Case Report

Vision preserving role of radiotherapy in orbital plasmablastic lymphoma

Aiswarya Raveendran[1], A Ravichandran[2], Christopher John[3], K Satish Srinivas[4]

From [1]Junior Resident, [3]Associate Professor, [4]Professor and Head, Department of Radiation Oncology, [2]Associate Professor, Department of Medical Oncology, Sri Ramachandra Medical College, Chennai, Tamil Nadu, India

ABSTRACT

An uncommon but aggressive variant of diffuse large B-cell lymphoma is plasmablastic lymphoma (PBL). Although they primarily involve the gastrointestinal tract, lymph nodes, and oral mucosa of immunocompromised males, orbital presentation is rare. In this article, we report a case of an elderly, immunocompetent gentleman presenting with progressive proptosis, ptosis, and diminished vision of the right eye with a palpable mass in the right upper eyelid. Imaging and biopsy confirmed the rare diagnosis of orbital PBL. After obtaining institutional tumor board concurrence, the patient was delivered 45 Gy external beam radiation therapy followed by six cycles of chemotherapy. Post-therapy, he is in remission with improvement in vision and is currently on maintenance chemotherapy.

Key words: Chemotherapy, Orbital lymphoma, Radiotherapy, Vision

Plasmablastic lymphomas (PBLs) are a belligerent but uncommon variant of diffuse large B-cell lymphoma, accounting for about 1–2% of all lymphomas. The characteristics of PBL cells are the presence of B immunoblast tumor cells which exhibit positive immunohistochemistry markers as that of plasma cell lineage with nil or diminished B-cell markers [1,2]. PBL was first described and is still commonly diagnosed in immunocompromised males [2]. Orbital presentations of PBL are extremely rare and with scarcely any published literature references [3]. PBLs similarity to plasma cell myeloma makes it a challenging diagnostic for the pathologists, at the same time its recurrent nature and poor survival rates pose a trial to most clinicians [1,4].

Here, we are reporting a case of Orbital PBL in an immunocompetent elderly male.

CASE REPORT

A 75-year-old gentleman was evaluated in the Department of Ophthalmology for complaints of progressive proptosis, ptosis, and diminished vision in the right eye for 3 months (Fig. 1a).

General examination including vitals were normal. Upon evaluation, a non-tender, firm-to-hard mass was palpable within the substance of the right upper eyelid, with no involvement of the skin or palpebral conjunctiva. His best corrected visual acuity (VA) was observed to be hand movements close to the face. Systemic examination including cardiothoracic, perabdominal, and the remainder of the neurological examination was normal.

Evaluation with magnetic resonance imaging (MRI) Orbit revealed an ill-defined T1 isointense 3.3 cm × 2 cm lesion showing diffusion restriction in the right lateral peri-ocular region, molding the temporal surface of the globe from 6 to 2 o'clock, involving the lacrimal gland and lateral rectus muscle. The lesion showed heterogenous enhancement on contrast administration and was also associated with retinal detachment (Fig. 2). Biopsy of the lesion showed poorly differentiated malignant round cells arranged in a trabecular pattern (Fig. 3). Immunohistochemistry was done, which showed positivity for CD45, CD138, Kappa, CD20, and CD3 paving the way to the diagnosis of PBL. Bone marrow biopsy showed reactive marrow and the immunofixation study was negative for monoclonal gammopathy, thus ruling out multiple myeloma. The serological evaluation confirmed the immunocompetent status of the patient. Staging positron emission tomography with computed tomography (PET-CT) revealed a well-defined F-18 fluorodeoxyglucose (FDG) avid enhancing 3.8 cm × 2.8 cm × 3.1 cm lobulated lesion with standardized uptake value max −20.34 in the superior and lateral aspect of the right orbit, causing inferomedial displacement of the right globe. The right lacrimal gland was not separately visualized. The lesion was abutting the skin over the upper eyelid, infiltrating the lateral rectus muscle, with no significant perilesional fat stranding or any obvious bony erosions (Fig. 4). There were no other FDG avid lesions anywhere else in the body.

