Raguraman *et al.*
The Egyptian Journal of Otolaryngology (2023) 39:66
https://doi.org/10.1186/s43163-023-00419-x

The Egyptian Journal of Otolaryngology

CASE REPORT

Open Access

Proptosis under evaluation—a rare case report on pseudolymphoma of orbit

Sharulatha Raguraman, Akshat Kushwaha* and Lokesh Kumar Penubarthi

Abstract

Background Orbital pseudolymphoma is one of the rare diseases affecting the ocular adnexa and causing various symptoms requiring histopathological evidence for diagnosis and appropriate treatment with oral steroids or immunosuppressive agents. However, there is less literature available on this disease, thus adding to it will help in the early diagnosis and treatment of the patients.

Case presentation A 48-year-old female from a village in South India came to our tertiary care centre with the only complaints of right eye protrusion for 1 year. With a working diagnosis of intraorbital mass, radiological investigations like contrast-enhanced CT and MRI were taken. It showed well-capsulated mass involving the right inferior rectus extending into the intraconal space just abutting the optic nerve. Trans-antral endoscopic biopsy under general anaesthesia was taken and reported as pseudolymphoma proven with CD 3 and CD 20 immunohistochemistry markers. The patient was treated with oral corticosteroids and tapered over a period of 3 months. During the treatment period, the patient developed no diminution of vision or new complaints.

Conclusion Orbital pseudolymphomas are benign lymphoproliferative diseases affecting the ocular adnexal tissue. Patients can present with painless ptosis, proptosis, diplopia, or eyelid swelling. Prompt diagnosis and treatment result in the remission of the disease.

Keywords Painless proptosis, Pseudolymphoma of orbit, Proptosis, Lymphoproliferative disease, Benign lymphoid hyperplasia

Introduction

Orbital pseudolymphoma is one of the rare diseases affecting the ocular adnexa causing various symptoms. It is not a tumour per se but an inflammatory response to a known or unknown cause. Diagnosis requires histopathological evidence and appropriate treatment with oral steroids or immunosuppressive agents.

Background

Pesudolymphoma is a benign inflammatory response that can mimic lymphoproliferative disorder or malignancy. It is composed of highly reactive cells of lymphoid origin. Orbital pseudolymphomas are rare clinical pathology that can occur in adnexal structures like the lacrimal glands, orbital soft tissue, and extraocular muscles. Orbital lymphomas are usually associated with IgG4-related diseases. Here, we present a case of orbital pseudolymphoma, which is not associated with IgG4-related disease.

Case presentation

A 48-year-old woman from a low socioeconomic status in a village in Tamil Nadu presented to the Otorhinolaryngology Outpatient Department in our tertiary care centre in South India with complaints of insidious onset and gradually progressing painless protrusion of the right eye for 1 year. There was no increase in swelling on lifting weights, coughing, or straining. The patient denied any complaints of visual disturbance,

*Correspondence:
Akshat Kushwaha
akshatk16@gmail.com
[1] Department of Otorhinolaryngology, JIPMER, Puducherry, India

nasal obstruction, or bleeding from the nose. She did not give any history of headaches, vomiting, and seizures. No history of facial pain or decreased sensation on the face. No history of trauma. She did not have a history of fever, weight loss, or any other associated illnesses or immunocompromised states or any surgeries in the past. Physical examination revealed abaxial proptosis with the eyeball pushed supero-laterally. No chemosis or congestion was noted. With the help of the Ophthalmology Department, complete vision and fundus examination were done. The patient had a normal distant and near vision along with normal extraocular movements in both eyes. The bilateral fundus showed normal findings. Ear, nose, and throat examination was unremarkable. Systemic examination revealed no other lymphadenopathy, organomegaly, or swelling in the body.

Differential diagnoses such as orbital lymphoma and metastasis from unknown primary and neurogenic tumours of the orbit were kept in mind, but most of them could not be ruled out based on clinical examination alone. Hence, a contrast-enhanced CT was taken (Fig. 1). It showed a 3×2.5 cm brilliantly enhancing mass arising from the inferior rectus muscle extending intraconally. Magnetic resonance imaging was taken in T1 isointense and T2 hyperintense mass arising from the inferior rectus muscle just abutting the optic nerve (Fig. 2). No infiltration to adjacent structures or bony erosion was noted. As per the imaging study, the mass did not have any intracranial extension.

