CASE REPORT

Rapid Orbital and Optic Nerve Infiltration In a Lady With Aggressive Diffuse Large B-cell Lymphoma

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ABSTRACT

Hyperkalaemia, an excessive level of potassium in the blood, can arise from increased intake, reduced renal clearance, or a combination of both. Severe hyperkalaemia is a medical emergency with life-threatening implications, particularly when diagnosis and treatment are delayed. Numerous factors can contribute to hyperkalaemia, and although it is relatively rare, consuming dokong fruit in the presence of impaired kidney function may be a contributing factor.This article presents a case study involving a woman who experienced a life-threatening episode of hyperkalaemia following the consumption of a substantial amount of dokong fruit. The incident was accompanied by a hypertensive crisis, leading to acute pulmonary oedema, and occurred in the context of underlying Chronic Kidney Disease (CKD). The patient received successful treatment and was discharged in good condition. This case highlights an unusual but important dietary trigger of life-threatening hyperkalaemia, emphasizing the need for early recognition, prompt management, dietary counseling, and timely planning for dialysis in patients with advanced CKD.

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INTRODUCTION

Lymphoma is a malignancy arising from lymphocytes, subdivided into Hodgkin or Non-Hodgkin lymphoma (NHL). Ocular manifestations are seen in individuals diagnosed with NHL, but optic nerve involvement is uncommon.(1) Patients with Diffuse Large B-cell lymphoma (DLBCL) have a lower risk of central nervous system (CNS) metastasis, around 10%; and only 5% risk of optic nerve infiltration.(1) The recognition of malignant encroachment of the optic nerve is challenging due to a similar presentation to other optic nerve pathologies, namely compressive, inflammatory, infectious, ischemic, or toxic-metabolic. We report a case of aggressive DLBCL with involvement of the brain, orbit, and optic nerve. The patient was initially treated with Sulfamethoxazole/trimethoprim (Bactrim) for right Bell's palsy with Jensen papillitis secondary to Presumed Ocular Toxoplasmosis due to her similar presentation to infectious cause and her exposure to cats at home. The new onset of Bell's palsy raised the suspicion of central nervous system (CNS) Lymphoma.

CASE REPORT

A 50-year-old lady, newly diagnosed with Diffuse Large B-Cells Lymphoma, was on an ongoing Rituximab Cyclophosphamide, Hydroxydaunomycin, Oncovin and Prednisolone (R-CHOP) regime. She presented with floaters for three weeks duration and painless blurring of vision over her right eye. Both symptoms progressively worsened, and thus referred to the ophthalmology department. However, she denied vomiting, headache, or any limb weakness. Of note, it was associated with right-sided facial asymmetry. The only positive social history was that she had multiple pet cats at home.

Visual acuity of the right was 6/9 and 6/6 over the left. Pupillary examination showed negative relative afferent

pupillary defect (RAPD). Her pupils were regular size. Lids were normal, and no ptosis was noted. A dilated fundoscopy examination revealed a diffusely edematous optic disc with an inferior peripapillary subretinal mass resembling a granuloma around two discs diameter in size (Figure 1a). Her Left eye was of normal findings. Figure 1 shows the sequential fundus photo showing the progression of ocular manifestations of her disease.

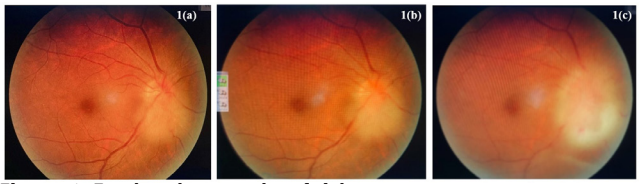

Figure. 1: Fundus photography of right eye:
1(a) at presentation; 1(b) one month later ;1(c)Two months later

Magnetic Resonance Imaging (MRI) scan of the brain and orbit showed no evidence of central nervous system involvement. Fundus Fluorescein Angiogram showed the presence of a hot disc with no leakage(Figure 2). Given the preserved optic nerve function and the presence of hyperfluorescent optic disc on FFA, Jensen papillitis was a more probable diagnosis than compressive optic neuropathy. She was treated for right Bell's palsy with Jensen papillitis secondary to Presumed Ocular Toxoplasmosis with negative serology. Although her blood parameters showed negative Toxoplasmosis serology, Oral Bactrim 960mg bd was commenced, due to her exposure to domestic cats.

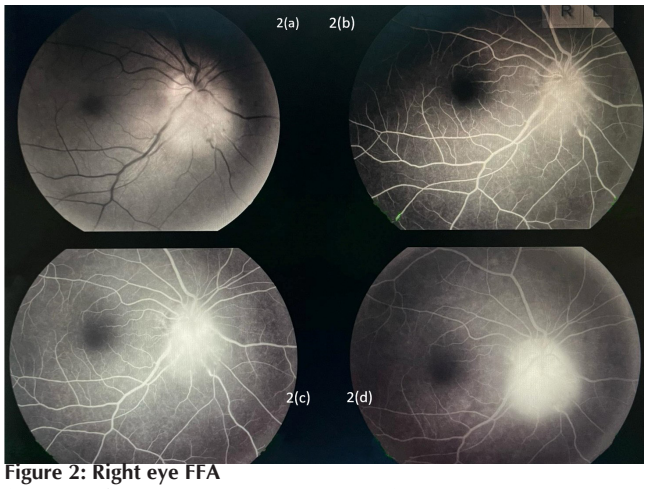

Figure 2: Right eye FFA
(a): Arterial;
(b): Early venous;
(c): Late Venous;
(d): Late phase

Improvement of floaters was seen with oral Bactrim and concurrent RCHOP chemotherapy regimen. A multidisciplinary approach from medical and ophthalmology in deciding further management for this patient. Lumbar puncture with intrathecal methotrexate injection was performed in suspicion of CNS Lymphoma due to the new onset of Bell's palsy. However, cerebrospinal fluid was sent and analyzed as negative for malignancy.

