Bilateral primary intraocular lymphoma, a case report

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Introduction

Primary intraocular lymphoma can occur as an isolated eye tumor or involve both the globe and the central nervous system [1]. Secondary lymphoma may also involve the eye as a result of a systemic tumor [2–5]. The syndrome was first recognized in 1951 by Cooper and Riker [6], and numerous cases have been reported since [1–27]. The initial term for this non-Hodgkin’s lymphoma was reticulum cell sarcoma [1-27]. Rappaport reclassified these large cell tumors as histiocytic lymphomas [28]. Recent data demonstrate that these tumors are predominantly large cell lymphomas and consist of either malignant lymphoma or their precursors [29–31]. Most commonly, this malignancy occurs in older patients; however, patients as young as 27 years of age have been described [32]. Central nervous system involvement has been reported in approximately 75% of cases and is the usual cause of death [2,3,13,18,32]. A case of lymphocytic lymphomas of both eyes is reported; no such cases have been reported in Thailand before.

Case report

An 88-year-old Thai man was referred to Maharaj Nakorn Chiang Mai University Hospital 23 April 1990. His past history included chronic uveitis of unknown etiology in both eyes, 4 months before, which received topical and systemic steroid therapy, but the patient was lost to follow-up. At this time, he presented with visual loss in the left eye progressing to blindness over a 3-month period with progressive exophthalmos and gradually decreased vision in the right eye.

Physical examination revealed an old thin man (Fig. 1), with body weight of 28.7 kg, blood pressure 120/70 mmHg, heart rate 70/min.

The eye examination revealed a mass in the left orbit size of approximately 6 × 8 cm. The left eye was in the centre of the mass and was phthisis bulbi (Fig. 2). The right eye was slightly proptosis, with best corrected visual acuity of 6/60; intraocular pressure was 10 mmHg by applanation tonometer. Slit lamp examination of the right eye showed minimal anterior chamber cells with moderate cells in the vitreous cavity. The lens showed mild nuclear sclerosis and the fundus was unremarkable.

Initial laboratory data showed a white blood cell count of 6500/mm³, with 76% neutrophils, 20% lymphocytes and 4% eosinophils. The hemoglobin was 12.2g/dl the hematocrit was 39% and the platelets were adequate. Urinalysis was unremarkable.

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Other laboratory findings included: creatinine 1.1 mg/dl, total protein 7.4 gm/dl, albumin 3.7 gm/dl, globulin 3.7 gm/dl alkaline phosphatase 122 U/L, AST (GOT) 31 U/L, ALT (GPT) 22 U/L, cholesterol 212 mg/dl, direct bilirubin 0.06 mg/dl, total bilirubin 0.33 mg/dl, calcium 8.9 mg/dl, inorganic P 4.4 mg/dl and uric acid 5.7 mg/dl.

Neurologic examination showed no abnormality. CSF cytologic examination was negative. Electrocardiogram showed poor R progression in V1-V5. Chest film revealed small calcific density in both lungs with thickening of the lung markings. No definite infiltration was seen (Fig. 3). Orbital roentgenograms showed a soft tissue mass overlying at the left orbital region and no evidence of calcific density or definite bony destruction. (Fig. 4).

Pre- and postcontrast enhancement scans of the orbit and brain were performed in axial and coronal sections of 5 mm. The study revealed a big mass in the left orbit with engulfment of the left globe. The left optic nerve and lateral rectus muscle were enlarged. There was a soft tissue mass in the lateral aspect of the right orbit with obliteration of the right lateral rectus muscle and right optic nerve (Fig. 5). Moderate homogenous enhancement was seen in these lesions. Proptosis of the right globe was also noted. No bone destruction was seen. Mild ventricular dilatation and prominent sulci were seen without definite intracranial lesion.
Fig. 3. Chest film of the patient, revealing small calcific density in both lungs. No definite infiltration was seen.

