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Case Report

Bilateral Idiopathic Orbital Inflammation Syndrome in an adult patient: A rare case report

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Abstract

Bilateral Idiopathic Orbital Inflammation Syndrome (IOIS) is rare in adults. Most commonly it affects the young adults and children. It is mostly unilateral condition. This is a case of 50 years old male coming with the complaints of painless proptosis on both the eyes for 1 year. The history of diplopia was also found. On examination, it was found that there was restriction of movements in Right eye superiorly. Any history of associated systemic illness was not found. Based on the clinical examination, laboratory reports and contrast enhanced computed tomography (CECT) the diagnosis of Bilateral IOIS was made. Patient was treated with Intravenous methyl prednisolone followed by oral Prednisolone in tapering doses for a period of 2 months. After 2 month of regular follow up the patient was found to have a favorable outcome with improved visual acuity and reduction in the size of the proptosed eye. Patient was followed up for another 6 month to observe any recurrence or relapse.

Keywords: Diagnosis, Proptosis, Pseudotumor, Treatment

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Introduction

Idiopathic orbital inflammation syndrome (IOIS), formerly known as pseudotumor orbita, is named after a group of benign non-infectious, non-granulomatous inflammation disorders of the orbit most commonly affecting young adults and children.[1] It is rare condition where no local or systemic cause can be identified. They constitute a broad category of orbital inflammatory diseases with varying degree of involvement of orbital contents. Sometime optic nerve involvement has been seen as a part of the posterior segment process.[2] Systemic associations such as lymphoproliferative disorders in adults and rhabdomyosarcoma, leukaemia, neuroblastoma, etc in children need to be ruled out before a diagnosis of Idiopathic orbital inflammation syndrome (IOIS) can be made.[3] Identifying the aggressive nature of this entity is of utmost importance so that aggressive medical management can be initiated and unnecessary surgical intervention can be avoided. The aim of our study was to summarize the clinical features, histopathology, radiological evaluation and treatment outcomes in a rare case of bilateral IOIS who had a favorable outcome with corticosteroid treatment.

Case report

A 50-year-old male patient attended the outpatient department of Agartala Government Medical College, Agartala, Tripura with the complaint of painless proptosis involving both eyes for the past year (Fig. 1). Proptosis was more prominent in the right eye and rapidly it was increasing over the last 9 months which was associated with diplopia. In addition chemosis, excessive tearing, itching and burning sensation was also complained. Patient did not give any history of floaters, flashes, ocular trauma or any intraocular surgery. There was no history of weight loss, palpitations, tremor, increased appetite, headache, vomiting. Vitals was found to be normal. Patient was not suffering from any kind of systemic illness like thyroid, diabetes melitus, tuberculosis, or any other chronic illness. On examination, there was restric-

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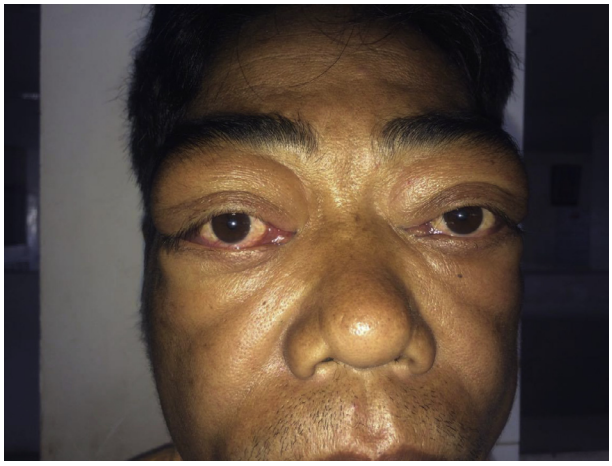

Fig. 1. Patient showing bilateral IOIS.

tion of upward gaze in Right eye. Decreased in visual acuity was noticed in both eyes (6/36 OD and 6/24 OS). Pupil was normal and reactive towards light. There was mild lid edema but no palpable mass on either side. There was proptosis of 22 mm in OD and 20 mm in OS measured on hertel exopthalmometer. On palpation, orbital margins were intact. Proptosis was non compressible and non-reducible. There was no audible bruit on auscultation in any of the eyes. Intraocular pressure measured on applanation tonometry was 19 mm hg in OD and17 mm hg OS. Fundal examination after pupillary dilatation with tropicamide and phenylephrine eye drops showed mild disc edema and venous congestion in both eyes (see Figs. 2 and 3).

