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Primary Non-hodgkin's Lymphoma of the Orbit: A Case Report

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Abstract:

Primary orbital lymphomas are a rare subset of tumors constituting 1–2% of non Hodgkin's lymphoma. They are mostly indolent B cell lymphomas presenting with gradual progressive proptosis, decreased visual acuity, restricted ocular mobility, and diplopia. In this paper we report a case of early stage orbital lymphoma in a Muslim male aged 75 years, who presented a left orbital swelling with significant proptosis. Ophthalmological examination complemented by CT has objectified orbital mass, whose biopsy was in favour of a non Hodgkin's lymphoma (NHL). Patient was treated successfully with multimodality treatment, including surgical excision as a major treatment modality. The role of surgery is mainly for obtaining a biopsy. Most of these tumors require multimodality treatment including chemotherapy, radiation, or both, which have major role.

Keywords: *Non-Hodgkin's lymphoma, orbital lymphoma.*

Introduction:

Primary orbital lymphoma is a rare condition and site for non-Hodgkin's lymphoma. The most common type of NHL is diffuse large B-cell lymphoma. NHL can have extra- nodal presentation in 25% to 35% of patients. However, extra-nodal lymphoma of the head is very rare[1]. Orbital lymphomas represent 8 to 10% of all extranodal NHL, and only 1% of all lymphomas[2]. Orbital lymphoma usually presents with proptosis, periorbital swelling, conjunctival ("salmon- pink") swelling, diplopia, and conjunctival redness and irritation. It can remain indolent for a long period of time before the patient notices the periorbital abnormality often seen are proptotic cases in association with various degrees of ptosis[3–7].

Case Report:

An old man aged 75 years, who present a left eyelid swelling with gradually increasing volume and forward bulging of the eyeball for six months. Ophthalmological examination disclosed an orbital mass, developed outside the eyeballs with extension to the upper eyelid (Figure-1). Levo-version, levo-elevation and levo-depression restricts in ocular movement of left eye. CT scan (Figure-2) of the orbit revealed a definite soft-tissue mass in the superior orbit. Orbital MRI was performed and disclosed a lesion measuring 2x1 cm at upper outer quadrant of left orbit adjacent to the lacrimal gland and the upper eyelid, which is iso to hypointense in T1WI and iso to hyperintense in T2WI in comparison to muscle(Figure 3). Biopsy with histopathology study was for low grade non-hodgkins lymphoma. Plans for treatment with chemotherapy and radiation were initiated.

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Fig 1: *Patient presented with left eyelid ptosis.*

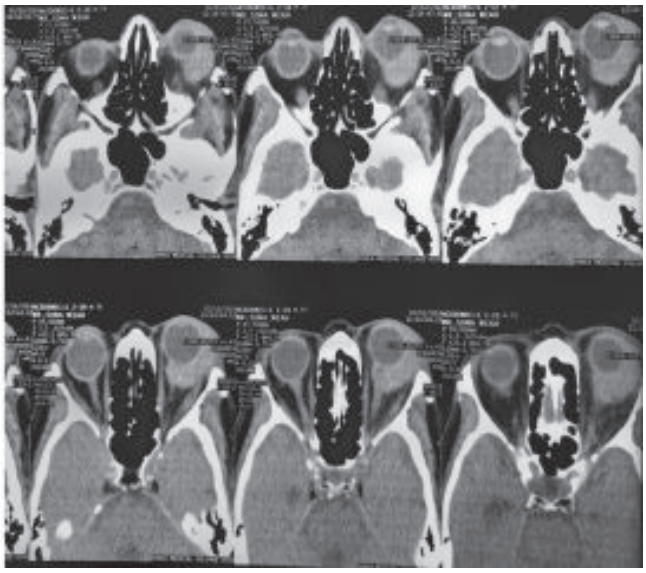

Fig 2: *CT scan of the orbit revealed a definite soft-tissue mass in supero-lateral orbit*

