International Journal of Research in Medical Sciences
Swer D et al. Int J Res Med Sci. 2024 Jul;12(7):2660-2663
www.msjonline.org
pISSN 2320-6071 | eISSN 2320-6012

DOI: https://dx.doi.org/10.18203/2320-6012.ijrms20241931

Case Report

Non-Hodgkins lymphoma of bilateral eyelids with basal cell carcinoma of nose: a rare case report

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Received: 13 May 2024
Accepted: 12 June 2024

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ABSTRACT

Malignant tumours affecting the eyelid mainly include basal cell carcinoma. Non-Hodgkin's lymphoma may occur in almost any part of the body and should be considered in the differential diagnosis of extra lymphoid tumours. Non-Hodgkin's lymphoma of the eyelid is rare. Rare diseases should be considered in differential diagnoses because they have the tendency for rapid systemic involvement and they may require treatments that are different from the other common diseases. Basal cell carcinoma occurs most frequently at sun-exposed sites, most commonly in the head and neck regions. The aim of any therapy selected for BCC treatment involving the head and neck is to ensure complete removal, functional preservation, and a good cosmetic outcome. Here, we reported a case of a 70-year-old female with non-Hodgkin lymphoma of eyelids followed by basal cell carcinoma of the nose treated with multimodality approach.

Keywords: Non-Hodgkin lymphoma, Basal cell carcinoma, Eyelids, Skin, Nose

INTRODUCTION

Lymphomas can arise from any lymphatic tissue e.g., lymph nodes, spleen, thymus gland, or mucosa (mucosa-associated lymphoid tissue). The eyelid is a rare site for lymphoma, but a few isolated cases have been reported.[1] These lymphomas should not be confused with other cancers that can affect the eyelid such as basal cell carcinoma, squamous cell carcinoma, sebaceous carcinoma, melanoma, mesenchymal tumours and Kaposi sarcoma, all of which have different modes of treatment.[1]

Ocular adnexal lymphoma is a rare type of extra nodal non-Hodgkin lymphoma. Its incidence is about 0.28 cases per 100,000 patients and comprises only about 8% of all extra nodal lymphomas. It commonly affects patients in the 5th and 6th decade of life with a female predominance.[2,3]

Basal cell carcinoma (BCC) is a malignant neoplasm originating from basal layer of the epidermis non-keratinizing cells and is the most common type of skin cancer with cumulative exposure to ultraviolet radiation being the important risk factor. BCC is the most common cutaneous malignancies worldwide. Its incidence is estimated to be about 770 per 100,000.[4] Most frequent in geographic location with greater UV exposure.[5] Several treatment options such as surgical excision and nonsurgical procedures are available. The aim of any therapy selected for BCC treatment involving the head and neck is to ensure complete removal, functional preservation, and a good cosmetic outcome. The choice of treatment should be determined based on the histological subtype of a lesion, size, location, patient age, general condition and performance status of the patient, treatment availability, and the patient's wishes.[6]

We reported a case of a 70 years old female with non-Hodgkin's lymphoma of bilateral eyelids followed by basal cell carcinoma of the nose.

CASE REPORT

A 70 years old female patient came to the out-patient department (OPD) of ophthalmology, Regional Institute of Medical Sciences (RIMS), Imphal with complain of increased lacrimation and itching of bilateral eyelids for the past 3 months for which she was managed conservatively. She later developed thickening of the eyelids followed by swelling of soft tissue of both eyelids, for which she underwent contrast enhanced computed tomography (CECT) of bilateral orbit which showed lobulated faint hyperdense attenuation in bilateral upper and lower eyelids, more in left upper lid preseptal region. The maximum thickness of the involved area in the left upper lid was 8.2 mm. The involvement in left appeared to extend to the region of the lacrimal sac. No associated calcification was noted (Figure 1).

