

**Case report:**

**Primary Non-Hodgkin's Orbital lymphoma of diffuse large B-cell type: a rare presentation**

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**Abstract:**

Primary Non-Hodgkin's lymphoma of the orbit is a rare presentation and the diffuse large B-cell lymphoma (DLCL) type of histology is much less commoner than the mucosa associated lymphoid tissue (MALT) and follicular lymphoma. A 70 years female patient presented with palpable mass arising from right orbit and proptosis of the right eye. CT scan suggested homogenous enhancing soft tissue mass affecting right lacrimal region without any bony destruction. Biopsy confirmed it to be a case of Primary non-Hodgkin's lymphoma of diffuse large B-cell type with strongly positive CD 20. She was given 6 cycles of chemotherapy with R-CHOP after surgery. The patient is now asymptomatic one year after the last cycle of the chemotherapy.

**Key-words:**

Non-Hodgkin's lymphoma (NHL); diffuse large B-cell lymphomas (DLCL); orbit; chemotherapy

**Key Messages:**

Primary Non-Hodgkin's lymphoma of the orbit is rare and DLCL type of histology is much rarer. It can occur in orbit without any systemic features. It must be diagnosed early and can be treated successfully by surgery and R CHOP chemotherapy

*Bangladesh Journal of Medical Science Vol. 15 No. 04 October '16*

**Introduction:**

Lymphomas are malignant neoplasm of lymphoreticular system and mainly involve lymph nodes, spleen and other non-haemopoietic tissues. Eighty percent of lymphomas are B-cell type, while 14% are T-cell type, with natural killer type (NK) forms only six percent <sup>1</sup>. Primary orbital Non-Hodgkin's Lymphoma is a rare presentation of extra-nodal non-Hodgkin's lymphoma accounting for less than one percent of NHL <sup>2</sup>. It affects primarily the lacrimal glands, conjunctiva, eyelids and orbits <sup>3</sup>. Diffuse large B-cell lymphoma (DLCL) type of histology is much less commoner types of primary orbital NHL.

**Case History**

A seventy years old female patient presented with a gradually increasing painless mass over the right eyelid for last five months before admission. She was unable to open her right eye, decreased vision and proptosis of the right eye. There was no history of fever, night sweats, significant weight loss, weakness, anorexia, past history of tuberculosis or contact and neither she was immunocompromised nor on any immunosuppressive drugs. Prior history of exposure to radiation was absent. On general examination, she had mild pallor. The swelling was 5.0 x 3.0 cm, oval, involving mainly the upper and outer aspect of the right eye, and extended to involve the major part of the eyelid (Figure 1).

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Fig 1- Patient before treatment

It was firm in consistency, non-tender, attached to superficial skin and underlying structures without any discharging sinus or overlying ulcer. Superficial temperature was not raised, no thrill or pulsation was noted and no bruit was found. On systemic examination, there was no organomegaly without any lymphadenopathy, neither mediastinal dullness nor any features of superior vena caval obstruction. There was no cranial nerve involvement. Peripheral blood smear showed no abnormal cells. Haemoglobin was 9.8 g/dl, serum LDH was 677 U/l. Other routine laboratory tests like blood glucose, urea, creatinine, liver function test, uric acid, calcium, chest X-ray, echocardiography, and upper gastrointestinal endoscopy were normal. HIV serology was nonreactive. She was further investigated by Contrast enhanced CT Scan of Orbit along with the Paranasal Sinuses which revealed an extraconal lobulated homogeneously enhancing soft tissue mass in the right orbit in lacrimal region. The lesion appeared to be exophytic, protruding to superolateral aspect of the right orbit, medially extending along the superior aspect of the right orbit investing the anterior wall of right globe involving superior oblique muscle and superior rectus – levator palpebrae muscle complex without significant mass effect over right globe (Figure 2, 3, 4).



