Peripheral T-cell lymphoma of the eyelid

Makoto Ishikawa
Hiroshi Watabe
Masahiro Hayakawa
Takeshi Yoshitomi

Department of Ophthalmology, Akita University Faculty of Medicine, Akita, Japan

Purpose: To report a case of a 25-year-old woman with previously treated peripheral T-cell lymphoma (PTCL) presenting with a recurrent lower eyelid lesion.

Patients and method: Case report.

Results: A 25-year-old young woman with previously treated PTCL noted an induration in the skin of her left lower eyelid. Upon diagnosis of a chalazion, antibiotic eye drops and ointments as well as steroid eye drops were administered. However, the condition worsened rapidly and swelling of the lower eyelid became remarkable. An excisional biopsy revealed that the palpebral lesion was diagnosed as recurrence of PTCL. Electron irradiation was applied as a radical treatment, and the prognosis was satisfactory without recurrence at 10 months after the last irradiation.

Conclusions: Although lymphoma isolated to the ocular adnexa is rare, it should always be included in the differential diagnosis of any patient presenting with progressive swelling of the eyelid or ocular region.

Keywords: peripheral T-cell lymphoma, chalazion, eyelid

Introduction
Peripheral T-cell lymphoma (PTCL) represents a broad spectrum of disease with several different manifestations. PTCL often occurs in the extremities, but rarely in the eyelid. Periocular involvement by PTCL usually occurs in the setting of the Sézary syndrome or mycosis fungoides, which is a variant of PTCL.1–4 We report a case of a 25-year-old young woman with previously treated PTCL presenting with a recurrent lower eyelid lesion.

Case report
The patient’s history dates back to 2006 when she had multiple skin lesions that were diagnosed as PTCL and subsequently excised. Submandibular and parotid lymph nodes were palpable, but there was no swelling of the liver or spleen. Blood tests were negative for adult T-cell leukemia antibody and human immunodeficiency virus (HIV) antibody and soluble interleukin-2 receptor was within normal range. No other lesion was detected by gallium scintigraphy or systemic computed tomography. A bone marrow trephine biopsy did not show the involvement of lymphoma. Immunostaining revealed the cells to be positive for CD3 and CD5, but negative for CD30, CD10, and CD75. Based on the findings, the patient was classified as PTCL Stage IVB according to the new World Health Organization classification.5 As the patient was chemorefractory...
against initial treatment with combined cyclophosphamide, deoxorubicin, vincristine, and prednisone, she underwent allogenic stem cell transplantation from a matched non-
sibling donor in March 2007 after salvage treatment with
cyclophosphamide, cytosine arabinoside, etoposide, and
dexamethasone. Transplant-conditioning therapy was cyclo-
phosphamide, 2900 mg/day for three days, plus total body
irradiation for three days (total 12 Gy). Graft-versus-host
disease prophylaxis was provided with cyclosporine A and
prednisolone. Cyclosporin levels were maintained between
300 and 400 µg/L until tapering was initiated. Initial dose
of prednisolone was 17.5 mg per oral.

In January 2008, the patient presented to our department
to check for any ocular manifestations of the graft-versus-host
disease. At that time, significant superficial punctate keratopathy was noted in both eyes. Systemic skin and eyelids
were macroscopically normal. Systemic lymph nodes were
not palpable, and there was no swelling of the liver or spleen.
The superficial punctate keratopathy significantly improved
with eyedrops containing hyaluronan.

On July 11, 2008, the patient noted an induration in the
skin of her left lower eyelid (Figures 1A, B). Conjunctival
congestion and pain were present. Upon diagnosis of an acute
chalazion, conservative management was used. However, the
condition worsened rapidly, and swelling of the lower eyelid
became remarkable (Figures 1C, D).

On August 25, 2009, the patient underwent an
excisional biopsy of the palpebral lesion after informed
consent. A retention cyst in chalazion was not detected during
the intraoperative microscopic examination. Pathological
examination demonstrated that medium-sized lymphoma
cells with round nuclei (Figures 2A, B) had infiltrated
subcutaneously. Immunostaining revealed the cells to be
positive for CD3 (Figures 2C). Considering these results, the
palpebral lesion was diagnosed as recurrence of PTCL.

As the lesion was limited to the left lower eyelid, 25.2 Gy
electron irradiation was applied as a radical treatment. Globe
shielding was used to help prevent radiation retinopathy. The
eyelid tumor was dramatically improved after irradiation
(Figures 2D, 2E). The prognosis was satisfactory without
recurrence at 10 months after the last irradiation.

Discussion

Ocular manifestations of PTCL can include both intraocular
and extraocular conditions. Cook and colleagues reported
that eyelid ectropion was the most common findings in their
patients with PTCL, though other ocular manifestations,
including eyelid thickening or edema, placoid tumor, tight
skin, blepharitis, and corneal abnormalities, were also found.
Stenson and Ramsay found eyelid tumors in eight of 30
consecutive patients with mycosis fungoides diagnosed by
biopsy. Isolated PTCL of the eyelid is rare. Gilbertson
and colleagues reported a case of a 38-year-old acquired
immunodeficiency syndrome (AIDS) patient with isolated
eyelid PTCL, and found only eight previous reports of
isolated PTCL, most diagnosed as mycosis fungoides. In this
type of lymphoma, relapses are common and can produce
an indolent course. Kirsch and colleagues reported a patient
with multiple recurrences of systemic T-cell lymphoma after
initial presentation with only an eyelid mass that was treated
with systemic chemotherapy and local radiation.

The present case was a single episode of lymphoma of
the eyelid with clinical symptoms resembling a chalazion.
However, an excisional biopsy of the current lesion con-
firmed the recurrence of the actual tumor and so ruled out
a chalazion. The current case still requires close follow-up
observation, particularly of the skin around the lesion. When
radiation therapy is used for eyelid lesions, great care must
be taken with ocular side effects, including xerophthalmia
and radiation retinopathy. Although lymphoma isolated to
the ocular adnexa is rare, it should always be included in the
differential diagnosis of any patient presenting with progres-
sive swelling of the eyelid or ocular region, particularly with
a history of lymphoma.
Disclosures
The authors report no conflicts of interest in this work.

References

Figure 2 A) Light micrograph of hematoxylin and eosin stained eyelid biopsy specimen. Lymphoma cells were infiltrated into the subcutaneous tissue. P, palpebral skin; Bar = 200 µm. B) High magnification of the lymphoma cells infiltrated in the orbicular muscles (arrows). Bar = 100 µm. C) Light micrograph of eyelid biopsy specimen immunostained with CD3 antibody. CD3 positive lymphoma cells were distributed in the subcutaneous tissue. Bar = 100 µm. Photographs of eyelids before D) and after irradiation E) The eyelid tumor was dramatically improved after irradiation.