Original Article

# Extranodal marginal zone B cell lymphoma of mucosa-associated lymphoid tissue type of the ocular adnexa: Retrospective single institution review of 95 patients

*Su-Ho Lim, Minkyu Kang[1], Junhyuk Son*

**Context:** There are few reports on the management of extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue type (MALT), which are based on the experience of a single institution, as opposed to large multicenter randomized trials. **Aim:** The aim of this study was to report on the clinical experience of a single institution. **Materials and Methods:** A retrospective review was undertaken of records of 95 patients with MALT lymphoma of the ocular adnexa. Histologic diagnosis of MALT lymphoma was made according to established criteria, and clinical staging was carried out to determine treatment modalities. All patients were treated by external beam irradiation (30.6–45.0 Gy) after biopsy. Additional chemotherapy was performed in accordance with the clinical stage of the disease. All the patients were treated by the same hemato-oncologist and radio-oncologist. **Results:** Almost all patients showed complete response, except for four patients who showed partial response. In two of 95 patients, contralateral eye showed recurrence, and they were salvaged by additional radiotherapy. The 3-year overall survival and event-free survival rates were 100 and 97%, respectively, by Kaplan–Meier survival analysis. Moreover, there were no serious radiation-associated complications. **Conclusions:** Radiotherapy alone can be an important treatment modality for the local control and survival in patients with localized MALT lymphoma of ocular adnexa. Systemic chemotherapy should be considered in patients with advanced stage disease.

**Access this article online**

**Website:** www.ijo.in

**DOI:** 10.4103/0301-4738.81993

**Quick Response Code:**

**Key words:** Lymphoma, mucosa-associated lymphoid tissue, radiotherapy

Orbital lymphoma is rare and comprises about 10% of all orbital neoplasms. Among the orbital lymphomas, non-Hodgkin's lymphoma (NHL) of the ocular adnexa constitutes less than 1% of all cases of nodal and extranodal NHL[1] and accounts for about 8% of all extranodal NHL.[2] The majority of NHLs of the ocular adnexa are extranodal marginal zone B-cell lymphomas of mucosa-associated lymphoid tissue (also known as MALT lymphoma).[3-6] As a disease entity, it was first described in 1983 by Isaacson and Wright,[7] and it is now well established in the revised European-American lymphoma (REAL) classification of lymphoid neoplasms[8] and the more recently published World Health Organization classification system.[9]

MALT lymphoma of the ocular adnexa is characterized by an indolent natural history and by slightly different clinical and biological characteristics compared to MALT lymphoma in the gastrointestinal tract. For example, the most impressive proof of chronic antigenic stimulation was the identification of a causative role of *Helicobacter pylori* (HP) in the development of gastric MALT lymphoma.[10] Regression of primary low grade B-cell gastric lymphoma of MALT after eradication of HP was also known. Extragastric lymphoma did not regress after eradication of HP. Also, ocular adnexa lymphomas were associated with *Chlamydia psittaci* infection in some countries.[11]

Excellent local control and long-term survival can be achieved using radiotherapy alone.[5-6,12] However, several studies have reported a rather high incidence of local or distant failure after radiotherapy alone.[7,13] Efficacy of chemotherapy as a first-line treatment in ocular adnexal extranodal marginal zone B-cell lymphoma was excellent. But recurrence or disease progression was observed in up to 33.3% at a median follow-up of 58 months.[14]

Besides radiotherapy and chemotherapy, a variety of new treatment options have emerged in the management, especially immunotherapy including interferon alpha, monoclonal antibody therapy and antibiotic therapy against *C. psittaci* which has been associated with the pathogenesis of ocular adnexal lymphomas in some parts of the world.[15]

Because of the rarity of this tumor, most of the previous literature consists of single institutional retrospective reviews on ocular lymphomas consisting of a variety of histologic subtypes. Thus, in this report, we present the experience of a single institution (clinical features, treatment outcomes, and complications) with MALT lymphoma of the ocular adnexa.

## Materials and Methods

A retrospective review of records from the Department of Ophthalmology at a tertiary care teaching hospital, from October 1997 to January 2009, was performed and all cases of MALT lymphoma that presented primarily in the ocular adnexa were included in the study.