Fig. 4. Orbital film showing a soft tissue mass in left orbit and no evidence of bone destruction.

Fig. 5. CT scan of the orbit and brain revealing a big mass in the left orbit, enlarged left optic nerve and left lateral rectus muscle. There was soft tissue mass in the lateral aspect of right orbit with obliteration of the right lateral rectus muscle and right optic nerve.
The patient refused any diagnostic or therapeutic operation in his right eye; however, the incisional biopsy specimen from the mass of the left eye was performed under local anesthesia. Histopathology study revealed low grade malignant lymphoma, small lymphocytic (working formulation) or diffuse well-differentiated lymphocytic (Rappaport) (Fig. 6).

The immunologic results showed almost all tumor cells gave positive staining with anti-CLA (B and T lymphocyte marker), but negative with anti-S-100 (a melanoma marker); besides that, kappa to lambda staining showed reverse ratio which might be a reflection of abnormal clonal production.

Metastatic work-up including bone marrow aspiration and biopsy showed no evidence of extraocular lymphoma.

Orbital irradiation of 20,000 rads was delivered in 400-rad fractions. The mass responded well to local radiotherapy (Fig. 7). After treatment, the media of the right eye became clear, but vision did not improve because of a dense cataract. The patient refused to receive lens extraction of the right eye, was discharged from the hospital after admission for 2 months, and then lost to follow-up.

Discussion

Ocular lymphoma is a highly malignant multicentric form of non-Hodgkin’s lymphoma which most commonly involves the uvea, retina, vitreous and central nervous system [2]. Although relatively uncommon, this disease should be consid-
Fig. 7. External appearance of both eyes after radiotherapy, showing disappearance of proptosis in right eye and regression of mass in left eye.

ined in any adult patient with vitreitis or chorioretinitis of unknown etiology, particularly if the inflammation does not respond to the usual therapy [19]. Painless blurring of vision is the most frequent initial symptom of disease [3]. Posterior pole involvement includes vitreous cellular reaction, retinal infiltrates, chorioretinal lesions, and subretinal masses [19].

Vitrectomy with cytologic examination of cells could offer an opportunity for earlier diagnosis of intraocular lymphoma. However, it was demonstrated that there appeared to be cellular degradation when specimens were obtained with vitrectomy and aspiration rather than biopsy with a syringe and needle, and also, a single vitreous biopsy specimen was not always adequate to establish the diagnosis of this condition [33]. Another study has showed that transscleral choroidal biopsy offered a promising alternative diagnostic approach to lesions that had yielded negative vitreous biopsies [27].

The origin and derivation of the cells which produce intraocular lymphoma are not clear [31], but it was shown that these cells are derived from different lymphocytic populations which often admixed with normal inflammatory cells [33]. It was also shown that cytologic evaluation was more accurate than lymphocyte surface marker analysis to differentiate lymphoma from uveitis [33].

Radiation therapy causes effective amelioration of ocular symptoms [3,32]. The use of chemotherapy for ocular lymphoma is less clear. The experience of Char et al. [2] with three patients given combination chemotherapy, including intrathecal
treatment, was favorable. Baumann et al. [26] and Siegel et al. [32] successfully treated patients with intravenous high-dose cytosine arabinoside (Ara-C).

In summary, a case of bilateral primary intraocular lymphoma has been reported. Despite that this patient did not permit vitreous aspiration of the right eye or enucleation of the left eye, however, a history of chronic uveitis in people over 60, particularly not responding to the usual therapy heralded this malignancy disease. The incisional biopsy specimen from the mass of the left eye and the vitreitis in the right eye that disappeared after radiotherapy strongly suggested the disease. This diagnosis should be considered in all adults with posterior uveitis, vitreitis or chorioretinitis of unknown etiology, particularly when seen in association with neurologic symptoms [19]. The importance of proper diagnosis lies in the radiosensitivity of these tumors and the improvement in visual acuity and prolonged survival that frequently accompany therapy.

References