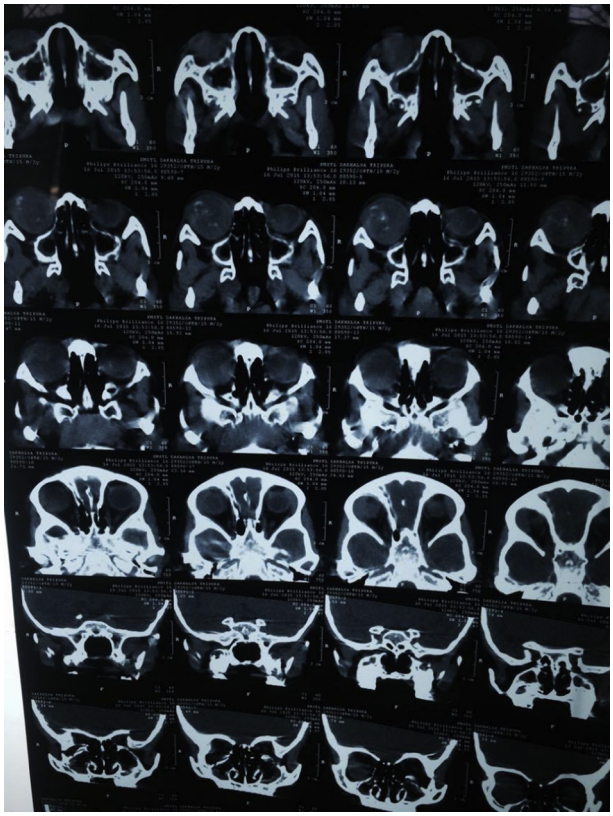

Fig. 2. CT scan of orbit showing bilateral lesion with large lesion in right side.

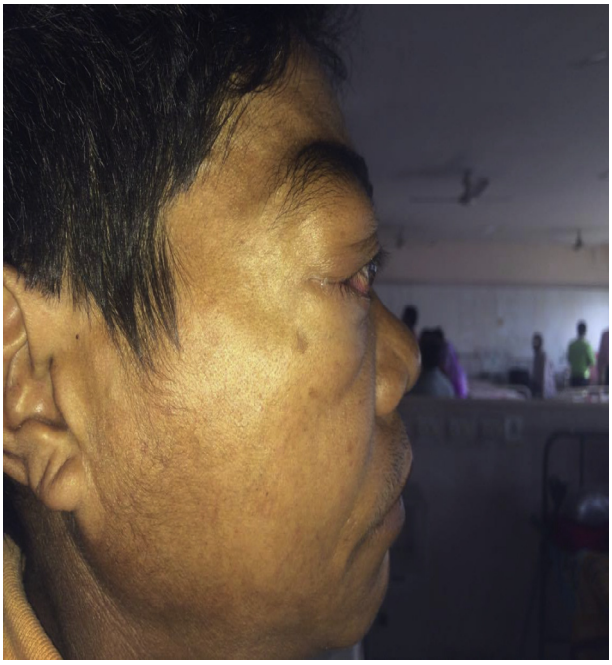

Fig. 3. Proptosis reduced after corticosteroid therapy.

Laboratory work up revealed haemoglobin-12.2 gm/dl, red blood cell count was normal and white blood cell count- 7200 cumm.[2] Erythrocyte sedimentation rate was mildly elevated (48 mm/hr). Peripheral blood smear showed normocytic normochromic red blood cells. Thyroid function test, kidney function test and liver function tests was normal. Serum levels of antinuclear antibody was normal. Contrast enhanced Computed tomography of the orbits showed ill defined soft tissue mass lesion measuring 29 × 19 mm in superotemporal aspect of right eye and similar type of lesion measuring 24 × 17 mm on the left eye. Orbital walls, extraocular muscles and the optic nerve was seems to be normal. There was no signs of any bony destruction or intracaranial extension of the lesion. Contrast enhanced computed tomography of Brain revealed normal study. Biopsy of the lesion was performed and histopathological examination was suggestive of non specific inflammation. Considering the biopsy result as well as the CECT scans and blood workup, diagnosis of axial proptosis due to IOIS was established. The patient was started on Intraveous Methyl prednisolone 1gm per day for 3 consecutive days followed by oral prednisolone 1 mg/kg body weight per day on tapering doses for 8 weeks. Topical prednisolone and tear substitute like carboxy methyl cellulose 1% was also prescribed. Before starting Intravenous methyl prednisolone a complete physical examination by a internal medicine specialist was done and his blood sugar level was also checked. After 2 months of treatment with oral steroids patient slowly started showing improvement in ocular movements and proptosis also reduced (16 mm OD and 13 mm OS). Visual acquity has also improved quite a bit (6/15 OD and 6/12 OS). The patient was put on a maintenance dose of steroids and was kept under observation with monthly follow-up up to six months period to rule out local recurrence or disease progression.