Histologic diagnosis of MALT lymphoma was performed according to the criteria outlined by Isaacson.[16] Immunologic phenotyping on paraffin sections was done to demonstrate κ and λ light chain restriction and the phenotype $CD20^{+}CD5^{-}CD10^{-}$

Departments of Ophthalmology and [1]Radiation Oncology, Yeungnam University College of Medicine, Daegu, Korea

Correspondence to: Dr. Junhyuk Son, #317-1 Daemyung-dong, Nam-gu, Daegu, 705-717, Korea. E-mail: sjh@med.yu.ac.kr

Manuscript received: 05.01.10; Revision accepted: 22.12.10

cyclinD1$^-$, which, in the context of microscopic appearance, is consistent with MALT lymphoma. Clinical staging was undertaken according to the Ann Arbor classification[17] and on the basis of the following studies: meticulous history and physical examination, complete blood count, biochemistry profile ($\beta_2$-microglobulin and lactate dehydrogenase), computed tomography (CT) scans (orbit, chest, abdomen, and pelvis) and bone marrow biopsy. Simultaneous bilateral orbital involvement without any other lesions outside the orbits was classified as stage $I_{EE}$ according to the criteria of other investigators.[18-20]

All patients were treated by external beam irradiation with lens shielding after biopsy of the lesion in our hospital. Treatments were given 5 days/week using a fraction size of 180 cGy. The total dose given varied between 30.6 and 45.0 Gy. Instead of curative surgical treatment, maximal excision was performed in some patients for diagnosis and supportive treatment before referral to the radiation oncologist and hemato-oncologist.

Additional chemotherapy was performed according to patient's clinical stage. All patients initially received chemotherapy with cyclophosphamide, doxorubicin, vincristine, and prednisone (CHOP). Two patients with a history of cardiovascular disease were deemed to be at a greater risk for doxorubicin cardiomyopathy and they were omitted from the CHOP regimen. The two patients were then referred and put on cyclophosphamide, vincristine, and prednisone (COP) regimen.

All patients were followed up regularly after treatment. Serial follow-up visits were scheduled at 1, 3, 6, 12 months and every 6 months thereafter. Initial response was evaluated 4–6 weeks after the last cycle of chemotherapy. For patients who underwent radiotherapy, a new staging was performed one month after completion of treatment. Complete response (CR) was defined as the complete disappearance of all measurable and evaluable disease. Partial response (PR) was defined as diminution of >50% of all the initial masses. No response was defined when the disease remained virtually unchanged in the size and number of lesions. Generally, a less than 50% decrease or a slight increase in size is described as no response. Recurrence was defined as an increase in size or number of lesions on treatment during the follow-up period.

## Results

There were 95 patients with MALT lymphoma of the ocular adnexa, who were identified and included in this study. Table 1 lists the characteristics of the patients. The table shows the following: male dominance, median age 44 years, symptom to presentation time (10.5 months). Tumors were located most commonly in the conjunctiva (65.2%) and were unilateral (82/95).

The patients had varying symptoms and signs. The most common presenting symptoms and signs were palpable mass in 49 (51.6%), discomfort in 18 (18.9%), conjunctival injection in 9 (9.5%), proptosis in 7 (7.4%), chemosis in 4 (4.2%), peri-orbital swelling in 4 (4.2%), and ptosis in 4 (4.2%).

In histopathologic findings, extranodal marginal zone B-cell lymphoma of MALT type (also known as MALT lymphoma) is characterized by poorly defined follicular appearing areas that are composed of monocytoid B cells that feature enlarged nuclei. An important diagnostic feature of MALT lymphoma is lymphoepithelial lesions (LEL), defined by the infiltration and distortion of epithelial structures by aggregates of neoplastic lymphoid cells. Immunochemically, the lymphocytes in LEL were CD20 (corresponding B-cell marker) positive and CD3-negative (corresponding for T-cell marker). So, the presence of a monotonous population of CD20-positive intraepithelial lymphocytes supports a diagnosis of MALT lymphoma [Fig. 1].

