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Bilateral Idiopathic Orbital Inflammatory Syndrome, Associated With Unilateral Anterior Uveitis and Review of Literature

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Abstract

Idiopathic orbital inflammatory syndrome (IOIS) is a non-neoplastic, nonspecific inflammatory pathology of the orbit without a known local or systemic cause. It is the third most common orbital disease. It is typically a unilateral disease. We report a case of 76 year old male who presented with complaints of gradual, progressive proptosis, redness and diminution of vision in both eyes for 2 months. He was diabetic and hypertensive. On examination patient had asymmetrical proptosis of 22 and 24mm right and left eye respectively. On slit lamp evaluation severe chemosis and keratinization of the conjunctiva was noted in both eyes. Hirschberg's test showed 30 degree of left exotropia. There was presence of anterior chamber (AC) reaction and posterior synechiae in right eye, while there was relative afferent pupillary defect (Grade 3) in left eye with a quiet AC. On dilated fundus examination, right eye examination was unremarkable but there was optic atrophy in left eye. MRI orbit showed bulky extra-ocular muscles with thickened tendinous insertions suggestive of IOIS. The optic nerve appeared to be diffusely thickened along its orbital course on left side. Patient was managed with intravenous, oral and topical steroids, cycloplegics as well as lubricants. In a course of 3 months patient had significant reduction in proptosis and improved visual acuity. Due to rarity of anterior uveitis associated with IOIS, we are reporting this case.

Keywords: Idiopathic orbital inflammatory syndrome; Anterior chamber; Asymmetrical; Cycloplegics; proptosis

Abbreviations: IOIS: Idiopathic Orbital Inflammatory Syndrome; AC: Anterior Chamber; PR: Projection of Rays; PL: Perception of Light

Introduction

Idiopathic orbital inflammatory syndrome (IOIS) is a non-neoplastic, nonspecific inflammatory pathology of the

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orbit without a known local or systemic cause and is also known as "orbital pseudotumor" [1,2]. It is typically a unilateral disease [3]. The incidence of the disease is maximum in middle ages and no gender predilection has been noted for the disease [4]. The pathogenesis remains unclear but may include autoimmune disorder, inflammation and abnormal healing [5]. Symptoms are non-specific but often patients with IOIS present with triad of proptosis, pain and ophthalmoplegia [6]. Patient may also present with periorbital edema, ptosis, diplopia and other uncommon findings such as scleritis and exudative retinal detachment. It may be associated with systemic diseases such as rheumatoid arthritis, SLE, Crohn's disease etc [7]. IOIS is a diagnostic challenge and is a diagnosis of exclusion. Investigation of choice for orbital pseudotumor is MRI orbit [8]. Blood investigations to rule out systemic diseases (complete hemogram, ESR, Rheumatoid factor, ANA.) must be performed. A biopsy of the lesion is required in very few cases in which doubt persists and in those which do not respond to the treatment but histopathological findings showed lymphoid hyperplasia [9]. Systemic corticosteroids are considered as the mainstay of treatment in IOIS [10]. Anterior uveitis has been rare manifestation of IOIS seen in few pediatric patients but on literature search only one adult patient has been reported so far with unilateral IOIS with panuveitis [11]. In this report we describe one elderly patient with bilateral IOIS and unilateral anterior uveitis.

Case Report

A seventy six years old male, farmer by occupation presented with complaints of protrusion associated with pain, redness and diminution of vision in both eyes for two months. The symptoms were gradual and progressive. He had no history of fever or chills. Patient was taken to various eye institutes but got no relief with treatment and then he presented to us for further management. Patient was a known diabetic but uncontrolled glycemic status on regular oral hypoglycemic agents and a hypertensive for which patient was not taking any treatment for last two years. The patient refused any history of trauma, similar episode in the past, and any other long term ocular or systemic disease.