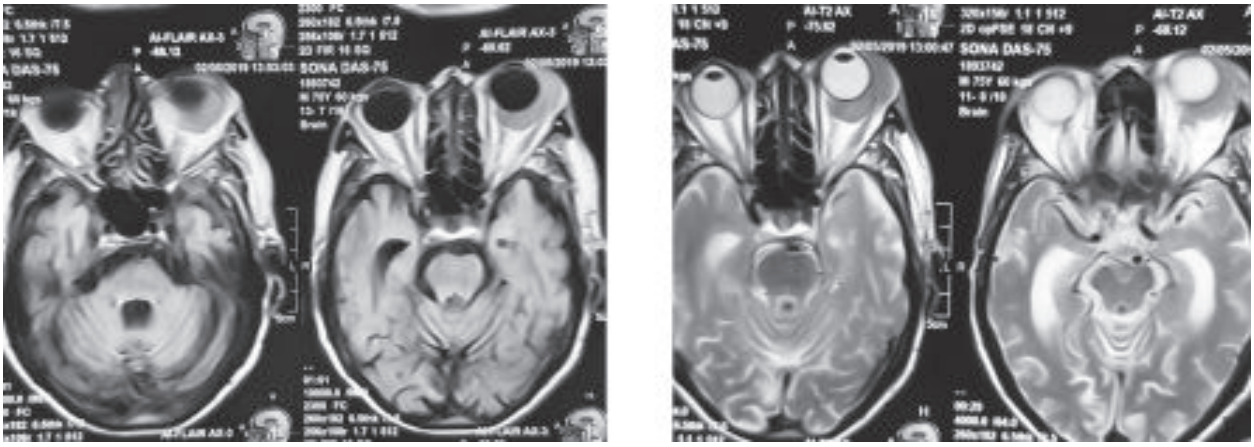

Fig 3: *MRI scan of the orbit revealed a definite soft-tissue mass in supero-lateral orbit*

Discussion:

Orbital lymphoma is reported as the most common malignant tumor of ocular adnexa constituting 55% of all orbital tumors. The occurrence of primary orbital lymphoma, on the other hand, is exceedingly rare and comprises approximately 1% of non-Hodgkin's lymphoma and 8% of extranodal lymphoma. Such patients usually do not give any history of prior lymphoma and do not show any manifestation of systemic lymphoma. Majority of them are B-cell lymphomas, and marginal zone lymphomas represent the most common subtype.[8]

Non-Hodgkin's lymphoma is a heterogeneous group of malignant lymphomas that reflect the development stages of lymphocytes, with the majority arising from a B-cell origin. Even though primary orbital lymphomas represent 1% of NHLs and 8% of extra-nodal NHLs, POLs make up about half of adult primary orbital malignancies.[2,9]

The mean duration of symptoms of a POL at presentation is approximately 18 months (range: 10 days to 10 years). Less than 20% of patients present with any kind of visual acuity loss, and complete vision loss is rare, especially without other common symptoms.

MRI usually shows hypointensity particularly on T1- weighted images and homogenous enhancement.[9] Most orbital lymphoid tumors tend to conform to structures around them without bony erosions. The MRI images of our patient do agree with some of these findings such as

hypointensity on T1-weighted imaging; however, our patient's lesion did show intense contrast enhancement.

Conclusion:

This case highlights the important role of diagnostic imaging for achieving successful diagnosis and treatment plan for patients having an early stage orbital lymphoma.

References:

1. Newton R, Ferlay J, Beral V, Devesa SS. The epidemiology of non- Hodgkin's lymphoma: comparison of nodal and extra-nodal sites. *Int J Cancer* 1997; 72(6):923-30.
2. Freeman C, Berg JW, Cutler SJ. Occurrence and prognosis of extranodal lymphomas. *Cancer* 1972; 29: 252-60.
3. Van der Gaag R, Koornneef L, van Heerde P, et al. Lymphoid proliferations in the orbit: malignant or benign? *Br J Ophthalmol* 1984; 68:892–900.
4. Sarah EC, Michael H, Harald S. Ocular adnexal lymphomas: five case presentations and a review of the literature. *Survey Ophthalmol* 2002; 47:470–90.
5. Tranfa F, Di Matteo G, Strianese D, et al. Primary orbital lymphoma. *Orbit* 2001; 20:119–24.
6. Coupland SE, Krause L, Delecluse HJ, et al. Lympho-proliferative lesions of the ocular adnexa. Analysis of 112 cases. *Ophthalmology*. 1998; 105:1430–41.
7. McKelvie PA, McNab A, Francis IC, et al. Ocular adnexal lympho-proliferative disease: a series of 73 cases. *Clin Exp Ophthalmol* 2001; 29:387–93.
8. Stefanovic A, Lossos IS. Extranodal marginal zone lymphoma of the ocular adnexa. *Blood* 2009; 114:501-10.
9. Volpe NJ, Gausas RE. Optic nerve and orbital tumors. *Neurosurg Clin N Am*. 1999; 10(4): 699-715.