Ocular adnexal lymphoma in the Northeast Indian population

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We present the clinical profile of biopsy and immunohistochemistry-proven ocular adnexal lymphomas in the Northeast Indian population. Nineteen patients between October 2004 and June 2006 with ocular adnexal lymphoma were analyzed retrospectively. Histopathological classification was done according to international working formulation. Most of the tumors (89%) were treated with radiotherapy followed by chemotherapy.

Key words: Histopathology, immunohistochemistry, non-Hodgkin's lymphoma, ocular adnexa

Indian J Ophthalmol 2008;56:153-5

Lymphoid tumors are amongst the common neoplasms of ocular adnexa encountered by ophthalmologists. Lymphomas are malignant neoplasms characterized by propagation of cells natural to the lymphoid tissue. Ocular adnexal lymphoid tumors may involve the eyelids, conjunctiva, orbital connective tissue or lacrimal gland. Lymphoma is broadly classified into main two types - A) Hodgkin's lymphoma and B) Non-Hodgkin's lymphoma (NHL). The onset is generally in the sixth and seventh decades of life and it is uncommon in children. The detailed descriptions of ocular adnexal lymphoma in Northeast India have not been previously reported. Few studies on lymphoma in India are reported in the literature. We present this retrospective hospital-based study on ocular adnexal lymphomas from the Northeastern part of India.

Materials and Methods

Case records from the Ocular Pathology Laboratory of 19 patients who presented between October 2004 and June 2006 were retrospectively analyzed. The modes of presentation were studied. All the cases were biopsy and immunohistochemically proven. Following a diagnosis of lymphoma, all patients were evaluated by a medical oncologist for systemic involvement. A detailed history followed by Snellen's visual acuity, slit-lamp biomicroscopy, applanation tonometry, fundus evaluation by indirect ophthalmoscopy and detailed proptosis evaluation was done for all patients. B scans, ultrasound and computed tomography (CT) scan was done in all cases.

| Case No. | Age/sex of the patients | Presentation | CD20, IHC | CD3, IHC |
|----------|-------------------------|--------------|-----------|----------|
| 1        | 70 Y/M                  | Proptosis    | Positive  | -        |
| 2        | 68 Y /F                 | Lacrimal gland swelling | Positive | -        |
| 3        | 54 Y /M                 | Proptosis    | Positive  | -        |
| 4        | 77 Y /F                 | Conjunctival mass | Positive | -        |
| 5        | 80 Y /M                 | Proptosis    | -         | Positive |
| 6        | 64 Y /F                 | Upper lid mass | Positive | -        |
| 7        | 22 Y /M                 | Proptosis    | Positive  | -        |
| 8        | 38 Y /F                 | Upper lid mass | Positive | -        |
| 9        | 42 Y /M                 | Proptosis, bilateral | Positive | -        |
| 10       | 56 Y /F                 | Upper lid mass | Positive | -        |
| 11       | 74 Y /M                 | Conjunctival mass | Positive | -        |
| 12       | 37 Y /M                 | Proptosis    | Positive  | -        |
| 13       | 28 Y /F                 | Conjunctival mass | Positive | -        |
| 14       | 59 Y /M                 | Lacrimal gland swelling | -        | -        |
| 15       | 78 Y /M                 | Proptosis, bilateral | Positive | -        |
| 16       | 64 Y /M                 | Upper lid mass | Positive | -        |
| 17       | 70 Y /F                 | Proptosis    | Positive  | -        |
| 18       | 85 Y /M                 | Upper lid mass | Positive | -        |
| 19       | 62 Y /M                 | Proptosis    | Positive  | -        |

Note: M - Male, F - Female, IHC - Immunohistochemistry, Y - Age in years

Table 2: Various presentations of Non-Hodgkin's lymphomas with percentage

| Presentation               | No. of cases | Percentage |
|----------------------------|--------------|------------|
| Proptosis                  | 9            | 47.36      |
| Upper lid mass             | 5            | 26.31      |
| Conjunctival mass          | 3            | 15.78      |
| Lacrimal gland swelling    | 2            | 10.52      |
[Fig. 2 and Fig. 3] and lymphoepithelial lymphoma in one case (5%) [Fig. 4]. Immunophenotypic analysis has shown that most of the ocular adnexal lymphoid tumors are monoclonal proliferation of B-lymphocytes consistent with NHL. Histologically, orbital lymphomas were low to intermediate in grade with predominance of the low-grade variety. The range of follow-up was from three months to two years. Eight patients (42%) had local radiotherapy in doses ranging from 25Gy to 46Gy, two (11%) had CHOP (cyclophosphamide, doxorubicin, vincristine and prednisolone) chemotherapy alone and one had combination with local radiation (28-30Gy) and chemotherapy COP (cyclophosphamide, vincristine and prednisolone). None of the patients had systemic involvement and local recurrence.

