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AN UNUSUAL PRESENTATION OF ORBITAL PSEUDOTUMOR

Ophthalmology

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ABSTRACT

The term orbital pseudotumor refers to a broad category of non-specific idiopathic inflammations of the orbit, which may affect any orbital structure. A 28-year-old female presented with swelling in the right upper eyelid for 1 month associated with pain and mild redness of the eye. She was previously been seen by other ophthalmologists and diagnosed as Hordeolum Internum prescribed oral antibiotics and advised incision and drainage. She visited our OPD for secondary opinion and surgical treatment. Best Corrected Visual Acuity- R/E- 6/9, LE -6/6P. No Proptosis. No Limitation of Extra Ocular Movements. Considering the clinical presentation of the patient Computed Tomography orbit was advised, revealed abnormal soft tissue density in lateral aspect of right orbit within extraconal compartment suggestive of Orbital Pseudotumor. The patient was started on oral steroids and had a favourable outcome with no recurrence till date.

KEYWORDS

Extraocular movements, Orbital Pseudotumor, Recurrence

INTRODUCTION

Idiopathic orbital inflammatory syndrome (IOIS), also known as orbital pseudotumor is a non-granulomatous inflammation of orbital soft tissue for which no cause is found after local and systemic evaluation.[1] It is a diagnosis of exclusion based on clinical and radiographic findings. [2] It may be present acutely, sub acutely or chronically in one orbit. [3] Unilateral periorbital pain, cranial nerve palsies and dramatic response to corticosteroid therapy are the hallmarks of clinical presentation in IOP. [4] The aim of this study was to diagnose the clinical features, radiological evaluation and treatment outcomes in IOIS who had a favourable outcome with corticosteroid treatment.

Case Report-

A 28-year-old female presented with swelling in the right upper eyelid for 1 month. Swelling was associated with pain and mild redness of the eye. It was gradual increasing in size, progressive and more on lateral aspect of upper lid. No History of Ocular trauma or systemic disease. She was previously been seen by other ophthalmologists and diagnosed as Hordeolum Internum prescribed oral antibiotics and advised incision and drainage. She visited our OPD for secondary opinion and the surgical treatment. On Examination Best Corrected Visual Acuity R/E- 6/6, LE -6/6P R/E- Upper Lid- 1.5 X 1 cm localised, firm oval shaped swelling on lateral one third of lid, with smooth overlying skin and no changes in surrounding area. (Figure1)

Both eyes:

Anterior Segment-With in Normal limits, Reaction to Light – Present (NO RAPD), No Protrusion of eyeball, No Chemosis, Extra Ocular Movements- Full Range no restriction. (Figure2). On Fundus- Central fundus and disc – Normal (Figure3). On neurological examination, the cranial nerves were normal. Considering the clinical presentation of the patient following investigations were advised to confirm the diagnosis.

Investigations-

Routine blood test was With in Normal Limits. CT orbit revealed 23 X 21 X 20 mm size well defined, hyper intense, abnormal soft tissue density in lateral aspect of right orbit within extraconal compartment abutting lateral rectus muscle, eyeball, lacrimal gland and lateral wall of right orbit suggestive of Orbital pseudotumor.

Treatment-

Oral corticosteroids with the dose of 1 mg per kg body weight and tapered over a period of one and half month in the follow up done at an interval of 15 days (Figure4 and 5).Along with topical treatment using eye ointment chloremphenicol and eye drop Nepafenac. Case showed prompt response to steroid as the swelling subsided.Pseudotumor Orbit was the final diagnosis for the case, with no recurrence till date.

DISCUSSION-

Idiopathic Orbital Pseudotumor is the third most common orbital disease after thyroid orbitopathy and lymphoproliferative disorders.[5] It compromises 10% of the orbital mass leisons. Patient presents with proptosis, diplopia,conjunctival chemosis, visual disability and restriction of extraocular movements. [6]

However in our case patient presented only with unilateral upper eyelid swelling with no proptosis or ophthalmoplegia. Peak incidence appears to be predominantly in the adult population, typically in the middle-aged persons, and there is no sex predilection.[7]The disease has been reported in all ethnic groups around the globe.[2] Pathogenesis and etiology are currently unknown. [8]

The clinical course ranges from mild and self-limiting to devastating orbital sclerosis with blindness. [1] The diagnosis of orbital pseudotumor is usually clinical and confirmed by prompt response to steroids. Only a minority of patients do well without treatment.[9] Around 37% of patients treated with steroids showed failure to resolve. In these situations, methotrexate, cyclophosphamides and other antineoplastic agents can be used.

Radiotherapy is another modality of treatment. An alternative mode of treatment in recurrent and recalcitrant cases is with infliximab (TNF-alfa blocker).[10] In our case patient responded well to steroids with no recurence aftercompleting the course of treatment.

CONCLUSION-

Orbital pseudotumor represents a diagnostic and often therapeutic challenge for ophthalmologists, as in this case it presented in unusual manner. So, complete and detailed medical history and imaging studies are important before concluding towards final diagnosis. It is a diagnosis of exclusion that is associated with many other disorders. Although steroids are the mainstay of therapy, other therapeutic modalities and new immune suppresive medications may offer additional options in future.

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Figure 1: Image of Case with swelling in Right upper eyelid.

Figure 2: Extra Ocular Movements during Visit 1

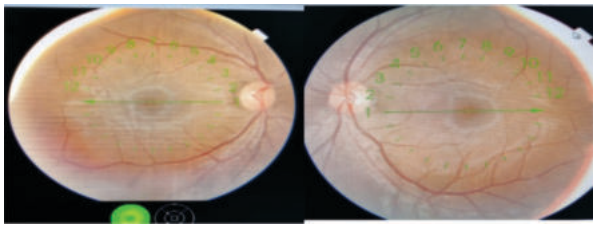

Figure 3: Fundus Photograph of both the eyes

Figure 4: Extra Ocular Movements in Visit 2 (after 15 days)

Figure 5: Extra ocular Movements in Visit 3 (After 15 days from Visit 2)

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