PRIMARY ORBITAL LYMPHOMA : A RARE PRESENTATION OF NHL

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Summary

Non Hodgkins Lymphoma (NHL) is a malignant neoplasm of lymphoid tissue representing 5% of all malignant tumors and one of the commonest haematological malignancy in our country. The disease can involve in any organ commonly in lymph nodes, liver, spleen, brain, eye, testis, thyroid. Orbital lymphoma is comprised about 1% of all NHL. This is a case report of a patient age of 50 years from Daudkandi, Comilla admitted in Chittagong Medical College Hospital with history of gradual non tender swelling in the left eye with no other organ involvement or any B symptoms. The objective of this study to consider orbital lymphoma as differential diagnosis of orbital swelling as it can occur almost any part of the body and is to minimize the delay of diagnosis as well as to improve prognosis. We analyzed the history, examination, investigations & histopathology and immunohistochemistry of orbital growth that showed Non Hodgkin Lymphoma (Low Grade). The Patient was treated with consecutive six cycle R-CHOP (Rituximab, Cyclophosphamide, Adriamycin, Vincristine, Prednisolone) resulting in complete remission of lymphoma.

Key words

Orbital lymphoma; Imaging and histopathology; R-CHOP.

Introduction

Non-Hodgkin's lymphoma (NHL) is a malignant neoplasm of lymphoid tissue representing 5% of all malignant tumors. It comprises 90% of all Lymphomas [1]. Orbital lymphoma is estimated to represent 1% of NHLs, 1% of intracranial tumors, and less than 1% of intraocular tumors. It is common in Asia and Europe typically affecting elderly with median age at presentation is older than 60 years. The survival rate is approximately 60% after 5 years [5].

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The usual sites of involvement are lymph nodes but all ocular tissues and adnexa may become affected [2]. Lymphomas of the eye and its adnexa are frequently of B cell lineage but may rarely be non-B-cell NHL [3]. The lack of pathognomonic features, high clinical variability, and the limited value of imaging techniques and histopathological measures often lead to delays in diagnosis [4,6].

Lymphoma may present as a localized eyelid mass or nodule [3,5]. These mass lesions are usually small and may cause Proptosis. The patient may present as a diagnostic dilemma. Imaging like CT scan or MRI of orbit & incisional biopsy with immunohistochemistry confirms the final diagnosis. R-CHOP (Rituximab, Cyclophosphamide, Adriamycin, Vincristine, Prednisolone) therapy is the curative option for orbital lymphoma [6].

Case report

A 50 years old male school teacher from Comilla presented in Haematology department of CMCH with gradually increasing swelling of the left eye for 3 years duration. There was no history of pain, discharge, trauma, or B symptoms (Fever, night sweats, weight loss).

He had first presented to a local hospital from where he was referred to a Ophthalmology center to rule out intracranial lesions. He underwent cranial computed tomography (CT) scanning which showed that the mass had no intracranial connection or extension and there is an orbital growth measuring (4.5 X 2.4 X 3.3) cms (Fig 1).

He was then referred to our hospital. On ophthalmologic examination, there was Proptosis of left eye with chemosis (Fig 2).

Visual acuity was 6/6 in both eye. Opthalmoscopic & Slit lamp examination was normal. Examination of the right eye was unremarkable; the pupil was round, central and reactive to light. Tonometry reveals normal pressure in both orbit. There was no palpable preauricular, postauricular, submandibular, cervical or peripheral lymphadenopathy at the time of presentation. Neurological and other systemic examinations were normal. Complete blood count and peripheral blood film study showed no abnormality . Other routine examinations (Chest X ray, ECG, S.Creatinine, Urine R/E, RBS) including S. LDH and Ultrasonography of whole abdomen also reveled no abnormality . Hepatitis B, C and HIV screening was negative. Considering CT scan findings, an incisional biopsy on the mass done.

Histology of the biopsy specimen showed fibrocollagenous & fibrovascular tissue revealing

diffuse & perivascular infiltration of small lymphocytes which is suggestive of Non hodgkin's Lymphoma, small lymphocyte type (low grade) (Fig 3). Immunohistochemistry for CD 20 was also positive. A diagnosis of Non-Hodgkin's lymphoma was made.

The chemotherapy was commenced after a complete workup. He was placed on R-CHOP regimen (Rituximab 375mg/m2 day1, Cyclophosphamide 650 mg/m^2 IV, days 1 and 8; Adriamycin 45 mg/m^2 IV, days 1 and 8; Vincristine 1.5 mg/m^2 IV, days 1 and 8 and Prednisolone 20 mg PO, TDS). The mass was considerably reduced in size after the second course of chemotherapy, after six courses it resolved completely.

