Case Report

Orbital lymphoma - A diagnostic challenge

Anamika H Agrawal[1], Vidhya D Khobragade[2*], Ankita A Patil[3], Anjali D Nicholson[4]

[1]Additional Professor, [4]Professor and HOD, Department of Ophthalmology, TNMC, Mumbai -400008, Maharashtra, INDIA.
[2,3]Medical Officer, KEM Hospital and Seth G. S. Medical College, Parel, Mumbai-400012, Maharashtra, INDIA.
Email: vidhyak510@gmail.com

Abstract

We report a case of young female with subconjunctival mass that was reported to be inflammatory on radiological imaging but did not respond to local and systemic steroids given initially. So after being reviewed by radiology, medicine and hematology an excision biopsy was planned which revealed non hodgkins lymphoma on histopathological examination. She was referred to oncology department where she was successfully managed by radiotherapy without any recurrence on follow up. Here we highlight the diagnostic challenge to an ophthalmologist in a case of lymphoma and the need of multidisciplinary approach for successful management of such cases.

Keywords: Subconjunctival mass, histopathology, lymphoma, radiotherapy.

[*]**Address for Correspondence:**
Dr. Vidhya D Khobragade, Medical Officer, KEM Hospital and Seth G. S. Medical College, Parel, Mumbai-400012, Maharashtra, INDIA.
Email: vidhyak510@gmail.com

Received Date: 10/01/2016 Revised Date: 14/02/2016 Accepted Date: 12/03/2016

Access this article online

Quick Response Code:

Website: www.statperson.com

DOI: 02 April 2016

INTRODUCTION

Subconjunctival masses could be due to degenerative conditions like pingecula, pterygium benign epibulbar tumours like conjunctival naevus, conjunctival papilloma, dermoid, dermolipoma malignant lesions as melanoma ocular surface squamous neoplasia and lymphoma. An experienced ophthalmologist can differentiate these conditions based on their appearance, age presentation and careful slit lamp examination. Lymphoid proliferations can affect the eye in various ways. Non-Hodgkin's lymphoma (NHL) is the commonest type of lymphoma involving ocular and orbital structures. Primary non-Hodgkin's lymphoma (NHL) of the orbit is a rare presentation, representing 8-10% of extranodal NHL and only 1% of all NHL.[1-2] Ocular NHL can either occur in isolation or in association with CNS or systemic involvement. Orbital lymphoma may be unilateral or bilateral and up to 20% bilateral presentation is noted. [3] Ocular lymphoma is probably the most elusive intraocular tumor to diagnose. It frequently masquerades as other more benign ocular lesions. Diagnosis can be difficult and it is frequently delayed as the clinical condition can mimic a number of other ocular conditions. Correct diagnosis thus depends on a high index of suspicion and frequently requires radiologic imaging, and histopathological analysis. Furthermore, ocular manifestations of lymphoma are generally rare events. Thus it is thus important to suspect and review the ocular manifestations of lymphoma and investigate it accordingly for prompt diagnosis and treatment.

CASE REPORT

A 38 year old healthy unmarried female presented to our outpatient department with history of painless, slow growing mass in her left eye for the past six months .She did not have any visual or systemic complaints. On examination the best corrected visual acuity in both eyes was 6/6. Left eye showed 4 x 6 mm, oval, soft, non tender, non mobile mass at medial aspect of bulbar conjunctiva extending below plica with normal overlying conjunctiva. Rest of the anterior segment examination was within normal limits. There was 14pd of exodeviation capable of central fixation with normal extra ocular movements. The anterior segment examination of right eye was within normal limit. The pupillary reaction and fundus examination in both eyes was within normal limits. All systemic examination was within normal limits. All routine blood investigations including thyroid functions were normal. Peripheral blood smear did not show any abnormal cells or any parasitic cyst. USG B Scan showed well defined hypoechoic mass lesion over

How to site this article: Anamika H Agrawal, Vidhya D Khobragade, Ankita A Patil, Anjali D Nicholson. Orbital lymphoma - A diagnostic challenge. *International Journal of Recent Trends in Science and Technology*. April 2016; 18(3): 391-393. http://www.statperson.com (accessed 04 April 2016).

