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Clinical Presentation and outcome in Cases of Orbit Lymphoma: A Prospective Interventional Study

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Abstract

Context: The origin of orbital lymphoma can be variable. It may arise from conjunctiva or likely to arise from eyelid or may happen to arise from orbit or lacrimar glands. Diagnosis is most likely to be delayed as the clinical features are not very specific.

Aims: To study clinical presentation and outcome in orbital lymphoma cases.

Settings and Design: It was a Prospective Interventional Study carried out at Sarojini Devi Eye, Hospital, Hyderabad for a period of two years.

Methods and Material: A total of 26 cases were included in the study. Only lymphoma orbit cases were included. Appropriate intervention was done depending upon the need i.e. stage, histological type, patient age and any other patient characteristics. Outcome was studied specific to the intervention carried out.

Statistical Analysis: The data was entered in Microsoft Excel Worksheet and analyzed using proportions.

Results: Male predominance was observed in present study i.e. males were 69% and females were 31%. Majority of the study participants had proptosis (23.8%) as main clinical presentation. About 19.23% had orbital & lid mass. Small Lymphocytic lymphomas were observed in 50% of the study participant, Diffuse mix small & large cell was seen in 30.7%, Diffuse large cell in 11.5% and Burkitt's lymphoma in 7.6% of cases.

Conclusion: Small Lymphocytic lymphoma was the most common orbit lymphoma found in the present study almost in half of the cases. Next most common histologic type was Diffuse mix small & large cell in about one third of the cases. Incisional biopsy & radiotherapy was the most commonly adopted intervention by us.

Keywords: Orbit; Lymphoma; Proptosis; Lid.

Introduction

Malignant Lymphomas are neoplastic transformations of cells that reside predominantly within lymphoid tissues. Lymphoid tumours are the most common primary orbital malignancy [1,2], constituting approximately 10% of all orbital tumours, 40 to 60% of lympho-proliferative disease in the orbit [3]. The majority of orbital lymphomas are Non-Hodgkin's type and are seen primarily in adults in the 50-70 years age group.

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, downward displacement of the globe, eyelid edema, a palpable non tender orbital mass and ptosis. The non-Hodgkin's lymphomas (NHL) are a group of neoplasms characterized by proliferation of malignant lymphocytes. Patients with NHL have a wide variety of presenting signs and symptoms, depending largely on the site of involvement and aggressiveness of the disease [3]. Therefore the present study was conducted with the objectives to study incidence of orbital Lymphoma, and to study clinical presentation in orbital lymphoma cases.

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The origin of orbital lymphoma can be variable. It may arise from conjunctiva or likely to arise from eyelid or may happen to arise from orbit or lacrimal glands. Diagnosis is most likely to be delayed as the clinical features are not very specific. The site of origin may be at one place and the appearance of signs and symptoms may be a bit different. As for example almost one fourth of the cases occur in conjunctiva but the presentation may be with red patches [4].

Diagnosis of orbital lymphoma and its types is biopsy guided which gives perfect clue of staging also. But for staging, CT scan or if required MRI scan should be done. This imaging will help ophthalmologist to know the extent of invasion of tumor not only locally but also systemic involvement. Other tumors of orbit which are malignant or inflammatory as well as "thyroid associated orbit disease" should be ruled out to reach a final diagnosis [5].

The outcome or prognosis of lymphoma of the orbit is dependent on many factors like age of the patient, histological type of the tumor, whether the tumor was local or invasion took place and the stage of the tumor. Naturally younger the patient, histology of low grade, local tumor and early diagnosis favor the prognosis [6].

Studies of this kind i.e. clinical profile of orbit lymphoma are few. Such studies throw light on the clinical presentation, histological types, type of intervention and related outcome. This knowledge is useful.

Materials & Methods

Study Design

Prospective Interventional Study.

Study Area

Sarojini Devi Eye Hospital, Hyderabad.

Study Duration

The study was done for period of 2 years.

Ethical Consideration

The study was started after taking permission from institutional Ethical Committee

Sample Size

A total of 26 cases were included in the study.