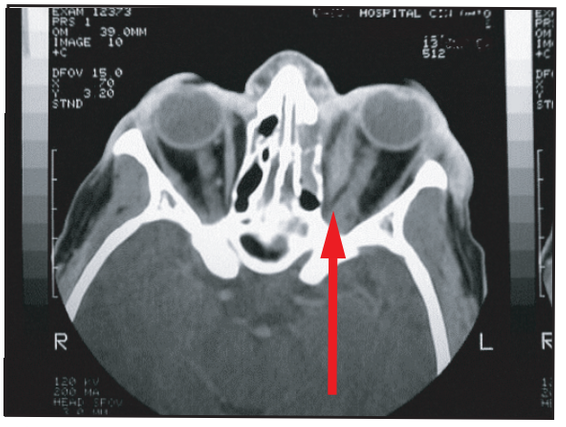

Fig 1 : CT scan Orbit showing left eye involvement

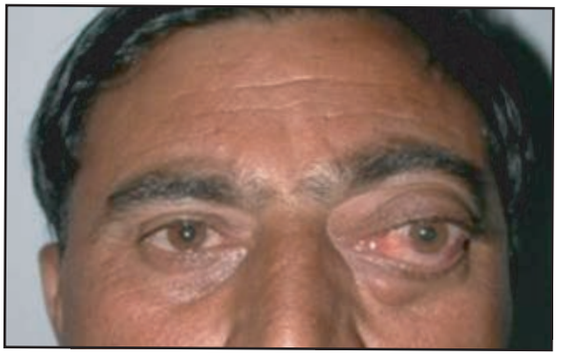

Fig 2 : Unilateral exophthalmos with chemosis

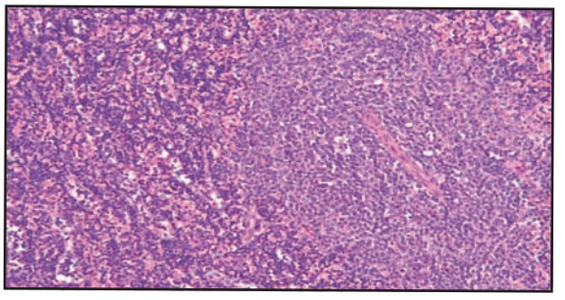

Fig 3 : Biopsy specimen showed diffuse & perivascular infiltration of small lymphocytes .

Discussion

Non-Hodgkin's lymphoma has a wide variety of ophthalmic and neuro-ophthalmic presentations [2]. This particular case was seen by a general practitioner, an ophthalmologist and a medicine specialist, but none of them suspected lymphoma. It was only after the incisional biopsy that the true nature of the mass was revealed and he was referred to a haematologist for treatment.

Ocular adenexal lymphomas represent the malignant end of the spectrum of lymphoproliferative lesions that occur in the conjunctival eyelids, lacrimal glands & orbit. Orbital lymphoma is a rare presentation of extranodal non hodgkin's lymphoma.

Non Hodgkin's lymphoma can involve any part in the body. In annals of oncology 2003(14) A krol, S. Cessie, M kluin, E M Nurdijk showed a study of 1168 patients that how it involves in every part of body [7].

Table I : Primary extranodal involvement of NHL

Organs involved	Percentage %. n=1168
Lymph node	72
Head Neck	41
Intrathoracic	12
Intraabdominal	35
Axilla/ arm	22
Inguinal/ leg	26
Spleen	17
Weldeyer's ring	09
Bone marrow	35
Salivary Glands	01
Oral Cavity	01
Stomach	01
Intestine	11
Colon/ rectum	04
Liver	03
Pancreas	06
Nasal cavity/ sinus	01
Lungs/ pleura	04
Bones	03
Skin/ Soft tissue	02
Breast	03
Uterus/ Cervix	01
Testis	01
Urinary bladder	01
Eye/ lacrimal gland	01
Brain	02
Spinal cord	02
Thyroid gland	01
Suprarenal gland	01
Ill defined sites	01

On differential diagnosis of primary orbital Non hodgkin's Lymphoma, we have to take in consideration some more common orbital tumors seen in adults such as cavernous haemangioma, lymphoid lesions of orbit (benign reactive lymphoid hyperplasia lymphoma atypical lymphoid hyperplasia), optic nerve meningioma, orbital metastasis, neurofibroma, neurilemmoma, Fibrous histiocytoma, haemangiopericytoma, lymphangioma, mucocele & thyroid disease. To confirm the diagnosis we need complete ophthalmologic examination, Blood count & blood film study, ultrasound, CT orbital image, cytology & histopathology examination.

The treatment depends on the type & extension of tumor. Our experience showed that R-CHOP protocol has good clinical response. The patient was in good clinical condition through out the period of chemotherapy & the tumor mass disappeared after the 12th cycle. The patient was followed up for 3 more years & there was no sign of tumor recurrence. Although usually demonstrating & indolent course, non hodgkin's lymphoma are known for recurrence at extranodal sites including other ocular adenexal sites. Long term followup with six monthly examination is therefore recommended. The major prognostic criteria for ocular adenexxal lymphomas include patient age, anatomical location of the tumor, stage of the disease at first presentation, serum LDH level at the time of diagnosis, lymphoma subtype as determined according to WHO lymphoma classification & tumor cell growth rate.

Conclusion

Although primary orbital Non Hodgkin's Lymphoma (NHL) is a rare diagnosis it should be taken in consideration when finding orbital masses because the prognosis of newly diagnosed cases of NHL depends on tumor staging.

Disclosure

All the authors declared no competing interest.

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