CASE REPORT

A Unique Presentation of Diffuse Large B-Cell Lymphoma Involving the Central Nervous System

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ABSTRACT

Diffuse large B-cell lymphoma is the most common type of non-Hodgkin lymphoma, but ophthalmic presentations of this type of lymphoma constitute a small subset of these cases. An even smaller subset of cases involves the central nervous system at the time of presentation. The authors report a unique case of diffuse large B-cell lymphoma with central nervous system involvement initially presenting with ophthalmic manifestations and altered mental status.

Keywords: Altered mental status, diffuse large B-cell lymphoma, orbital cellulitis

INTRODUCTION

Diffuse large B-cell lymphoma (DLBCL) is the most common type of non-Hodgkin lymphoma, constituting about 25–31% of all lymphoma cases.^{1,2} Of the lymphomas that are found in the ocular adnexal tissue, DLBCL makes up only 8–13%³ and most commonly presents with swelling and diplopia.⁴ Cases of DLBCL masquerading as orbital cellulitis have previously been described in the literature.^{5,6} We present a unique case of DLBCL involving the central nervous system (CNS) presenting as orbital cellulitis and altered mental status.

CASE

A 59-year-old female was transferred from an outside hospital with progressively worsening left periorbital swelling. She reported that she may have scratched her eyelid recently but denied any other trauma. Her past medical history included hypertension, hyperlipidaemia, and a previous hip fracture. Her past ocular history was unremarkable. Her medications included ibuprofen, hydrochlorothiazide, enalapril, aspirin, simvastatin, fluoxetine, methadone, and morphine. She had no known drug allergies.

Her family history was only remarkable for type 2 diabetes mellitus. Her social history was positive for tobacco use and a history of intravenous heroin use. Her review of systems was positive for foot numbness.

At the time of presentation, the examination was limited due to her waxing and waning mental status. Her visual acuity was at least counting fingers at 6 feet in the right eye and counting fingers at 5 feet in the left eye. Her pupils were equal at 2 mm but both were minimally reactive. Her extraocular motility showed full motility of the right eye and mild limitation to all fields of gaze of the left eye. There was mild resistance to retropulsion of the left eye. Her external examination showed significant oedema and erythema to the left lower lid. The patient's initial computed tomography (CT) scan from the outside hospital showed fat stranding with a possible mass within the left orbit and mild mucosal thickening of the paranasal sinuses. The patient was tentatively diagnosed with orbital cellulitis and thus admitted to the medicine service for intravenous vancomycin and piperacillin/tazobactam.

The patient's altered mental status continued throughout her hospital stay. The patient's initial infectious work up (blood cultures, urinalysis, urine culture, orbital culture, human immunodeficiency

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virus [HIV]) was negative and she remained afebrile with a normal white blood cell count. The patient's methadone was stopped to rule out the possibility of oversedation from narcotics. An electroencephalogram showed moderate to marked degree of diffuse slow-wave abnormality indicating a significant generalized cerebral involvement.

A repeat CT of orbit, sella, and ear with and without contrast showed enlargement of the mass in the left orbit (Figure 1). CT of the brain was normal, without any haemorrhage or infarction. Without improvement on broad-spectrum antibiotics and a trial of steroids, the patient was taken to the operating room for an anterior orbitotomy. In the operating room, a mass without any frank purulence was found in the anterior aspect of the left lower eyelid within the orbicularis muscle. Several biopsies were sent for pathological analysis, and swabs of the orbit during surgery were sent for culture. Biopsies were consistent with non-germinal centre type of DLBCL (CD10–, MUM1+, BCL-6–).

FIGURE 1 CT of the orbits (coronal view) showing left inferior orbital mass.

Magnetic resonance imaging (MRI) of the brain showed infiltration involving the parietal gyri mainly on the left, basal nuclei of the diencephalon, brainstem, and cerebellum (Figure 2). These findings on MRI were concerning for CNS involvement of the DLBCL. Flow cytometry of cerebrospinal fluid from a lumbar puncture confirmed CNS involvement of DLBCL.

CT of the chest, abdomen, and pelvis with contrast obtained for staging showed mild hepatomegaly with indeterminate hypodense splenic lesion and permeative osseous changes of the left hip with compression fracture of the left acetabular roof, both suspicious for lymphomatous infiltration. A bone marrow biopsy showed a small 2% population of DLBCL on flow cytometry. In addition, a left axillary lymph node was biopsied and pathology was consistent with DLBCL.

After approximately 1 month of systemic and intrathecal chemotherapy with little improvement, the patient's family decided upon hospice care.

DISCUSSION

In summary, the case presented here shows an unusual presentation of DLBCL involving the CNS. The incidence of CNS involvement of DLBCL at the time of initial diagnosis is reported to be approximately 1%.⁷ An ophthalmic presentation of DLBCL with CNS involvement is even more rare. The only case in the literature of an ophthalmic manifestation of DLBCL involving the CNS is of a patient with DLBCL in remission presenting with retinal findings as the first sign of CNS relapse.⁸ However, our case is unique because it displays an orbital process with altered mental status as the first presentation of CNS involving DLBCL.

Lymphoma involving the orbit can have various clinical presentations. DLBCL without CNS

FIGURE 2 MRI of the brain showing infiltration involving the parietal gyri mainly on the left (A), the basal nuclei of the diencephalon (B), and the brainstem and cerebellum (C). Fluid attenuated inversion recovery sequence.

involvement has been reported to masquerade as orbital cellulitis. Additionally, other types of lymphoma such as T-cell, natural killer T-cell, and plasmablastic lymphoma presenting with orbital signs can be found in the literature.^{9–11} The characteristic signs and symptoms of orbital cellulitis, which included proptosis, chemosis, eyelid swelling and erythema, decreased vision, and pain are very similar to those found with any orbital process. Thus, a high level of suspicion with a broad differential diagnosis is warranted. Furthermore, an orbital process with altered mental status is a potentially serious condition that warrants a thorough evaluation to rule out sequelae of orbital cellulitis (i.e., meningitis and brain abscesses) and other disease processes involving the CNS as displayed in the case above.

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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