In order to obtain a histopathological diagnosis, an initial fine needle aspiration was thought of but avoided owing to the deeper location of the mass and the potential risk of bleeding into the orbit. The patient was then worked up for intraorbital biopsy under general anaesthesia via the Caldwell-Luc approach and infraorbital approach. Although more invasive as compared to the endoscopic approach, it was selected to avoid inadvertent injury to the medial rectus muscle as endoscopic surgery would require a breach in the lamina papyracea to gain access to the tumour. Intraoperatively, a well-encapsulated firm tumour was noted in the inferior part of the orbit. A biopsy was taken and sent for histopathological examination.

During the postoperative period, the patient had no deterioration of vision or development of new symptoms (Fig. 3). Intra-op biopsy was reported as pseudolymphoma with reactive nature of lymphoid cells proven by CD3 and CD20 IHC. Ki67 highlighted the germinal centre. No increase in IgG/IgG4 ratio ruling out the possibility of IgG4-related disease.

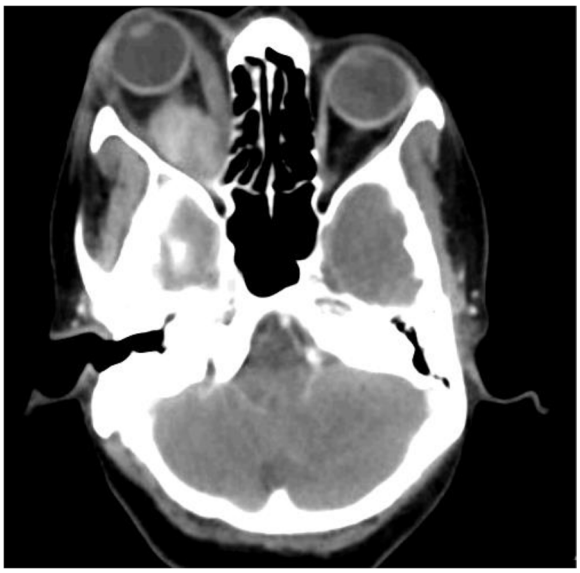

Fig. 1 Contrast-enhanced CT showing a 3×2.5 cm brilliantly enhancing mass arising from the inferior rectus muscle with intraconal extension

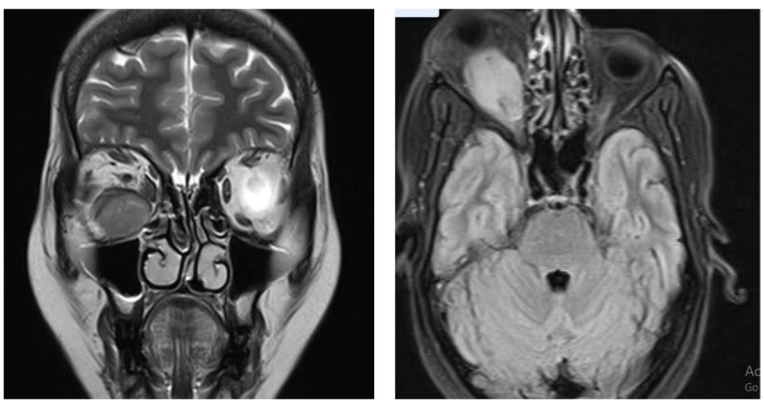

Fig. 2 Contrast-enhanced MRI showing mass arising from the inferior rectus muscle just abutting the optic nerve

Fig. 3 Pre-op picture showing right eye proptosis and post-op picture

The patient was explained about the modalities of treatment available, namely oral corticosteroids and monoclonal antibody against CD 20. The patient opted for a low cost-effective treatment, and she was started on oral steroids initially at 1 mg/kg per day for 2 weeks. During the follow-up, the patient's proptosis decreased significantly. Oral steroids were then tapered gradually over a period of 3 months. During the follow-up period, the patient had no new complaints and developed no new symptoms or recurrence of the disease.