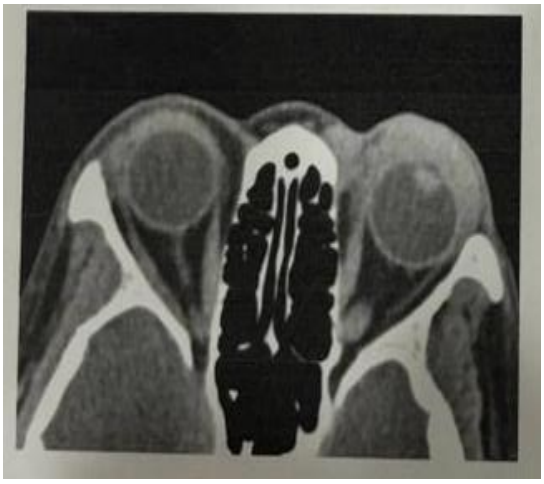

Figure 1: CECT of bilateral orbit showed lobulated faint hyperdense attenuation in bilateral upper and lower eyelids, more in left upper lid preseptal region.

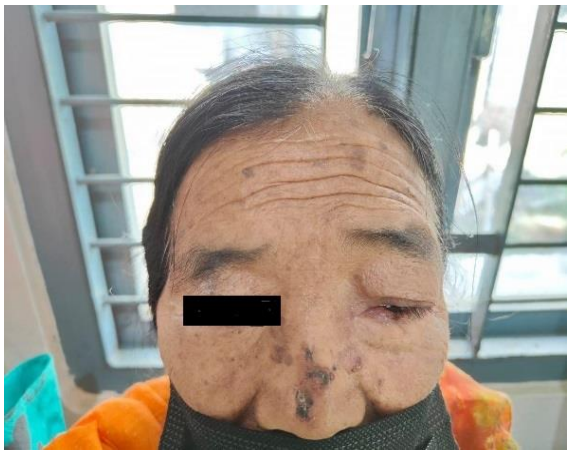

Figure 2: Irregular lesion with punch out edges.

Figure 3: Patient receiving brachytherapy.

Figure 4: Patient on follow up with no evidence of any malignancy.

The patient was then advised for incisional biopsy from soft tissue mass of eyelids and the specimen was sent for histopathological examination (HPE) which showed diffused infiltration of predominantly small atypical lymphoid cells with irregular nuclear margin, dispersed chromatin, inconspicuous, nuclei, and scant pale cytoplasm admixed with scattered histiocytes and atypical monocytic cells, infiltration and destruction of glandular tissue and the features were suggestive of non-Hodgkin's lymphoma B cell type with IHC marker showing strong expression of CD20 positive and CD45 positive and Ki 67% proliferation index at 23%.

The patient then attended the OPD of radiation oncology, RIMS, Imphal, with the HPE report. Complete history and thorough physical examination were done which showed a 0.5×0.4 cm soft to firm round nodule, with smooth surface, 2 in number, in bilateral eyelids with normal visual field and acuity. No other lymph node were clinically palpable. Patient had no history of surgery or similar illness in the past. No family history of any similar illness in the past. The patient general condition was good with Karnofsky performance status (KPS) being 80%, and BSA of 1.48 m^2. All routine baseline investigation were within normal limits. Metastatic work up showed no evidence of metastasis. The patient was

then started on systemic chemotherapy with RCVP regime (rituximab, cyclophosphamide, vincristine and prednisolone) on three weekly basis for a total of 6 cycles. 1 month after the completion of chemotherapy, positron emission tomography-computed tomography (PET-CT) was done which showed no evidence of residual disease or any metabolically active lesion in the body and the patient was kept on 2 monthly follow up with monitoring of serum LDH level.

The patient was on regular follow up with normal LDH levels but 7 months later from the last chemotherapy the patient had a suspicious lesion on the dorsum of the nose, around 3×2 cm in size, irregular in shape, 3 in number and blackish pigmentation patchy lesion with punched out edges. It was not associated with any discharge, bleeding or pain (Figure 2).

The patient underwent punch biopsy of the lesion on the dorsum of the nose, left and right-side wall of the nose with HPE showing a predominantly nodular pattern intermixed with superficial spreading, infiltrative, adenoid, pigmented, basosquamous pattern suggestive of BCC mixed type. Considering the indolent nature of BCC, the patient underwent wide local excision with reconstruction of flap. The HPE of the excised specimen showed focal areas of squamous differentiated within the islands of nasoloid cells are also seen as basosquamous pattern with high-risk category (WHO 2023), distance of tumour from closest margin base at 1.7mm. Numerous mitotic figures 20 /mm^2 and apoptotic figures were seen suggesting BCC mixed type. The patient was then planned for adjuvant RT to the nose due to high-risk category (WHO 2023), with mitotic rate at 20 /mm^2 with the distance of tumor from closest margin base at 1.7 mm. The patient was treated with surface mould brachytherapy (BT) by Flexitron HDR BT machine using Iridium 192 source. A total dose of 33 Gray in 7 fraction was given, twice weekly at a depth of 0.5 cm. With the 1st to 6th fractions being 5 Gray each and the last fraction of 3 Gray (Figure 3).