the nasal conjunctiva with no calcification or infiltration. CT Orbit revealed well defined peripherally enhancing cystic lesion medial to left globe in contact with bulky enhancing medial rectus muscle with adjacent fat stranding but no evidence of any calcification .Mild thickening of coats of left globe were noted .Rest of the extra ocular muscles were normal. Further MRI orbit was adviced that revealed ill defined enhancement and fat stranding in the region of medial canthus of left orbit and retro orbital portion with bulky medial rectus muscle and thickened posterior coats of the globe. These findings were reported to be suggestive of inflammation. Based on this the differential diagnosis of parasitic cyst, or orbital pseudotumor or ocular lymphoma was presumed. So the patient was treated with tab*let al*bendazole 100 mg twice a day for 3 days and tab Wysolone 50 mg / day with tapering. However the mass did not show any signs of regression. Hematology opinion was taken to rule out any lymphoma. As the patient had no systemic complaints or weight loss, nor any palpable lymph node with normal blood counts and peripheral blood smear USG Abdomen was suggested which also was normal. FNAC was adviced which yielded a dry tap. Finally we planned an excision biopsy of the mass under local anaesthesia. Peritomy was done nasally 1 mm beyond the limbus with relaxing cuts given at both ends. Sub conjunctival dissection was done around the mass. Medial rectus muscle was hooked to avoid any inadvertent damage to it. As the medial extent of the mass could not be traced it was excised approximately without damaging the MR muscle. Conjunctiva was closed. Post operatively the patient was treated with steroid antibiotic and lubricating eye drops. Histopathological examination of the mass showed monotonous lymphoid cells arranged in diffuse sheets with features of pleomorphism and mitosis suggestive of non hodgkins lymphoma which was further confirmed on immunohistochemical analysis. Bone marrow biopsy was performed that showed normocellular marrow. She was further referred to oncology department for radiotherapy where she was treated with 4500 c Gy over 30 days. She tolerated radiotherapy well. She was followed up every three months and she did not show any recurrence.

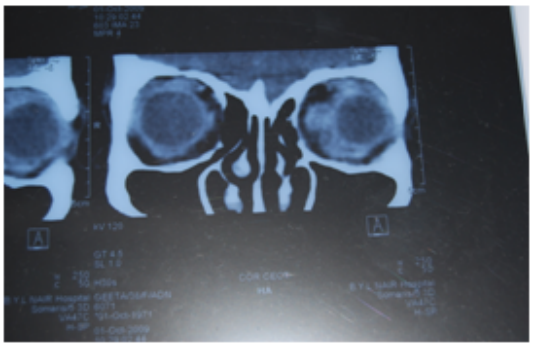

Figure 1:

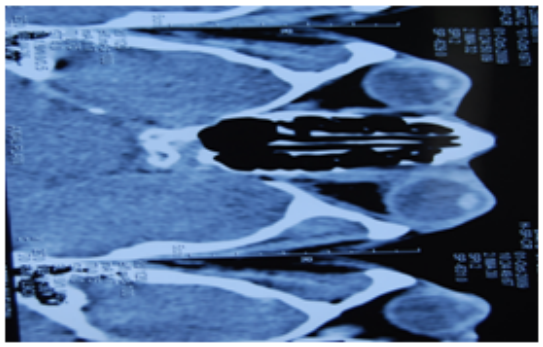

Figure 2:

DISCUSSION

Almost all the ocular tissues may be affected by lymphoma including the orbit, adnexial structures, anterior and posterior segment. Lymphomas of the eye and its adnexae are frequently of B lineage but may rarely be affected by non-B-cell non-Hodgkin's lymphoma. Lymphoproliferative disease of the orbit usually presents later in life and causes symptoms due to gradually increasing mass effect .No sex predilection was noted for ocular lymphomas in some studies.[4] However, in cases of intraocular lymphoma, women are known to be affected up to twice as often as men. Orbital lymphoma was found to have a female preponderance.[5] *Chlamydia psittaci* is associated with ocular adnexal mucosa-associated lymphoid tissue (MALT) lymphoma. Orbital lymphomas are predominantly of mucosa associated lymphoid tissue (MALT) histology (57%), but they also include other histological subtypes, such as follicular lymphomas (19%), diffuse large B-cell lymphomas (DLCL) and mantle cell lymphomas.[6] Recent molecular studies demonstrating viral DNA in the ocular lymphoma cells suggest a role for infectious agents in the pathogenesis of intraocular lymphoma. The ocular symptoms may be due to inflammation of the infiltrated tissues and this is largely responsible for the variability of ophthalmic manifestations ranging from typical uveitis to retinitis and vasculitis. Infiltration of the eyelids may result in periorbital swelling and ptosis or may also present as a localized eyelid mass or nodule. Infiltration of the conjunctiva may give rise to conjunctival swellings or chronic follicular conjunctivitis which may mimic the clinical picture of infectious or allergic conjunctivitis, and may result in diagnostic difficulties. Orbital and conjunctival lymphomas can be solitary or associated with systemic disease. Occasionally they can have