On general examination, he was conscious and well oriented. His blood pressure was 160/97mmHg without antihypertensive medication. His best corrected visual acuity was counting fingers at two meters and perception of light (PL) with accurate projection of rays (PR) in right and left eye respectively. On examination patient had asymmetrical proptosis of 22 and 24mm right and left eye respectively. (Figure 1) Intraocular pressure for both eyes was 18 and 20mm Hg. Both lids were swollen, chemosed and baggy. On Hirschberg's test left eye showed 30 degree of exotropia which may be because of fact that it was nonfunctional eye because of blindness. The orbital pressure of both eyes was raised on retropulsion. On slit lamp evaluation severe chemosis, diffuse congestion and keratinization of the conjunctiva was noted in both eyes. Right eye showed temporal pseudopterygium and left eye showed severe chemosis in area of lacrimal gland. Dilated and tortuous conjunctival vessels could be appreciated in left eye. The right eye showed evidence of exposure keratitis in interpalpaberal fissure. On fluorescein staining superficial puntate keratitis was visible in interpalpaberal fissure in right eye without any evidence of stromal in filtrates. There was presence of anterior chamber (AC) reaction with AC cells grade 2+ and posterior synechiae in right eye, while there was relative afferent pupillary defect (Grade 3) in left eye with a quiet AC. Patient had immature senile cataract with nuclear sclerosis grade 3 in both eyes. (Figure 2) On dilated fundus examination, right eye showed hyperemic disc but there was optic atrophy in left eye. (Figure 3)There was no evidence of diabetic or hypertensive retinopathy in both eyes. MRI orbit of the patient was ordered which was suggestive of bilateral proptosis with bulky extra-ocular muscles with thickened tendinous insertions which suggested a diagnosis of IOIS.

The optic nerve appeared to be diffusely thickened along its orbital course on left side. (Figure 4) Patient was known diabetic and had a random blood sugar level of 315 mg/dl at presentation with HbA1C as 12.5% despite oral hypoglycemic medication. The thyroid profile was within normal limits. White blood cells and neutrophil counts were $12.4x10^{9}/L$ and $9.1x10^{9}/L$ respectively. Serum creatinine was 0.75 mg/dl. The patient was started on intravenous methyl prednisolone 1gm/kg body weight for three days followed by oral steroids (oral Prednisolone 1 mg/kg/day) under supervision of endocrinologist for glycemic control with injectable subcutaneous insulin. Systemic and topical antibiotics were started. Topical lubricating eye drops and cycloplegic were added to take care of exposure keratitis and uveitis in right eye. Topical steroids were started for left eye. Patching was advised at night time for right eye after putting ointments. The patient showed significant reduction in chemosis and proptosis in both the eyes by third day and the improvement was progressive since then. The oral steroids were tapered completely till six weeks along with strict glycemic control. After three months patient was symptomatically relieved with best corrected visual acuity of 20/60 in right eye while for left eye it was status quo.

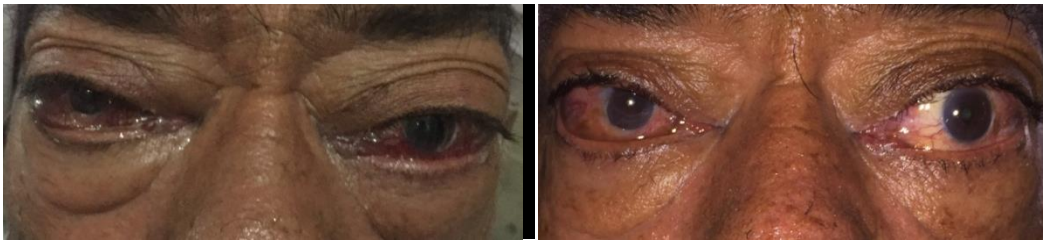

Figure 1: a) Showing both eyes proptosis, periorbital edema and severe conjunctival chemosis at presentation b) Both eyes showing clinical resolution at 3 months of follow up.

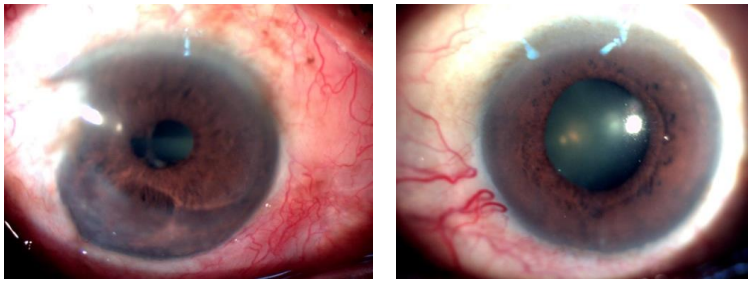

Figure 2: On slit lamp examination. a) Right eye shows conjunctival chemosis, congestion, exposure keratitis and posterior synechiae. b) Left eyes shows dilated and tortuous conjunctival vessels and middilated pupil (RAPD).