**Table 1: Characteristics and tumor characteristics of 95 patients**

| Characteristics | No. of patients |
|---|---|
| Males/females | 57/38 |
| Median age (range) | 44 years (21–80) |
| Median duration of symptoms before diagnosis | 10.5 months (1–54) |
| Localization | |
| Conjunctiva | 62 (65.2) |
| Orbital soft tissue | 15 (15.8) |
| Eyelid | 13 (13.7) |
| Lacrimal gland | 5 (5.2) |
| Laterality | |
| Right eye | 48 (50.5) |
| Left eye | 34 (35.8) |
| Both eyes | 13 (13.7) |

Figures in parentheses are in percentage

At diagnosis, of the 95 patients who completed the staging evaluation, 83 patients presented with stage $IA_E$, 7 patients with stage $IA_{EE}$, 3 patients with stage $IIIA_E$, and 2 patients with stage IVA disease. Treatment modalities included radiotherapy ($n$ = 73), radiotherapy combined with chemotherapy ($n$ = 4), radiotherapy combined with maximal excision ($n$ = 14), and radiotherapy combined with maximal excision and chemotherapy ($n$ = 4) [Table 2].

The initial response rate was 100%, with 91 patients achieving a complete response and 4 patients achieving a partial response. There were two patients who developed recurrent tumor in the soft tissue of the contralateral orbit at 28 and 32 months after completion of radiotherapy, and they were salvaged by additional radiotherapy. Thus, 95 patients obtained ocular adnexa tumor control at the latest follow-up. Three-year overall survival was 100% and event-free survival was 97% by Kaplan–Meier survival analysis [Fig. 2].

Severe chronic complications such as corneal ulcer, cataract, or radiation retinopathy were not reported in our study. There were 17 patients who developed acute radiation conjunctivitis with spontaneous resolution after therapy. Periorbital erythema occurred in 13 patients and phimotic punctum developed in 1 patient. Filamentary keratitis followed by dry eye syndrome developed in one patient.

## Discussion

In this study, we evaluated the clinical features, clinical course, and results of treatment in a relatively large series of patients with MALT lymphoma of the ocular adnexa, who were observed for a mean period of 42.2 (range 6–73) months.

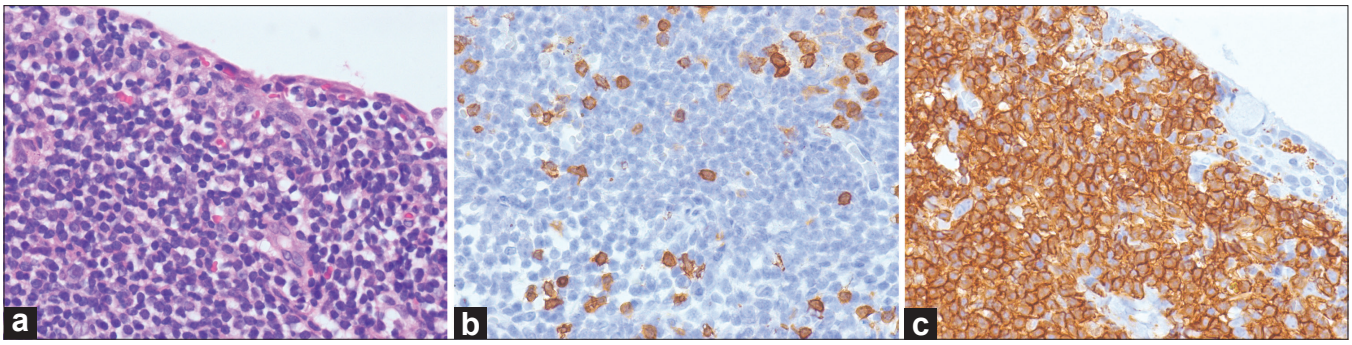


**Figure 1:** These slides show conjunctival extranodal marginal zone B cell lymphoma of mucosa-associated lymphoid tissue (also known as MALT lymphoma). MALT lymphoma is characterized by poorly defined follicular appearing areas that are composed of monocytoid B cells that feature enlarged nuclei. (a) An important diagnostic feature of MALT lymphoma is lymphoepithelial lesions (LEL), defined by the infiltration and distortion of epithelial structures by aggregates of neoplastic lymphoid cells (H and E, ×400). Immunochemically, the lymphocytes in LEL were CD20-positive and CD3-negative. (b) The figure corresponding to T-cell marker shows negative immunostain with CD3 (×400). (c) Positive CD-20 immunostain (×400). So, the presence of a monotonous population of CD20-positive intraepithelial lymphocytes supports a diagnosis of MALT lymphoma. (b, c)

