Case Report

A Case of Idiopathic Orbital Inflammation And Third Nerve Palsy: Interesting Case

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Abstract

Purpose: *This case is a unique presentation of idiopathic orbital inflammation and 3rd nerve palsy (superior division involvement).*

Case report: *A 30 years female patient presented with chief complaints of diplopia for two days and drooping of right upper eyelid for one day. Patient also complained of ocular pain and headache for preceeding ten days. Considering the clinical presentation, a differential diagnosis of thyroid orbitopathy, ocular myasthenia, orbital neoplasm and idiopathic orbital inflammatory syndrome (IOIS) was considered. Laboratory investigations revealed nothing significant. MRI orbit was suggestive of IOIS.The patient was started on oral steroids and had a favorable outcome.*

Conclusion: *Understanding of the clinical features of patients with IOIS, differentiating it from other clinical conditions and timely implementation of available treatment may help to prevent visual loss and other associated morbidity from this condition.*

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Keywords: *Orbital pseudotumor, Orbital myositis, extraocular movements*

Introduction

Idiopathic orbital inflammatory syndrome (IOIS), also known as orbital pseudotumor, is a non-infectious inflammation of the orbital soft tissues for which no cause is found after local and systemic evaluation.[1] It can be diffuse or localised.[2] The localized type of idiopathic orbital inflammation is further subdivided into myositis, periscleritis, perineuritis and dacryoadenitis.[3] We present a case of orbital myositis with unique presentation.

Case Report

A 30 years female patient presented in OPD with chief complaints of diplopia for two days and drooping of right eyelid for one day. Diplopia was binocular and when looking towards right side and upwards. Patient also complained of ocular pain and headache for preceeding ten days. The headache was acute in onset ,severe in intensity and intermittent in nature and was referred to right eye. For this pain, patient took some oral analgesic from local practitioner.There was no history of fever, sorethroat ,neck rigidity, ocular trauma, any preceding infection or any other systemic illness. There was no significant past history, treatment history and family history.

General physical examination was normal. On ocular examination ,complete ptosis was present in right eye (Figure 1a). Visual acuity was 6/12 (with PH 6/6) in right eye and 6/6 in left eye on Snellen,s visual acuity chart. It was obtained in right eye by lifting upper eyelid manually. Bilateral Pupils were normal in size and reacting normally to light. Intraocular pressure and colour vision was normal in both eyes. In extraocular movements abduction and elevation was restricted in right eye (Figure 1b & 1c), adduction and depression was normal in right eye (Figure 1d &1e) and the extraocular movements were full in left eye in all gazes. Diplopia was present in dextroversion and elevation, but

Figure 1: *(a) shows complete ptosis in right eye*

Figure 1: *(b) shows limitation of abduction in right eye*

Figure 1: *(c) shows limitation of elevation in right eye*

Figure 1: *(d) shows normal adduction in right eye*

Figure 1: *(e) shows normal depression in right eye*

more in elevation. There was no lid oedema, chemosis or any localised conjuctival congestion. On Slit lamp examination, anterior and posterior segment revealed no abnormality. There was no audible bruit on auscultation over the right eye. The remainder of the neurological examination was normal.

A differential diagnosis of thyroid ophthalmopathy, Tolosa Hunt syndrome, ocular myasthenia, orbital neoplasm and lymphoproliferative infiltration was considered. Laboratory investigations and radiological investigations were done. Complete hemogram, blood sugar level, liver function test, renal function tests and thyroid function tests were normal. Rheumatoid factor and antinuclear antibody were negative. Ice pack test was negative.

MRI Orbit (Plain&contrast) showed enlarged right lateral rectus muscle (measuring upto 7.5 mm in thickness,on left side it measured 2.9 mm in thickness) with normal anterior tendinous insertion & thickening of posterior tendinous insertion causing crowding at the apex of orbit with focal intense postcontrast enhancement in the enlarged right lateral rectus muscle (Figure 2a, 2b). MRI Brain was normal.

After clinical evaluation , laboratory and radiological investigations, diagnosis of idiopathic orbital inflammation was considered and treatment was started. Oral prednisolone in the dose of 60mg/day (body weight-58 kg) was started initially for two weeks and dramatic improvement in symptoms was noted . Ocular pain was relieved, ptosis in right eye was improved, abduction was almost full and slight improvement in elevation right eye was also seen. There was no diplopia on dextroversion , but was present on elevation. After two weeks dose of oral prednisolone was decreased to 50mg/day and then tapered slowly later on over two months. After one month of treatment patient,s eyes were almost looking normal except for mild limitation of elevation and mild diplopia on elevation(Figure 3a,3b,3c). Patient was on weekly follow-up for the first month and then biweekly for the next one month and subsequent follow-up was advised if patient had symptoms and risk of recurrence of disease was also explained to her. Patient recovered completely after ten weeks of treatment.