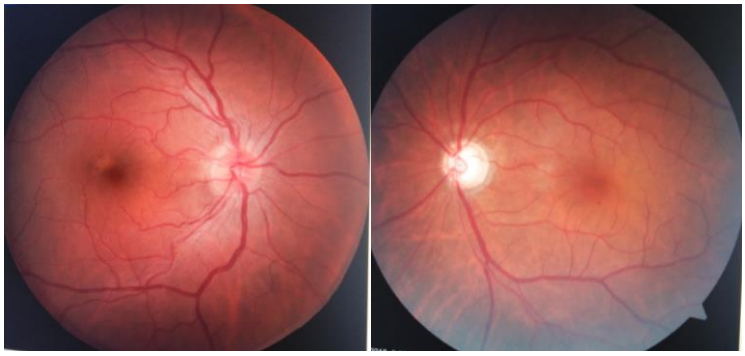

Figure 3: a,b) Fundus shows Right eye hyperemic disc and Left eye disc pallor.

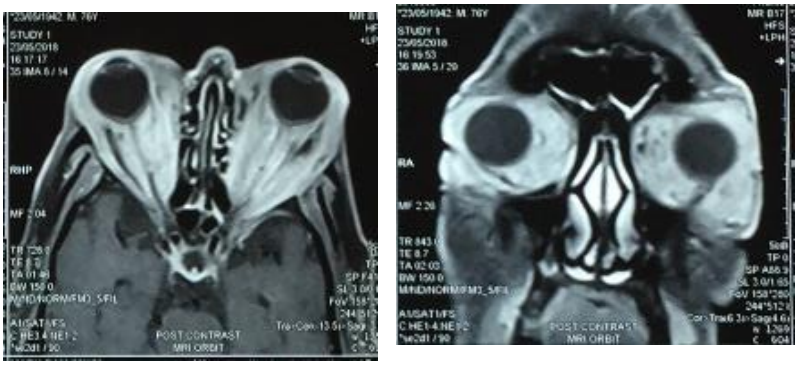

Figure 4: a and b) MRI On TI Axial and coronal with contrast shows heterogenous diffuse hypertrophy of extra ocular muscles with tendinous insertions.

Discussion

Grave's ophthalmopathy and lymph proliferative disorders of the orbit are the first and second most common. IOIS is the third most common orbital disease [12]. It comprises of 5-8% of all the orbital masses [13]. Investigation of choice for IOIS is imaging of the orbit. It shows diffuse orbital mass, sclera and choroidal thickening, extra ocular muscles thickening with tenon's space enhancement on contrast imaging and optic nerve enhancement [14]. The differential diagnoses include thyroid orbitopathy, orbital cellulitis, sarcoidosis, lymphoid tumors and metastatic carcinomas [15]. The case described a rare incidence of bilateral orbital pseudotumor in association with uveitis. Abdulkadir Shehibo reported a case of bilateral orbital pseudotumor in a 3 year old [16]. Recently Sandip Sarkar also described a rare case report of bilateral IOIS in an adult patient [17]. To the best of our knowledge, there is only few case reports of bilateral IOIS associated with anterior uveitis as present in current case. The association of anterior uveitis and IOIS has been reported in pediatric patients [18,19].

Xu et al reported a case of unilateral IOIS with panuveitis in adult female [11]. Still bilateral IOIS with anterior uveitis has not been reported in adults so far. Various features which were notable in this case were loss of vision in left eye since last two months to the extent of PL present. On fundus examination pale disc was seen which showed that the extra ocular muscle were so bulky along with the soft tissue surrounding the optic nerve which led to optic nerve compression and eventually optic atrophy. Another possibility of optic neuritis can be also there. In right eye despite anterior uveitis, exposure keratitis and cataractous lens patient had visual acuity of counting fingers at two meters which improved up to 20/60. After starting the treatment for right eye exposure keratitis healed, uveitis resolved and patient retrieved good visual acuity. The differential diagnosis of lymphomas was kept in mind which was ruled out by the clinical course of the disease. Cataract surgery with posterior chamber intraocular lens will be planned under cover of oral and topical steroids when required.

Conclusion

IOIS is a diagnosis of exclusion, and presents commonly in middle aged individuals. It is typically a unilateral disease with common recurrences. Bilateral disease has been reported but is rare. Patient presents with triad of pain, proptosis and ophthalmoplegia. Atypical presentations include pediatric IOIS, optic nerve IOIS, scleritis and exudative RD. Lesions are usually restricted to the orbit but may extend to adjacent retro-orbital regions. MRI orbit is the choice investigation while biopsy is rarely required for non-responsive cases. Early diagnosis and treatment with systemic corticosteroids is the key to successful management of the disease.

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