**Table 2: Stage at diagnosis and treatment modality according to Ann Arbor stage[17]**

| Treatment modality | Stage | | | | | Total |
|---|---|---|---|---|---|---|
| | IAE | IAEE | IIAE | IIIAE | IVA | |
| Radiotherapy | 68 | 5 | | | | 73 |
| Radiotherapy + Maximal excision | 13 | 1 | | | | 14 |
| Radiotherapy + Chemotherapy | 2 | 1 | | | 1 | 4 |
| Radiotherapy + Chemotherapy + Maximal excision | | | | 3 | 1 | 4 |
| Total | 83 | 7 | | 3 | 2 | 95 |

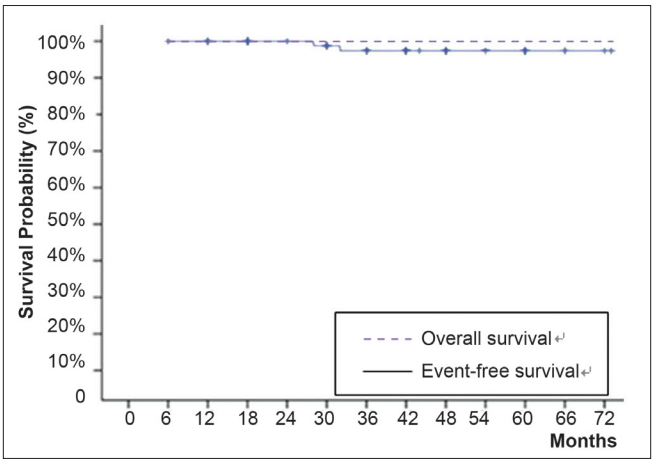


**Figure 2:** Kaplan–Meier survival rate. The 3-year overall survival and event-free survival rates were 100 and 97%, respectively

The presenting symptoms depend on the site of involvement in the ocular adnexa and its stage. The most common presenting symptom was a slowly growing orbital mass. This reflected the involvement of anterior structure (conjunctiva, lacrimal gland, and lid) in the majority of patients, which is consistent with earlier reports.[5,12,21] Especially, the initial symptoms and signs of onset of the disease lasted 10.5 months (range 1–54 months) because of the slow development of a palpable mass.

The issue of bilaterality in orbital lymphoma is interesting. It is somewhat difficult to apply the Ann Arbor staging system to MALT lymphoma in paired extranodal organs such as orbits and parotid glands. According to the studies of Bhatia *et al*,[12] Coupland *et al*.[22] and Tsang *et al*,[13] bilateral orbital lymphoma was regarded as stage $I_E$ disease. Some authors have categorized bilateral low-grade lymphoma limited to both orbits as stage $IA_{EE}$.[18-20]

In our study, MALT lymphoma of the ocular adnexa was usually, predominantly involving the right eye. Bilateral orbital involvement was found in 13 patients (13.7%). Our study also categorized simultaneous bilateral orbital lymphoma without lymph node involvement as stage $IA_{EE}$. However, patients with bilateral lesions have a course similar to those with unilateral lesions and should be considered as having stage I disease. In other words, whether bilateral orbital MALT lymphoma was considered stage $I_E$ or $I_{EE}$, curative radiotherapy alone was delivered without chemotherapy for the majority of patients, and there was no difference in event-free survival or overall survival between stage $I_E$ and $I_{EE}$ after curative radiotherapy alone. Of the 90 patients with stage $IA_E$ or $IA_{EE}$ who received a surgical resection or biopsy with consecutive radiotherapy, the overall complete response rate was 100%.

Overall, in this study, most disease stages were early. Of the 95 patients with MALT lymphoma, 90 presented with stage I, 3 presented with stage III, and 2 presented with stage IV. This finding is characteristic of extranodal lymphoma. These lymphomas remain confined to the orbit probably because they share features of homing to their tissue of origin[16] or because there are no lymphatic drainage channels in the orbit, which prevents dissemination of neoplastic cells.[23] However, one of the largest studies on MALT lymphoma of ocular adnexa by Jenkins *et al*,[24] reported that 18% of the patients with MALT lymphoma of ocular adnexa had previous or concurrent systemic disease at the time of diagnosis. Fung *et al*,[3] also reported that 19% of the patients with MALT lymphoma of ocular adnexa had stage III or IV disease at diagnosis. These

findings underscore the importance of adequate staging evaluation in MALT lymphomas of ocular adnexa.