Discussion

Orbital pseudotumor is not a single entity but rather a spectrum of idiopathic orbital inflammations, excluding infectious, neoplastic, and systemic inflammatory or immunological etiologies.2 The term 'orbital pseudotumor' was first coined by Birch-Hirschfeld in 1905 to include all the non-specific or idiopathic obital inflammatory syndromes (IOIS) . IOIS is the third most common orbital disease after thyroid orbitopathy and lymphoproliferative disorders.[4] It represents approximately five to eight percent of all

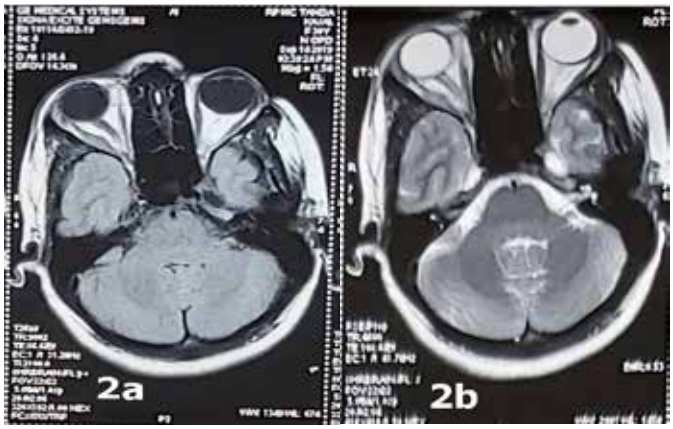

Figure 2: *MRO orbit plain (2a) and contrast (2b) showed enlarged right lateral rectus muscle (measuring upto 7.5 mm in thickness,on left side it measured 2.9 mm in thickness) with normal anterior tendinous insertion & thickening of posterior tendinous insertion causing crowding at the apex of orbit*

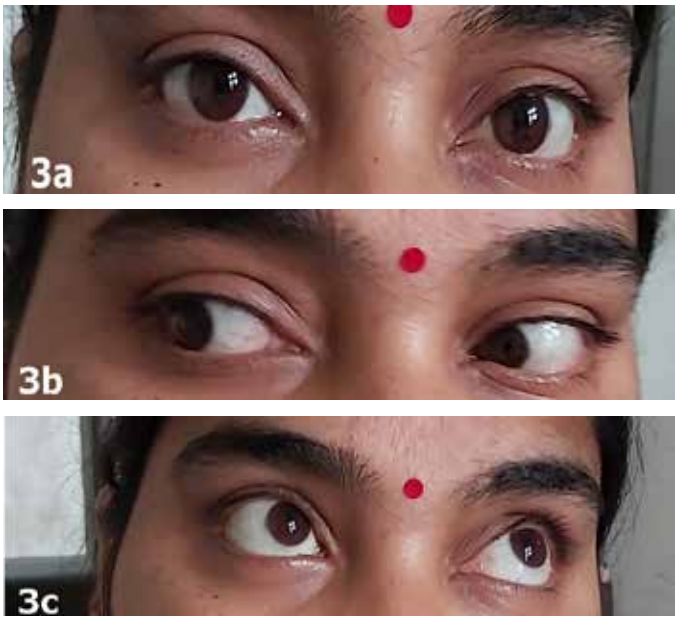

Figure 3: (a) *shows ptosis completely disappeared in right eye after one month of treatment*
Figure 3: (b) *shows normal abduction in right eye after one month of treatment*
Figure 3: (c) *shows improved elevation in right eye after one month of treatment*

orbital masses.[5] Disease usually occurs in adults, but may also affect children.[6] There is higher incidence of bilateral orbital involvement in children.[7] Peak incidence is seen predominant typically in the middle-aged persons and there is no sex predilection.[8] The disease has been reported in all ethnic groups around the globe.[9]

This entity has been further divided into localised and diffuse type. The localized type of idiopathic orbital inflammation was further subdivided into myositis, periscleritis, perineuritis and dacryoadenitis. In orbital myositis ,the inflammatory process may involve one or more extraocular muscles.[10] Usually monocular involvement is there, but it could be bilateral also with involvement of one or more muscles in both orbits.[11]

The clinical course of IOIS ranges from mild and self-limiting

to devastating orbital sclerosis with blindness. Disease relapse is common. The clinical presentation of the disorder may be acute, subacute or chronic. The lesion is most commonly restricted to the orbit; however, extension into adjacent retro-orbital structures is also known. The cause of idiopathic orbital inflammation is unknown. Neoplastic, infectious, and systemic inflammatory or immunological syndromes must be excluded. The differential diagnosis of IOIS include Thyroid- associated orbitopathy, Sarcoidosis, Wegener's granulomatosis, Tolosa-Hunt syndrome, Lymphoproliferative orbital disease, Metastatic orbital disease and Orbital cellulitis. The laboratory investigations advised in these cases include Complete blood count, sedimentation rate ,electrolytes, thyroid function tests, antinuclear antibodies, antineutrophil cytoplasmic antibodies, angiotensin converting enzyme level and rheumatoid factor etc.

