Primary Orbital Lymphoma

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Abstract
Primary orbital lymphoma is a rare condition involving primarily the lacrimal glands. We present the second Indian case of right-sided primary orbital lymphoma in an elderly male who presented with the typical features and showed a good response to chemotherapy.

INTRODUCTION
Lymphomas are neoplastic proliferation of cells of the lymphoid series. This condition primarily involves the axial lymph node groups; although the non-Hodgkin’s type, which is more heterogeneous may also involve extra-lymphatic tissues like nasopharynx, tonsils, GIT, brain, heart, skin etc. Ten percent of systemic lymphomas have orbital involvement, however, in even less than one percent cases lymphoma develops primarily in the orbit. Primary orbital lymphoma usually presents as a painless periorbital swelling with low-grade proptosis and diplopia in a patient in his 6th or 7th decade. The disease is commonly extracranial, located anterogradely involving primarily the lacrimal glands and rarely the extraocular muscles. Early disease is curative with either radiotherapy or chemotherapy and has a favourable prognosis with long disease-free interval.

There is only one report of primary orbital lymphoma from India to our knowledge as yet. We present a rare case of a right-sided primary orbital lymphoma in an elderly male who showed a good response to chemotherapy.

CASE REPORT
A 51 years Muslim male cultivator presented at the ophthalmology out-patient department with a history of gradually progressive protrusion of the right eyeball since one and a half-month (Fig. 1). It was not associated with visual abnormality. However, he experienced watering from the right eye and there was associated occasional early morning pain with heaviness of the head. The patient did not have any constitutional or other significant symptoms.

There was no significant past or family history.

On general physical examination, pulse was found to be 78/min., regular and blood pressure was 110/70 mm of Hg.

Fig. 1: Right sided proptosis at presentation

There were no palpable lymph nodes or any other significant findings. No viscera could be palpated on examination of the abdomen and no abnormality could be detected on examination of the CVS, chest and the CNS.

Ocular examination of the right eye revealed proptosis with mild lid edema. Conjunctiva, cornea and lens were normal. No abnormality in movement was noted. On performing refraction, near vision was found to be N 36 and distant vision was 6/18 and the IOP was 14.4 mm of Hg. Fundoscopy showed choroidal folds in the macular areas with a dull foveal reflex. The left eye was normal in all aspects including refraction and IOP.

Investigations and Differential Diagnosis
Routine blood investigation revealed a total count of 7,200/cumm with a DLC of 58% polymorphs, 34% lymphocytes, 2% monocytes and 6% eosinophils. Hemoglobin was 11.1 gm % and ESR was 12 mm. RBS and blood urea were 120 mg% and
24 mg% respectively. Platelet count was 2,00,000/cumm and reticulocyte count was 1.5%.

Plain X-ray of the skull and the orbit showed a normal diploic space with an intact orbital margin and normal overlying soft tissue. CECT scan of the brain and orbit detected an irregular homogeneously enhancing lesion in the right retroorbital region displacing the globe inferolaterally. No intra-ocular invasion or orbital erosion was noted and the global volume and contour were maintained (Fig. 2), raising the possibilities of right-sided orbital secondaries or a primary orbital lymphoma.

On performing incision biopsy and HPE of the mass, sheets of monomorphic lymphoid cells were demonstrated, giving rise to the possibility of a non-Hodgkin’s lymphoma. Repeat HPE of the tissue showed sheets of neoplastic lymphocytes with prominent nucleus and nucleoli surrounded by inconspicuous cytoplasm. No evidence of other specific pathology was noted, thus providing a definitive diagnosis of diffuse non-Hodgkin’s lymphoma (Fig. 3).

USG of the whole abdomen including KUB and prostate and PA view X-ray of the chest (Fig. 4) were done to detect any possible primary or other extralymphatic site of origin, but no positive findings could be detected. ECG was normal. Bone marrow aspiration cytology was within normal limits.

Considering the age and mode of presentation of the patient, the common differential diagnoses considered were Grave’s disease, benign reactive lymphoid hyperplasia, pseudotumour, orbital secondaries and multiple myeloma; and each of these were carefully excluded. Furthermore, HPE of the mass proved beyond doubt the lesion to be non-Hodgkin’s lymphoma in this patient.

**Follow-up**

A diagnosis of primary orbital lymphoma was made and the patient was put on CHOP regimen. After the first cycle, there was definite clinical improvement evidenced by a 50% reduction of the proptosis and alleviation of symptoms. After the second cycles 100% reduction of proptosis was noticed. The patient was given a total of six cycles of chemotherapy. The patient did not develop any new symptoms pertaining to involvement of other sites during the course of treatment. Repeat CBC, PA view X-ray chest and USG abdomen were normal; but repeat CT scan could not be done due to financial restraints of the patient.

**DISCUSSION**

Primary orbital lymphoma is an extremely rare condition and usually presents as a painless periorbital swelling with low-grade proptosis and diplopia in a patient in his 6th or 7th decade, the median age being 61 years. The disease is largely extraconal with the lacrimal glands being primarily involved, and the proliferated lymphoid tissue assumes a well defined round to oval shape, molding itself along the globe margin without involving bony or neural tissue. However, the disease may also be intraconal and may also involve the extra-ocular muscles and even the entire orbital space. The condition runs an indolent course in contrast to the aggressive course
of the orbital involvement of the systemic disease. The disease is monoclonal B cell in origin and usually shows intermediate grade histology and can be diagnosed early owing to its early symptomatology. Early disease can be effectively treated with either radiotherapy or chemotherapy and a radiation dose of 30-40 Gy has been found to be curative.³ The disease has a favourable prognosis, since 100% local control can be achieved with adequate therapy with an overall 5-year survival rate of 73.6% and relapse-free survival of 69.5% respectively.³ Rarely, there can be a contralateral relapse.⁵ Primary orbital lymphomas may be associated with macroglobulinemia⁶ and Sjogren’s syndrome secondary to rheumatoid arthritis.

Our case presented with the typical features of a primary orbital lymphoma and it holds a good prognosis since it has shown good response to chemotherapy. However, he has to be kept under regular follow up in order to pick up relapse.

REFERENCES

Announcement

3rd International Congress on Cardiovascular Disease, Taipei, Taiwan, to be held from November 26-28, 2004 and Meeting of the Board of Directors of the International College of Cardiology to be held on April 25, 2004.

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