Complete surgical excision usually is not possible without causing severe functional deficiencies.[1] This surgical limitation is associated with high rates of local relapse after surgical excision.[18] In our study, maximal excision was done not for curative treatment, but for supportive treatment and diagnosis in 18 patients as much as possible. Like NHL in other sites, surgery alone should not be employed as the main treatment modality in the treatment of MALT lymphoma of the ocular adnexa.

Radiotherapy with a dose of 30–35 Gy has been reported to be sufficient to provide local control and cure in localized orbital MALT lymphoma.[20,25-26] Most of the studies including a variety of lymphoma subtypes have documented the short- and long-term efficacy and side effects of this therapeutic modality. However, there is no universally accepted radiation schedule for patients with ocular adnexa marginal zone lymphoma. Thus, controversy exists regarding the optimal radiation dose and fractionation. Our data also demonstrated that excellent local control and survival can be achieved for patients with MALT lymphoma of ocular adnexa by radiotherapy alone with a median dose of 30 Gy. Only in two cases, contralateral orbital relapse developed out of 95 patients. What if there are residual tumors after completion of radiotherapy of around 30 Gy? We delivered an additional 10 Gy or 15 Gy to the patients with residual disease, 4 weeks after the end of initial radiotherapy, to enhance loco-regional control. The two persons with recurrence were salvaged by additional radiotherapy. These results confirm the effectiveness of radiotherapy as primary treatment in the management of extranodal marginal zone B-cell lymphomas.

Bennett *et al*,[27] reported that chemotherapy was effective in primary orbital lymphoma of high grade and/or advanced stage. In our study, eight patients received additional chemotherapy (COP or CHOP regimens). Two patients were stage IVA with atypical lymphocytic infiltration of the bone marrow; one patient was stage $IIIA_E$ with involvement of an inguinal lymph node; two patients were stage $IIIA_E$ with involvement of an abdominal lymph node; and the other patients were stage $IA_E$, who had received treatment at other hospitals. It has been reported that combination chemotherapy is effective in ocular MALT lymphoma.[4,28] But, there was small number of patients who receive chemoradiotherapy in our study, and none of the patients received chemotherapy alone. So, it is difficult to comment on the results of chemotherapy for MALT lymphoma. Further studies are needed to know the efficacy of combination chemotherapy and the role of anti-CD20 monoclonal antibody treatment. The results of literature survey of treatment and outcome of patients with MALT lymphoma of ocular adnexa are summarized in Table 3.

In our study, most complications were mild, requiring no intervention. Important side effects such as cataract development have not yet been observed in this series of patients treated with doses ranging from 30.6 to 45.0 Gy; however, Minehan *et al*,[29] reported a high rate of complications, where complications developed in 12 of 22 patients. The absence of cataract development can be explained only partly by the low-dose radiation therapy used in almost all our patients except for one, together with the relatively young age of our patients. In addition, lens shielding is an important aspect of this treatment method. Especially two out of four patients who received 40 Gy or more suffered from complications later like phimoptic punctum or dry eye syndrome.

Several retrospective studies,[3-6,13] most of which include a variety of lymphoma subtypes, have documented the short- and long-term efficacy and side effects of this therapeutic modality. However, there is no universally accepted radiation schedule for patients with ocular adnexa marginal zone lymphoma. Thus, controversy exists regarding the optimal radiation dose and fractionation. However, patient population is too small to produce a generalized conclusion. The previous case series on "Radiotherapy as first-line treatment in patients with ocular adnexa marginal zone lymphoma" particularly had a very small sample size ($n$ = 15–58). In our study, the number of patients with radiotherapy only as their first-line treatment was 73. Although a larger sample size does not

**Table 3: Result of literature survey of treatment and outcome of patients with MALT lymphoma of ocular adnexa**

| Authors | No of patients with MALT lymphoma | Stage | Subtype studied | Treatment | Complete response rate (%) | Overall survival rate |
|---|---|---|---|---|---|---|
| Fung[3] | 48 | I: 39<br>III: 3<br>IV: 6 | NHL | RT (23.1–45 Gy)<br>CTx for stage IV<br>Surgery only (1) | 100 for RT | 94% (5 years) for stage I, 39% for stage III–IV |
| Galieni[4] | 15 | I | MALT | RT 36–45 Gy<br>CTx | 93 | 100% cause specific survival |
| Hasegawa[5] | 20 | I: 19<br>IV: 1 | NHL | RT 20–40 Gy<br>CTx + RT for stage IV | NA | 100% (5 years) |
| Uno[6] | 50 | I: 50 | MALT | RT (20–46 Gy) | 98 | 91% (5 years) |
| Tsang[13] | 19 | I | MALT | RT 25 Gy<br>RT 30 Gy for PR | 95 | NA |
| This study | 95 | I 90<br>III 3<br>IV 2 | MALT | RT (30.6–45 Gy)<br>RT + surgery (14)<br>CTx + RT (1)<br>Surgery + CTx + RT (3) | 93.7 | 100% (3 years) |

