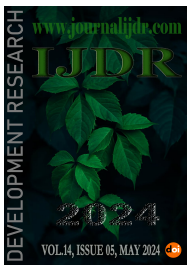
ISSN: 2230-9926

Available online at http://www.journalijdr.com

IJDR

International Journal of Development Research
Vol. 01, Issue, 05, pp. 65576-65578, May, 2024
https://doi.org/10.37118/ijdr.26949.05.2024



RESEARCH ARTICLE **OPEN ACCESS**

CLINICAL AND EPIDEMIOLOGICAL FEATURES OF LYMPHOPROLIFERATIVELESIONS IN THE ORBIT AND OCULAR ADNEXA AT ASOCIACIÓN PARA EVITAR LA CEGUERA EN MÉXICO

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ARTICLE INFO

Article History:

Received 02nd February, 2024
Received in revised form
20th March, 2024
Accepted 26th April, 2024
Published online 30th May, 2024

KeyWords:

Lymphoma, Lymphoproliferative lesions.

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ABSTRACT

Purpose: To evaluate all lymphoproliferative lesions of the orbital and adnexa diagnosed at our hospital during 11 years. Methods: Observational, Retrospective descriptive análisis. For statistical analysis we used SPSS package. Results: 74 cases with lymphoproliferative lesions of the orbital and adnexa were identified. 45 Female and 29 Male, with median age of 60 years. Most common presenting symptoms were increase of volumen and proptosis. Pathologic and immunohistochemistry diagnosis coincided in 71.6 % of cases. All specimens represented B cell non-Hodgkin's lymphomas. The orbit and conjunctiva were the most commonly affected sites. We found 16.2 % of bilateral involvement. The majority of patients were send to a cancer treatment center for follow up. A variety of therapeutic regimens was administered, the main form of treatment being radiotherapy. Conclusions: The majority of our results correlate with literature. In our experience orbital and adnexal region lymphoproliferative lesions were present in elderly women. We had an increase and posterior decrease in lymphoma incidence in general.

Citation: ***Linda Guakil Sakruka and Andrea Portilla Demichelis. 2024.*** "Clinical and Epidemiological features of lymphoproliferativelesions in the orbit and ocular adnexa at Asociación Para Evitar la Ceguera en México". *International Journal of Development Research*, 14, (05), 65576-65578.

INTRODUCTION

Lymphatic neoplasms are tumors composed of cells of lymphoid origin and can be found in lymph nodes or extranodal tissues. This lesions are derived from lymphocytes and its precursor cells, expressing B or T cell phenotype (9). Lymphomas are divided in 2 large groups: Hodgkin and non Hodgkin lymphoma and then subdivided in several groups (5). Speaking of lymphoma of the ocular adnexa, it develop as primary or secondary tumors in the conjunctiva, eyelid, orbit, lacrimal gland, and lacrimal sac. (2,3). Ocular adnexal lymphomas develop as primary or secondary tumors in the conjunctiva, lid, orbit, lacrimal gland, or lacrimal sac (2,3). Most patients presents with localized disease between 6th and 7th decade (3,4,5). 1 – 2 % of all lymphoma and aproximmately 8% of all extranodal lymphoma arise in ocular adnexa (1). Age, gender and anatomic location doesn't seem to have any prognostic impact (4,10). 60% are secondary. There are predisposing factors such as genetics, infectious diseases and autoimmune among others (4). Presentation tends to be insidious, from months to years, with painless proptosis, sometimes diplopia and low vision (1,9). Aproximately 40 to 50 % presents in the orbit, 20-30% in the lacrimal gland, and 10-20% in the eyelids and conjunctiva (2). Diagnosis starts with clinical data and image studies, even tough the gold estándar is the histopathological and immunohistochemical study (2,4,8). Management depends on extension of disease using radiotherapy and chemotherapy among other treatments (1). The aim of our study is to evaluate epidemiologic and clinic features in patients with limphoproliferative lesions in the orbital and adnexal region at tertiary treatment center Asociación Para Evitar la Ceguera en Mexico.

MATERIAL AND METHODS

Case series of patients with histopathological diagnosis of limphoprolipherative lesions of the orbita and ocular adnexa in the departments of pathology and oculoplastics at the Asociación Para Evitar la Ceguera en México. The data obtained were age of presentation, gender, onset symptoms, presence or absence of pain, evolution, histopathological and immune his to chemical diagnosis, location, bilateral involvement and eye affected. Incisional or excisional biopsy was performed in all patients, with several approaches depending on the location of the lesion. It is a retrospective, observational study, with descriptive analysis, using SPSS statistical package.

RESULTS

There were 74 patientfiles included with lymphoproliferative lesions of the orbita and adnexa, diagnosed by incisional or excisional biopsy

at our institution. 45 women (60.8%) and 29 men (39.2%) with a presentation age of 60 years old, showed in Table 1. Onset symptom presented more frecuently were mass/bulking at the affected area in 64.9% of cases, followed by proptosis in 10.8 % of cases, showed in Table 2.