No single laboratory test is available to diagnose IOIS. The diagnosis is based on the clinical picture and verification of enlargement of extraocular muscle by orbital imaging (CT& MRI), as well as on the exclusion of any specific disease. Biopsy is not indicated in all cases and should be reserved for the cases with an atypical course6 or cases suspicious for an orbital malignancy or when a poor or equivocal response to corticosteroids is seen.

Corticosteroids are the mainstay of therapy, inducing a rapid and dramatic resolution of symptoms within few days after starting treatment.[12,13] The prompt response to steroids is critical also in establishing the diagnosis of idiopathic orbital inflammation, as other conditions are not expected to exhibit such a rapid and dramatic response.[14] As per literature it is recommended that oral prednisone should be initially used at doses of 60 to 80mg per day over two to three weeks.14 Then, steroids should be tapered slowly, usually over six weeks to three months ,in order to prevent exacerbation or recurrence of inflammation. The adjunctive use of immunomodulatory agents or radiotherapy must be reserved for selected recalcitrant cases.[15] This patient had complete ptosis with limitation of movements of abduction and elevation The lateral rectus was markedly enlarged and showed postcontrast enhancement presumably because of myositis. Oculomotor nerve divides in the anterior cavernous sinus and the superior and inferior division enters the orbit through the superior orbital fissure. The superior division of third nerve was involved in the inflammatory process which was well corroborated by the MRI findings of crowding at the apex of orbit. The involvement of superior division of third nerve lead to complete ptosis and limitation of elevation while involvement of lateral rectus caused restriction of abduction. The patient improved as the inflammation was controlled by the use of oral corticosteroids

References

1. Bijlsma WR, van Gils CH, Paridaens D, Mourits MP, Kalmann R. Risk factors for idiopathic orbital inflammation: A case — control study. Br J Ophthalmol 2011;95:360-4.
2. Kennerdell JS, Dresner SC. The nonspecific orbital inflammatory syndromes. SurvOphthalmol 1984;29:93-103.
3. Rootman J, Nugent R. The classification and management of acute orbital pseudotumours. Ophthalmology 1982; 89:1040-1048.
4. Li Y, Lip G, Chong V, Yuan J, Ding Z. Idiopathic orbitalinflammation syndrome with retro-orbital Involvement: A Retrospective study of eight patients. PLoS One 2013;8:e57126.
5 .Szabo B, Szabo I, Crişan D, Stefănuţ C. Idiopathic orbital inflammatory pseudotumor: Case report and review of the literature. Rom J MorpholEmbryol 2011;52:927-30.
6. Khawaja KS, InamUlHaq. Idiopathic orbital inflammation (pseudotumor). Pak Armed forces Med J 2010;60:151-3.
7. Partab R, Syed Imtiaz AS, Syed Aftab HS, Azizullah J, Imdad Ali A. Presentation of idiopathic, non-specific, orbital inflammation(pseudotumor)- study of 46 cases. Medical Channel; October-December 2010;16:594-9.
8 Yuen SJ, Rubin PA. Idiopathic orbital inflammation, distribution clinical features and treatment outcome. Arch Ophthalmol2003;121:491-9
9. Chaudhry IA, Shamsi FA, Arat YO, Riley FC. Orbital pseudotumor: Distinct Diagnostic Features and Management. Middle East Afr J Ophthalmol 2008;15:17-27.
10. Weinstein GS, Dresner SC, Slamovits TL et al. Acute and subacute orbital myositis. Am J Ophthalmol 1983;96:209.
11. Ludwig I, Tomsak RL. Acute recurrent orbitalmyositis. J ClinNeuroophthalmol 1983; 3: 41–47-217.
12. Jacobs D, Galetta S. Diagnosis and management of orbital pseudotumor. CurrOpinOphthalmol 2002; 13:347-351.
13. Rubin PA,Foster CS. Etiology and management of idiopathic orbital inflammation. Am J Ophthalmol 2004;138:1041-1043.
14. Karesh JW, Baer JC, Hemady RK. Noninfectious orbital inflammatory disease. In: Tasman W, Jaeger EA (ed). Duane's Clinical Ophthalmology. Philadelphia, New York, Baltimore: Lippincott Williams & Wilkins,2004;vol.2,ch.35:1-45
15. Taylor PC. Anti-tumour necrosis factor therapies.Curr Opin Rheumatol 2001;13: 164-169.

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