CTx: chemotherapy, RT: radiotherapy

always guarantee more significantly meaningful data, it can reduce statistical error at the same confidence level. Moreover, although this study has limited significance compared to large multicenter randomized trials, the over 10-year experience of a single institution, which includes long follow-up of its patients, should provide meaningful data regarding the use of radiotherapy in MALT lymphoma of the ocular adnexa. Hemato-oncologist and radiation oncologist, being the same person, can reduce bias. But relatively small numbers of patients to produce meaningful data to Kaplan–Meier survival analysis is another limitation.

In conclusion, MALT lymphomas constitute the majority of NHLs of ocular adnexa. Complete staging evaluation including abdominal and chest CT scans and a bone marrow study is needed to select an adequate treatment modality. Radiotherapy alone can produce excellent local control and survival in patients with stage $I_E$ disease irrespective of bilaterality. Systemic chemotherapy should be considered in patients with advanced disease or systemic manifestation.

## References


1. Keleti D, Flickinger JC, Hobson SR, Mittal BB. Radiotherapy of lymphoproliferative diseases of the orbit. Surveillance of 65 cases. Am J Clin Oncol 1992;15:422-7.
2. Freeman C, Berg JW, Cutler SJ. Occurrence and prognosis of extranodal lymphomas. Cancer 1972;29:252-60.
3. Fung CY, Tarbell NJ, Lucarelli MJ, Goldberg SI, Linggood RM, Harris NL, *et al*. Ocular adnexal lymphoma: Clinical behavior of distinct World Health Organization classification subtypes. Int J Radiat Oncol Biol Phys 2003;57:1382-91.
4. Galieni P, Polito E, Leccisotti A, Marotta G, Lasi S, Bigazzi C, *et al*. Localized orbital lymphoma. Haematologica 1997;82:436-9.
5. Hasegawa M, Kojima M, Shioya M, Tamaki Y, Saitoh J, Sakurai H, *et al*. Treatment results of radiotherapy for malignant lymphoma of the orbit and histopathologic review according to the WHO classification. Int J Radiat Oncol Biol Phys 2003;57:172-6.
6. Uno T, Isobe K, Shikama N, Nishikawa A, Oguchi M, Ueno N, *et al*. Radiotherapy for extranodal, marginal zone, B-cell lymphoma of mucosa-associated lymphoid tissue originating in the ocular adnexa: A multiinstitutional, retrospective review of 50 patients. Cancer 2003;98:865-71.
7. Isaacson P, Wright DH. Malignant lymphoma of mucosa-associated lymphoid tissue. A distinctive type of B-cell lymphoma. Cancer 1983;52:1410-6.
8. Harris NL, Jaffe ES, Stein H, Banks PM, Chan JK, Cleary ML, *et al*. A revised European-American classification of lymphoid neoplasms: A proposal from the international lymphoma study group. Blood 1994;84:1361-92.
9. Isaacson PG, Berger F, Muller-Hermelink HK, Nathwani BN, Piris MA, Swerdlow SH, *et al*. Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma). In: Jaffe ES, Harris NL, Stein H, Vardiman JW, editor. World Health Organization classification of tumours; Tumours of haematopoietic and lymphoid tissues. Lyon: IARC Press; 2001. p. 157-60.
10. Wotherspoon AC, Doglioni C, Diss TC, Pan L, Moschini A, de Boni M, *et al*. Regression of primary low-grade B-cell gastric lymphoma of mucosa-associated lymphoid tissue type after eradication of *Helicobacter pylori*. Lancet 1993;342:575-7.
11. Ferreri AJ, Guidoboni M, Ponzoni M, De Conciliis C, Dell'Oro S, Fleischhauer K, *et al*. Evidence for an association between Chlamydia psittaci and ocular adnexal lymphomas. J Natl Cancer Inst 2004;96:586-94.