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Correspondence to: Dr. K. Satish Srinivas, Department of Radiation Oncology, F0, Sri Ramachandra Medical College, Porur - 600116, Chennai, Tamil Nadu, India. E-mail: dr_satishsrinivas@sriramachandra.edu.in

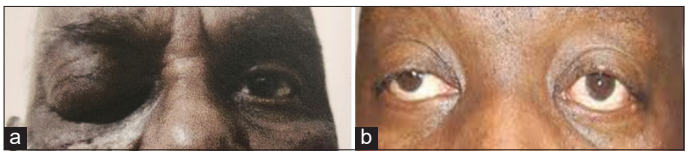

Figure 1: (a) Initial clinical presentation showing ptosis and increased thickness of right eyelid; (b) Clinical picture post-chemotherapy

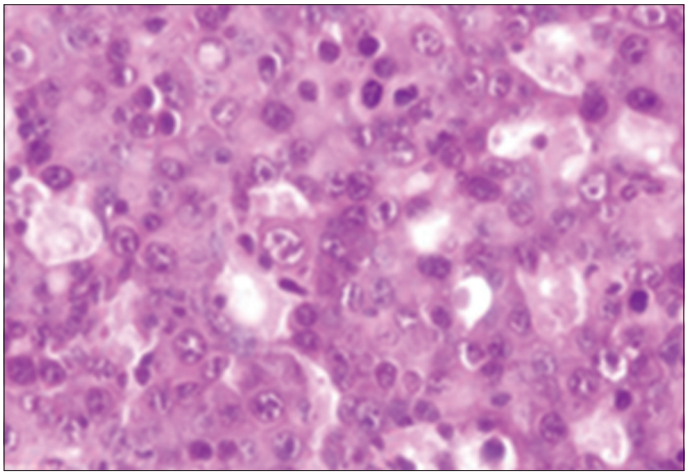

Figure 2: Poorly differentiated malignant round cells arranged in a trabecular pattern

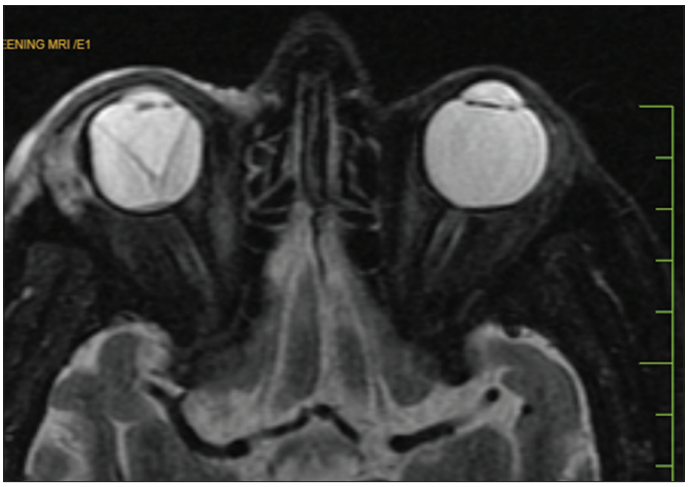

Figure 3: Initial magnetic resonance imaging orbit showing an ill-defined lesion in the right lateral peri-ocular region, molding the temporal surface of the globe from 6 to 2 o'clock, involving lacrimal gland and lateral rectus muscle, associated with retinal detachment

The recommendation of the institutional multi-specialty tumor board after careful analysis of the case was to proceed with radiotherapy. The patient was given 36 Gray (Gy) external beam radiation therapy with photons in 25 fractions using the intensity-modulated radiotherapy technique to the right orbit, followed by a 9 Gy electron boost in 5 fractions over a period of 1 month (Fig. 5a and b). The patient then proceeded to receive six cycles of Rituximab, Cyclophosphamide, and Oncovin along with Prednisolone. Post-therapy clinical evaluation showed a significant decrease in the degree of proptosis with an improvement of corrected vision in his right eye to VA 6/60 (Fig. 1b). Response assessment PET-CT showed no residual uptake in the right orbit. The patient was followed up every 3 months clinically, every 6 months with MRI, and annually with PET-CT. The patient is currently on maintenance Rituximab at 17 months post-radiotherapy, in clinical and radiological remission.