Discussion

IOIS is an idiopathic, non-neoplastic, non infectious inflammatory condition of the orbit. It was first described in the year 1905 by Birsch-Hirschfield.[4–6] It is the third most common disease of the orbit after Grave's ophthalmopathy and lymphoproliferative disorders which represents approximately 8–11% of cases.[7] It is most commonly seen in young adults and children.[8] Pediatric IOIS encompasses about 6–16% in children, mostly bilateral orbital involvement without evidence of underlying systemic disease.[9] Peak incidence appears to be predominantly in the adult population, typically in the middle-aged persons, and there is no sex predilection.[6] In adults, IOIS tends to be unilateral. It is a diagnosis based on exclusion criteria, clinical symptoms, blood tests and imaging reports. This disease is reported in all ethnic groups around the globe. In adults, IOIS tends to be unilateral.[10] Bilateral IOIS is common in children and extremely rare in adults. The clinical course of IOIS ranges from mild and self-limiting to devastating orbital sclerosis with blindness. Disease relapse is common.

The most important risk factors for developing this disease are lower socioeconomic status, high body mass index, young age, use of oral bisphosphonate, lithium and chemotherapies.[10] IOIS can be found in certain rheumatological disorders such as Wegener's granulomatosis, sarcoidosis, giant cell arteritis, systemic lupus erythematosus, dermatomyositis and rheumatoid arthritis.[11] Clinically it may present with periorbital oedema, erythema, proptosis, ptosis, diplopia or painful eye movements. Imaging modalities used for diagnosis are Computed tomography (CT) and magnetic resonance imaging (MRI); however, serological tests may be required to exclude a systemic cause. On CT and MRI there may be a diffuse mass involving the retro bulbar tissues, infiltration of the intraconal fat, orbital inflammation, and thickening of extraocular muscles and optic nerve.[12,13] Inflammation of extraocular muscle or myositis can be acute, subacute or recurrent with involvement of the medial rectus being common. Here the extraocular muscles were normal. This can be differentiated from Grave's disease where there is fusiform enlargement of muscle with sparing of the tendon insertion.[14] All these changes including infiltration of adjacent fat, periocular involvement or discrete mass help in making a diagnosis of IOIS.

Biopsy is not mandatory in all cases and should be reserved for those with an atypical course or cases suspected orbital malignancy or poor response to corticosteroids is seen.[15] It is suggested that systemic causes should be ruled out and a biopsy be performed. In our bilateral case, we did the biopsy on the right side which revealed non-specific inflammation suggesting IOIS. When in doubt, you may always go ahead and biopsy the other side.

IOIS is a diagnosis of exclusion, after other identifiable local or systemic causes have been eliminated. However, identifying the true nature of this condition and recognizing it from other malignant pathologies is important to avoid unnecessary surgical intervention. Only a small number of patients responds well to the treatment.[16] Corticosteroids are the mainstay of treatment. Around 37% of patients who were treated with steroids, failed to resolve. In our case we got an excellent response with steroids. Other treatment options that can be given as monotherapy or in addition to corticosteroids are immunosuppressives, radiotherapy, and surgical excision. Immunosuppressives like methotrexate, cyclophosphamides and other antineoplastic agents can be useful. Radiotherapy is another modality of treatment. An alternative mode of treatment in recurrent and recalcitrant cases is with infliximab (TNF-alfa blocker).[16] Invery few case with localized lesion, surgery may be used.[17]

A total of 63% of IOIS patients show complete remission after treatment. In 35% patients, only partial relief is obtained with persistent motility dysfunction, pain or visual loss. With all the available treatment modalities 2% patient experience no remission. We were lucky to provide remission with an 8 week therapy with corticosteroids and there was no relapse for 6 months. Of course, this is a short duration to consider total remission and we keep following-up the patient.

Understanding the clinical features of IOIS and differentiating it from alike conditions is of supreme importance, especially when the disease is bilateral. Early diagnosis and appropriate treatment with corticosteroids are very important for better visual prognosis and resolution of proptosis.

Conflict of interest

The authors declared that there is no conflict of interest.

Patient consent

Informed consent was taken from the patient before enrolling him for the study.

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