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Long-term follow-up results of no initial therapy for ocular adnexal MALT lymphoma

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Background: The majority of lymphomas in the ocular adnexa are low-grade B-cell lymphomas of mucosa-associated lymphoid tissue (MALT lymphoma). Although radiotherapy is the most frequently applied management, cataract and dry eye are problematic complications.

Patients and methods: Between 1973 and 2003, the clinical features of 36 patients with ocular adnexal MALT lymphoma with no symptoms who were managed with no initial therapy after biopsy or surgical resection were retrospectively analyzed.

Results: The median patient age was 63 years (range 22–84) and all patients had stage I disease, consisting of 31 unilateral cases and five bilateral cases. With a median follow-up of 7.1 years, 25 (69%) did not require treatment. The median time until the initiation of treatment in the remaining 11 patients (31%) was 4.8 years. Six patients (17%) died, and among them only two (6%) died due to progressive lymphoma. Seventeen patients (47%) progressed, but histologic transformation was recognized in only one (3%). The estimated overall survival rates of the 36 patients after 5, 10 and 15 years were 94%, 94% and 71%, respectively.

Conclusions: In selected patients with ocular adnexal MALT lymphoma, no initial therapy might be an acceptable approach, because 70% of patients remained untreated at a median of 8.6 years, and their survival was comparable to that of reports on immediate therapy.

Key words: MALT lymphoma, ocular adnexa, no initial therapy, prognosis

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introduction

An extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma), first described in 1983 by Isaacson and Wright, was recognized in 1994 as a distinct entity of low-grade B-cell lymphoma in the revised European–American lymphoma (REAL) classification among marginal zone B-cell lymphomas, as well as in the most recent classification of the World Health Organization (WHO) [1–3]. Generally, the majority of MALT lymphomas have an indolent natural history and two-thirds of patients with MALT lymphoma have localized disease [4, 5]. For localized MALT lymphoma, radiotherapy is the most frequently applied management, and most patients show good response to radiotherapy, although several recent reports suggest that radiotherapy alone may not provide for a superior outcome [6–10].

For the management of lymphomas in the ocular adnexa, especially for localized disease, radiotherapy is a safe and effective form of local treatment [11–16]. Histology according to the REAL or WHO classification can be used to accurately predict the prognosis of lymphomas in the ocular adnexa, and the MALT type has a more favorable prognosis than do malignant lymphomas of differing histology [17–20]. Although there have been few analyses of large numbers of MALT lymphomas in the ocular adnexa, its prognosis is thought to be better.

In general, low-grade lymphoma spontaneously regresses on several occasions. Conservative management by deferring treatment until disease progression is an acceptable option for selected patients with follicular lymphoma [21, 22]. This watch-and-wait strategy was initially applied to advanced-stage patients, but a recent retrospective study from Stanford University suggested that this conservative approach is applicable to selected patients with Stage I or II follicular lymphoma [23]. For the radiotherapy of ocular adnexal lymphoma, cataract and dry eye are problematic complications in some patients. From these points, it is controversial whether all patients with MALT lymphoma in the ocular adnexa should be treated with radiotherapy. There is only one report on a small number of patients according to the watch-and-wait approach for conjunctival MALT lymphoma [24]. Therefore, we conducted a retrospective analysis of

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