid therapy. Adequate knowledge and understanding of the clinical course of this condition will help prevent visual loss and associated morbidity.
Bibliography