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Case Report

Orbital pseudotumor: A masquerading presentation

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Abstract

Purpose: We describe the clinical presentation of a 65yr old male patient with orbital pseudotumor of the right eye with secondary Ocular Surface Squamous Neoplasia, not responding to conventional steroid therapy.

Observation: Reporting a case of Pseudotumor of the right orbit with secondary Ocular surface Squamous Neoplasia in a 65yr old diabetic male presenting with painless proptosis and redness of the right eye since 6months. CT orbit scan was suggestive of a Thyroid orbitopathy. Patient initially responded well to steroid therapy along with oral thyroxine and proptosis decreased, but relapsed in few months. Patient was readmitted and a Contrast enhancemed MRI of orbit was done-suggestive of right eye Orbital pseudotumor. A detailed blood investigation and whole-body workup, Fine Needle Aspiration Cytology studies were done to rule out secondaries to the eye and confirmed to have orbital pseudotumor. Patient was started on immunomodulators Oral Methotrexate 7.5mg once a week, after obtaining clearance. Excision biopsy of the proptosed exposed conjunctiva was done and on histopathological examination diagnosis of Orbital pseudotumor tumor was confirmed with Ocular surface squamous neoplasm due to chronic conjunctival exposure secondary to the proptosis.

Conclusion: Patients with orbital pseudotumor typically respond to steroid therapy. However, in patients who are unresponsive or have recurrences, diagnosis should be revised or reconfirmed. Radiotherapy and immunomodulators are modes of treatment of unresponsive cases. Diagnosis and management of atypical cases pose a major clinical challenge.

Keywords: Biopsy, Steroids, Methotrexate, Orbital pseudotumor, Proptosis, Thyroid ophthalmopathy,

Introduction

Orbital pseudotumor, is a benign, noninfective, nonspecific inflammatory condition of the orbit. It has no identifiable local or systemic causes.[1] It is the third most common orbital disease with overall incidence of 4.3% to 7.3%. Corticosteroids are the corner stone of therapy, however refractory or persistent cases may require radiotherapy and immunomodulator therapy, Refractory cases pose a diagnostic and therapeutic challenge.[1]

Case Report

A 65-year-old diabetic male patient presented with history of redness, painless swelling and outward protrusion of the right eye since the last 6 months. He had no visual symptoms. Ocular examination revealed a 5 mm upward and 3 mm out ward displacement of the right eye with lid edema, diffuse conjunctival congestion and grade 4 chemosis (Fig. 1). There was no local rise of temperature, no tenderness, no palpable mass could be felt, fingers could be insinuated all around the orbital margin and retropulsion test was negative. Visual acuity was 20/200 in right eye and 20/20 in left eye. Pupil were 3 mm round regular briskly reactive to light in both eyes, both direct and consensual light reflex present. Right eye lens was senile cataract – nuclear sclerosis with posterior subcapsular cataract. Extraocular movements of right eye were mildly restricted due to chemosis. Fundus examination of right eye revealed a crowded hyperemic disc. Left eye fundus was normal.

Initial Computed tomography of the orbit (CT) showed proptosed right globe with increased periorbital fat contour with muscle enlargement suggestive of a thyroid orbitopathy. Thus, patient was started on Tablet Levothyroxine 50 µg OD as advised by physician and oral prednisolone 60 mg OD (1 mg/kg body) weight with gradual tapering. Initially the proptosis and chemosis reduced with steroids. However, on attempts of tapering steroids recurrence was noted. Over the span of 6 months, 3 such recurrences were noted on attempts of tapering steroids suggestive of a steroid dependence.

On second admission a Contrast Enhanced Magnetic Resonance Imaging of orbits and Paranasal Sinuses was carried out which showed subtle fat stranding in right retroorbital perineural region with subtle contrast enhancement which was in favor of orbital pseudotumor (Fig. 2 and 3).

Basic blood investigation and other parameters including complete hemogram, erythrocyte sedimentation rate, C-reactive protein level, antinuclear antibody and antineutrophil cytoplasmic antibodies were done, which were within normal limits except for high blood sugars, for which patient was started on Insulin. A full body workup was done to rule out metastasis and secondaries to the eye.

