

## Orbital Lymphoma: Clinical Profile, at a tertiary care hospital, Bangladesh

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### Abstract

**Purpose:** To explore the baseline clinical profile of orbital lymphoma, at a tertiary care in Bangladesh.

**Materials and Method:** As part of routine delivery services, we evaluated data from January 2010 to June, 2017 of the orbital lymphoma patients included in the study. Initially they are confirmed by CT scan and histopathological study reports. Their clinical characteristics were evaluated.

**Results:** A total of 123 patients were evaluated. Of them 116(94.30%) were Non-Hodgkin's Lymphomas and Male: Female ratio was 2.23:1 and age range was 8 to 82 years with median age 50 years. Among all cases 82(66.7%) got orbital involvement, conjunctival and eyelid involvement was 32.5% and 37.4% respectively. Bilateral cases were found in 17.1%. In this study Intermediate Grade of tumours was predominant 59(48.0%).

**Conclusion:** Orbital lymphoma is a disease of the elderly. It tends to be localized to the orbit at the time of diagnosis. Geographically it is common disease of Asia and in our country its occurrence is also not so uncommon.

### Introduction

Malignant Lymphomas are neoplastic transformations of cells that reside predominantly within lymphoid tissues. Lymphoid tumours are the most common primary orbital malignancy,<sup>(1,2)</sup> constituting approximately 10% of all orbital tumours, 40 to 60% of lymphoproliferative disease in the orbit.<sup>(3)</sup> The majority of orbital lymphomas are non-Hodgkin's type and are seen primarily in adults in the 50-70 years age group, only 1% of all Non-Hodgkin's Lymphomas. Most of the orbital lymphomas are extranodal marginal-zone B-cell lymphomas of mucosa associated lymphoid tissue type. Approximately 85 to 90% of orbital lymphomas are low-grade, diffuse proliferations of small, monoclonal B-cell lymphocytes. The remaining 10-15% have follicular or nodular characteristics. Orbital lymphomas are usually unilateral but may involve both orbits and demonstrate a predilection for the lacrimal gland. Patients with orbital lymphoma usually present with painless proptosis of insidious onset, downwards displacement of the globe, eyelid edema, a palpable non tender orbital mass and ptosis. Imaging studies usually confirm the presence of a mass, most commonly in the superior and anterior orbit but less commonly deep in the orbital apex. As there is no available information regarding orbital lymphoma in Bangladesh at present. So our purpose was to see the scenario of clinical presentation and diagnostic findings of the orbital lymphoma patients at a tertiary care hospital in a developing country like Bangladesh.

### Materials and Method

A retrospective observational study, comprising of 123 cases of diagnosed orbital lymphoma at Oculoplastics Department of National institute of ophthalmology and Hospital, Dhaka, Bangladesh, from 2010 to 2017 was done. All these patients' diagnosis was confirmed by histopathological study after an excision

biopsy. A pre structured questionnaire was used to collect data from available investigations like CT scans of Orbit, whole body bone scan and blood test, etc and from the register book. Prior that, Ethical clearance was obtained from Ethical Review committee of National Institute of Ophthalmology and Hospital to perform the study.

**Classification:** Histologic classification of lymphoma has evolved and improved as newer techniques are utilized to elucidate the antigenic expression, cytogenetic features, and molecular characteristics of lymphomas. In the past, the most commonly used system of classification for lymphoma was the National Cancer Institute's Working Formulation, which divided lymphomas into low, intermediate and high grades based on their histological characteristics and morphologic features.

**Investigation:** An imaging study of the orbit is critical in delineating the extent of orbital involvement. A characteristic diffuse to moderately well-defined orbital mass with molding to the globe, optic nerve and orbital bones strongly suggests the diagnosis of orbital lymphoma. On computed tomography, lymphomas appear homogenous in texture and isodense to muscle, showing mild enhancement with contrast.

### Results

Total 123 patient's information was obtained from registry book of hospital. Of all, 6 were pseudolymphoma, 1 HL and others were NHL. Among them Male: Female ratio was found 2.23:1 and age range was 8 to 82 years. Duration of disease from symptom appearance to consult with doctor was also varies high, from 20 days to 24 years. Among the all cases 17.1% patients got bilateral lymphoma, others are unilateral. Most of the cases 82(66.7%) got orbital involvement. Metastasis was present in 32.5% cases. Conjunctival and

eyelid involvement was 32.5% and 37.4% respectively. 32.5% patients got systematic metastasis.



