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Case Report

Pseudotumor orbit-bilateral v/s unilateral: a case study of two orbital pseudotumor cases

Sunanda Nandi*, Jawahar Jyoti Kuli

Department of Ophthalmology, Assam Medical College & Hospital, Dibrugarh, Assam, India

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***Correspondence:**
Dr. Sunanda Nandi,
E-mail: sunanda.amc@gmail.com

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ABSTRACT

Orbital pseudotumor is a non-granulomatous inflammation of orbital soft tissue for unknown etiology. In the first case, a 35-year-old female presented with bilateral painless upper eyelid swelling since 3years.No proptosis, no extra ocular movements restriction of both eyes was noted. Best corrected visual acuity both eye: 6/6, N6. USG B-Scan and CECT revealed bilateral lacrimal gland enlargement with bulky muscles and tendons. USG guided FNAC confirmed pseudotumor which showed prompt response to steroids but recurred after 1month of stopping of steroid therapy. In the second Case, a 15year old female presented with unilateral proptosis left eye for 3 months with painful extraocular movements and restricted in all gazes. Best corrected visual acuity in L/E - 6/12, N6, R/E-6/9, N6. USG B-scan and CECT revealed soft tissue lesion encasing Optic nerve. CEMR revealed bulky lacrimal gland, muscles and lesion extending up to orbital apex. Prompt response to steroid with no recurrence till date.

Keywords: Bilateral, Extraocular movements, Lacrimal gland, Orbital Pseudotumor, Recurrence

INTRODUCTION

Idiopathic orbital pseudotumour (IOP), also known as idiopathic orbital inflammatory syndrome (first described by Birch-Hirschfeld in 1905), is a benign, non-infective inflammatory condition of the orbit without identifiable local or systemic causes.[1]

IOP is a diagnosis of exclusion based on a combination of clinical and radiographic findings.[2] It may present acutely, subacutely or chronically usually in one orbit. Presentation may vary according to the specific location and degree of inflammation.[3]

IOP can affect people of virtually any age,with no sex predilection. Unilateral periorbital pain, cranial nerve palsies and a dramatic response to corticosteroid therapy are the hallmarks of clinical presentation in IOP.[4]

CASE REPORT

In the first case, a 35 year old female presented bilateral swelling of the upper eyelids since three years. Swelling over the right upper eyelid was since three years and left eye since 1.5 years. It was painless and gradually progressive, more in the lateral aspect of the eyelids with no pain in extraocular movement. BCVA: R/E- 6/6,N6; L/E-6/6,N6. No history of protrusion of eyeball or drooping of eyelids and no history of trauma or any drug intake (Figure 1 and 2).

In the second case, a 15 year old female presented with Proptosis of L/E - 22mm by luedde exopthalmometer. Pupil of both eyes were within normal limit. Anterior and posterior segment findings of B/E were WNL. Extraocular movements were painful and restricted in all

gazes. BCVA: R/E-6/9, N6; L/E-6/12,N6. No history of trauma or any drug intake (Figure 3 and 4).

Investigation

In first case with bilateral puffiness of eyelid routine blood test was wnl, thyroid profile was wnl. USG B-SCAN revealed bilateral rectus muscles and tendons to be bulky. Lacrimal glands were enlarged taking increased vascularity in doppler.

In CECT BRAIN it showed heterogenous enhancement in lacrimal gland with hypertrophied muscle belly and tendon. USG guided FNAC revealed presence of mixed population of lymphoid cells, mature lymphocytes, immunoblasts and occasional plasma cells suggestive of benign lymphoid pseudotumour (Figure 5 and 6).

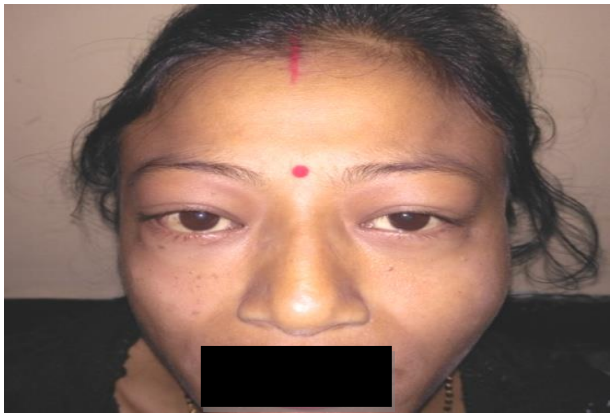

Figure 1: Picture of first case.

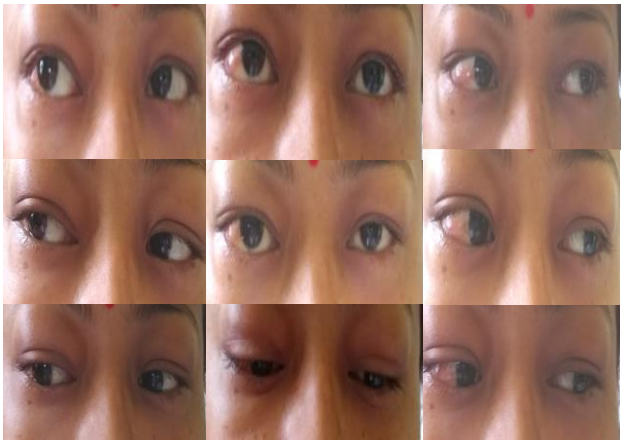

Figure 2: Picture of EOM in case 1: EOM unrestricted in all gazes.

