A Typical Presentation of Orbital Pseudotumor Presenting As A Large Medial Canthal Sub Conjunctival Cyst in A Young Adult Male

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Abstract :

Background : Pseudotumor of orbit is an unusual clinical entity with incidence of about 4-6%.

Case Report : A 26-year-old man presenting with redness, cystic swelling and pain at the medial aspect of the right eye since 1 week but was not associated with proptosis and ptosis. Considering the age and clinical presentation of the patient, all differential diagnosis of conjunctival cystic and inflammatory lesions were considered. Relevant laboratory investigations were performed but were normal except eosinophilia and raised ESR. Contrast enhanced magnetic resonance imaging reports were suggestive of IOIS. The patient was managed conservatively with favourable outcome.

Conclusion : All conjunctival cystic lesions need not to be operated unless confirmed otherwise.

Keywords : Idiopathic orbital inflammatory syndrome(IOIS),orbital pseudotumor, eosinophilia, immune mediated, conjunctival cystic lesions, proptosis, case report.

Introduction:

Orbital pseudotumor also known as Idiopathic Orbital Inflammatory Syndrome (IOIS) is a idiopathic, benign, noninfective inflammatory condition of the orbit which was first described by Birch-Hirschfield in 1905, as 'idiopathic orbital inflammatory syndrome.¹³

It comprises of 4.1–6.3% of orbital disorders and typically occurs in the adult population. Orbital Magnetic Resonance Imaging (MRI) is the single most important diagnostic test, but serological studies and incisional biopsy can be necessary to exclude a systemic cause.⁴

The aim of this study was to describe the clinical presentation, radiological features and favourable outcomes with corticosteroids in the rare cases of IOIS.

Case Report:

A 26-year-old man presented on 05-09-2019 with the chief complaints of redness, swelling and pain at the medial canthus of right eye since week. The swelling was associated with lid edema and restriction of ocular movements superiorly and medially since tweek. There was no history of fluctuation of symptoms or diminution of vision. There was no history of fever, rash, joint pain, any other systemic symptoms, preceding infection or trauma to the eye. There was no history of intake of any medications .Also there was no family history of similar complaints.

His general examination was normal. On neurological examination, the cranial nerves were normal except extraocular

movements. On ocular examination the extraocular movements were restricted superiorly and medially in the right eye. Visual acuity, pupillary reaction and fundoscopy were normal. There was no audible bruit on auscultation over the right eye. The intraocular tension in both eyes was normal. A possibility of inflammatory swelling was considered and patient was prescribed an antibiotic steroid combination eye drop along with systemic anti-inflammatory drugs and was asked to follow up after 3 days.

Due to certain personal reasons patient presented after 1 week with a localised cystic swelling near the medial canthus. The symptoms still persisted but the pain and redness was slightly reduced. Swelling was approx. 6mm X 7mm in size, well defined superior, inferior and lateral margins, cystic in constistency, non reducible, non mobile, lateral margin was 1mm away from the limbus but the medial margin was merged with the caruncle and was not clearly defined. The overlying conjunctiva was congested but was free however; cyst was adherent to the underlying tissues. (figure 1)



Figure 1 : Pre-treatment Photograph of the Patient Showing Right Eye Conjunctival Cystic Lesion in the Young Adult Male

All differential diagnosis of conjunctival cystic and inflammatory lesions were considered. From the battery of lab tests that were advised TLC was normal except with eosinophilia. The erythrocyte sedimentation rate was elevated. Liver, renal function, thyroid functions and stool examinations were normal.

A Computed tomography (CT) with intravenous contrast showed bulky medial rectus muscle with fluid filled hypodense cystic lesion near the medial rectus with focal bulge in the conjunctiva suggestive of pseudotumor of the right orbit (figure 2 & 3). Considering the radiological features and negative blood reports, the patient was diagnosed as Idiopathic orbital inflammatory syndrome.

