 DOI: 10.18231/2455-8478.2018.0003

# An overview of the management of orbit and adnexal lymphoma (OAL)

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Orbit and adnexal lymphoproliferative lesions are comprised of 94.2% Non-Hodgkin lymphoma (NHL), 0.8% Hodgkin lymphoma (NHL) and 4% benign reactive lymphoid hyperplasia (RHL).[1] Orbital and adnexal lymphoma includes 1-2% of all lymphomas and 8% of all extra-nodal lymphomas and comprises 20-30% of all orbital tumors. Male is preponderance than female (M: F= 2.23:1). Though orbital lymphoma is a disease of elderly but affecting the younger people in a recent trend and age ranges from 8 to 88 years with median age 60 years.[1-4] The ocular adnexal lymphoma is involving the orbit, 37%; conjunctiva, 29%; lacrimal, 20%; and eyelid, 14%. Unilateral OAL is 83% and primary orbit and adnexal lymphoma is about 72%. Extra-nodal marginal zone lymphoma (ENMZL) is the most common primary Orbit and adnexal lymphoma that is 62% of all primary OAL followed by follicular lymphoma (FL), 17% and diffuse large B-cell lymphoma (DLBCL) at 10%. The most frequent secondary lymphoma is mantle cell lymphoma which accounts 22% of all secondary lymphomas. Clinical features may include gradual painless proptosis, mechanical ptosis, palpable rubbery mass in the eyelid, diplopia due to motility defects, and salmon patch appearance of the conjunctiva. The painful lesion may found in 10-20% of follicular lymphoma (FL) and 20-30% of diffuse large B cell lymphoma (DLBCL). Gastric H. pylori positivity is associated with 32% patients with ENMZL.[2,3] A CT scan of the orbit is the gold standard imaging technique and is using to identify the extent and location of the lesion and helps in planning the surgical approach. CT scan may present as well circumscribed, homogeneous, Hyper dense to Iso dense lesion which molds around the eyeball and other orbital structures may form the pancake-like configuration in the orbit.[3,5] Bone erosion is seen in DLBCL and Mantle Cell Lymphoma. The recent modality imaging technique FDG PET-CT scan can be performed to detect the early onset of malignancy, assess the effectiveness of treatment plan, and is also used to determine how far cancer has spread to other organ systems.[2,5] Surgery plays a dual role both diagnostic and therapeutic. Histopathology followed by convenient immune-histochemical analysis are the mainstay to confirm the diagnosis of lymphoma in our context. The combination of IRTA1 with t-bet has been described as a suitable immune-histochemical marker for the positive identification of ENMZL. Follicular lymphomas are BCL2, BCL-6, and CD10 positive in 88% of cases. Diffuse large B-cell lymphomas should also be tested for MYC, BCL2, and BCL6 rearrangements.[2,6]

**Fig.1: Salmon Patch appearance of Conjunctiva**

**Fig. 2: CT scan shows bilateral hyper to iso-dense lesion which molds to the globe,**

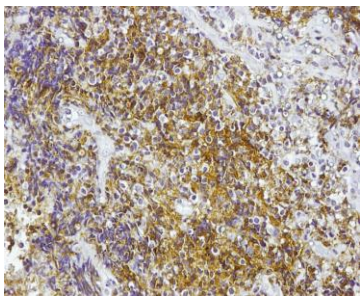

**Fig.3: Immunohistochemistry shows CD 20 positive**

TNM Staging is used for documentation of the disease, prognostication, and planning out the treatment and follow-up. Poor prognosis is associated with higher T staging, the involvement of lymph nodes and distant metastasis.[7] Treatment of OAL is progressing day by day and depends on the TNM staging, histological types, immuno-histochemical analysis, patient's physical condition, and potential ocular toxicity of treatment.

Orbital Radiotherapy is the proved treatment modality to control the low grade localized OAL like ENMZL/MALT associated lymphoma and success rate is more than 90% of patients up to 10 years follow up. Radiotherapy may be used in combination with chemotherapy and immunotherapy in the cases of secondary lymphoma or with systemic involvement. The effective external beam radiotherapy (EBRT) doses are 25-30 Gy for low-grade OAL and 30-40 Gy for high-grade OAL.2 Chemotherapy is given indicated in the cases of OAL with systemic involvement or high-grade malignant lymphomas like DLBCL, mantle cell lymphoma, and small lymphocytic lymphoma. The most common chemotherapy regimen is CHOP therapy. When chemotherapy is given with local radiotherapy to orbit has a synergistic effect to control the OAL. A most common regimen is CHOP.[2,3,5] Newer treatment modalities are immunotherapy and radio-immunotherapy. Immunotherapies are interferon α 2b and anti-CD20 antibody rituximab. Sub Conjunctival or perilesional injection of IFN-α 2b is the effective treatment option to control localized ENMZL in patients without the systemic involvement and its disease control is 85% at 5 years follow up period. Rituximab can be used as alone or in combination with CHOP therapy known as R-CHOP therapy for DLBCL and others high-grade lymphoma. Radio-immunotherapy consists of combining radioisotope with rituximab.[3,8,9] Antibiotic may be used for positivity for Chlamydia psittaci in ENMZL.[2] Treated patients need long-term follow up to assess complications and recurrence. Follow up may be scheduled for every three months in the 1st year, every four months in a 2nd year, every six months for the next 3 years, and then every year.

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