As patient was an uncontrolled diabetic, Mucor mycosis was also suspected. Otolaryngologist opinion was taken and Mucormycosis was ruled out. Neurologist opinion was sought for, who by previous clinical experience suggested

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http://doi.org/10.18231/j.ijooo.2019.038

Fine Needle Aspiration Cytology to rule out of slow growing fungal etiology.

Fine Needle aspiration Cytology was done, studies of the tissue showed granular necrotic material and areas of fibrin with sparse mixed inflammatory cell infiltrate. Microbiological studies of KOH, Periodic Acid Schiff were negative and culture revealed sterile sample (Fig. 4).

Patient was started on immunomodulators after obtaining physician clearance. Tablet Methotrexate 7.5 mg once a week under cover with folinic acid was started. The proptosis reduced however chemosis was persistent after 4 doses of oral Methotrexate (Figure 5).

An incisional biopsy was then planned. Specimen consisting of exposed conjunctiva, subtenons tissue and muscle was sent for Histopathological and microbiological studies (Fig. 6). Histopathological diagnosis which was conclusive of a full thickness conjunctival dysplasia with tenon and muscle showing fibrofatty tissue. Microbiological studies of KOH, Periodic Acid Schiff were negative and culture revealed sterile sample.

Excision biopsy of exposed proptosed conjunctiva was done to provide symptomatic relief (Fig. 7) and specimen was sent for thorough histopathological analysis which was conclusive of Ocular Surface Squamous Neoplasia.

Patient was continued on Oral Methotrexate therapy, after 4 doses, an impression cytology was done (Fig. 8) and patient was free of dysplasia.

Fig 1: Patient at presentation with right Globe proptosis and comjunctival chemosis

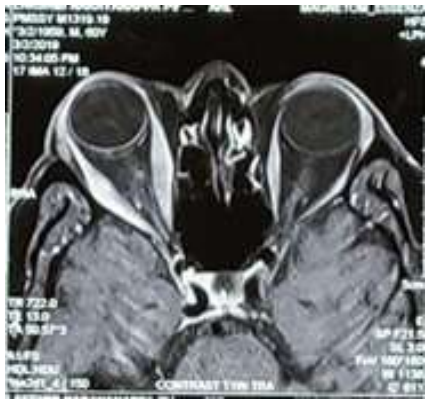

Fig. 2: MRI SCAN of Orbit and Brain

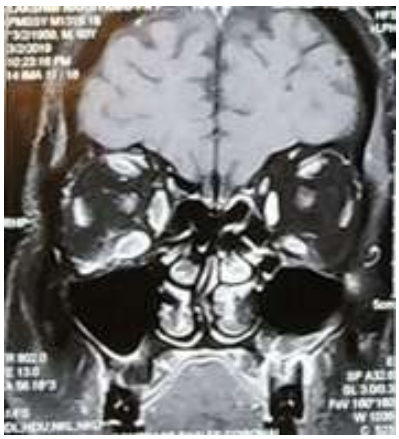

Fig. 3: MRI Scan of Orbit and Brain

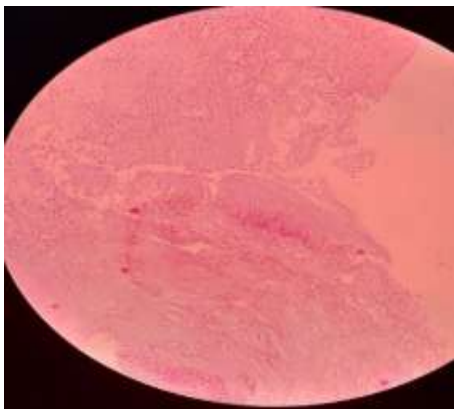

Fig 4: Fine Needle Aspiration cytology slide

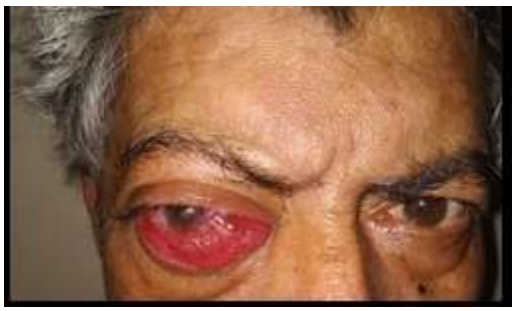

Fig 5: Patient with increasing chemosis despite medical therapy

Fig. 6: Incision biopsy samples from conjunctiva, tenons and lateral recti muscle

