CASE REPORT

Relapsed Burkitt Lymphoma Presenting as an Isolated Infiltrative Optic Neuropathy

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ABSTRACT

We report a case of relapsed sporadic Burkitt lymphoma (BL) presenting as an isolated infiltrative optic neuropathy. This is a single-patient, retrospective case report of a patient seen by the ophthalmology service at our institution. To our knowledge, our case is the first to report isolated infiltrative optic neuropathy associated with sporadic BL as the sole manifestation of recurrence. The possibility of disease relapse should be considered for patients with a history of lymphoreticular malignancy who present with acute visual symptoms suggestive of optic neuropathy.

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Burkitt lymphoma; non-Hodgkin lymphoma; ocular lymphoma; optic neuropathy

Introduction

Burkitt lymphoma (BL) is a highly aggressive, mature B-cell non-Hodgkin lymphoma (NHL) that occurs in three major variants (sporadic, endemic, and immunodeficiency-related). The sporadic variant occurs in non-endemic areas where it typically manifests as an abdominal tumour with bone marrow involvement, but rarely occurs with ocular or orbital involvement.^{1,2} We report a case of relapsed sporadic BL presenting as an isolated infiltrative optic neuropathy.

Case report

A 59-year-old Caucasian female presented with floaters and a rapidly progressing central scotoma in the left eye over the preceding 2 weeks. Her past medical history was significant for sporadic BL involving the bone marrow and spleen diagnosed 6 months prior. Staging at that time revealed no central nervous system (CNS) or ocular involvement. She was human immunodeficiency virus (HIV) and Epstein-Barr virus (EBV) negative. The patient underwent one cycle of chemotherapy with a standard CODOX-M/

IVAC regimen including cyclophosphamide, vincristine, doxorubicin, methotrexate, ifosfamide, etoposide, and cytarabine. Bone marrow biopsies and staging workup 2 months later revealed no evidence of residual disease.

On examination, her visual acuity was 20/20 in the right eye and counting fingers at 3 feet in the left eye, with a brisk relative afferent pupillary defect of the left eye. Anterior segment examination was unremarkable. Posterior segment examination of the right eye revealed equatorial drusen. The left eye revealed a 3+ vitritis with strands of inflammatory debris extending from the disc into the vitreous. The entire left disc was elevated and infiltrated with white material and surrounded with haemorrhage (Figure 1A). There were equatorial drusen, but no peripheral lesions. Fluorescein angiography of the left eye showed staining and late leakage of the disc with pooling into the macula (Figure 1B). Spectral-domain optical coherence tomography demonstrated sub-retinal fluid surrounding the disc extending into the fovea (Figure 1C).

Magnetic resonance imaging (MRI) of the brain and orbits without contrast revealed a focal hypointense T2 signal within the posterior aspect of the left globe associated with restricted diffusion (Figure 2A)

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Figure 1. (A) Colour fundus photograph montage of the left eye shows a vitritis with strands of inflammatory debris extending from the disc into the vitreous. The entire left disc is elevated and infiltrated with white material and surrounded by haemorrhage. (B) Fundus fluorescein angiogram montage of the left eye shows staining and late leakage of the disc with pooling of dye into the macula. (C) Spectral-domain optical coherence tomography shows sub-retinal fluid surrounding the disc and extending into the fovea.

and non-specific soft tissue density at level of posterior globe/optic nerve insertion (Figure 2B), which was not seen on MRI performed 6 months prior. The patient underwent diagnostic vitrectomy of the left eye. Flow-cytometric analysis of the vitreous fluid demonstrated kappa-restricted CD10+, CD20+, and CD19+ B-cell populations. Cytopathology showed Burkitt cells (Figure 2C). All studies were consistent with relapsed BL. Repeat bone marrow biopsy, lumbar puncture with cerebrospinal fluid (CSF) analysis, and positron emission tomography-computed tomography (PET-CT) revealed no evidence of systemic recurrence outside of the left eye. No further treatment was initiated because the patient developed septic shock in the setting of pre-existing multi-organ failure. The patient expired 3 days later and no post-mortem examination was indicated.

Discussion

Optic nerve infiltration in NHL is rare and is typically accompanied by active CNS involvement or

pathologic bone marrow.² Isolated optic nerve infiltration, as the sole manifestation of relapsed systemic NHL, is very rare, with only three cases reported in the literature. In 1986, Kay reported the first case of isolated optic neuropathy occurring in a patient with NHL remission.³ In 2002, Lee et al. reported a 56-year-old male with acquired immunodeficiency syndrome (AIDS) and regressed Burkitt-like, EBV-positive systemic NHL, who developed lymphomatous infiltration of the optic nerve and occlusion of the central retinal vein in both eyes.⁴ More recently, Kim et al. reported a 2-year-old female with diffuse large B-cell lymphoma, which primarily occurred as a suprapubic mass and relapsed in the form of isolated optic nerve infiltration.⁵

To our knowledge, our case is the first to report isolated infiltrative optic neuropathy associated with sporadic BL as the sole manifestation of recurrence. The possibility of disease relapse should be considered for patients with a history of lymphoreticular malignancy who present with acute visual symptoms suggestive of optic neuropathy.

Figure 2. (A) MRI of the brain and orbits without contrast revealing a focal, mildly lobulated hypointense T2 signal within the posterior aspect of the left globe, near the insertion site of the left optic nerve, measuring approximately $3 \times 5 \times 6$ mm, and associated with restricted diffusion. (B) MRI of the brain and orbits shows a non-specific soft tissue density at level of posterior globe/optic nerve insertion. (C) Cytopathology of vitreous fluid from the left eye shows Burkitt cells identified by open chromatin, cleaved nuclei, and a thin rim of blue-grey cytoplasm with cytoplasmic vacuoles.

Declaration of interest

The authors report no conflicts of interest. The authors alone are responsible for the content and writing of the paper.

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