Five weeks later, she developed right axial proptosis and ptosis affecting the visual axis of her right eye. Her right visual deterioration to counting fingers with positive RAPD, complete ophthalmoplegia. Anterior segment examination showed exposure keratopathy and marked worsening of optic disc swelling. Contrast Computed Tomography (CT) imaging revealed an intracranial mass extending to bilateral optic nerves, intraconal and cavernous sinus space. (Figure 3) shows the sequential anterior segment photos of the patient at earlier and later presentations.

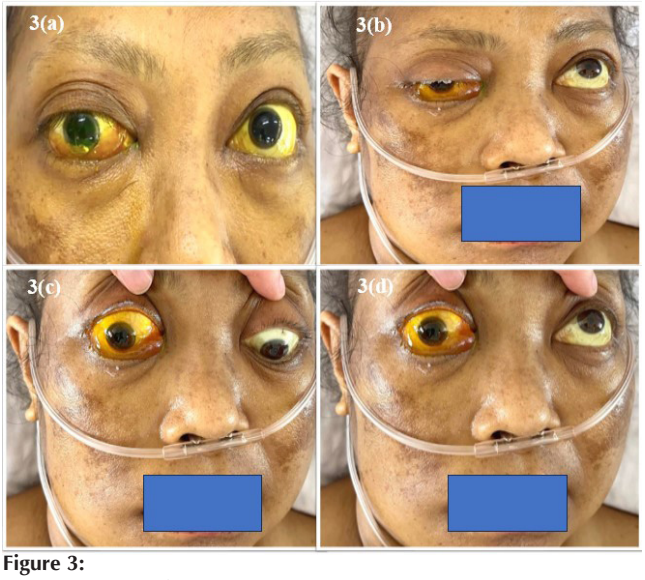

Figure 3:
3(a): Patient's earlier presentation
3(b): Patient's presentation 5 months later: Right eye proptosis, chemosis & Bell's palsy
3(c) Downward gaze
3(d) Upward gaze

Her condition worsened rapidly with systemic metastases. The patient and family subsequently opted for palliative treatment, and she succumbed to her illness shortly after.

DISCUSSION

In non-Hodgkin lymphoma (NHL), several different mechanisms of optic nerve involvement, including the compressive effect from orbital lymphoma and central artery occlusion secondary to hyperviscosity. In comparison, primary optic nerve malignancy is less commonly seen as secondary infiltration from systemic disease(1).

Identifying optic nerve involvement secondary to systemic illness, in this case, lymphoma, is a challenging dilemma for physicians in excluding other etiologies of neurologic emergencies, such as infective, inflammatory, demyelinating, and ischemic optic neuropathies. The correct diagnosis of the disease has serious implications for patients' management, as different treatments may be initiated for the etiologies.

Suspicion of ocular toxoplasmosis with Jensen papillitis was raised based on the presence of a whitish inflammatory lesion on the optic nerve, typically seen in toxoplasmosis-related optic neuropathy, and the patient's high-risk exposure to Toxoplasma gondii. However, certain features were against the diagnosis of ocular toxoplasmosis, including the absence of hyperemic optic disc and lack of peripapillary retinal haemorrhages.(2) The patient further showed only a minimal response to systemic Bactrim, with mild reduction of blurry vision and floaters.

Suspicion of central nervous system metastasis from lymphoma complicated with Bell's palsy, a minority of 4% individuals with Non-Hodgkin Lymphoma (NHL) suffered CNS lymphomatous meningitis with multiple cranial nerve pathologies, primarily facial nerve, oculomotor and followed by trigeminal nerves, as asserted by Van Horn A et al.(3) Thus, lumbar puncture with intrathecal methotrexate was given.

Proposed theories for metastatic spread of the optic nerve include dissemination via the bloodstream, perineural or cerebrospinal fluid, and direct tumour cell invasion. Lee LC et al further reinforced the theory that meninges, cranial, and spinal nerves may be encroached upon by malignant cells or related to orbital lymphomatosis with leptomeningeal involvement.(4) A classification of optic nerve lymphoma was put forward by Kim JL et al (5): primary involvement of optic nerve; CNS or systemic disease complicated with optic nerve manifestation; optic nerve invasion in primary intraocular lymphoma. The case in this literature is categorized into CNS disease complicated with optic nerve manifestation of vision reduction, and concurrent CNS neuroimaging findings. Patients who have shown established CNS involvement of lymphoma with optic nerve involvement may present with decreased vision or diplopia.(3) Among patients with lymphoma, the varied onset of visual symptoms, such as blurry vision or diplopia, ranged from one month after treatment initiation to nearly a decade post-diagnosis, as reported in a study in Georgia.(5)

Confirmation of the lymphomatous invasion of the optic nerve via optic nerve biopsy could be replaced with CSF samples analysis, as asserted by Kim JL et al(5). However, due to the possibility of false negative results, several lumbar punctures should be done for those patients with likely lymphomatous CNS infiltration.

There is no established standard treatment for lymphoma with CNS involvement. The treatment for each patient should be tailored to the severity and staging of systemic lymphoma. Generally, a combination of intrathecal and systemic chemotherapy would be the therapy directed to patients with CNS involvement.

CONCLUSION

While uncommon, optic nerve, orbital involvement in DLBCL should be considered in patients presenting with optic disc swelling. It poses a diagnosis dilemma between optic nerve neoplastic infiltrations and infective, inflammatory, and demyelinating optic nerve conditions.

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