Fig. 7: Post Operative picture of the patient

Fig. 8: Impression Cytology being done

Discussion

Orbital pseudotumor is a benign, noninfective, nonspecific, non inflammatory clinical conditions of the orbit without identifiable local or systemic causes.[1,2] The term "orbital pseudotumor" has been used to describe inflammatory lesions of the orbital tissues of unknown aetiology simulating a neoplasm of the orbit, The name is misleading because the condition is an inflammatory tumefaction and not a neoplastic one.[2] It was first described in 1903 by Gleason and by Busse and Hochheim and characterized as a specific clinicopathological entity in 1905 by Birch-Hirschfeld.[3]

It is the third most common orbital disease, following Graves orbitopathy and lymphoproliferative diseases. Which accounts for 4.7% to 6.3% of orbital disorders.[1,4,5,6] It is a diagnosis of exclusion.[4,6] The clinical picture may be mimicked by thyroid myopathy[2] as initially diagnosed in our case. Other differential diagnosis includes orbital cellulitis, Vasculitis, foreign body, sarcoidosis and neoplasms.[1,4]

Unilateral disease is the rule in adults. In children it may present bilaterally.[2,4,6,7] The peak incidence of the condition is middle age[2] or in fourth and fifth decade but it can also occur in children. There is no sex predilection [1,4]

Immune mediated processes have been linked to development of this condition, including upper respiratory tract infection, flu like viral infection Crohns disease, Systemic lupus erythramatosus, Diabetes mellitus, Myasthenia Gravis, ankylosing spondylitis, rheumatoid arthritis have been associated with Idiopathic orbital inflammation.[1] It is mediated by both B and T lymphocytes. Successful treatment of the condition with corticosteroids and other immunosuppressive agents suggests an autoimmune mechanism.[1,4]

Serologic studies are necessary to exclude a systemic cause.[6] Which include complete hemogram, erythrocyte sedimentation rate, C-reactive protein level, antinuclear antibody and antineutrophil cytoplasmic antibodies.[4,8] In our case all the above parameters were within normal limits.

Clinically, acute orbital pseudotumor typically have a rapid onset and frequently produce proptosis, lid swelling, chemosis, pain, and limitation of ocular movement. Papilledema and optic neuropathy may also occur.[2,4,8,9]

It may present as dacryoadenitis, sclerotenovitis or as a diffuse anterior soft tissue inflammation.[7] In our case on 2 months follow up, patient developed papilledema due to Central Retinal vein occlusion with Macular edema.

On Computed tomography, the pseudotumor reveals a broad range of pathological changes. It may present as contrast enhancing diffuse or focal mass. Common manifestations are infiltration of retrobulbar fat, proptosis, extraocular muscle enlargement, muscle tendon or sheath enlargement whereas tendons are spared, apical fat oedema, optic nerve thickening, uveal scleral thickening, oedema of tenons capsule and lacrimal gland infiltration.[1]

On MRI it appears hypointense to fat on T1 weighted images and isointense or hypointense to fat on T2 weighted images with marked gadolinium enhancement.[8] MR imaging now provides prognostic significance.

Patients who present with multiple recurrences, or those unresponsive to therapy, should have biopsy samples taken to rule out leukaemia or lymphoma [1,6,9] Histopathological diagnosis can be arrived at by fine needle aspiration and cytology (FNAC) or incisional biopsy.[4] The histological picture varies between lymphocytic developments, true granulomatous infiltration, dense hyalinization, and the involvement of a particular orbital structure. No difference can be established between the histological groups as far as clinical course, symptomatology, or prognosis is concerned.[2] In our case Fine Needle aspiration Cytology was done, studies of the tissue showed granular necrotic material and areas of fibrin with sparse mixed inflammatory cell infiltrate. Incisonal biopsy was also done, Specimen consisting of exposed conjunctiva, subtenons tissue and muscle was sent for Histopathological examination which was conclusive of a full thickness conjunctival dysplasia with tenon and muscle showing fibrofatty tissue.

