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Case Report

Orbital pseudolymphoma: a benign condition mimicking malignancy

Nirupama Kasturi, Salwa Kaidakath*, Mary Stephen, Jayasri P.

Department of Ophthalmology, JIPMER, Puducherry, India

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***Correspondence:**
Dr. Salwa Kaidakath,
E-mail: sksalwaibrahim@gmail.com

ABSTRACT

Orbital lymphomas are not a rare condition that causes systemic and ocular morbidity, in which vision loss often occurs due to compressive optic neuropathy, and whose treatment is complex, involving chemotherapy and radiotherapy. We report a 54-year-old male with protrusion of the right eye associated with mild diminution of vision for one year, limitation of extraocular movements, and a relative afferent pupillary defect with optic disc edema. Imaging revealed a well-defined soft tissue lesion filling the retro-orbital intraconal space, suggestive of orbital lymphoma. Incisional biopsy and fine needle aspiration cytology revealed the lesion to be a reactive lymphoid hyperplasia. Following the administration of oral steroids, the patient showed significant improvement. To our knowledge, a benign lesion with an aggressive presentation is rare, and histopathology plays an important role in diagnosis and management.

Keywords: Orbital pseudolymphoma, Proptosis, Compressive optic neuropathy, Disc edema, Steroids

INTRODUCTION

Orbital tumours are not uncommon in adults, and lymphoproliferative disorders, which can be divided into reactive lymphoid hyperplasia, atypical lymphoid hyperplasia, and malignant lymphoma, all have similar clinical presentations.[1] Ocular morbidity occurs through a variety of mechanisms, including compressive optic neuropathy, exposure keratopathy, and elevated episcleral venous pressure. Even though imaging techniques have advanced, histopathological analysis is still required for a definitive diagnosis and treatment to be initiated. In our report, an adult male presented with proptosis of the right eye and compressive optic neuropathy, which was initially diagnosed as orbital lymphoma based on clinical picture and imaging, but was later found to be a pseudolymphoma that was well managed with steroids.

CASE REPORT

A 54-year-old male with no known comorbidities presented with an insidious onset, painless, progressive, protrusion of the right eye for 1 year, associated with diminution of vision in the right eye. During the examination, the right eye's visual acuity was 6/9 and the left eye's was 6/6, as measured by Snellen's chart. He had right eye axial proptosis with Hertel's exophthalmometer reading of 22 mm in the right eye and 13 mm in the left eye at 101 mm, with limitation of extraocular movements in the right eye, conjunctival chemosis, and grade I relative afferent pupillary defect also found. Fundus examination revealed disc edema in the right eye and normal fundus in the left eye (Figure 1a). The patient had impaired colour vision and contrast sensitivity in the right eye. Contrast enhanced MRI images showing a relatively well-defined soft tissue lesion filling the retro-orbital intraconal space on the right side showed homogenous post-contrast enhancement and diffusion restriction, indicating orbital lymphoma (Figure 1d). Despite the fact that the clinical picture and imaging suggested malignancy, incisional biopsy and fine needle aspiration cytology were performed under local anaesthesia after the medial rectus muscle was disinserted intra-operatively. It appeared as a homogenous, pink fleshy mass with a lobulated surface. The procedure

was not sono-guided. Histopathologic examination showed a mixture of reactive T and B cells highlighted with CD3 and CD20 respectively, with no atypical cells suggestive of reactive lymphoid hyperplasia (Figure 1e). The patient was started on high dose oral steroids-Tab Prednisolone 1mg/kg with close follow-up. After two weeks, the patient was symptomatically better with a best corrected visual acuity of 6/6 in the right eye and a three mm decrease in proptosis of the right eye, with significant improvement in extraocular movements and resolution of the disc edema in the right eye (Figure 1b). Patient missed medication and the need for long-term steroids was explained; the patient was kept on close watch with slow steroid tapering over a period of 6 months.

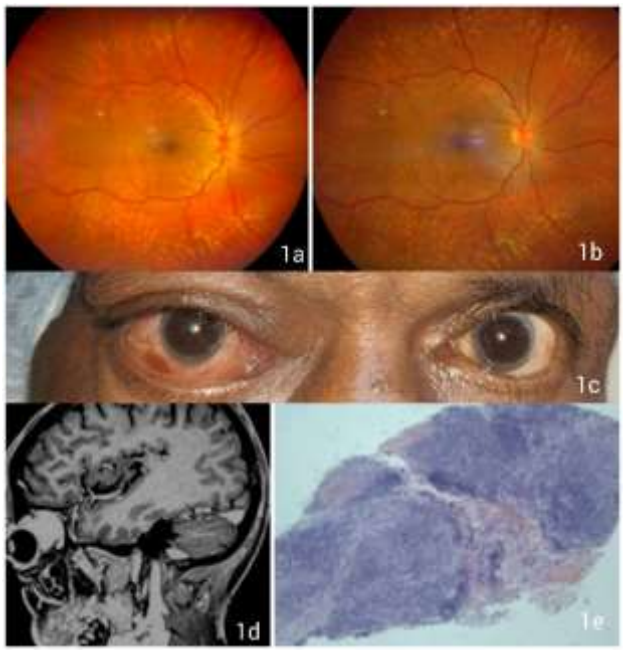

Figure 1: a) Colour fundus photograph of patient right eye showing 360 degree blurring of optic disc margin with obliteration of cup disc ratio and pigmentary abnormality around arcade, b) Colour fundus photograph of patient right eye showing normal optic disc appearance (Post steroid treatment) with similar pigmentary lesion around arcade, c) Clinical photograph of patient showing presence of axial proptosis and inferior scleral show, d) MRI image of axial section showing presence of mass lesion in retroconal space pushing the globe anteriorly, e) Histopathology examination of orbital intraconal mass showing CD3+ paracortical cells (H&Ex100), f) Histopathology examination of orbital intraconal mass showing CD20+ germinal cells (H&Ex200).

