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Bilateral Lacrimal Gland Lymphoma: A Rare Case Report

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Abstract

Lymphomas of the lacrimal gland are rare, but account for 37% of all malignant tumors of the gland. We report a case of a 40-year-old female with painless gradually progressing mass in both upper eyelids for last 6 months. The CT and MRI scans revealed heterogeneously enhancing symmetrical extra and intra conal lesion involving the superior and lateral rectus muscle on both sides,. The patient underwent excisional biopsy, which revealed features of small round cell tumor most probably Lymphoma. The final diagnosis was bilateral lacrimal gland lymphoma, a rare entity. The patient received chemotherapy and achieved complete remission.

Keywords: Lymphomas; Chemotherapy; Non-Hodgkin Lymphoma (NHL)

Introduction

Lymphomas of the lacrimal gland are rare, accounting for 37% of all malignant tumors of the gland. Diffuse large B cell lymphoma (DLBCL) is the most common histologic subtype of non-Hodgkin lymphoma (NHL) [1]. We report a rare case of bilateral lacrimal gland lymphoma in a 40-year-old female.

Case Report

A 40-year-old female presented with painless gradually progressing mass in both upper eyelids for last 6 months. The mass was measuring approximately 55*33*40 mm on the right side and 42*32*25 mm on the left side, which seemed to arise from the superolateral orbital region and was lobulated and predominantly firm in consistency bilaterally, not adherent to skin. Pulsation, bruit, crests, and transillumination all were negative. Multislice CT scan of orbit showed heterogeneously enhancing symmetrical extra and intra conal lesion involving the superior and lateral rectus muscle on both sides, which was highly more suggestive of Idiopathic inflammatory thyroid eye disease. MRI brain and orbit showed bilateral enlarged lacrimal glands with bilateral enlarged submandibular and cervical lymphadenopathy, consistent with lymphoma. The patient underwent an excisional biopsy, which revealed features of small round cell tumor most probably Lymphoma. The final diagnosis was bilateral lacrimal gland lymphoma, a rare entity. The patient received chemotherapy and achieved complete remission.

Discussion

Lymphomas of the lacrimal gland are extremely rare [1]. Diffuse large B cell lymphoma (DLBCL) is the most common histologic subtype of non-Hodgkin lymphoma (NHL). The presentation is usually as painless progressive proptosis and/or eyelid swelling, similar to the presentation in our patient [2].

Lymphoma of the lacrimal gland is a rare and challenging diagnosis. A high index of suspicion is required to establish the diagnosis [3]. The differential diagnosis include idiopathic orbital inflammation, pseudotumor, sarcoidosis, and other malignancies [4]. Imaging studies, such as CT and MRI, play a crucial role in the diagnosis and staging of the disease [5]. In our case, the CT and MRI

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scans revealed bilateral symmetrical extra and intra conal lesion involving the superior and lateral rectus muscles on both sides, and enlarged lacrimal glands with bilateral enlarged submandibular and cervical lymphadenopathy, which were consistent with lymphoma.

The diagnosis of lacrimal gland lymphoma is confirmed by histopathological examination. Fine needle aspiration cytology (FNAC) is often inadequate to establish a definitive diagnosis, and an excisional biopsy is preferred [6]. The histologic subtypes of lymphoma vary widely and include DLBCL, follicular lymphoma, mantle cell lymphoma, and others. DLBCL is the most common subtype of NHL and accounts for approximately 60% of cases [7]. In our case, the biopsy revealed features of a small round cell tumor most probably Lymphoma.

The treatment of lacrimal gland lymphoma depends on the extent and stage of the disease. Early-stage disease can be treated with radiation therapy, while advanced-stage disease requires systemic chemotherapy [8]. Surgery is rarely indicated in the treatment of lacrimal gland lymphoma, except in cases of localized disease that is not amenable to radiation therapy or chemotherapy [9]. In our case, the patient received chemotherapy and achieved complete remission.



Figure 1: Upper Lid Mass measuring 55*33*40 mm on the right side and 42*32*25 mm on the left side.

Conclusion

Bilateral lacrimal gland lymphoma is a rare entity that can present as painless gradually progressive mass in both upper eyelids. A combination of imaging, clinical examination, and histopathology is necessary for the diagnosis. Treatment options include radiation therapy, chemotherapy, and surgery, depending on the extent and stage of the disease.

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