In the second case with unilateral proptosis , routine blood test was wnl with thyroid profile wnl. USG B-scan L/E revealed left extra-ocular muscles and lacrimal gland to be bulky. CECT brain showed heterogeneously enhancing soft tissue attenuated lesion in the left retro-bulbar region encasing the left optic nerve with proptosis of L/E. CEMRI brain and orbit showed heterogeneously enhancing ill-defined T1 iso-intense and T2 hyperintense lesion in intra and extraconal space of left orbit and causing infero-medial displacement of optic nerve. Features suggestive of inflammatory mass extending upto the orbital apex of left orbit (Figure 7).

Figure 3: Picture of second case.

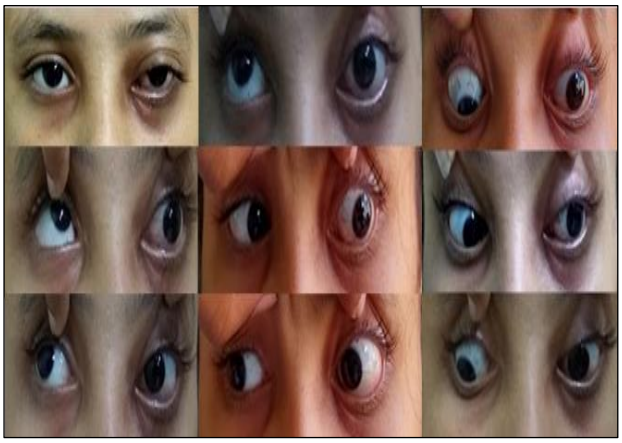

Figure 4: Picture of EOM in case 2; EOM restricted in all gazes.

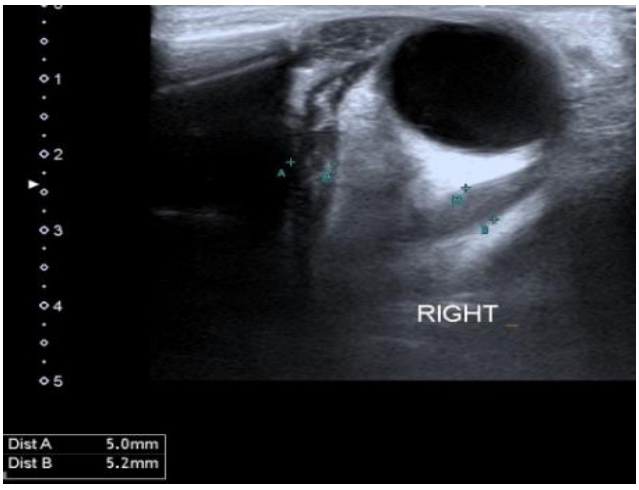

Figure 5: USG B SCAN of case 1 ; bulky rectus muscle.

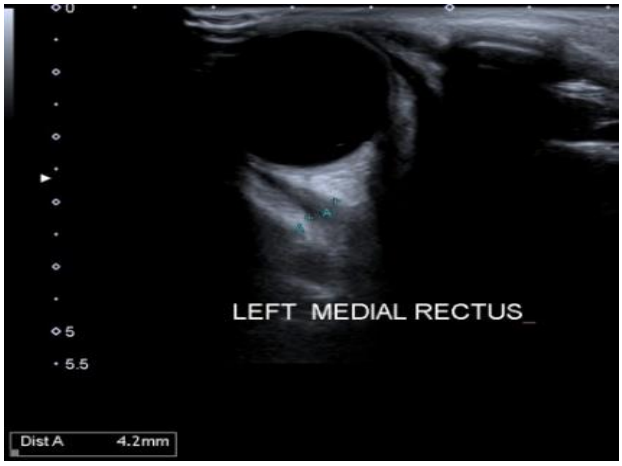

Figure 6: USG B SCAN of case 1; bulky medial rectus.

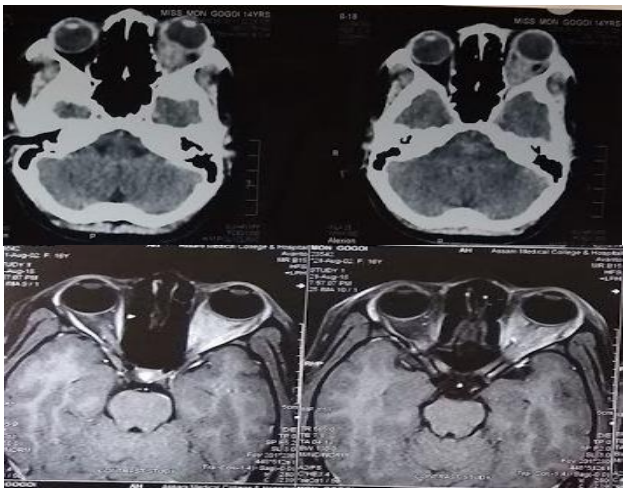

Figure 7: CECT (above) ; CEMRI (below) of case 2.