One month following BT, the patient was examined and no mass and lymph node were clinically palpable. PET-CT was done which showed no evidence of residual diseases and no metabolically active lesion in the body with normal LDH levels. The patient is currently under follow up for the past 6 months with no evidence of any malignancy (Figure 4).

DISCUSSION

Ocular adnexal lymphomas accounts for 8% of all extra nodal lymphomas and eyelid involvement is usually due to forward extension of the tumour.[7] The most common sites are the superior orbit, followed by the conjunctiva, lacrimal gland and eyelids. Most primary ocular adnexal lymphomas are low-grade lymphomas with marginal zone lymphoma being the most common type. High-grade B-cell lymphomas only occasionally involve the ocular adnexa and T-cell lymphoma, natural killer-cell lymphoma, and Hodgkin's lymphoma are rarely encountered in this site.[8] Systemic chemotherapy remains the mainstay for the treatment of NHL along with radiation therapy. Surgical excision is not usually recommended because tumour micro-infiltration into the surrounding tissue may be present. However, biopsy is required to establish a diagnosis and for subtyping the lesion.[9] Patients with low-grade lymphomas are treated with local radiotherapy whereas those with high-grade lymphomas require single-agent or combination chemotherapy. It has been noticed that a more aggressive treatment regimen has not clearly changed the overall survival rate. Immunotherapy for treating non-Hodgkin's lymphoma is showing promising results.[10] Biopsy is essential because the diagnosis changes the treatment modality; surgical excision usually performed for more common tumours of the eyelid is not successful in such patients. If not diagnosed correctly, lymphoma of the eyelid may lead to dissemination of the disease.

BCC is the most common cutaneous malignancy worldwide with an ever-increasing incidence rate worldwide. The most common sites are the face especially the nose, cheeks, forehead, and nasolabial folds and eyelids. Surgical excision of lesions is the treatment for BCC. If surgery is contraindicated, radiation therapy is a primary option. Topical therapy with 5-FU cream and imiquimod 5% is FDA-approved for superficial BCC.[6] The incidence of BCC is positively associated with malignancies with immune suppression like non-Hodgkin's lymphoma, a haematological malignancy. Exposure to ultraviolet rays is recognized as a critical factor in the pathogenesis of basal cell carcinoma, presumably partly because of immune suppression. It could therefore be hypothesized that an association would exist between BCC and malignancies associated with immune suppression such as haematological neoplasm.[11,12]

CONCLUSION

Patients with primary non-Hodgkin's lymphoma of the eyelid, as with other non-Hodgkin's lymphomas, must undergo full screening investigations to determine the extent of the disease, to select a treatment protocol and predict the prognosis. When confronted with an unusual eyelid mass, non-Hodgkin's lymphoma should be considered and a diagnostic biopsy should be performed. Patients who present with isolated orbital disease should be re-examined periodically for systemic disease. Radiation therapy is also used in the treatment of primary BCC or in cases where postsurgical margins are positive for cancer. Specifically, radiation treatment using high-dose rate brachytherapy (HDR-BT) has become increasingly used over the past decade for the treatment of early-stage skin cancer e.g., BCC, with high local control rates with series reporting local control between 90% to 98% and good-excellent cosmetic outcomes. HDR-BT is a growing treatment option for BCC due to

its less invasive nature and higher tolerability profile in elderly patients. Adequate treatment promises superior local disease control for BCC. Thus, doctors in all specialties need to become more aware of BCC, and accurate and early diagnoses need to be made by them. We reported a rare case of a 70-year-old female with non-Hodgkin lymphoma of eyelids followed by BCC of the nose. This patient was treated with multimodality approached with chemotherapy, surgery, radiation therapy. The patient showed significant improvement and is currently disease free and on follow up.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Swer D, Mahawar R, Devi YS, Sangtam D. Non-Hodgkins lymphoma of bilateral eyelids with basal cell carcinoma of nose: a rare case report. Int J Res Med Sci 2024;12:2660-3.