Orbital Non-Hodgkin Lymphoma (NHL) - A rare case report

*Polak Kumar Das,[1] Md Mofazzal Sharif,[2] Nazmul Alom,[3] Khaleda Parvin Rekha[4]

ARTICLE INFO

Article history:

Received: 12 April 2019

Accepted: 04 June 2019

Online:

www.nbmc.ac.bd

Keywords:

Lymphoproliferative tumuors, Orbital NHL, MRI

ABSTRACT

Lymphoproliferative tumuors of the ocular adnexa encompass a wide spectrum of lesions that range from reactive benign hyperplasia to malignant lymphoma which comprises 6-8% of orbital tumors and 10-15% of adnexal lesions. An 84 years old lady was presented in the Department of Ophthalmology, Khwaja Yunus Ali Medical College and Hospital, Enayetpur, Sirajganj with complaints of swelling and outwards bulging of left eye ball for last six months. On examination, salmon or flesh-pink colored lesion was found in superolateral aspect of eyeball. MRI revealed minimal enhancing T1WI isointense, T2WI hyperintense lesion in lateral aspect of eye ball, encasing lacrimal gland, superior, lateral recti muscles, causing proptosis and was diagnosed as lymphoma with differential diagnosis of malignancy of lacrimal gland and Sjogren syndrome. Finally, biopsy from the lesion confirmed left orbital non-Hodgkin lymphoma (NHL). Although this was a rare case for 84 years old female patient, the importance of this case report is to make aware the ophthalmologists as well as radiologists and oncologists to keep lymphoma in differential diagnosis, if such type of patient comes in day to day practice.

1. *Associate Professor (cc), Department of Ophthalmology, KYAMCH, Enayetpur, Sirajganj, Bangladesh*
2. *Associate Professor, Department of Radiology and Imaging, KYAMCH, Enayetpur, Sirajganj, Bangladesh*
3. *Assistant Professor, Department of Oncology, KYAMCH, Enayetpur, Sirajganj, Bangladesh*
4. *Assistant Professor, Department of Radiology and Imaging, KYAMCH, Enayetpur, Sirajganj, Bangladesh*

Corresponding author: ✉ dr.pkdas69@gmail.com

INTRODUCTION

Although uncommon, lymphoproliferative disease of the orbit usually presents later in life and causes symptoms due to gradually increasing mass effect. Proptosis and visible conjunctival mass are the common modes of presentation. It tends to be localized to the orbit at the time of diagnosis and responds well to local or systemic therapy.[1] Where as, Hodgkin lymphoma very rarely causes ocular disease, non-Hodgkin lymphoma (NHL) is the most common type of ocular lymphoma. Depending on the site of involvement, ocular lymphoma can be either intraocular or orbital and adnexal. Over the years, different systems have been used to classify lymphomas, including the Rappaport Classifica-

tion (used until the 1970s), the Working Formulation, the National Cancer Institute Working Formulation, and the Revised European-American Lymphoma Classification (REAL). In 2001, a modern comprehensive classification system was published under the auspices of the World Health Organization (WHO); this represents the first worldwide consensus document on the classification of lymphoma.[2] The lifetime risk of being diagnosed with non-Hodgkin lymphoma (NHL) is 2.08%. From 1975-2001, a rapid and steady increase occurred in the incidence of ocular NHL, with annual increases of 6.2% and 6.5% among white males and white females, respectively.[3] Intraocular lymphoma is rare, with fewer than 200 cases being reported. This type of lymphoma is estimated to represent 1% of NHLs, 1% of intracranial tumors, and less than 1% of intraocular tumors. However, over the past 20 years, a steady rise has occurred in the number of reported cases in both immunocompetent patients and immunocompromised patients.[4,5] Incidence of ocular lymphoma increases with advancing age. Intraocular lymphoma typically affects elderly patients; with reported series having mean ages in the seventh decade of life.[3] The median age at presentation for orbital and adnexal lymphoma is older than 60 years. In a study conducted in the United States, malignant lymphoma was the most common orbital tumor in the elderly age group, accounting for 24% of cases.[6] No sex predilection was noted for ocular lymphomas in some studies. 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. During 1992–2001, ocular NHL rates per 100,000 person-years for both sexes were highest among Asians/Pacific Islanders, lower in whites, and still lower in blacks. Orbital and ocular adnexal lymphoma has an insidious onset and can progress slowly for over a year before producing symptoms. Symptoms are usually secondary to pressure effects on surrounding structures. Clinical features include painless proptosis with or without motility disturbances, double vision, ptosis, and, rarely, decreased vision. The lesions can be unilateral or bilateral. Lymphomatous lesions can involve the preseptal portion of the eyelid.[3,4,6] Orbital lymphomas present with painless proptosis, the lesions being more common in the anterior superior orbit. The mass is usually rubbery to firm on palpation with no palpable bony destruction. The lacrimal gland, lacrimal sac, and extraocular muscles can also be similarly involved. Conjunctival lymphoma has a characteristic salmon-pink appearance. It may be an extension of orbital or intraocular lymphoma. The presence of cervical or preauricular lymphadenopathy, parotid gland swelling, or an abdominal mass can signify systemic disease. Hence, a thorough physical evaluation should be carried out in all patients with ocular lymphoma.[2,4,6] Ocular lymphoma typically affects elderly patients, with reported series having mean ages in the seventh decade of life.[3] B-scan ultrasonography can show the presence of an intraocular mass. In addition, retinal detachment may be seen. Both computed tomography (CT) and magnetic resonance imaging (MRI) have a low sensitivity for intraocular lymphoma and do not facilitate differentiating the diagnosis against uveitis or ocular melanoma. On CT scan (Figure 3, after Flanders et al.[7]) of the orbits, orbital lymphomas are seen as well-defined, lobulated or nodular, homogeneous masses of relatively high density and sharp margins. The lesions mold themselves to pre-existing structures without eroding the bone. The lesion is usually extraconal but can extend intraconally as well. Lacrimal gland disease may involve both orbital lobes and palpebral lobes. The lacrimal sac and extraocular muscles may also be involved. A streaky appearance may be seen, which represents