DISCUSSION

The most common primary orbital tumours in older adults are lymphoproliferative lesions, of which 67-90% are orbital lymphomas and only 8-19% are reactive lymphoid hyperplasia.[2] Benign lymphoid hyperplasia is an idiopathic lymphocytic cellular response to some inflammatory stimulus, which could be an exogenous or endogenous antigen. Lymphoid hyperplasia does not have any gender or racial predilection. It is seen in adults in the fifth to seventh decades. Patients present with lid edema, proptosis, ptosis, and extraocular movement limitations like any other orbital mass lesion.[3] Orbital lymphoid hyperplasia is predominantly extraconal, and reactive lymphoid hyperplasia is difficult to differentiate from orbital lymphoma clinically and radiologically. Hence, a tissue biopsy is mandatory in all cases. Though benign, there are rare chances of developing non-Hodgkin's lymphoma, which makes follow-up for at least five years necessary. The diagnostic criteria for reactive lymphoid hyperplasia include the presence of a solitary or multiple orbital mass with radiological features suggesting homogenous internal architecture and no bony erosion, a biopsy showing mature lymphocytes, well-defined architecture, scant or no fibrosis, polyclonal immunotyping, no features of alternate diagnosis, and no systemic features or blood investigations suggestive of malignancy.[4] On contrast imaging, orbital lymphoid hyperplasia will be a well-defined mass lesion that shows diffuse contrast enhancement. The lesion will usually mould to the surrounding solid structures such as the globe, orbital wall, and optic nerve, causing less globe displacement. Histopathology reveals a mixture of reactive T and B lymphocytes, with T cells accounting for more than 40% of the total, with their specific cell markers, CD 20 for reactive B cells and CD3/CD 43 for reactive T cells.[5] The dense lymphocytic infiltrates are usually organised into secondary follicles with well-defined zonal architecture. Ki67 staining will be restricted to germinal centres that have vigorous mitotic activity. Immunohistochemical staining will show an orderly compartmentalization of B and T cells. B cells are positive for CD20, CD10, BCL6, and retinoblastoma protein. T cells are positive for CD3, CD5, and BCL2. The first line of management is oral corticosteroids, but they will not decrease the risk of its transformation into lymphoma.[6] If the lesion is limited to one orbit or in recurrent cases where long term steroids cannot be used, radiotherapy is the treatment of choice. The monoclonal antibody Rituximab can be used in refractory cases. Though benign, they can cause complications like compressive optic neuropathy, as in our case, which is an atypical presentation. Our patient, despite having defaulted on medications for a brief time, responded very well to treatment and showed good visual recovery.

CONCLUSION

The lymphoproliferative disorders have similar clinical presentations which put forward diagnostic challenges both clinically and radiologically. Hence the tissue biopsy becomes mandatory. Despite initial clinical and imaging indications of orbital lymphoma in this case, subsequent histopathological analysis identified a pseudolymphoma, effectively managed with high-dose oral steroids, resulting in substantial symptomatic improvement and visual recovery. This case highlights the significance of biopsy in

differentiating between reactive lymphoid hyperplasia and malignancy, given their overlapping clinical features. The distinct histopathological criteria, along with treatment modalities emphasizing corticosteroids and radiotherapy when warranted, are crucial in managing these benign yet potentially complicated lesions, such as compressive optic neuropathy, as evidenced in this atypical presentation.

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REFERENCES

1. Olsen TG, Heegaard S. Orbital lymphoma. Surv Ophthalmol. 2019;64(1):45-66.
2. Vogele D, Sollmann N, Beck A, Haggenmüller B, Schmidt SA, Schmitz B, et al. Orbital Tumors-Clinical, Radiologic and Histopathologic Correlation. Diagnostics. 2022;12(10):2376.
3. Montejano-Milner R, López-Gaona A, Fernández-Pérez P, Sánchez-Orgaz M, Romero-Martín R, Arbizu-Duralde A. Orbital metastasis: Clinical presentation and survival in a series of 11 cases. Arch Soc Esp Oftalmol. 2020:20:30317-8.
4. Garner A. Orbital lymphoproliferative disorders. Br J Ophthalmol. 1992;76(1):47-8.
5. Li EY, Yuen HK, Cheuk W. Lymphoproliferative Disease of the Orbit. Asia Pac J Ophthalmol. 2015; 4(2):106-11.
6. Olsen TG, Holm F, Mikkelsen LH, Rasmussen PK, Coupland SE, Esmaeli B, et al. Orbital Lymphoma-An International Multicenter Retrospective Study. Am J Ophthalmol. 2019;199:44-57.

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