Study Participants

- *Inclusion Criteria:* All histopathologically confirmed cases of orbital lymphoma irrespective of age and sex included.
- *Exclusion Criteria:*
  - In cases where histopathology is inconclusive even after immuno-histochemistry
  - Cases who did not report for follow up.

Methodology

A pre-designed, pre-structured questionnaire was used. The study participants were explained about the purpose of the study. A written consent was taken before the start of the study. A detailed history was taken. A detailed clinical examination was done. Detailed ocular examination such as slit lamp examination of anterior segment, cover tests, ocular motility was done. Hertel's exophthalmometry and fundus evaluation was done. Routine examination like CBP, ESR were done. CT scan brain and orbit was done. Ultrasound abdomen was done. Bone marrow aspiration with peripheral smear examination was done. Incisional/Excisional biopsy was done and sent for histopathological examination. Immunohistochemistry was done where ever needed. The patients were followed till 12 weeks.

Statistical Analysis

The data was entered in Microsoft excel sheet and analysis was done using proportions.

Results

Table 1 shows sex distribution of study participants. Male predominance was observed in present study. Males were 69% and females were 31%.

Table 2 shows distribution of study participants with side of eye involvement. Nearly about 54% of the study participants had lymphoma in right eye and 46 % had in left eye. No bilateral involvement observed.

Table 3 shows distribution of study participants with clinical presentation. Majority of the study participants had proptosis (23.8%) as main clinical presentation. About 19.23% had orbital & lid mass followed by orbital mass (11.54%), lid mass (11.54%), orbital & lid mass (11.54%) respectively

Table 4 shows distribution of study participants with site involved. Orbit-retro bulbar area and lacrimal gland was involved in 23.08% of the study participants which was followed by orbit &

conjunctiva in 15.38%, orbit & lid (15.38%), lid (11.54%) & conjunctiva (11.54%).

Table 5 shows distribution of study participants with histological type. Small Lymphocytic lymphomas were observed in 50% of the study participant, Diffuse mix small & large cell in 30.7%, Diffuse large cell in 11.5% and Burkitt's lymphoma in 7.6%.

Table 6 shows distribution of study participants with type of treatment incisional. Biopsy & radiotherapy was done in majority of the study participant's (73.07%), followed by Incisional biopsy & chemotherapy in 19.23%. Excision was done in 7.69% of study participant's.

Table 7 shows comparison between pre operative and post operative mean values of axial displacement of proptosis. It was found that their mean values of axial displacement of proptosis came to normalcy post operatively as compared to the pre operative

Table 1: Sex wise distribution of study participants

Sex	Frequency	Percentage
Male	18	69
Female	08	31
Total	26	100

Table 2: Distribution of study participants with side of eye involvement

Eye	Frequency	Percentage
Right	14	54
Left	12	46
Total	26	100

Table 3: Distribution of study participants with clinical presentation

Clinical Presentation	Frequency	Percentage
Proptosis	6	23.8
Orbital mass	3	11.54
Orbital & lid mass	5	19.23
Orbital & conjunctival mass	3	11.54
Lid mass	3	11.54
Conjunctival mass	3	11.54
Lacrimal mass	3	11.54
Total	26	100

Table 4: Distribution of study participants with site involved

Site involved	Frequency	Percentage
Orbit-Retrobulbar	6	23.08
Lacrimal gland	6	23.08
Orbit & conjunctiva	4	15.38
Orbit & lid	4	15.38
Lid	3	11.54
Conjunctiva	3	11.54
Total	26	100

Table 5: Distribution of study participants with histological type

Histological Type	Frequency	Percentage
Low Grade Lymphoma		
• Small Lymphocytic lymphomas	13	50
Intermediate Grade Lymphoma		
• Diffuse mix small & large cell	08	30.7
High Grade Lymphoma		
• Diffuse large cell	03	11.5
• Burkitt's lymphoma	02	7.6
Total	26	100

Table 6: Distribution of study participants with type of treatment

Type of treatment	Frequency	Percentage
Excision	02	7.69
Incisional biopsy & radiotherapy	19	73.07
Incisional biopsy & chemotherapy	05	19.23
Total	26	100

Table 7: Comparison between pre operative and post operative mean values of axial displacement of proptosis

mean values of axial displacement of proptosis	Pre operative Mean±SD	Post operative at 4 weeks Mean±SD	T value	P value
	17.28±1.96	15.5±1.52	3.6593	0.0006

values. The mean decreased from 17.28 to 15.5 and this reduction was found to be statistically significant. At 12 weeks all patients had normal values.