Fig. 1-3: Clockwise- Unilateral Orbital Lymphoma, Bilateral Orbital Lymphoma, Salmon Patch



Fig. 3-5: From Left to right.-Pre-treatment state, CT scan Findings and Post treatment state of an Orbital Lymphoma Patients

Table 1: Patients profile of Orbital Lymphoma

Variables	Number	Percentage
Total patients	123	
Male: Female	85:38	2.23: 1
<b>Age</b>		
Median	50 years	
Range	8-82	
Duration of disease	20 days – 24 years	
<b>Type of Lymphoma</b>		
NHL	116	94.3
HL	1	0.8

Histopathology report was not available to 16 cases but their immunocytochemistry were done for them. Among the rest 107 cases 42(34.1%) low grade, 59(48.0%) intermediate grade and 6(4.9%) were high grade orbital lymphoma.

Table 2: Clinical and systematic involvement of orbital lymphoma

Variables	Number	Percentage
<b>Site</b>		
Left Eye	53	43.1
Right Eye	49	39.8
Bilateral	21	17.1
<b>Orbital Involvement</b>		
Yes	82	66.7
No	41	33.3
<b>Conjunctival Involvement</b>		
Yes	40	32.5
No	83	67.5
<b>Metastasis status</b>		
Present	40	32.5

Absent	83	67.5
<b>Eyelid Involvement</b>		
Yes	46	37.4
No	77	62.6

Table 3: Distribution of Orbital Lymphoma according to Grading

Variables	Number	Percentage
Low grade	42	34.1
Intermediate grade	59	48.0
High grade	6	4.9

**Discussion**

The incidence of orbital lymphoma has been reported to account for between 1 in 10% of NHL.<sup>(4)</sup> Although it has been known to present between 15 and 70 years, more clusters around seventh decades of life. In our study, median age of the patients was 50 years, ranging 8 to 82 years. In this study male patients were more 69% around, though from different studies it was said that historically female preponderance of this disease.<sup>(5)</sup> May be, due to social factors female patients neglect it, until and otherwise too advance.

Clinical presentation of this is nonspecific and depends mostly on its location. Usually optic nerve involvement or globe infiltration is very rare, vision remains normal most of the cases. In the current study 66.7% patients present with orbital involvement. Orbital presentation is usually present with a painless mass in the superolateral quadrant. It may lead to proptosis, ptosis, diplopia or difficulties in ocular movement.

Patients typically demonstrate with a pink or red ‘salmon patch’ of swollen conjunctiva or hyperemia at

conjunctiva. Here, 40(32.5%) patients present with this complaints.

Among all patients 37.4% got eyelid involvement. Usually swelling and prolapsed of the eyelid occurred on the affected eyelid.

From the histopathological study of the patients, only 4.9% were found rapidly aggressive high grade. In this study intermediate grading was found more predominant, 48.0%. About 13% patients' didn't do histopathological study, but for them immunocytochemistry reports were available as a confirmation of orbital lymphoma.

### Conclusion

Orbital lymphoma is a disease of the elderly. It tends to be localized to the orbit at the time of diagnosis. Geographically it is common disease of Asia and in our country its occurrence is also not uncommon.

### References

1. Jakobiec FA, Font RL. Orbit: lymphoid tumors. In: Spencer WH, Font RL, Green WR, et al, eds. *Ophthalmic Pathology: An Atlas and Textbook*. Vol. 3. 3<sup>rd</sup> ed. Philadelphia: WB Saunders; 1986:2663-2711.
2. Ellis JH, Banks PM, Campbell RJ, Lisegang TJ. Lymphoid tumors of the ocular adnexa: clinical correlation with the working formulation classification and immunoperoxidase staining of paraffin sections. *Ophthalmology* 1985;92:1311-1324.
3. Bairey O, Kremer I, Rokowsky E, et al. Orbital and adnexal involvement in systemic non-Hodgkin's lymphoma. *Cancer* 1994;73:2395-2399.
4. Eckardt A M et al. Orbital Lymphoma: diagnostic approach and treatment outcome. *World j Surg Oncol*.2013;11:73.
5. Ahmed S et al. Orbital Lymphomas: a clinicopathologic study of a rare disease. *Am J Med Sci*.2006;331(2):79-83.