

CASE REPORT

RETRO-BULBAR PRIMARY NON-HODGKIN'S LYMPHOMA: A RARE CASE REPORT

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ABSTRACT: A 65 years old male patient presented with gradually increasing painless swelling of both eyes (left eye more than right eye) associated with diminution of vision in left eye for last one year. Patient underwent thorough examination, proper investigation and diagnosed as non-Hodgkin's lymphoma. Patient was put on chemotherapy and the immediate response was dramatic.

KEYWORDS: Non-Hodgkin's lymphoma, Chemotherapy.

INTRODUCTION: An increasing number of cases of ocular non-Hodgkin's lymphoma is being reported while ocular involvement in Hodgkin's lymphoma is rare.¹ There are two broad forms of non-Hodgkin's lymphoma: primary oculo-central nervous system non-Hodgkin's lymphoma and systemic non-Hodgkin's lymphoma with secondary metastatic ocular involvement.^{2,3} Previously non-Hodgkin's lymphoma was called ocular reticular cell sarcoma but now it is known as large cell non-Hodgkin's lymphoma. Primary ocular non-Hodgkin's lymphoma mainly involves retina and vitreous. Systemic spread is common and central nervous system being affected first.

There are 60-90% chances of primary non-Hodgkin's lymphoma to spread to CNS while primary CNS lymphomas are associated only in 12-15% of cases with ocular involvement.²⁻⁴ Ocular non-Hodgkin's lymphoma is estimated to occur as oculo-CNS disease in 6%, ocular in 17%, oculo-visceral in 17% and oculo-visceral-CNS in 5% of the cases.²⁻⁴ Primary ocular non-Hodgkin's lymphoma usually occur in 6th -7th decades of life with no sexual and racial predilection. Patients usually present with unilateral or bilateral painless loss of vision. Primary ocular non-Hodgkin's lymphoma typically masquerades as uveitis.^{2,3}

Most of the time patients are misdiagnosed as intermediate or posterior uveitis and receive steroid treatment. Early diagnosis is only possible by careful clinical examination and a high degree of suspicion. Diagnosis is achieved by vitreous aspiration and histopathological examination of the specimen. Recent protocol of treatment of non-hodgkin's lymphoma is chemotherapy followed by radiotherapy. Recurrence can occur at the same site or elsewhere which eventually leads to fatal.

CASE REPORT: The patient was a 62 years old male barber, resident of South Tripura district, India. He was admitted in eye ward of Agartala Government Medical College & Govinda Ballabh Pant Hospital, Agartala, Tripura with chief complaints of proptosis of both eye for one year. It was more in the left eye, gradual in onset and slowly progressive in nature with diminution of vision both eye. His past history, family history and allergic history were insignificant. His general and systemic examinations were also within normal limits. In local examination, there was proptosis of both eyeballs. It was 6 mm in right eye and 14 mm in left eye. It was axial, non-compressible and non-

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pulsatile with normal movement of both eyeballs. Visual acuity was 6/9 in right and 6/12 in left eye. IOP and differential IOP were within normal limits. In posterior segment examination, macular edema in left eye was found.

After 3 days visual acuity in right eye was same but it was found to be decreased to 6/18 in left eye. This vision was further decreased to 6/12 in right and 6/60 in left eye. All routine investigations were within normal limits. HIV, HBV, HCV and VDRL tests were non reactive. Chest x-ray and ultrasonography of abdomen were normal. But in B-scan ultrasonography of orbit retro-bulbar mass was found in both eyes. CECT of orbit also showed retro-bulbar mass in both eyes suggestive of probable non-Hodgkin's lymphoma, pseudotumor or secondary involvement with no intra-ocular involvement. CECT of brain also revealed no intra cranial involvement. Histopathology of ultrasound guided biopsy showed features suggestive of non-Hodgkin's lymphoma. In immunohistochemistry the mass was positive of markers CD21 and CD79.

These are markers of large B-cell lymphoma which is a type of non-Hodgkin's lymphoma. Vitreous aspiration was planned but the patient refused. After confirming the diagnosis the patient was transferred to cancer wing of the hospital. Patient was put on cyclophosphamide, doxorubicine, vincristine, azathioprine and prednisolone. On 25th day of chemotherapy the patient had complete regression of proptosis in both the eyes and vision also became 6/6 in right and 6/9 in the left eye. Macular oedema of left eye was also found to be subsided. On 45th day of follow up CECT of orbit was also found to be within normal limits.

DISCUSSION: The patient was 65 years old. Usually ocular non-Hodgkin's lymphoma mainly involves the elderly.⁵ It is not always true that the disease always spares the younger. Recently a 15 yrs old patient has reported.² Here the patient was male, which is similar with the observation of Whitcup SM et al.⁴ In patients of ocular CNS non-Hodgkin's lymphoma ocular symptoms precedes CNS involvement.^{2,3,4} But in visceral non-Hodgkin's lymphoma patients already have signs and symptoms of systemic diseases before ocular involvement.^{3,4} Ocular non-Hodgkin's lymphoma typically masquerades uveitis.⁶ This is often associated with multiple small chorioretinal lesions.^{7,8} These cases are initially diagnosed as birdshot retinopathy or multifocal pigment epitheliopathy.⁹

If cases of chronic uveitis do not respond to steroids there may be possibility of lymphoma. A high degree of suspicion should be kept in mind if a case of chronic uveitis does not respond to steroids. Diagnosis is confirmed by vitreous aspiration and histopathological examination.¹⁰ Recent protocol for treatment of non-Hodgkin's lymphoma is chemotherapy followed by radiotherapy. The usual dose of radiotherapy ranges between 40-50 Gy.³ Patients with ocular non-Hodgkin's lymphoma has limited life expectancy. Only a few patients survive more than 3 years. Typically the patients respond favorably during the initial phase of treatment. However, most of them show recurrences at the same site or elsewhere and these eventually turn out to be fatal.

CONCLUSION: Primary ocular non-Hodgkin's lymphoma is difficult to diagnose. Diagnosis is often delayed and is only considered after a variable period of uveitis which is not responding to steroid therapy. A high index of suspicion is the key stone of diagnosis. Treatment of choice is chemotherapy followed by radiotherapy. Initial response to treatment is usually dramatic but recurrence may occur at the same site or elsewhere which is eventually fatal.

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Before chemotherapy:



Pic. 1



Pic. 2

After chemotherapy:



Pic. 3



Pic. 4

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