Fig 2- CECT orbit Axial view



Fig 3- CECT orbit Oblique view



Fig 4- CECT orbit Coronal view

She was provisionally diagnosed as a case of Primary Orbital lymphoma and underwent for debulking surgery for removal of the mass. The tissue sample was sent for histopathological examination, which revealed Non-Hodgkin's lymphoma of diffuse large B cell type and immuno-histochemistry confirmed the diagnosis with CD 20 strongly positive; CD 23, CD 3 & CD 5 negative. She was prescribed with 6 cycles of chemotherapy with R-CHOP (considering her height 4' 11", body weight 50kgs, body surface area 1.44m<sup>2</sup>); Inj. Rituximab (375mg/m<sup>2</sup> = 540mg), Inj. Cyclophosphamide (750mg/m<sup>2</sup> = 1080mg), Inj Doxorubicin (40mg/m<sup>2</sup> = 60mg), Inj. Vincristine (1.4mg/m<sup>2</sup> = 2mg) and Tablet Prednisolone (100mg). She had been asymptomatic thereafter and no further recurrence noted on one year follow-up (Figure 5)



Fig 5- Patient after treatment

**Discussion:**

Orbital lymphoma is a lymphoma occurring in the conjunctiva, lacrimal gland, eyelid and ocular musculature. Primary non-Hodgkin's lymphoma (NHL) of the orbit is a rare presentation, representing 8-10% of extranodal NHL<sup>3</sup> and only 1% of all NHL<sup>2</sup>. Generally, it has an indolent course. Orbital and adnexal lymphoma is associated with systemic lymphoma in 30-35% of cases. Hence, all patients with ocular lymphoma should have a complete workup to rule out systemic lymphoma. The usual presentation of intraocular lymphoma is decreased vision with nonresolving uveitis, along with proptosis and visible conjunctival mass. It may be of following histology: mucosa associated lymphoid tissue (MALT)

histology (57%), follicular lymphomas (19%), diffuse large B-cell lymphomas (DLCL), mantle cell lymphomas, B-cell chronic lymphocytic leukemia, peripheral T-cell lymphoma, and natural killer cell lymphoma. Out of which ocular adnexal mucosa-associated lymphoid tissue (MALT) lymphoma (57%) is the commonest one, and it's usually associated with Chlamydia psittaci 2, 4, 6. Majority of the orbital lymphomas are of low-grade variety (84%) and only 16% are of high-grade histology<sup>5</sup>. Orbital lymphoma may be unilateral or bilateral and up to 20% bilateral presentation is noted. Although intraorbital lymphoma is rare, the number of cases per year is rising, affecting mainly people in their seventies<sup>7, 8</sup> and immunocompromised patients<sup>9, 10</sup>. A recent study has shown that ocular lymphoma is more prevalent in women than men<sup>11</sup>. The survival rate is approximately 60% after 5 years. Total or subtotal excision of the mass followed by radiotherapy and/ or chemotherapy has a better outcome in these patients. However, radiation exposure predisposes to the development of cataracts after 3 – 8 years. As MALT lymphoma has the best prognosis, DLCL prognosis can be improved by early prompt diagnosis and combination chemotherapy. The role of radiotherapy in DLCL is unclear. Combination chemotherapy with CHOP is quite efficacious, while addition of intravenous rituximab (anti CD 20 monoclonal antibody) in the regimen helps in rapid remission with good results<sup>12, 13</sup>. As the patient is CD 20 strongly positive, we had chosen rituximab as the chemotherapeutic agent along with other chemotherapeutic agents. The patient is asymptomatic even after one year of last dose of chemotherapy.

**Conclusion:**

Lymphoma can occur in the orbit without any systemic manifestations. A palpable painless mass of an eye particularly in elderly female must be diagnosed early in case of an orbital lymphoma before any bony destruction starts and that will improve the prognosis of the patient. In case of DLCL though a rare form, can be treated with the surgical debulking followed by combination chemotherapy. R-CHOP chemotherapy regimen is quite efficacious in treating the case, without any recurrence at 1 year follow-up.

**Conflict of interest** :None

**Acknowledgement:**

Department of Tropical Medicine, School of Tropical Medicine

**Ethical approval:**

This case report was published with prior ethical approval from the authority of School of Tropical Medicine

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