Burkitt Lymphoma Leading to Isolated Conjunctival Relapse

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Dear editör,

Lymphoma is a malignancy that most frequently causes ocular involvement. Lymphomas that cause orbital involvement are considered in three groups. The first group includes primary ocular adnexal lymphomas; the conjunctiva, lacrimal gland, eyelid, orbital soft tissue, and extraocular muscles are involved in 1% to 2% of all non-Hodgkin lymphomas (NHL), and these structures are involved in 5% to 15% of lymphomas that cause extranodal involvement. The second group includes the ocular adnexal lymphomas that are associated with diffuse involvement by lymphoma determined at diagnosis or relapse. The third group of intraocular lymphomas are cases with involvement of the eye that generally occur in diffuse large B-cell lymphoma; they are considered in the category of aggressive primary central nervous system (CNS) lymphomas. When eye involvement is present, lymphomas cause proptosis and swelling due to the pressing effect of the mass, and also minimal pain and inflammation; salmon colored areas in the conjunctiva, ptosis, exophthalmus, loss of vision, and epiphora may be more widely seen.[1,2]

In the present study, we have discussed our patient with Burkitt lymphoma (BL) occurring with isolated conjunctival relapse, and reported this uncommon case.

The 8-year old patient attended with a complaint of abdominal mass in April 2011, and was diagnosed as BL [CD20(+), CD10(+), CD3(-), bcl-6(+), bcl-2(-)] in stage 3 (no bone marrow or CNS involvement); the patient underwent LMB 89 treatment protocol.[3] The patient did not undergo radiotherapy, and was accepted as cured in January 2012. The case was routinely followed-up, and 34 months following remission and in November 2014, when she was 11 years old, she applied with a complaint of ptosis in the right eye. Magnetic resonance imaging (MRI) of the eye and brain revealed thickening in the pharyngeal walls, and a mass lesion of soft tissue in the upper right palpebra in dimensions of 41 x 21 x 13 mm, which showed heterogenous and intensive trapping of contrast that was hypo-intense in T1A, and hyperintense in T2A; these signs were reported as involvement by lymphoma (Figure 1). Malign cells were not visualized in the biopsy specimen undertaken by the nasopharyngeal route, however investigation of the conjunctival biopsy material was reported as a BL [CD20(+), CD10(+), CD79a(+), CD3(-), bcl-6(+), bcl-2(-)] originating from the conjunctiva. The patient did not have involvement of the bone marrow, CNS, bone, thorax and abdomen, and in January 2015 she started to receive the relapse NHL protocol (Carboplatin, ifosfamide, Etoposide (ICE)).[3] ICE plus CNS prophylaxis as intrathecal methotrexate three doses was given.

doi: 10.4999/uhod.161388

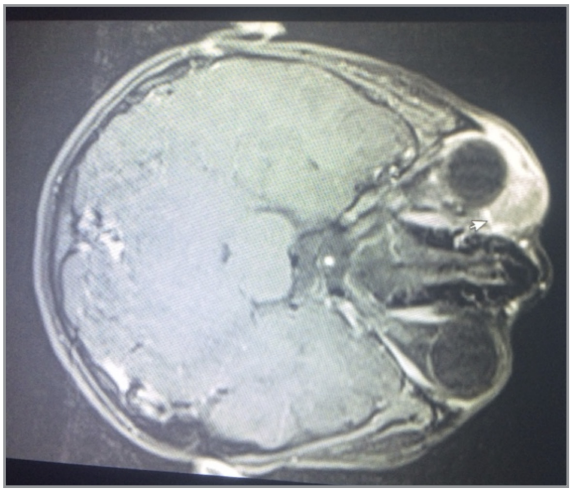

Figure 1. Magnetic resonance imaging (MRI) of the eye; a mass lesion of soft tissue in the upper right palpebra, which showed heterogenous and intensive trapping of contrast that was hypo-intense in T1A (arrow).

In the evaluation of patient following threecycles of chemotherapy, the signs in the cranial and eye MRs were found to improve, and positron emission tomography was normal. Treatment of patients completed six cycles.

One year after diagnosis, there has been no local recurrence and in the patient is complete remission now.

The high-intensity and multiple-agent chemotherapy regimens currently applied increases the success rate of treatment in BL to more than 90%. The incidence of relapse in cases with BL or resistance to treatment indicates that they have a poor prognosis.[4,5]

The marginal zone B-cell lymphoma, follicular lymphoma, diffuse large B-cell lymphoma, and mantle-cell lymphoma are the types of lymphoma that cause the most frequent ocular involvement. The T-cell/natural killer-cell lymphoma and BL cause rare orbital involvement.[1,2] The BLs that lead to orbital involvement occur sporadically in children, and they are therefore reported as case reports.[6] Our patient had abdominal involvement at diagnosis, and there was no conjunctival involvement. The disease was in remission, and isolated conjunctival relapse was determined after 34 months.

The first case with BL presented with conjunctival involvement was reported in 1995 when the patient was 16 years old. This patient presented with orbital mass, and the result of the conjunctival biopsy revealed BL at stage 1E. The patient received combined treatment and responded well.[7] In another study, a total of 1,014 cases of lymphoma with conjunctival involvement were screened; in this study, B-cell lymphomas were found to account for 98% of the involvements. In these patients, who were screened, only one case of BL was determined to occur with involvement, and this was the case reported in 1995. Moreover, conjunctival lymphomas existed mostly in elderly patients.[6]

Afanas et al.[8] screened 21 children with BL diagnosed between the years 2008 and 2011. The most common sites of involvement were found to be as follows: abdomen (13 cases), tonsils (three cases), orbita (one case), CNS (two cases), tongue (one case), and nasopharynx (one case).

When conjunctival lymphoma is considered, careful ophthalmologic examinations, and a tissue specimen sufficient for histopathological diagnosis, are essential. Conjunctival lymphomas generally has a good prognosis; however, these cases have to be followed-up for several years because of the risk of relapse.[1]

Orbital lymphomalar is associated with an increased risk of CNS dissemination either by hematogenous route or by direct infiltration through the skull bones. Cancer Network guidelines recommend prophylaxis of intrathecal methotrexate and/or cytarabine given during primary chemotherapy.[1,6] We have given to our patient CNS prophylaxis with intrathecal methotrexate during primary chemotherapy.

Radiotherapy is currently used as consolidation therapy after primary chemotherapy in patients with limited stage orbital lymphoma. However, radiotheraphy-related xerophthalmia, corneal ulceration, cataract formation,and vasculopathy of the retina and the optic nerve may occur.[1,6] We didn't need to give radiotherapy to our patient because of complete complete remission after chemotherapy.

Conjunctival involvement is relatively rare in patients with BL. However when the signs like conjunctival hyperemia and ptosis are determined, the patients have to undergo a detailed ophthalmologic examination, and radiological imaging of the eye must be conducted. A biopsy has to be made for a

definite diagnosis. It should be kept in mind that isolated conjunctival involvement may rarely be seen in the patients with BL, both in the course of diagnosis, and in the state of relapse. At the period of relapse, involvements of bone marrow, CNS, bone, thorax, and abdomen were not detected in our case. The case presented with isolated conjunctival relapse, and responded well to treatment. Nevertheless, the patient was in stage 3 at diagnosis, and occurred with relapse; we therefore consider that the case has to be closely followed-up.

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