

Lacrimal Gland Lymphoma with Systemic Involvement-A Case Report

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Abstract: Lymphoid tumors are amongst the common tumors occurring in the ocular adnexa. Orbital lymphoma appearing in the lacrimal gland region accounts for about 11% of all orbital lymphomas. Non-Hodgkin's lymphomas (NHLs) are among the most common ocular adnexal primary tumors. We present a 48-year-old male with bilateral superotemporal orbital mass and mechanical ptosis. Orbital mage obtained by computed tomography scanning showed bilateral bulky lacrimal glands and histopathology demonstrated monomorphic small lymphocytic cells suggestive of Non-Hodgkin's lymphoma.

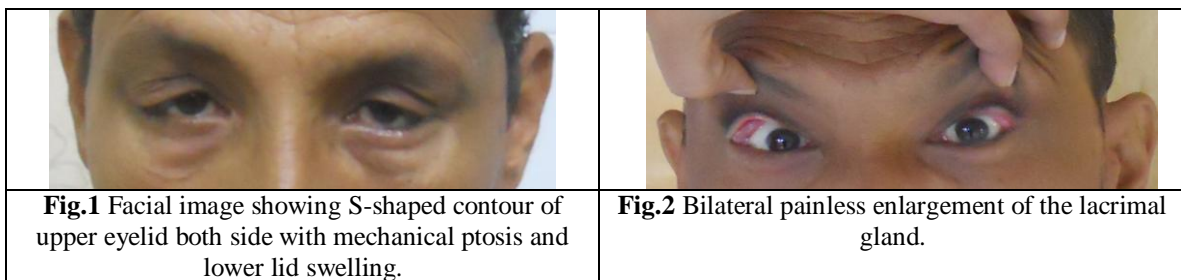
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I. Introduction

Lymphomas are malignant neoplasms characterized by propagation of cells natural to the lymphoid tissue.¹ They are broadly divided into Hodgkin's and Non-Hodgkin's lymphoma. Ocular adnexal lymphomas (ie, lymphomas of the orbit, eyelids, conjunctiva, lacrimal gland and lacrimal sac) constitute 2% of all extranodal lymphomas² and are the most common malignant tumours of the orbit³. Lacrimal gland lymphomas are relatively rare, representing 7% to 26% of ocular adnexal lymphomas.⁴⁻⁶ Lymphoma of the lacrimal gland presents as a painless mass involving the lacrimal fossa. Both bilateral and unilateral presentations are possible. The treatment depends on the type and extension of the tumour. If there is no systemic involvement, currently recommended therapy is radiotherapy^{7,8,9}; disseminated disease is treated with chemotherapy.^{7,9,10}

II. Case Report

A 48-year-old male presented to us with complaint of painless gradually progressive mass in superotemporal orbit of both eye for the last 5 months. The mass was rubbery in consistency, lobulated and mobile, not fixed to the skin. During this period the patient also experienced weight loss and fatigue.



Ocular examination revealed normal visual acuity in both eyes and mechanical ptosis due to the mass pushing on the lid. Soft tissue swelling of the lower eye lid was present in both eyes. The rest of the anterior and posterior segment examination were normal. There was no restriction of extraocular movements.

The patient had generalized lymphadenopathy and gave past history of tuberculosis for which he underwent treatment (Category I DOTS regimen) and recovered. His present report of sputum for AFB is negative.

Chest X-ray(PA view) showed hyperlucency and hyperinflation of both lung fields with mild flattening of both hemidiaphragm suggestive of emphysema.(FEV₁-99% of predicted □ FVC-96% of predicted)



Fig.3 Chest X-ray impression suggestive of Emphysema.

USG abdomen revealed Conglomerating and necrotic peripancreatic and periportal lymphadenopathy. Computed tomography scan showed bilateral bulky lacrimal gland showing mild homogenous enhancement on post contrast study and there was no erosion of lateral walls of orbits.

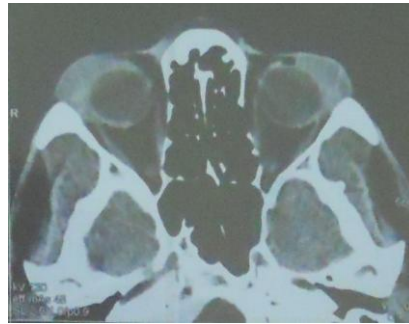


Fig.4. CT scan head and orbit showing bilateral bulky lacrimal gland.

Biopsy taken from the right inguinal lymph node demonstrated lymph node structure diffusely infiltrated with monomorphic small lymphocytic cells suggestive of Non- Hodgkin's lymphoma.

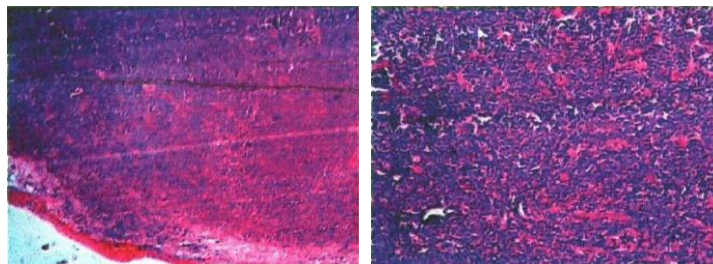


Fig.5. Biopsy specimen showing monomorphic small lymphocytic cells.

In consultation with a haematologist, the patient was started on Chemotherapy according to the CHOP protocol [Cyclophosphamide, Doxorubicin (or Adriamycin), Vincristine (or Oncovin) and Prednisone]. There is good clinical response and the patient's physical condition also improved.

III. Discussion

Ocular adnexal lymphoma represent the malignant end of the spectrum of lymphoproliferative lesions that occur in the conjunctiva, eyelids, lacrimal glands and orbit. Differential diagnosis of lacrimal gland pleomorphic adenoma, granulomatous disease like tuberculosis, adenoid cystic carcinoma and thyroid disease were considered. To confirm the diagnosis we need complete general and ophthalmic examination with ultrasound, CT orbital image and histopathological examination. Biopsy is important for diagnosis. CT scan can define the density and relative homogeneity of the lesion and determine the extent of the disease. If there is systemic involvement at the time of presentation, as in our patient, appropriate systemic chemotherapy should be administered.

IV. Conclusion

Non-Hodgkin's lymphoma is a systemic disease and various organs of the body could be affected including the eye and the orbit. Lacrimal gland lymphoma is a relatively rare disease but it should be taken in consideration when finding orbital masses. A thorough systemic evaluation and histopathological examination along with orbital imaging is required for proper diagnosis and appropriate management.

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