**Discussion**

Ocular adnexal lymphoid lesions show characteristic clinical features in most of the patients. They are most commonly seen in the sixth to seventh decade of life with a slight female preponderance. But the present study showed male predominance (63%) which is comparable with other studies from India. The most common site of involvement of lymphoid tumor is the superior orbit behind the orbital septum and 30 to 40% arise from the lacrimal gland. In our study 10% of the cases presented with lacrimal gland involvement. The lesion in this location results in proptosis with downward displacement of globe. Eleven patients (58%) presented with a mass in the superior orbit, either having lid mass or proptosis. L26 clone of B-cell marker CD20 and the UCHL-1 clone of T-cell markers CD45RO or CD 3 were used for immunohistochemistry analysis.

Most of the tumors were B-cell origin (89%) followed by T-cell origin (5%) and lymphoepithelial lymphoma (5%) [Fig. 4]. Jakobiec has stressed that more than 60% of ocular adnexal lymphoid infiltrates are composed of B-cell lymphomas. T-cell lymphomas are rare in the ocular adnexa and can be suspected on clinical and histopathological grounds. A tumor composed of small lymphocytes that are CD5+ and also express CD23 is probably small lymphocytic lymphoma (SLL), not a MALT lymphoma. CD5+ Mantle cell lymphoma is distinguished from SLL by positive staining for cyclin D1 and negative for CD23. None of the patients had Hodgkin’s lymphoma, which by itself a rarity in the orbit. Primary cases required surgical intervention consisting of excision and debulking.
Most of the patients required radiation with careful ocular shielding and some required chemotherapy based on working classification of NHL. None of the patients had a recurrence of the tumor or any systemic involvement. One of the most important prognostic factors was the extent of the disease discovered after thorough clinical staging. Long follow-up is required to comment on final outcome. All patients with ocular adnexal lymphoid tumor had a thorough systemic evaluation by an oncologist which included a complete blood count, bone marrow biopsy and ultrasonogram and CT scan of the body and abdomen. Patients were re-evaluated at three months interval.

Conclusion

Malignant ocular adnexal lymphomas are common neoplasm in the Northeast Indian population. In a 20-month period we saw 19 cases of NHL in our series. NHL (B-cell type) is the prime type encountered in the assortment of ocular adnexal lesions and the orbit is the foremost extranodal site of association. While the number of patients presented here is small, the clinical inference is that histological recognition of B-cell NHL in this part is a significant observation. Males are on the whole affected more than females. Superior orbit is the commonest site of the lesion. Correct histological diagnosis with immunohistochemistry ensures appropriate treatment.

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Direct aspiration of capsular bag material in a case of sequestered endophthalmitis

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Chronic recurrent endophthalmitis (delayed onset endophthalmitis) is an important cause of chronic, recurrent inflammation in pseudophakic eyes caused by organisms sequestered between the intraocular lens (IOL) optic and posterior capsular bag or at the equator. The clinical picture of the disease is highly variable and may be predictive of the disease. But the diagnosis is clinched through the microbiological isolation of the organism from the intraocular specimens. We describe a simple and effective technique to increase the yield of causative organism in such cases. This offers a chance to salvage the IOL, by delivering the appropriate antibiotics into the capsular bag.

Key words: Culture technique, delayed onset endophthalmitis, phacoantigenic uveitis, Propionibacterium acnes, sequestered endophthalmitis

Indian J Ophthalmo 2008;56:155-7

Chronic localized endophthalmitis (delayed onset endophthalmitis) can occur following uncomplicated cataract surgery with intraocular lens implantation secondary to organisms sequestered in the capsular bag. There is a need to identify these sequestered organisms to facilitate appropriate management. Frequently, specimens from the anterior chamber and vitreous cavity could be unyielding, especially in the early cases in which the vitreous is still uninvolved. This article highlights the technique of directly sampling the capsular bag material in the effective diagnosis of the organism, which facilitates the total cure by irrigation with appropriate antibiotics into the capsular bag.

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Manuscript received: 27.12.06; Revision accepted: 03.07.07

Case Report

A 64-year-old male presented to us six months after an uneventful phacoemulsification with IOL implantation, with