Diffuse bilateral non-specific orbital inflammatory disease: A case report

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Abstract
Non-specific orbital inflammatory disease (NSOID) is a inflammatory condition involving orbit and adnexa. It has is a recurrence with relapse & remission. It may cause irreversible damage and loss of function. On the contrary, prompt management may reduce the complications. Here we will present a case of a patient of non-specific orbital inflammation managed by pulsed corticosteroid therapy which reduces the disease progression and vision threatening condition in one eye. But, unfortunately he lost his vision in another eye.

Keywords: Non-specific orbital inflammatory disease, CT scan of orbit, Pulsed steroid therapy.

Introduction
Non-specific orbital inflammatory disease NSOID is the third most common orbital disease following thyroid eye disease and orbital lymphoma and accounting–for 4.5% to 10% of all orbital lesions. It may associated with lower socio-economic status, higher BMI and use of oral bisphosphonates. NSOID are variable from diffuse involvement of orbital tissues to localized tissue, the extraocular muscles (orbital myositis), lacrimal gland (dacryoadenitis), sclera (scleritis), uvea (uveitis), superior orbital fissure and cavernous sinus (Tolosa-Hunt syndrome).[1-3] NSOID often mimic a malignant condition clinico-radiologically. The definitive diagnosis are based on clinical features and CT scan of the orbit. CT scan finding as a focal or diffuse mass, usually poorly demarcated and enhancing with contrast allows a definitive diagnosis in most cases. MRI imaging, however, is indicated for evaluation of the Tolosa-Hunt syndrome.[3-5] The histopathological spectrum of idiopathic orbital inflammation is typically non-diagnostic, wide, and diverse, ranging from the typical diffuse polymorphous infiltrate to the atypical granulomatous inflammation, tissue eosinophilia, and infiltrative sclerosis.[6,7] Treatment options are varied and can include steroids, chemotherapeutic agents, irradiation and surgery.[3]

Case Report
A 42 years old male patient presented with complains of gradual protrusion of both eyeballs for 3 months. At the same time he developed gradual dimness of vision in both eyes associated with redness, grittiness, watering & itching. He was smoker (about 5 sticks/day), non-diabetic, and non-hypertensive. He had no other complains like tremor, heat intolerance, visible swelling at neck.

On Examination, His visual acuity was no perception of light (NPL) in right eye & hand movement in left eye. He had bilateral severe proptosis (Fig. 1). Eyelids were swollen in both eyes. Conjunctiva in both eyes was hugely chemosed. Severe exposure keratopathy was found in right cornea raher than left cornea. Ocular motility was restricted in both eyes. Eyeballs were severely proptosed axially measuring 30 mm in right eye & 29 mm in left eye. CT scan of the orbit (Fig. 2) showed diffusely large, hyper dense lesion in both orbit extending from anterior to apical part surrounding both globes resulting bilateral proptosis of both eyeballs. Extraocular muscles & optic nerves on both sides could not be clearly isolated from the lesion. After IV contrast, mild enhancement of the lesion was noted. This suggested bilateral diffuse non-specific orbital inflammatory disease. Dysthyroid ophthalmopathy, and Non- Hodgkin's lymphoma were also in suspicion. Thyroid function tests, complete blood count, blood sugar profile, Chest X-ray and ECG revealed normal studies.

He was treated by intravenous injection of 1 gram methyl prednisolone in 500 ml isotonic saline over 30 minutes in alternate days under supervision of an internist. 6 doses were given. Then oral prednisolone was started in a dose of 1 mg/kg body weight with tapered dose up to 3 weeks and maintained dose 10-20 mg up to 4 months. Exposure Keratitis of both eyes was managed by cornea specialist.