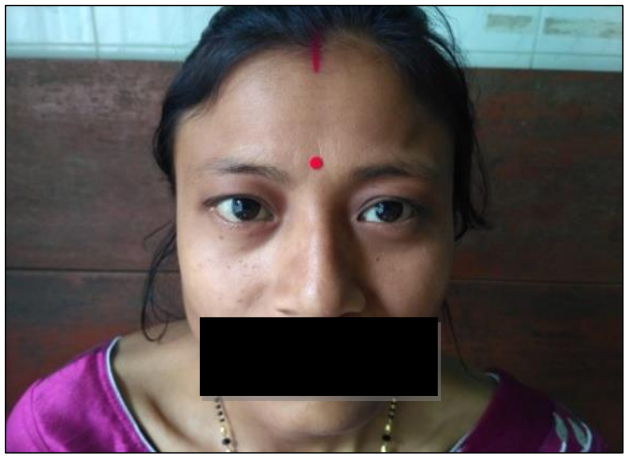

Figure 8: After treatment in 21 days in case 1.

Treatment

In both first and second case, Oral steroids at a dose of 1mg per kg body weight was given and tapered over a period of one and half month and in the follow up after 21 days both first and second case showed prompt response to steroid and the swelling subsided. Pseudotumor Orbit was the final diagnosis for both the cases. But however in the first case, it showed recurrence after one month of stopping of steroids which was not seen in 15 year old female presenting with unilateral pseudotumor orbit. Proptosis of L/E reduced and EOM were normal after 1 month with no recurrence till date (Figure 8 and 9).

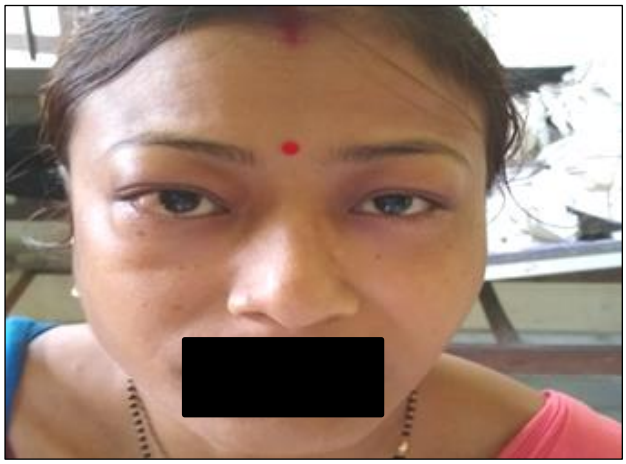

Figure 9: Recurrence after stoppage of steroid in case 1.

DISCUSSION

The earliest description of orbital pseudotumor can be traced back to 1905 when Birch-Hirschfield described a mysterious orbital syndrome with a clinical impression of orbital benign or malignant neoplasm in which, at the time of surgical exploration, only inflammatory tissue was found.[5] Patient presents with proptosis, diplopia, conjunctival chemosis, visual disability and restriction of extraocular movements.[6] However in our case patient presented only with bilateral upper eyelid swelling with no proptosis or ophthalmoplegia in first case and unilateral proptosis in the second case.

Commonly involved structures include orbital fat, lacrimal gland, extraocular muscles others being optic nerve, sclera and tenon.[7] In our case it involved bilateral enlargement of lacrimal gland in both the cases with involvement of orbital apex in the second case.

Idiopathic orbital pseudotumor comprises approximately 10% of all orbital mass lesions.[8] It is the most common cause of a painful orbital mass in adults and the third most common orbital disease after thyroid orbitopathy and lymphoproliferative disease.[9] Imaging findings are helpful and reflect the heterogenecity of the inflammatory process. In one study by Atlas et al, the MRI scan was abnormal in 10 of 10 patients with confirmed orbital pseudotumor.[10] The diagnosis of orbital pseudotumor is usually clinical and confirmed by prompt response to steroids. In our case patient responded well to steroids

however there was recurrence after stopping in first case. Relapse is common. A retrospective study by Mombaerts et al. of 32 patients found a modest response to prednisolone. Only 37% of patients cured, 78% had an initial response with a 52% recurrence rate.[11]

Radiotherapy has been demonstrated modest efficacy in several small studies and should be considered for patients who are steroid resistant or are unable to tolerate steroids.[12] Immunosuppresant agents such as cyclosporine, cyclophosphamide ,methotrexate have shown benefits in several small studies. However, more data are necessary to determine the most appropiate medication, dose and treatment duration.[13]

CONCLUSION

Orbital pseudotumor represents a diagnostic and often therapeutic challenge for ophthalmologists. Although it commonly presents with acute ocular findings consistent with inflammation and responds well to corticosteroids, it is a diagnosis of exclusion that is associated with many other disorders. Complete and detailed medical history, relevant serological tests and imaging studies are important to exclude systemic 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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