Discussion

Male predominance was observed in present study. Males were 69% and females were 31%. Nearly about 54% of the study participants had lymphoma in right eye and 46% had in left eye. No bilateral involvement observed. Majority of the study participants had proptosis (23.8%) as main clinical presentation. About 19.23% had orbital & lid mass followed by orbital mass (11.54%), lid mass (11.54%), orbital & lid mass (11.54%) respectively Orbit-retro bulbar area and lacrimal gland was involved in 23.08% of the study participants which was followed by orbit & conjunctiva in 15.38%, orbit & lid (15.38%), lid (11.54%) & conjunctiva (11.54%). Small Lymphocytic lymphomas were observed in 50% of the study participant, Diffuse mix small & large cell in 30.7%, Diffuse large cell in 11.5% and Burkitt's lymphoma in 7.6%. Biopsy & radiotherapy was done in majority of the study participant's (73.07%), followed by Incisional biopsy & chemotherapy in 19.23%. Excision was done in 7.69% of study participant's.

In Present study all 26 cases were Non-Hodkins Lymphoma's, among them 88.46% were B-cell lymphomas and 11.53% were T-cell Lymphomas. The present study findings were similar with Das D et al [7] study where 89% of the study participants were having B-cell lymphoma.

Male predominance was observed in present study. Males were 69% and females 31%. The present study findings were similar with other studies where males were more in number such as a study done by Dipankar Das et al [7] where they noted that nearly about 63% were males. In another study conducted by Narayan G et al [8] majority i.e. 68% were males.

Majority of the study participants had Proptosis (23.8%) as main clinical presentation. About 19.23% had orbital & lid mass followed by orbital mass (11.54%), lid mass (11.54%), orbital & lid mass (11.54%) respectively. In other studies too proptosis was the main presenting symptom. In a study done by Narayan G et al [8] it was noted that 100% of the study participants had proptosis as main clinical presentation. Present study findings were similar to Rey Porca C et al [9] study where 22% of the study participants had proptosis. Upper lid mass was seen in 26.3% of the study participants and conjunctival mass was seen in 15.78% in a study done by Das D et al [7].

In present study nearly about 54% of the study participants had lymphoma in right eye and 46% had in left eye. No bilateral involvement observed. In the study done by Narayan G et al [8] it had been observed that 52% of the study participants had mass in right and 8% had mass in both the eyes. In the study conducted by Tranfa F et al [10] it was found that 48% had mass in right eye. In the study carried out by Rosado MF et al [11] 90% had mass in right eye.

Orbit-retro bulbar area and lacrimal gland was involved in 23.08% of the study participants which was followed by orbit & conjunctiva in 15.38%, orbit & lid (15.38%), lid (11.54%) & conjunctiva (11.54%). In a study done by Rosado MF [11] 53% were having mass in orbit, 26% in conjunctiva and 19% in lacrimal apparatus. In a study done by Tranfa F et al [10] 64% of the study participants had mass in orbit.

Small Lymphocytic lymphomas were observed in 50% of the study participant, Diffuse mix small & large cell in 30.7%, Diffuse large cell in 11.5% and Burkitt's lymphoma in 7.6%. In the study by Tranfa F et al [10] study 50% of the lymphomas were proved as small lymphocytic lymphomas which is similar to present study. In a study by Rosado MF et al [11] study 3.2% were having small lymphocytic lymphomas.

It was found that their mean values of axial displacement of proptosis came to normalcy post operatively as compared to the pre operative values. The mean decreased from 17.28 to 15.5 and this reduction was found to be statistically significant. At 12 weeks all patients had normal values.

Conclusion

Small Lymphocytic lymphoma was the most common orbit lymphoma found in the present study almost in half of the cases. Next most common histologic type was Diffuse mix small & large cell in about one third of the cases. Incisional biopsy & radiotherapy was the most commonly adopted intervention by us.

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