choroidal involvement as well; however, most of the time they tend to be extraocular. Orbital lymphomas may have a molded appearance conforming to the shape of the globe and orbital walls or it can cause proptosis due to mass lesion with compression of surrounding tissues. Extraocular lymphomas are generally low-grade lymphomas. Orbital and adnexal lymphoma is associated with systemic lymphoma in 30-35% of cases.[6] Hence, all patients with ocular lymphoma should have a complete workup to rule out systemic lymphoma. Differential diagnoses for orbital lymphoma include idiopathic inflammatory pseudotumor, orbital lymphoid hyperplasia, orbital sarcoidosis, Wegener granulomatosis, and chronic dacryoadenitis. Diagnosis is based on a good history, clinical examination and investigations. Careful slit lamp examination is required to identify the anterior segment lesions. Fundoscopy and fluorescein angiography can identify the posterior segment lesions. Our patient had a slow growing sub conjunctival mass while rest of the ocular examination was normal. She did not have any systemic complaints. While investigating MRI was suggestive of inflammatory mass for which the patient was treated with systemic steroid and Albendazole with no resolution of mass. Orbital and intraocular mass lesions of lymphoma can be identified by radiological examinations such as ultrasound scan, computed tomography scan and magnetic resonance imaging. Definitive diagnosis is based on biopsy of the affected tissue. High suspicion of lymphoma has to be kept in mind as differential diagnoses in such cases. Due to the diagnostic dilemma we planned excisional biopsy for our patient. Immunohistochemical staining with CD markers helps to classify lymphomas. Diagnosis of the systemic disease is established based on tissue samples that are studied by histological examination of a surgical biopsy from an accessible lymph node site. Lumbar puncture with cytology of the cerebrospinal fluid is important in central nervous system involvement. Bone marrow aspiration is used for staging systemic lymphomas. CT scans of the chest and abdomen are obtained to rule out retroperitoneal lymphoma. Bone scans may also be done. Treatment of ocular lymphoma is by radiation therapy and this may be combined with chemotherapy in the presence of central nervous system involvement. Prognosis for visual recovery is good if diagnosis is made early and therapy is started on time. Compared with primary intraocular lymphoma, metastatic systemic lymphomas have a better prognosis and is less likely to create a diagnostic dilemma.

CONCLUSION

Almost all ocular structures can be involved in lymphoma and it frequently masquerades as other more benign intraocular conditions. The lack of pathognomonic features, high clinical variability, the limited value of imaging techniques and histopathological measures may lead to delayed diagnosis and result in fatal outcome. However, early diagnosis and treatment may be achieved by high index of suspicion with recognition of its modes of presentation and multidisciplinary approach.

REFERENCES

1. Freeman C, Berg JW, Cutler SJ. Occurrence and prognosis of extranodal lymphomas. Cancer 1972; 29:252-60.
2. Fitzpatrick PJ, Macko S. Lymphoreticular tumors of the orbit. Int J Radiat Oncol Biol Physics 1984; 10:333-40.
3. Smitt MC and Donaldson SS. Radiotherapy is successful treatment for orbital lymphoma. Int J Radiat Oncol Biol Physics 1993; 26:59-66.
4. Moslehi R, Devesa SS, Schairer C, Fraumeni JF Jr. Rapidly increasing incidence of ocular non-hodgkin lymphoma. *J Natl Cancer Inst*. 2006 Jul 5. 98(13):936-9. [Medline].
5. Ahmed S, Shahid RK, Sison CP, Fuchs A, Mehrotra B. Orbital lymphomas: a clinicopathologic study of a rare disease. *Am J Med Sci*. 2006 Feb. 331(2):79-83. [Medline].
6. Coupland SE, Hummel M, Stien H. Ocular adnexal lymphomas: Five case presentation and a review of literature. Surv Ophthalmol 2002; 47:470-90.

Source of Support: None Declared
Conflict of Interest: None Declared