Complete Remission of Bilateral Palpebral Malt Lymphoma with Hypofractionated Radiotherapy: Case Report

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Abstract

To report the efficacy of External beam Radiotherapy (EBRT) as a sole treatment for MALT primary Orbital lymphoma, we describe a technique of hypofractionated radiotherapy who allowed a spectacular complete response with very low toxicity in a 64 years old patient presenting with a bilateral lower eyelids and orbital MALT lymphoma.

Keywords: Orbital • Malt lymphoma • Radiotherapy • Remission • Case report

Introduction

Orbital or ocular adnexal lymphoma is a rare manifestation of non-hodgkin lymphoma, often characterized as low-grade malignancies, with approximately 50% classified as Mucosa-Associated Lymphoid Tissue (MALT) lymphomas. Clinically, these lesions typically progress slowly, with orbital involvement being the most common presentation, frequently resulting in exophthalmos. Diagnosis is primarily based on the histological examination of a biopsy taken from an accessible lesion. Accurate histological characterization and the assessment of systemic extension (present in about 30% of cases) are crucial for guiding treatment decisions. Various therapeutic options include localized radiotherapy and antibiotherapy, which may be combined with mono or polychemotherapy. Here, we report a case of bilateral lower palpebral MALT lymphoma in complete remission following hypofractionated radiotherapy with description of the radiation technique.

Clinical Case Report

Clinical presentation

A 65-year-old Moroccan man presented with a 4-months history of left ptosis, bilateral exophthalmos and decreased vision. Physical examination revealed painless swelling of the lower eyelids bilaterally (Figure 1). Orbital magnetic resonance imaging showed infiltration of the orbital fat bilaterally, extending to the lower eyelids, without any optic nerve abnormalities. An excision biopsy of the palpebral tumor confirmed the diagnosis of Mucosa-Associated Lymphoid Tissue (MALT) lymphoma.

PET FDG imaging demonstrated symmetrical basiorbital hyper metabolism, leading us to conclude a diagnosis of localized palpebral MALT lymphoma, classified as stage 2 according to the Ann Arbor classification.

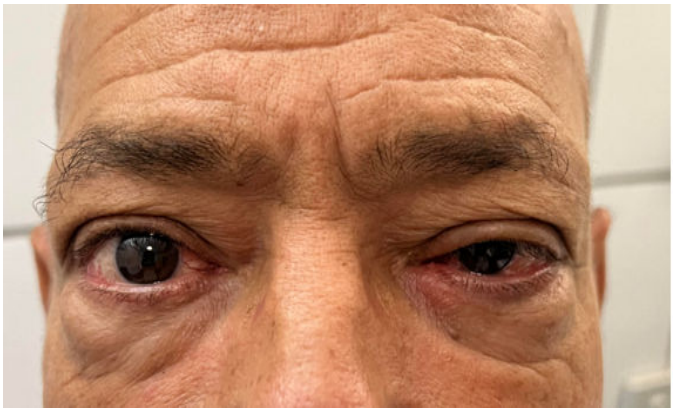

Figure 1. Bilateral lower eyelid tumefaction with exophthalmos.

Treatment radiotherapy technique

After consultation with hematologists, we decided to treat the patient with external beam radiotherapy following 3 weeks of tetracycline-based antibiotics. Initially, a CT scan with a thickness of 2.5 mm was performed in the supine position. A 1 cm bolus was placed over the closed eyes before immobilizing the patient with a standard thermoplastic mask (Figure 2).

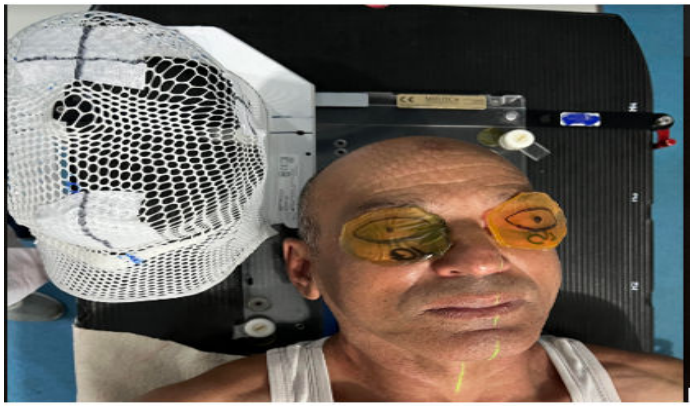

Figure 2. Patient positioning with bilateral bolus and immobilization with thermoplastic mask.

The Clinical Target Volume (CTV) was delineated to include the tumor, the lacrimal glands and the entire orbit bilaterally. The Planning Target Volume (PTV) was established with an isotropic automatic margin of 5 mm around the CTV. The Organs at Risk (OAR) included the eyeballs and lens. We prescribed a dose of 16 Gy in 5 daily fractions of 3.2 Gy, corresponding to an effective biological dose of 36 Gy. The dosimetric physician set up a rapid arc treatment plan using four half arcs and 6 MV energy. The next step involved optimization to achieve optimal coverage of the PTV while minimizing the dose to the OAR (Figure 3).