DISCUSSION

PBL although aggressive, is a rare and aberrant form of Diffuse Large B-cell Lymphoma. Although reports and published literature are emerging on PBL, its exact etiology and natural history still remain unclear. Plasmablasts resemble B-cell morphologically while exhibiting the immunophenotype of plasma cells. It is a challenging clinical diagnosis to clinch, especially in an immunocompetent individual. A strong immunoreactivity to CD138, VS38c with a weak/negative CD20, CD79a should guide the clinician to a diagnosis of PBL. With evolving genetic analysis and mutational analysis, MYC gene rearrangements have been implicated in immunocompetent individuals who develop PBL. On the other hand, human herpes virus - 8 and Epstein–Barr virus have been implicated among the etiological factors of PBL in immunocompromised individuals. Its brisk clinical progression, characterized by relapse while on treatment or untimely death makes both the patients and the clinician wary.

The overall survival of patients diagnosed with PBL, according to the available minimal literature, is less than a year, even in immunocompetent individuals [5,6]. Valenzuela *et al*. reported a case of aggressive PBL in the orbit of an immunocompetent individual, who quickly succumbed to the disease despite medical

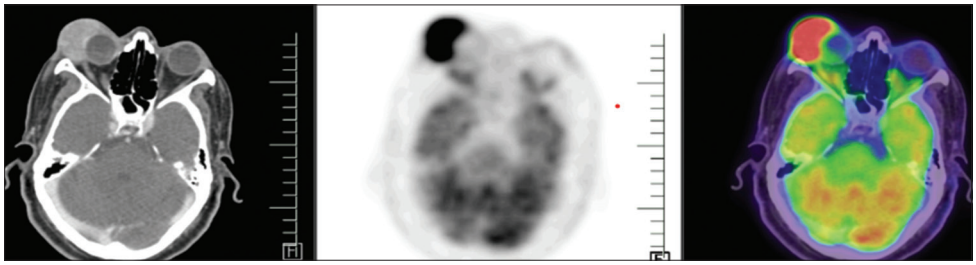

Figure 4: Staging positron emission tomography with computed tomography showing well-defined fluorodeoxyglucose avid enhancing 3.8 cm × 2.8cm × 3.1 cm lobulated lesion in the superior and lateral aspect of the right orbit, causing inferomedial displacement of the right globe and abutting the skin over the upper eyelid, infiltrating the lateral rectus muscle

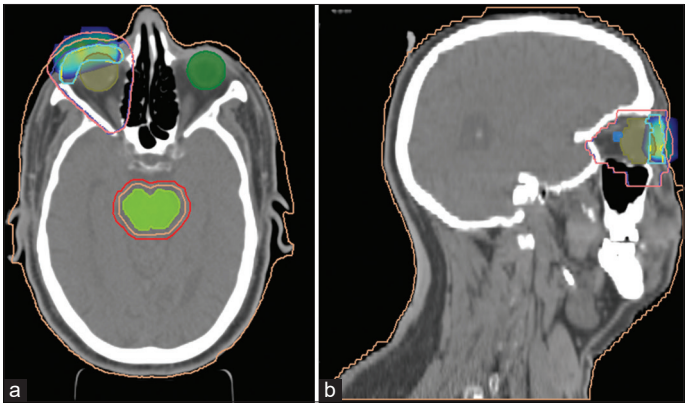

Figure 5: (a and b) Dose distribution of the radiation dose delivered over the right eyelid depicted using color wash

attention [7]. Even though most clinicians like Podder *et al*. follow CHOP-like regimens (with cyclophosphamide, oncovin, adriamycin, and prednisolone) for the management of PBL, like any other lymphoma, there are no available validated guidelines for the treatment of PBL [1,8].

Radiation therapy may be used as the primary modality of treatment in early-stage orbital lymphomas. The presence of a radiosensitive lens, lacrimal gland, and retina in the vicinity or within the target volume poses a challenge to the radiation oncologist. Electron beams can be used for superficial small lesions limited to the conjunctiva or lid. When a dose range of 25–30 Gy radiation is delivered as 1.8–2 Gy in every fraction for 5 days a week over 5 weeks, with meticulous planning and proper dose prescription, it can achieve up to 95% local control, while minimizing radiation toxicities [5,6,9].

CONCLUSION

Our patient has received a combination of radiotherapy and chemotherapy and has shown good clinical response to the treatment while maintaining good general health. Formulating a standard treatment protocol involving systemic therapy and local therapy, such as radiotherapy, and prognosticating these patients is prudent. Early diagnosis and individualization of therapeutic management may prove to be key in treating patients with Orbital PBL.

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