Discussion

Lymphoproliferative diseases account for 15% of all space-occupying lesions in orbit [1]. Distinguishing them as benign or malignant lesions is always a diagnostic challenge. Primary orbital lymphoid lesions are categorised into three groups: idiopathic inflammation/inflammatory pseudotumor, reactive lymphoid hyperplasia/pseudolymphoma, and malignant lymphoma [2].

Pseudolymphomas are usually idiopathic and are distributed in the orbit, lacrimal gland, conjunctiva, eyelid, and uvea, in that order of frequency [3]. The most frequent symptoms of orbital pseudolymphoma are painless swelling, proptosis, ptosis, diplopia, or disturbance in vision. Computed tomography and magnetic resonance imaging do not show any specific findings for pseudolymphoma. Thus, histopathological examination becomes inevitable to get a diagnosis.

Fine needle aspiration and biopsy play a greater role in diagnosing this spectrum of disease. An infiltrate showing a heterogenous population of cells consisting of lymphocytes, histiocytes, and plasma cells and benign lymphoid follicles with reactive germinal centres are categorised as pseudolymphoma. It also has a higher proportion of B cells (but not more than 75%) in the background of T cells [4].

Benign lymphoid hyperplasia has the ability to undergo malignant transformation, thus prompt diagnosis and treatment of the illness become necessary [5]. Treatment of orbital pseudolymphoma has traditionally been oral steroids and external beam radiotherapy (EBRT) [6]. The literature search also shows the use of monoclonal antibody against CD 20 (rituximab) as a newer line of treatment [4]. Patients who do not tolerate systemic steroids and EBRT can be given a trial of rituximab.

Conclusion

Orbital pseudolymphomas are benign lymphoproliferative diseases affecting the ocular adnexal tissue. Patients can present with painless ptosis, proptosis, diplopia, or eyelid swelling. Prompt diagnosis and treatment with steroids result in the remission of the disease.

Acknowledgements
Not applicable.

Authors' contributions
All the authors have equally contributed to the case report. SR and AK are the major contributors in writing the manuscript. AK and LP participated in the writing, editing, and data interpretation. LP and AK are the operating surgeons. All authors have equally participated in writing the manuscript. The authors read and approved the final manuscript.

Funding
There was no funding required to take up the study.

Availability of data and materials
The datasets during and/or analysed during the current study are available from the corresponding author upon reasonable request.

Declarations

Ethics approval and consent to participate
The study was taken after obtaining an ethics committee approval from the Institution of Ethics Committee, JIPMER, Puducherry, India.

Consent for publication
Written informed consent for the publication of their clinical details and/or clinical images was obtained from the patient. A copy of the consent form is available for review by the editor of this journal.

Competing interests
The authors declare that they have no competing interests.

Received: 30 December 2022 Accepted: 23 March 2023
Published online: 18 April 2023

References

1. van der Gaag R, Koornneef L, van Heerde P, Vroom TM, Pegels JH, Feltkamp CA et al (1984) Lymphoid proliferations in the orbit: malignant or benign? Br J Ophthalmol 68(12):892–900
2. Subramanian R, Solo S, Mishra MM, Murugan P, Siddaraju N, Basu D et al (2007) Fine needle aspiration cytology of primary lymphoid lesions of the orbit. Acta Cytol 51(3):417–420
3. Ulivieri S, Motolese PA, Motolese I, Motolese E, Menicacci F, Gennari P et al (2009) Benign orbital pseudolymphoma. Case report. Il G Chir 30:299–301
4. Witzig TE, Inwards DJ, Habermann TM, Dogan A, Kurtin PJ, Gross JB et al (2007) Treatment of benign orbital pseudolymphomas with the monoclonal anti-CD20 antibody rituximab. Mayo Clin Proc 82(6):692–699
5. Sood N, Soni PB (2016) Orbital lymphoid lesions: short series with cytohistological correlation. Ann Pathol Lab Med 3:C72–77
6. Baldini L, Blini M, Guffanti A, Fossati V, Colombi M, LaTargia ML et al (1998) Treatment and prognosis in a series of primary extranodal lymphomas of the ocular adnexa. Ann Oncol 9(7):779–781

Publisher's Note