LETTER TO THE EDITOR

Ocular adnexal marginal zone B cell lymphoma presenting with orbital apex syndrome

Lymphome B de la zone marginale se présentant comme un syndrome de l’apex orbitaire

I write to present a case report of ocular adnexal marginal zone B cell lymphoma presented with orbital apex syndrome.

Ocular adnexal lymphomas (OALs) are primary or secondary lymphomas of orbit, conjunctiva, lacrimal gland or eyelid [1]. B-cell non-Hodgkin lymphomas (NHLs) constitute the majority of ocular adnexal lymphomas and they occupy approximately 1% of all NHLs [2]. OALs are generally limited to the primary tissues in which they occurred. Involving to the apex is not an usual clinical finding of ocular adnexal tumors and to our knowledge, such a case has not been previously reported in the literature. Here, we investigate a case of stage IA primary unilateral ocular adnexal marginal zone B cell lymphoma presented with orbital apex syndrome.

A 60-year-old male patient was admitted to our clinic with complaints of swelling of right upper eyelid, progressive vision loss and binocular diplopia. The right globe was nearly immotile. He has suffered from these complaints for 6 months. The medical history and family history of the patient revealed no abnormality. Ophthalmic examination demonstrated hyperemia and edema of the right upper eyelid, axial protrusion of the right globe and chemosis of the right eye (Figs. 1 and 2). Limitation of the right globe in all positions of gaze was observed. There was just a minimal motility in down gaze position. Best-corrected visual acuity of the right eye is counting fingers at 2 m and the left eye is 20/20. Cornea and anterior segments of both eyes are reported normal. The ratio of cupping to the optic disc of the right eye is 0.9 and the left eye 0.5. Central corneal thickness and intraocular pressure of both eyes are examined normal.

Magnetic resonance imaging (MRI) scans of the right orbit showed a mass which is not distinguishable from lacrimal gland and protruding to the eyelid while extending from superior side to the orbital apex (Fig. 3). Histopathological examination of an incisional biopsy showed infiltration by a lymphoid cell population composed of CD20 positive small-sized lymphocytes and the pathologists reported this mass as low grade B cell non-Hodgkin’s marginal zone lymphoma (Fig. 4). Bone marrow biopsy revealed normocellular. Computed tomography (CT) scans of larynx, thorax and abdominal regions did not demonstrate any images compatible with lymphoma. The oncology department graded the mass as stage IA marginal zone lymphoma and recommended 4 cures of rituximab therapy during 4 weeks.

Infectious, inflammatory, traumatic, vascular and neoplastic conditions may cause orbital apex syndrome [2,3]. Fifty cases of orbital apex syndrome and the common aetiology of this series is carotid-cavernous sinus fistula while the second frequent one is tumors, of which lymphoma is the most common cause in tumors group [3]. In our report, presenting symptoms of the patient were swelling of eyelid, proptosis, progressive vision loss, binocular diplopia and ophthalmoplegia. Ophthalmoplegia occurs due to the involving the cranial nerves of eye [3]. This uncommon but dramatical finding of ocular adnexal tumors is the most striking sign of orbital apex syndrome. MRI is the preferred

Figure 1. Hyperemia and edema of the right upper eyelid.

Figure 2. Axial proptosis of the right globe and chemosis of the right eye.
imaging technique for evaluating the location, intraorbital extension and orbital apex involvement of the tumor [4]. Clinical and imaging features do not distinguish the benign and malign lymphoid proliferations, therefore histopathological, immunophenotypical and molecular analyses are required for histological diagnosis. Immunophenotypic analysis of B-cell and T-cell markers, CD5, CD10, CD23, cyclin D1, and bcl-2 also help classifying lymphomas [5].

Management of OAL requires a multidisciplinary approach. In general, low-grade tumors are successfully treated with radiotherapy. However, high-grade lymphomas or disseminated systemic involvement require systemic chemotherapy [6]. Doses of 15–30 Gy up to 40 Gy have been recommended for mild to moderate tumors [6]. This case is also a low-grade tumor that we suggested to treat with local radiotherapy. However, the patient hesitates to take radiotherapy because of the side effects. Intensity modulated radiation therapy (IMRT) is used to reduce the toxic effects of conventional radiotherapy in patients with orbital lymphoma. IMRT has excellent outcomes similar to conventional radiotherapy and reduces dose to critical structures while providing excellent dose coverage of target volumes [7].

Monoclonal antibodies represent the current modern lymphoma treatment. Rituximab has been used to treat low-grade CD20 positive lymphoma by destroying B cells [8]. Antiangiogenic drugs which inhibit cancer cell in-growth are also being researched for NHL treatment. Since the

Figure 3. MRI scans of the right orbit demonstrate a mass which is not distinguishable from lacrimal gland and protruding to the eyelid while extending to the orbital apex.

Figure 4. Histopathological examination shows the diffuse proliferation of lymphoid cells (hematoxylin-eosin staining, ×100). Immunohistochemical staining demonstrates CD20 positive small-sized lymphocytes (brown staining).
residual tumors may remain after the treatment, clinicians should be aware of the location of lesions, histopathological findings and the clinical stage of the disease. Long-term follow-up is recommended not to miss any recurrence out.

Disclosure of interest
The author declares that he has no conflict of interest concerning this article.

References

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