12. Bhatia S, Paulino AC, Buatti JM, Mayr NA, Wen BC. Curative radiotherapy for primary orbital lymphoma. Int J Radiat Oncol Biol Phys 2002;54:818-23.
13. Tsang RW, Gospodarowicz MK, Pintilie M, Bezjak A, Wells W, Hodgson DC, *et al*. Stage I and II MALT lymphoma: results of treatment with radiotherapy. Int J Radiat Oncol Biol Phys 2001;50:1258-64.
14. Song EK, Kim SY, Kim TM, Lee KW, Yun T, Na II, *et al*. Efficacy of chemotherapy as a first-line treatment in ocular adnexal extranodal marginal zone B-cell lymphoma. Ann Oncol 2008;19:242-6.
15. Stefanovic A, Lossos IS. Extranodal marginal zone lymphoma of the ocular adnexa. Blood 2009;114:501-10.
16. Isaacson PG WD. Malignant lymphoma of mucosa-associated lymphoid tissue: A distinctive of B-cell lymphoma. Cancer 1994;52:7.
17. Carbone PP, Kaplan HS, Musshoff K, Smithers DW, Tubiana M. Report of the committee on Hodgkin's disease staging classification. Cancer Res 1971;31:1860-1.
18. Esik O, Ikeda H, Mukai K, Kaneko A. A retrospective analysis of different modalities for treatment of primary orbital non-Hodgkin's lymphomas. Radiother Oncol 1996;38:13-8.
19. Sasai K, Yamabe H, Dodo Y, Kashii S, Nagata Y, Hiraoka M. Non-Hodgkin's lymphoma of the ocular adnexa. Acta Oncol 2001;40:485-90.
20. Smitt MC, Donaldson SS. Radiotherapy is successful treatment for orbital lymphoma. Int J Radiat Oncol Biol Phys 1993;26:59-66.
21. Cho EY, Han JJ, Ree HJ, Ko YH, Kang YK, Ahn HS, *et al*. Clinicopathologic analysis of ocular adnexal lymphomas: Extranodal marginal zone b-cell lymphoma constitutes the vast majority of ocular lymphomas among Koreans and affects younger patients. Am J Hematol 2003;73:87-96.
22. Coupland SE, Krause L, Delecluse HJ, Anagnostopoulos I, Foss HD, Hummel M, *et al*. Lymphoproliferative lesions of the ocular adnexa: Analysis of 112 cases. Ophthalmology 1998;105:1430-41.
23. Garner A. Orbital lymphoproliferative disorders. Br J Ophthalmol 1992;76:47-8.
24. Jenkins C, Rose GE, Bunce C, Wright JE, Cree IA, Plowman N, *et al*. Histological features of ocular adnexal lymphoma (REAL classification) and their association with patient morbidity and survival. Br J Ophthalmol 2000;84:907-13.
25. Lee SW, Suh CO, Kim GE, Yang WI, Lee SY, Hahn JS, *et al*. Role of radiotherapy for primary orbital lymphoma. Am J Clin Oncol 2002;25:261-5.
26. Yahalom J. MALT lymphomas: A radiation oncology viewpoint. Ann Hematol 2001;80:B100-5.
27. Bennett CL, Putterman A, Bitran JD, Recant W, Shapiro CM, Karesh J, *et al*. Staging and therapy of orbital lymphomas. Cancer 1986;57:1204-8.
28. Fisher RI, Gaynor ER, Dahlberg S, Oken MM, Grogan TM, Mize EM, *et al*. Comparison of a standard regimen (CHOP) with three intensive chemotherapy regimens for advanced non-Hodgkin's lymphoma. N Engl J Med 1993;328:1002-6.
29. Minehan KJ, Martenson JA, Garrity JA, Kurtin PJ, Banks PM, Chen MG, *et al*. Local control and complications after radiation therapy for primary orbital lymphoma: A case for low-dose treatment. Int J Radiat Oncol Biol Phys 1991;20:791-6.


**Cite this article as:** Lim S, Kang M, Son J. Extranodal marginal zone B cell lymphoma of mucosa-associated lymphoid tissue type of the ocular adnexa: Retrospective single institution review of 95 patients. Indian J Ophthalmol 2011;59:273-7.

**Source of Support:** This research was supported by the Yeungnam University research grants in 2008, **Conflict of Interest:** None declared.