Conjunctival Malt Lymphoma: A Case Report

Tuna S (1), Karacetin D (2), Basaran M (1), Agaoglu F (3), Dogan O (4), Buyukbabani N (4)

Conjunctival MALT Lymphoma: A Case Report

INTRODUCTION

Extranodal marginal zone B-cell lymphomas of the mucosa associated lymphoid tissue are generally slow growing lesions. Ocular adnexal lymphomas of the MALT type, are encountered more rarely compared to gastric MALT lymphomas. These show indolent course and very seldom become fatal. They may root from the eyelids, lacrimal gland, conjunctiva, and orbita.

In gastric MALT lymphoma patients, H. pylori and infection are observed in a rate between 72 % to 90 % (1). In so much as that only antibiotics and H. pylori eradication lead to regression 70 % to 80 % of gastric MALT lymphoma cases (2). Whilst the role the infectious agents, except H.Pylori, on MALT lymphomas are relatively less known, Chlamydia psittaci existence has been shown in ocular adnexal MALT lymphomas (3). Borrelia burgdorferi infection has been observed in some skin lymphomas (4,5). Again, an association has been determined between Hepatitis C virus infection and extranodal lymphomas (6,7).

Case Report

43 year old female patient, working as a nurse in the eye clinic was examined two years ago for the sense of a foreign body in her right eye (figure 1). A swollen mass was identified right under the epithelium and forming intensive diffuse masses attracting notice Figure 2,3). It was diagnosed as conjunctiva containing small lymphocytic proliferation (figure 4). Histopathologically, apparent malignancy findings were not observed in the monitored sections, yet the report was prepared as “These findings do not retract MALT lymphoma. Clinic follow-up and further examinations are suggested”. Torax, Abdomen and pelvic CT (Computerized Tomography) and orbital/cranial MRI (Magnetic Resonance Imaging) analysis carried out that day did not show any metastasis and the case was evaluated as Pha-se IE for Ann Arbor classification.

The gastric biopsy taken in esophagogastroduodenoscopic inspection showed positive and it was diagnosed as chronic active gastritis. Yet, atrophy negative and malignancy or metaplasia were not identified. For further inspection, incisonal
biopsy was requested from conjunctiva lower fornix. Intensive lymphoid infiltration was identified starting right from under the epithelium, filling whole subepithelial area, more dominant then centrocyte like cells containing ambiguous follicular organization.

As a result of the immunohistological analysis carried out using anti-CD3, CD20, kappa, lambda and bcl-6 antibodies, nearly all of the cells forming infiltration showed positive reaction with CD20 (figure 1). Cell groups are observed which show positive reaction in small groups with CD3. Few cells showed reaction with bcl-6 antibody. In the analysis carried out with kappa and lambda light chain antibodies, many cells having plasma cell or plasmacytoid cell morphology in infiltration showed reaction with kappa (Figure 2) whereas very few cells showed reaction with lambda. Morphological and immunophenotypic findings were found coherent with low-grade B-cell lymphoma (MALT lymphoma), rooted from mucosa associated lymphoid tissue and the diagnosis was reported as; Right eye lower fornix, conjunctiva incisional biopsy: B cell neoplastic lymphoid infiltration showing kappa light chain monoclonal.

The patient was started subconjunctival intralesional interferon-α injection following this diagnosis. The patient showed no regression after 14 months of treatment, and an increase was observed in tumor border towards medial. In physical examination, salmon patch shaped typical lesion existed in the right eye conjunctiva lower fornix (Figure 3).

The patient was accepted as refractor for local interferon treatment, and was sent to our institution for evaluation in terms of radiotherapy. The patient did not have sight problem. Therefore, RT was not considered as the first treatment. For the beginning, being CD20 positive, a treatment was planned with rituximab which is a CD20 antibody. Considering patient’s history; she was feeding a bird (a kind of a small parrot) until two years before; 3 weeks doxycycline empiric antibiotic therapy treatment was added to the treatment. Rituximab 375 mg/m² per week for 4 weeks; and 100 mg Doxycyclin per day for 3 weeks was started.

Complete response was positive after treatment. The patient showed no recurrence during 3 months-interval follow up’s which lasted until May 2009.

**DISCUSSION**

Conjunctival MALT lymphomas show indolent clinic course, and generally have low spreading risk. In physical examination, orange or salmon-pink masses draw attention (9). For orbital adnexal MALT lymphomas, treatment options include only follow-up (no initial therapy) (10), Radiotherapy (11), Chemotherapy (12), Rituximab (13), antibiotics (Doxycycline) (14), subconjunctival (intralasonal) interferon— injection (15).

Radiotherapy has shown successful results, yet it has side effects like cataract and eye drying. Long-term follow-up with no initial therapy might be suitable option for selected cases since it has a slow course (10,16). Whilst chemotherapy is a very effective treatment option, systemized chemotherapy might be considered in acute cases as well as palliative treatment. Although the role of Rituximab treatment has not perfectly be clarified, it is a suitable option with high response rates and low toxicity. Bacteria-eradicating therapy model with Doxycycline is considered as a fast, safe and active treatment in ocular adnexal MALT lymphomas according to a multicenter prospective study. This treatment has been effec-
tive in patients who have received RT and progressed, and it has even been effective in Chlamydia psittaci-DNA negative cases (17).

As a result, each case requires appropriately in ocular adnexal MALT lymphomas before determining the most appropriate treatment option. In clinic application; wait and see policy, anti-CD20 antibody treatment and bacteria-eradicating therapy are likely to forge ahead whereas RT which once was considered as the first-line treatment, is likely to fall back.

REFERENCES