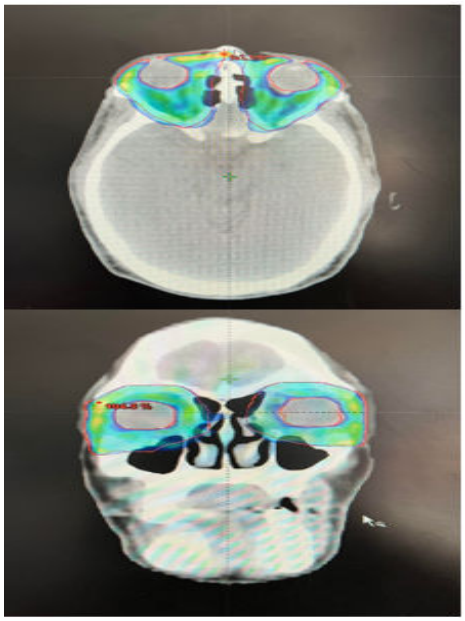

Figure 3. Treatment plan axial and coronal view.

Finally, treatment was delivered with a linear accelerator and positioning verification was conducted using Cone Beam CT (CBCT).

Follow up

During treatment, there was no acute toxicity. The evolution was marked by a complete response after the 5th treatment session, with total disappearance of the palpebral swellings and regression of the exophthalmos (Figure 4).

Figure 4. Complete remission at the last seance.

After 6 months of follow-up, the patient remains well-controlled and no severe late toxicity has been noted, aside from mild dryness syndrome, which was managed with the prescription of artificial tears.

Results and Discussion

Non-Hodgkin Lymphomas (NHLs) comprise a group of neoplasms characterized by monoclonal lymphocytic proliferation (B or T cells).

They can occur throughout the body, particularly in lymph nodes, but also in extranodal sites. Among these, involvement of the periocular region (including the conjunctiva, adnexa and orbit) is a rare primary or secondary localization of the disease, accounting for approximately 1% to 8% of NHL cases [1,2]. Typically, orbital and palpebral NHLs (or ocular adnexal lymphomas) are low-grade malignancies with an indolent course, whereas primary intraocular lesions tend to be high-grade NHLs [3].

Chronic infection by *Chlamydophila psittaci* has been identified as a potential causative agent, with other pathogens such as Hepatitis C virus and Helicobacter pylori also implicated. Orbital imaging plays a crucial role in assessing the locoregional extent of the disease, particularly for determining the orbital component, using Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) [4].

Diagnosis requires adequate tissue sampling for histopathological analysis. Further assessments for accurate staging and therapeutic planning include thorough history-taking, physical examination, laboratory studies, serum protein electrophoresis, serum LDH, β2-microglobulin, chest X-ray, bone marrow biopsy, CT of the chest, abdomen and pelvis and 18-Fluorodeoxyglucose-Positron Emission Tomography-CT (FDG-PET-CT) to complete patient staging [5]. The clinical stage is determined using the Ann Arbor staging classification and the American Joint Committee on Cancer Tumor, Node, Metastasis (TNM) system [6,7]. Approximately 85%-90% of patients present with stage I disease, nodal involvement is reported in 5% of patients and only 10%-15% have disseminated disease [8].

Various treatment strategies are available, depending on the initial stage of the neoplasm, including surgical excision, radiotherapy, chemotherapy, anti-chlamydial antibiotics (such as doxycycline and clarithromycin) and immunomodulatory therapy or combination therapies. Radiotherapy alone is typically the treatment of choice in early stages (I-IIE) [9]. Orbital and ocular adnexal lymphomas exhibit high radiosensitivity, with a median delivered dose of 30 Gy [10]. Numerous studies demonstrate significant effectiveness in terms of local response, with treatment response rates ranging from 86% to 100% and local recurrence rates of 0% to 15% [11].

Most patients experience mild acute toxicities during or immediately after radiotherapy, including skin erythema in the periorbital area, conjunctival hyperemia, chemosis, swelling and excessive tearing. These symptoms are generally self-limited or respond to a short course of lubricant gels or skin moisturizers, resolving within a few months after the completion of radiotherapy [12]. Long-term toxicities may include radiation-induced cataracts, persistent dry eyes and iris neovascularization.

Conclusion

The incidence of ocular adnexal lymphoma, particularly ocular adnexal MALT lymphoma, has risen worldwide over the last few decades. Primary radiotherapy of indolent lymphomas has shown a high response rate and optimal local control. However, there is no unanimous consensus about standard fractioning or dose, even if the most accepted is conventional radiation therapy (30 Gy) for localized disease with excellent local control.

Declaration

The patient consents to the publication of his case.

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