Lacrim sac lymphoma: a case report

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INTRODUCTION

Tumors of the lacrimal drainage system are rare, which can be classified into two types: epithelial and non-epithelial neoplasms. Epithelial tumors are more commonly found, with an incidence of 90%. Lacrimal sac lymphomas are even more rare and usually occur secondary to systemic lymphoma metastasis. Patients commonly present with epiphora, recurrent dacryocystitis, lacrimal sac mass and epistaxis. Early diagnosis and management can reduce the risk of disease recurrence and visual loss. Even though the lacrimal drainage system is located near the nasal area, the treatment of a malignant lymphoma of the lacrimal drainage system is slightly different from other tumors that located near the nasal area.

CASE REPORT

A 71-year-old woman presented with a painless mass below the right medial canthus. Computed tomography (CT) scan of the orbit revealed a mass invading the right lacrimal sac. An incision biopsy was obtained, and the pathologic findings suggested a diagnosis of primary diffuse large B-cell lymphoma of the lacrimal sac. The patient was treated with chemotherapy and intrathecal methotrexate. After completing eight cycles of chemotherapy, the patient was followed up by a CT scan, which revealed nearly total resolution of an ill-defined enhancing mass. At the time of this case report writing, the patient is in complete remission at six months with no other complications.

Abbreviations: CT, computed tomography; DLBCL, diffuse large B-cell lymphoma; MRI, magnetic resonance imaging

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A computed tomography (CT) scan of the head and neck showed an ill-defined enhancing mass, sized 1.8 × 1.6 × 2.9 cm, located at the right lacrimal sac extending to the right extraconal fat, right nasal cavity, and right maxillary sinus involving the medial maxillary wall. There were multiple cervical lymph nodes, sizes up to 1.7 cm (Fig. 1).

Fine needle aspiration of an enlarged cervical lymph node (right neck IIA level) was performed, and the pathologic studies showed polymorphous lymphoid cells, but were inconclusive.

Pathologic findings from the trans-nasal biopsy of the lacrimal sac mass showed atypical round cells (Figs. 2, 3). Immunohistochemistry studies found positive expressions for BCL-2, BCL-6, MUM1, c-MYC; negative for AE1/AE3, CD10, CD23, Cyclin D-1; CD3 of scattered small T-cell, CD20 of sheets of large atypical B-cells, Ki-67 of 80% proliferation index (Fig. 4).

Fig. 1. Enhanced computed tomographic image of a 71-year-old woman shows ill-defined enhancing mass located at the right lacrimal sac extending to the right extraconal fat, right nasal cavity and right maxillary sinus involving the medial site eyeball. (A) Axial and (B) coronal views.

Fig. 2. Lacrimal sac mass. Section showing fragments of the lacrimal glands and tumor cell clusters. The tumor cells are arranged in diffuse sheets without structure or nodular formation (H&E, ×200).

Fig. 3. Lacrimal sac mass. The tumor cells contain irregular-shaped nuclei with rather fine nuclear chromatin. The lesion also contains tingible body macrophages (H&E, ×400).

Fig. 4. Lacrimal sac mass with immunohistochemical stain. (A) CD3 was negative (×100). (B) CD20 was diffuse positive (×100). (C) Ki-67 showing a high proliferation index (up to 80%–90%) (×200).
From these findings, the mass was concluded as being a “diffuse large B-cell lymphoma (DLBCL) with non-germinal center phenotype.” However, the double expression of BCL-2 and c-MYC with a high proliferation index might indicate a poor prognosis.

There was no abnormal finding on the CT scan of her chest with whole abdomen and bone marrow aspiration. Thus, the patient was diagnosed with primary DLBCL of the lacrimal sac stage II.

The patient was treated with chemotherapy because the tumor was DLBCL that categorized as stage II. The patient received one cycle of CVP regimen, comprising cyclophosphamide, vincristine, and prednisolone, and eight cycles of R-CHOP regimen, comprising rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisolone. Also, intrathecal methotrexate was given to prevent the recurrence of neurological complications.

After completing eight cycles of chemotherapy, the follow-up CT scan revealed nearly total resolution of the mass. There was an overall decrease in size of the cervical nodes, sized up to 0.5 cm.

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After completing eight cycles of chemotherapy, the follow-up CT scan revealed nearly total resolution of the mass. There was an overall decrease in size of the cervical nodes (Fig. 5). Rhinoscopic examination showed no mass at the right nasal cavity and no palpable cervical lymph node upon neck examination. At the time of writing this report, the patient is in complete remission for 6 months with no other complications.

DISCUSSION

Lacrimal sac tumors are rare [1-4]. Approximately 55% of lacrimal sac tumors are malignant, and 71% originate from epithelial cells [1]. Malignant lymphomas account for approximately 6% of all lacrimal sac tumors [5]. They are commonly second-
tem lymphomas are the B-cell type, which has a good response to radiotherapy or chemotherapy [2], and therefore the prognosis appears favorable.

For primary lacrimal sac lymphoma, chemotherapy or radiotherapy is considered the primary treatment [19]. Extensive surgical interventions should be avoided to preserve the function and aesthetic of the eye [2]. In DLBCL, systemic chemotherapy that is CHOP regimens, which include cyclophosphamide, doxorubicin, vincristine, prednisolone, and adjunct immunotherapy therapy that is rituximab, are the treatment of choice. For DLBCL cells that have the CD20 antigen on the cell surface, immunotherapy with rituximab, which is a chimeric monoclonal anti-CD20 antibody, combined with chemotherapy has a significant advantage over chemotherapy alone in terms of overall survival and response rate [20]. As a result, we treated the patient with CHOP regimen chemotherapy, rituximab for adjunct immunotherapy and intrathecal methotrexate for preventing the recurrence of neurological complications.

In conclusion, primary lacrimal sac lymphoma is rare, with minimal and nonspecific signs and symptoms. In this case report, we present a patient with primary lacrimal sac DLBCL who showed a good response to R-CHOP regimens.

NOTES

Conflict of interest

No potential conflict of interest relevant to this article was reported.

Ethical approval

The study was approved by the Human Ethics Committee of Srinakharinwirot University (SWUEC/X-144/2564).

Patient consent

The patient provided written informed consent for the publication and the use of her images.

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