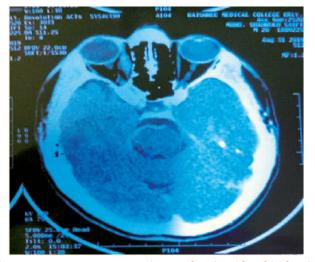


Figure 2: Pre –treatment CECT image of Brain with Orbit Showing Medial Rectus thickening in the Right Orbit.

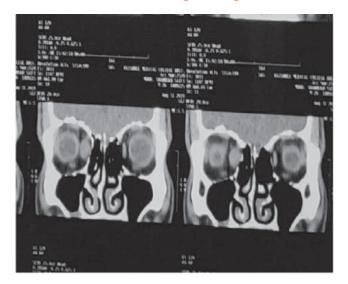


Figure 3: Pre-treatment CECT Orbit Showing Cystic Conjunctival Lesion the Medial Aspect of Right Eye.

He was started on high dose of oral prednisolone (1.5 mg/kg/day) for 15 days followed by tapering doses over 10–12 weeks. The patient started showing response after 48 h and, at the end of 2 months there was improvement in eye symptoms. There was progressive thinning of the cyst wall and conjunctival epithelium with spontaneous rupture and expulsion of the cyst through the conjunctiva(figure 4). The patient is on a maintenance dose of steroids and is kept under observation with monthly follow-up to rule out local recurrence or disease progression(figure 5).

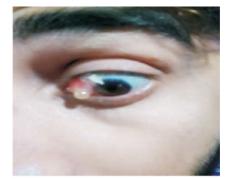


Figure 4: Spontaneous Rupture and Expulsion of the Cyst through the Conjunctival Layer.

Figure 5: 3weeks Post Treatment Image of the Patient Showing Slight Redness of the Overlying Conjunctiva with Remenant Inflammation.



Discussion :

IOIS is the third most common orbital inflammatory disease next to thyroid eye disease and orbital lymphoproliferative disease usually seen in adults but may be seen in children. Unilateral presentation is more typical but bilateral presentations are not uncommon.⁵

Pain is the most common symptom in adult IOIS and occurs 58-69% of the time followed by diplopia(31-38%).^{6,7}

Periorbital edema/swelling is the most common sign and occurs 75-79.2% of the time followed by proptosis (32-62.5%), EOM restriction (54.2%) red eye (48%), chemosis (29%), decreased vision (20.8%), and ptosis (16.7%)[8-9] Therefore, physical examination of patients with suspected IOIS involves lid assessment (retraction/lid lag/lagophthalmos), orbital assessment (proptosis), extraocular muscles (restriction),

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globe (injection/chemosis), and optic nerve function (visual acuity/color plates/relative afferent pupillary defect).

The etiology and pathogenesis of IOIS is currently not known but however infectious and immune-mediate etiologies have been implicated.

Observation for IOIS for mild cases of inflammation may be acceptable but if there is no clinical resolution or worsening of symptoms then additional therapy is indicated.

★ The details of the case are taken with due consent of the patient.

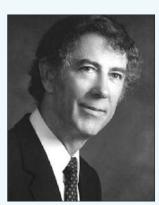
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LEGEND IN OPHTHALMOLOGY



Sir Charles William Simcoe

The Simcoe Cannula was developed about 40 years ago by C. William Simcoe MD, an ophthalmologist in Oklahoma, USA. Bill Simcoe was born in Stillwater on June 5, 1931 and passed away in his Tulsa home on October 22, 2017.

Dr. Simcoe also developed many innovations such as Simcoe irrigation and aspiration cannula &C-loop haptics. While examining and reshaping a paper clip, he had an idea of how to invent a much safer intraocular lens design the Simcoe open C loop which has become the industry standard in modern cataract surgery. He refused to patent any of his inventions and are widely used now in cataract surgery

A notive of Stillwater, Charles William Simcoe was a Korean War and Marine Corps

veteran. He was a graduate of the University of Oklahoma medical school. Through Project Orbis, a nonprofit dedicated to preventing blindness, Simcoe travelled the globe, teaching doctors how to perform safer, less costly cataract surgeries.