irregular infiltration of the microfascial structure of retrobulbar fat. Calcification is rarely seen. Heterogeneous lesions with bony destruction are indicative of high-grade lymphomas. Bilateral lesions are possible and can signify systemic disease. MRI of the orbits possesses good soft tissue definition; however, it lacks the ability to delineate bone destruction, which can be seen in high-grade lymphomas. MRI may miss conjunctival disease. Orbital lesions are usually hypointense or isointense on T1-weighted MRI and hyperintense on T2-weighted images. Gadolinium enhancement is seen on T1-weighted images. This is indicative of high cellularity. Fluorine-18 deoxyglucose PET (FDG-PET) can sometimes find systemic extranodal lymphomatous sites that are not detected with conventional imaging studies.[7,8] The prognosis for ocular lymphoma depends on the tumour's histologic type and stage, as well as on the treatment employed. In general, with modern treatment of patients with NHL, the overall survival rate at 5 years is approximately 60%.[2, 3, 5]

THE CASE

An 84 years old lady was presented in the Department of Ophthalmology, Khwaja Yunus Ali Medical College and Hospital, Enayetpur, Sirajganj with complaints of swelling and outwards bulging of left eye ball for last six months. She had no eye ache or visual disturbance. On examination salmon or flesh-pink colored lesion was found in superolateral aspect of eyeball, and photography was taken after informed written consent from the patient (Figure 1).

Visual acuity was normal for her age. Intraoccular pressure was within normal range for her age. Imaging study by MRI (Figure 2) showed minimal enhancing T1WI isointense, T2WI hyperintense lesion in lateral aspect of eye ball encasing lacrimal gland, superior, lateral recti muscles causing proptosis and diagnosed as lymphoma with differential diagnosis of malignancy of lacrimal gland and Sjogren syndrome. Finally, biopsy from the lesion confirmed left orbital non-Hodgkin lymphoma (NHL). Immunohistochemistry was advised, but patient could not do it due to financial constrain.

Figure 1: Salmon or flesh-pink colored lesion in superolateral aspect of eyeball on examination

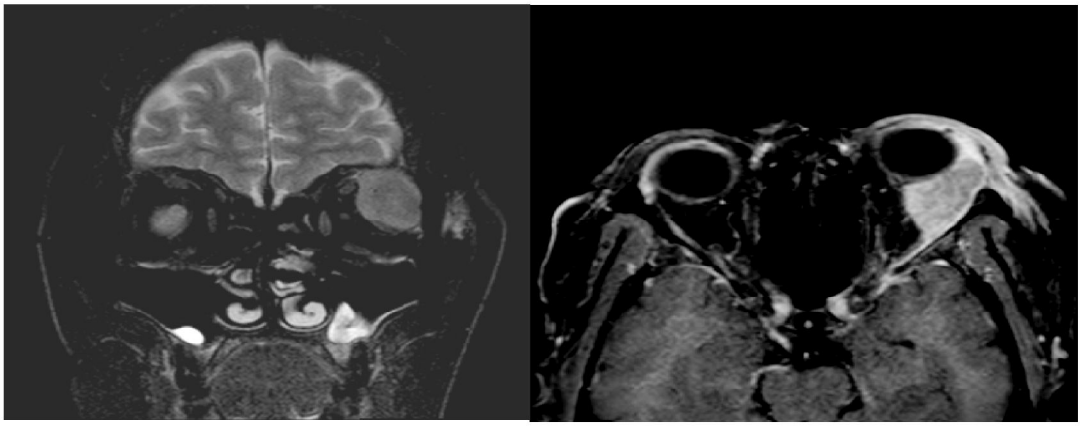

Figure 2: Coronal T2 weighted, axial post-contrast T1 weighted images revealed minimal enhancing T1WI isointense, T2WI hyperintense lesion in lateral aspect of eye ball encasing lacrimal gland, superior, lateral recti muscles causing proptosis

