A 71 yr old male presented to the department of Ophthalmology, Medical College Hospital, Calicut with swelling of the left lower eyelid of 6 months, progressive since 1 month. He had no pain, visual disturbance or constitutional symptoms.

On examination, he had generalized lymphadenopathy with preauricular, submandibular, cervical and inguinal nodes (Fig. 2). These nodes were 1-2 cm in size, firm, discreet and non tender. Systemic examination was within normal limits (Fig. 3).

Ocular examination revealed a non tender swelling in the left lower lid 0.5 cm x 1.0 cm, with the skin and lower palpebral conjunctiva freely mobile over it (Fig. 2). The pupillary light reflex was normal in both eyes and there was incipient lenticular opacities which accounted for a visual acuity of 6/24. Colour vision, fields and fundus examination were within normal limits in both eyes.

The patient underwent routine lab workup. Complete blood counts and peripheral smear were within normal limits.

Bone marrow trephine smear showed scattered erythroid, myeloid haemopoietic cells with normal maturation (Fig. 4).

FNAC of lid swelling was performed; Papanicoloau stain revealed sheets of uniform immature lymphoid cells with irregular chromatin clumps, irregular nuclear border with a cell size larger than mature lymphocytes. (Fig. 5 and 6).

Endonasal biopsy from a bulge in the nasopharynx showed sheets of uniform immature lymphoid cells (Fig 7 & 8). Both FNAC and biopsy confirmed low grade Non Hodgkins lymphoma (NHL).

Based on a diagnosis of low grade NHL, a management option of chemotherapy was given (CHOP regimen was completed in 5 cycles).

Discussion

Among all systemic lymphomas, ocular adnexal lymphomas constitute a small fraction only. Most of these tumors are NHL – B cell type.

Ocular Adenexal lymphoma constitute 6-8 % of orbital tumours and 6 % of extra nodal lymphoma. Ocular adnexal lymphoma is considered to be primary if it involves the ocular adnexa alone and secondary if it is accompanied by lymphoma of identical type at another site. The frequency of involvement of adnexa are as follows. Conjunctiva 20-33 %, Orbit 46-74 %, and eye lid 5-20 %.

The classification of lymphoproliferative disorders has been a major problem in diagnostic pathology. But with the development of immunohistochemical analysis clonality can be identified from the CD molecules these cells express. There are various classifications of
Fig. 1. Picture showing lower eye lid swelling

Fig. 2. Cervical lymphnode enlargement

Fig. 3. Chest X ray - Normal

Fig. 4. Bone marrow : Normal maturation of erythroid and myeloid series

Fig. 5. Lid swelling - Low power view (HPE)

Fig. 6. Lid swelling - High power view (HPE)

Fig. 7. Endonasal biopsy - Low power view (HPE)

Fig. 8. Endonasal biopsy - High power view (HPE)
lymphoma which are quite extensive and includes Revised European American Lymphoma (REAL) classification, Rappaport classification and NIH working formulation classification. Ocular Adnexal Lymphoma (OAL) exhibits a limited spectrum with majority being Non Hodgkins B cell. Types of OALs based on the Immunophenotypic analysis are Extra Nodal Marginal Zone Lymphoma (EMZL), Follicular Mantle cell, Lymphoplasmacytic (LPL) and Diffuse Large B Cell Lymphoma (DLBCL).

The diagnosis of OAL is made by the clinical and imaging studies followed by the combination of histopathologic, immunophenotypic and molecular genetic studies.

Management of OAL is a multidisciplinary approach first requiring a comprehensive staging evaluation. A complete blood count, hepatic enzyme levels, LDH, Chest Radiography, CT scan of abdomen and chest and Bone marrow studies are often required to stage the disease prior to the commencement of treatment. If conjunctival or eyelid OAL is identified a CT/MRI of the orbit is indicated for staging.

The factors that determine the prognosis in OAL are disease stage at presentation, type of lymphoma, site of disease, increased expression of tumor cells and age of the patients. Among the common OAL types, EMZL Follicular and LPL are considered low grade where as DLBCL and Mantle cell lymphoma are high grade.

Treatment of OAL is dependent on specific tumor type and its staging.

Surgery can be useful in certain cases of lymphoma and is appropriate for lesions localized to conjunctiva and orbit.

Radiation is the most frequently used modality for treating OAL in localized disease.

Chemotherapy is recommended in patients with Stage II or greater disease and this includes the use of standard regimens for systemic lymphoma including Cyclophosphamide ,Doxorubicin, Vincristine and Prednisolone and Chlorambucil.

Cases which are seen in outpatient department such as the one discussed, where eyelid involvement is present, may mimic benign tumours like chalazion and have to be taken care of, evaluated, and, properly managed.

References