Fig. 1: Bilateral diffuse and huge NSOID with exposure keratopathy

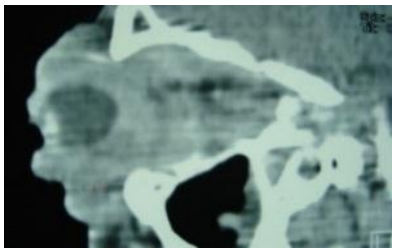

Fig. 2: CT scan of the orbit – bilateral diffuse and huge NSOID

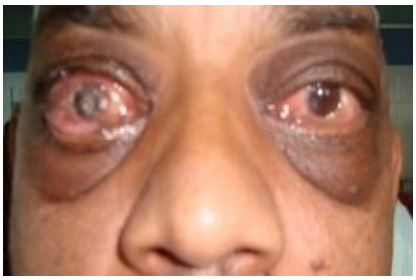

Fig. 3: Regressing the NSOID on treatment (Pulsed corticosteroid therapy)

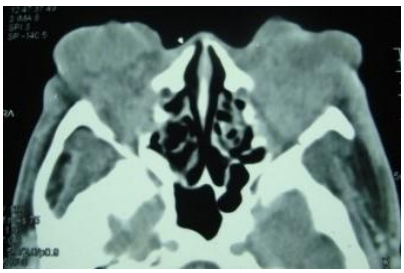

Fig. 4: Axial view of CT scan of the orbit- regressing NSOID

Fig. 5: Improving of the patient's condition on treatment

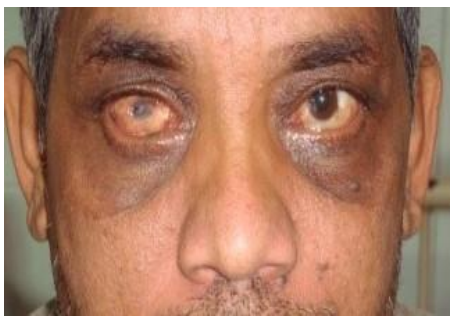

Fig. 6: After treatment of NSOID

Patient started improving day by day on treatment (Fig. 3 and Fig. 4). Proptosis and chemosis were reduced (Fig. 5). Cornea became clear in left eye, but in right eye its condition worsened (Fig. 6). On third week of management, he regained his vision. Words cannot say how happy he was. His visual parameters were improving gradually.

Discussion

Non-specific orbital inflammatory disease is a non-neoplastic, non-infective, space-occupying orbital lesion. The inflammatory process may involve any or all of the orbital soft tissues. Histopathological analysis reveals pleomorphic inflammatory cellular infiltration followed by reactive fibrosis.

Swamy et al. reported that 20.8% cases had remission.[8] Mannor et al. reported that NSAIDs could be used up to 3 weeks as long as clinical resolution was being observed, with steroids reserved for refractory cases.[2] It has been reported that systemic steroids are the mainstay of treatment for moderate to severe NSOID. Systemic prednisone can be started with an initial dosage of 60 mg to 100 mg per day for 1 to 2 weeks and a taper over 6 weeks. Yuen and Ruben were treated 69% cases with steroids alone, 12% with steroids and radiation therapy and 9% with steroids and NSAIDs. Twenty four patients had treatment failures with steroid dependence and steroid intolerance occurring 33% and 13% respectively.[6] Radiation therapy can be used in the treatment of NSOID resistant to or intolerant to corticosteroid therapy.[9] Immunosuppressive chemotherapy, consisting of either cyclophosphamide or chlorambucil combined with prednisone has been reported to be effective in the treatment of NSOID refractory to both steroid and radiation therapy.[7-8] Infliximab may be another therapeutic option in cases of recalcitrant or recurrent NSOID in which conventional treatment fails.[10] Surgical resection can be an effective form of treatment in NSOID refractory to treatment.[7-8] Pemberton and Fay reviewed seventeen articles with 56 biopsy-proven sclerosing NSOID. Regardless of 15 different treatment modality including steroids, radiation therapy and immunomodulatory drugs, the overall response was good in 19 (34%) patients, partial in 24 (43%), and poor in 13 (23%).[7] In our study, intravenous methylprednisolone followed by oral cortico-steroid therapy were the main regimens of treatment. Patient was improved dramatically day by day and restored the vision.

Conclusion

NSOID is a diagnosis of exclusion and diagnosis often made based on variable clinical presentations depending on the involved tissue, without pathologic confirmation. CT orbit play an important role in evaluation and distinguishing NSOID from other orbital lesions. Pulsed corticosteroid therapy are the treatment

of choice and can salvage the vision as well as the globe.

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