Corticosteroids are the mainstay of therapy and are administered for several months to ensure remission.[1,2,4] The response to steroids is so pathognomonic, that the diagnosis is often made retrospectively.[2,10]

Systemic corticosteroid therapy (1.5-2 mg/kg/day dose and tapered slowly over eight to ten weeks following resolution of signs and symptoms) is the mainstay of the treatment of "Pseudotumor" with the exception of a sclerotic variant. However, treatment failure with steroid therapy is common and is reported to occur in the range of 33 to 52%. Recurrence of disease on steroid taper and adverse effects of steroids are the common causes of treatment failure.[3]

In our case the patient was started on oral prednisolone 60 mg OD (1 mg/kg body) weight with gradual tapering. Initially the proptosis and chemosis reduced with steroids. However, on attempts of tapering steroids recurrence was noted. Over the span of 6 months, 3 such recurrences were noted.

Radiotherapy and immunomodulators are modes of treatment of unresponsive cases. Radiotherapy low dose external beam irradiation 15-20 Gy fraction over 10 days[1] may be used in patients who fail to respond to steroids or have a rapidly progressive course. In patients who are refractory to both corticosteroids and radiotherapy, anecdotal reports have suggested the use of chemotherapeutic agents such as cyclophosphamide 200 mg/day, methotrexate 7.5-12.5 mg/kg, and cyclosporine 2-5 mg/kg.[1,8,9] However, treatment with antimetabolites warrants close monitoring for complications like bone marrow suppression and liver dysfunction, especially as long-term treatment is required.[3]

In conclusion high-dose systemic steroid therapy is the first-line treatment, but refractory cases account for a significant proportion of treatment failures. In such recalcitrant cases, immunosuppressive therapy with antimetabolites (methotrexate and azathioprine) is a safe and effective treatment alternative to steroids.[3]

Conclusion

Patients with classic findings of orbital pseudotumor may be treated without a biopsy. These cases typically respond rapidly to steroid therapy. However, in patients who are unresponsive or have recurrences, diagnosis can be established with the help of biopsy. Radiotherapy and Immunomodulators are modes of treatment of unresponsive cases. Diagnosis and management of atypical cases pose a major clinical challenge.

Source of Funding

None.

Conflict of Interest

None.

References

1. Yuen, S. (2003). Idiopathic Orbital Inflammation. *Arch Ophthalmol* 121(4):491
2. Blodi, F. and Gas, J. Inflammatory pseudotumour of the orbit. *Br J Ophthalmol* 1968;52(2):79-93.
3. Priya, Y., Nithyanandam, S. and Reddy, M. Role of antimetabolites in recalcitrant idiopathic orbital inflammatory syndrome. *Oman J Ophthalmol* 2011;4(1):21.
4. Srinivasan R., Datta G. Orbital Pseudotumor. *Kerala J Ophthalmol* 2009; 21(2):127-31.
5. Zhu J, Zhan W, Wang L. and Wang Y. A Case Study on Idiopathic Orbital Pseudotumor: Surgery and Steroid Treatment. *Tropical J Pharm Res* 2015;14(11):2135.
6. Szabo B, Szabo I, Crişan D, Ştefănuţ C. Idiopathic orbital inflammatory pseudotumor: case report and review of the literature. Rom J Morphol Embryol 2011;52(3):927–30.
7. Gupta A, Sengupta S, Srinivasan R. and Suchi S. Chronic bilateral lacrimal gland pseudotumor in an adult. *Indian J Ophthalmol* 2008;56(1):86
8. Birch-Hirschfeld A *Handbuch der Gesamten Augenheilkunde*. Berlin, Germany Julius Springer1930;9
9. Nugent, R., Rootman, J., Robertson, W., Lapointe, J. and Harrison, P. (). Acute orbital pseudotumors: classification and CT features. *Am J Roentgenology* 1981;137(5):957-62.
10. Maurya, V. and Gautam, S. Orbital Pseudotumour. *Med J Armed Forces India* 2007;63(3):292-3.

How to cite this article: Anuradha A, Gupta P, Sultana N, Bekal D, Dogra M. Orbital pseudotumor: A masquerading presentation, *Int J Ocul Oncol Oculoplasty* 2019;5(3):146-9.