Table 1. Gender and age of Presentation

APEC	N.	%
Male	29	39.2
Female	45	60.8

Table 2. Onset symptoms

Symptom	
Increase in volume	48 (64.9%)
Proptosis	8 (10.8%)
Tearing	5 (6.8%)
Pain	4 (5.4%)
Asymptomatic	2 (2.7%)
Other	7 (9.4%)

Only 17% of patients presented with pain. Time of evolution of onset symptoms was 0 to 10 years, due to one patient that referred an evolution of symptoms of 10 years. We found a coincidence of 71.6% with the pathology report and immunohistochemistry report, wich shows the importance of including an immunohistochemistry study in this type of lesions. In most cases, the diagnosis was extranodal lymphoma in the marginal zone (52.7%). Every sample represented Non Hodgkin type B Lymphoma: Extranodal Marginal Zone Lymphoma (EMZL) and MALT (n=39), Diffuse Large B Cell Lymphoma (n=10), Mantle Cell Lymphoma (n=5), Follicular Lymphoma (n=4), others (n=5). We found 11 cases with lymphoid hyperplasia. As for Location, most frequently affected sites were the orbit (50 %) and the conjunctiva (35.1%), followed by the eyelid (9.5%) and lacrimal sac (5.4%). The most frequently found was lymphoma type MALT located in the conjunctiva, as shows in Table 3.

Tabla 3. Clinical Correlation

	Total	Site			
Diagnosis		Orbit	Conjunctiva	Eyelid	Lacrimal sac
Malt	39	15	19	3	2
Ldcgb	10	9	0	1	0
Lf	4	1	1	2	0
Mantle	5	2	1	0	2
Hl	11	5	5	1	0
Other	5	5	0	0	0

-LDCGB: B Cell Lymphoma
-LF: Folicular Lymphoma
-HL:Lymphoid Hyperplasia

There was a bilateral involvement in 16.2% of cases. The right and Left eye were envolved in 41.9% and 41.9 % of cases respectively. We observed an increased incidence from 2004 to 2008, a posterior decrease and a new increase in 2012 and 2013. Most patients (76.8%) were referred to a Cancer Center for followup. They use diverse types of treatment like radiotherapy and chemotherapy at the cancer center.

DISCUSSION

In this study we have evaluated the characteristics of 74 patients with lymphoproliferative lesions of the orbita and adnexa. Lymphoma was found more frequently in elderly women, which agrees with previous reports (Table 4). (1) Most frequently onset symptoms were mass/bulking, followed by proptosis. We found an absence of pain in most cases. This finding also agree with previous Studies. Table 5.1 y 5.2 (7). The orbit and conjunctiva were the most Commonly affected sites which also agrees with previous Studies. Table 6. (1,2,3)

Table 4.1 and 4.2 Gender and age comparatives

	N.	%
Male	29	39.2
Female	45	60.8
Luigi De Cicco (R.Oncology)	N.	%
Male	20	42.6
Female	27	57.4

Table 5.1 and 5.2 Onset symptom comparative

SYMPTOM	N
Volume increase	48 (64.9%)
Proptosis	8 (10.8%)
Tearing	5 (6.8%)
Pain	4 (5.4%)
Asymptomatic	2 (2.7%)
Other	7 (9.4%)

Volume increase	52 (33%)
Mass	41 (26%)
Diplopia	21 (13%)
Proptosis	20 (13%)
Pain	12 (8%)
Hyperemia	10 (6%)
Asymptomatic	4 (3%)

Table 6.1 and 6.3 Location comparison

Orbit	37	50
Lacrimal sac	4	5.4
Conjunctiva	26	35.1
Eyelid	7	9.5

Orbit	41	83.7
Lacrimal sac	1	2
Conjunctiva	3	6.1
Eyelid	4	8.2

Orbit	44	44.4
Conjunctiva	29	29.2
Eyelid	21	21.2
Caruncle	5	5

MALT	39	52.7
Diffuse B cell	10	13.5
Mantle cell	5	6.8
Lymphoid hyperplasia	11	14.9
Follicular	4	5.4
Other	5	6.8

En la mayoría de los casos, se trata de linfomas de células B de bajo grado, lo cual coincide con la literatura, sin embargo, se encontraron algunos casos de linfomas atípicos (células del manto, folicular, células grandes B), lo que difiere un poco con lo descrito en otros artículos, lo cual podemos observar en la Tabla 7.1, 7.2 Y 7.3. (1,2,3)

Table 7.1, 7.2, 7.2 Final diagnosis comparative

Luigi Di Cicco (R. ONCOLOGY)		
MALT	38	80.9
DIffuse B cell	5	10.6
Mantle cell	3	6.4
Burkitt lymphoma	1	2.1

Coupland	N.	%
MALT	63	63.6
Follicular	10	10.1
Diffuse B cell	9	9
Plastocytoma	6	6
Lymphoplasmocitic lymphoma	5	5
Mantle cell	2	2
Leukemia	1	1
T cell	3	3

In this study, we demonstrated the great importance of inmunohistochemistry to support the histopathological diagnosis and therefore to have more adequate treatment. We found an initial increase, then a posterior decrease and later on another increase in the incidence of this disease through time. Once we have a pathological diagnosis, we refer our patients to an oncologic treatment center. This makes our follow up difficult, because a lot of our patients do no return for ophthalmic consultation.

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