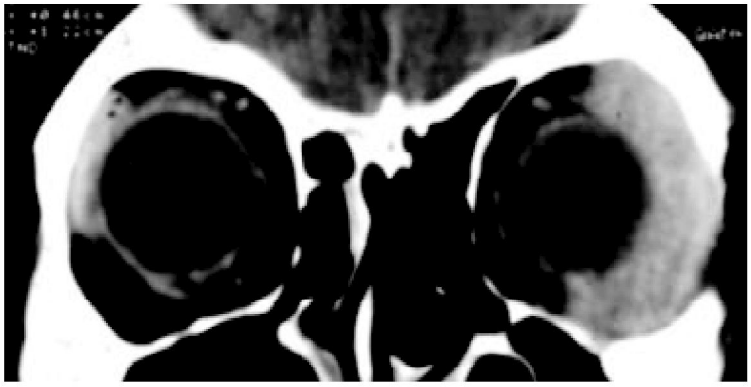

Figure 3: Coronal reformatted image showing homogeneous isodense enhancing sorbital mass molding around the lateral ocular surface (after Flanders et al.[7])

DISCUSSION

Owing to unspecific clinical symptoms, some diagnostic delay may occur in orbital lymphoma. If unspecific orbital symptoms are present, adequate imaging studies followed by early surgical biopsy would contribute to early diagnosis. Our present case of extra-ocular non-Hodgkin lymphoma (NHL) was 84 years old lady with no eye ache or visual disturbance and only gradual swelling of eye ball for last six months. Visual acuity and intraocular pressure was within normal range for her age. Examination revealed salmon or flesh-pink colored lesion in superolateral aspect of eyeball. Studies reported[4,5] increased incidence of ocular lymphoma with advancing age. No sex predilection was noted for ocular lymphomas in some studies. However, orbital lymphoma was found to have a female preponderanc.[4] This demographic finding is comparable with out current case. Researchers stated that many lesions of ocular NHL were asymptomatic but depending on the location of the mass, patients can complaint of exophthalmos, pain or diplopia, as well of conjunctival, eyelid, orbital or lacrimal gland mass. The differential diagnosis for orbital lymphoma include idiopathic inflammatory pseudotumor, orbital lymphoid hyperplasia, orbital sarcoidosis, Sjogren Syndrome, Wegener granulomatosis, and chronic dacryoadenitis.[9] MR imaging of the

patient showed minimal enhancing T1WI isointense, T2WI hyperintense lesion in lateral aspect of eye ball encasing lacrimal gland, superior, lateral recti muscles causing proptosis and diagnosed as lymphoma. Similar type of lesion on MRI went in favour of malignancy of lacrimal gland, Sjogren syndrome. In malignancy there might be bone destruction with more intra lesional in homogeneicity on T2WI. In Sjogren syndrome, MRI shows increased T2WI hyperintensity and homogeneous enhancement is observed in post contrast image. Radiotherapy is the most effective treatment for local disease either as the sole treatment for low-grade lymphoma or in combination with chemotherapy (CHOP or R-CHOP regimen) for intermediate- and high-grade lymphoma. Radiotherapy dose in range of 30-45 Gy administered in fractions are advised in treating the local orbital lymphomas.[10]

CONCLUSION

Success of treatment of ocular NHL depends on early diagnosis of disease. So, ophthalmologists as well as radiologists, oncologists should be aware of orbital lymphoma in older patient with orbital mass, specially female one.

Conflict of interest: None.

REFERENCES

1. Ahmed S, Shahid RK, Sison CP, Fuchs A, Mehrotra B. Orbital lymphomas: a clinicopathologic study of a rare disease. Am J Med Sci. 2006; 331(2): 79-83.
2. Russell-Jones R. World Health Organization classification of hematopoietic and lymphoid tissues: implications for dermatology. J Am Acad Dermatol. 2003; 48(1): 93-102.
3. Moslehi R, Devesa SS, Schairer C, Fraumeni JF Jr. Rapidly increasing incidence of ocular non-hodgkin lymphoma. J Natl Cancer Inst. 2006; 98(13): 936-939.
4. Levy-Clarke GA, Chan CC, Nussenblatt RB. Diagnosis and management of primary intraocular lymphoma. Hematol Oncol Clin North Am. 2005; 19(4): 739-749.
5. Schabet M. Epidemiology of primary CNS lymphoma. J Neuro Oncol. 1999; 43(3): 199-201.
6. Demirci H, Shields CL, Shields JA, Honavar SG, Mercado GJ, Tovilla JC. Orbital tumors in the older adult population. Ophthalmology. 2002; 109 (2): 243-248.
7. Flanders AE, Espinosa GA, Markiewicz DA, Howell DD. Orbital lymphoma. Role of CT and MRI. Radiol Clin North Am. 1987; 25(3): 601-613.
8. English JF, Sullivan TJ. The Role of FDG-PET in the Diagnosis and Staging of Ocular Adnexal Lymphoproliferative Disease. Orbit. 2015; 34 (5): 284-291.
9. Gao Y, Moonis G, Cunnane ME et-al. Lacrimal gland masses. AJR Am J Roentgenol. 2013; 201(3): 371-381.
10. Lenhard RE, Osteen RT, Gansler TS. Clinical oncology, Volume 1. USA: Wiley-Blackwell. 2000: p. 919.