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A RARE CASE OF EXTRAOCULAR ORBITAL LYMPHOMA IN A YOUNG PATIENT

Ophthalmology

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ABSTRACT

Orbital lymphoma refers to a lymphoma that commonly involves the conjunctiva, lacrimal gland, eyelid and ocular musculature.[1] Primary non-Hodgkin's lymphoma (NHL) of the orbit is a rare subset, representing only 1% of all NHL and 8-10% of extranodal NHL.[1] We report a case of 24 year old female presented with redness, swelling, an intermittent pain and gradual protrusion of the right eye (RE) since 6 months. Her vision in RE was 6/60 and in Left eye (LE) 6/6. Intraocular pressure (IOP) was 16 mmHg in Both Eye. On examination, RE was proptosed 30mm approximately. Extraocular movement were restricted in all gazes, conjunctiva was chemosed and congested. LE was within normal limits. No lymph nodes were palpable. Ultrasonography of the right orbit showed a mass in the lateral aspect in extraconal space extending to the superior, lateral and retrobulbar region. CT scan of the right orbit showed a mass encasing the extraocular muscles and displacement of the optic nerve. FNAC reports were suggestive of non-Hodgkin's Lymphoma of the right orbit. She was given 6 cycles of RCHOP regime which includes (rituximab, cyclophosphamide, hydroxydaunorubicin, oncovin and prednisolone). Follow-up during each cycle of chemotherapy was done and showed good response initially. **Conclusion-**Even though rare, primary orbital NHL should be kept in consideration in a young female patient with slow growing unilateral Proptosis. After imaging & diagnostic biopsy, patient should be immediately staged & treated. Early diagnosis and treatment improves survival.

KEYWORDS

non-Hodgkin's Lymphoma, proptosis, retrobulbar.

INTRODUCTION :

Orbital lymphoma refers to a lymphoma that commonly involves the conjunctiva, lacrimal gland, eyelid and ocular musculature.[1] Primary non-Hodgkin's lymphoma (NHL) of the orbit is a rare subset, representing only 1% of all NHL and 8-10% of extranodal NHL.[1] Most of them are primary extranodal lymphoma of the marginal zone of mucosa associated with lymphoid tissue (MALT type lymphoma).[2] Majority of the orbital lymphomas are of low-grade variety (84%) and only 16% are of high-grade histology.[2] Recent advances in molecular and cytogenetic establish that lymphoma can be associated with Chlamydia psittaci infection.[3]

Case Scenario

A 24-year-old female presented with redness, swelling and an intermittent pain in Right eye (RE) for 6 months. There was bulging of the RE since 1 year, which was of insidious onset and gradual progression [figure 1].

Figure 1: Unilateral Proptosis of RE at initial presentation.

There was loss of appetite and weight since 1 year. There was no history of feeling of pulsation within the orbit. She doesn't give any history of diplopia. Past history of thyroid disorder, associated systemic malignancies of lung, breast was absent. The family history was non-contributory.

On Examination

Her visual acuity was 6/60 in RE and 6/6 in LE uncorrected. There was protrusion of right eyeball more than 30 mm approx. from lateral orbital rim to corneal apex, measured Luedde's exophthalmometer. The Proptosis was eccentric, with downward and outward displacement of the right eyeball. Extraocular movement was restricted in all gazes in RE [figure 2].

Figure 3: Restricted motility of right eye in all gazes.

There was ptosis of RE due to fullness of right upper eyelid with absent lid crease. A superior orbital mass was palpable which was non-tender, firm and rubbery in consistency, displacing the eyeball infero-laterally [figure 3],

Figure 3: showing Proptosis of right eyeball.

however the posterior extension of the tumor mass couldn't be elicited. There was absent orbital pulsation/ thrill and the mass was not reducible. The orbital margins/ rim was rounded and regular. The tumor mass was not adherent to the overlying skin but was adhered to the underlying tissues Intraocular pressure was 16mmHg in both eyes. Conjunctiva was severely chemosed & congested in right eye with mucopurulent discharge. Exposure keratopathy was present in lower part of RE with fluorescein stain positive however the corneal sensation was intact. Pupils were round regular and reactive to light. Lens was clear, fundus was within normal limits. Left eye examination revealed normal findings. No organomegaly or lymphadenopathy was noted.

Investigation

USG B scan reports revealed ill-defined homogenous hyperechoic lesion in lateral aspect of RE in extraconal space extending to superior, lateral & retrobulbar region [figure 4].

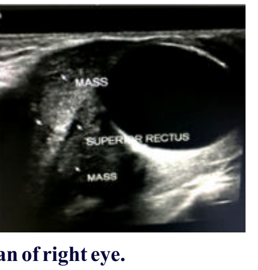

Figure 4: USG B-scan of right eye.

CT scan of orbit shows lobulated isodense soft tissue density mass in extraconal spaces of right orbit, encasing the nearby extraocular muscles & displacing the optic nerve [figure 5].

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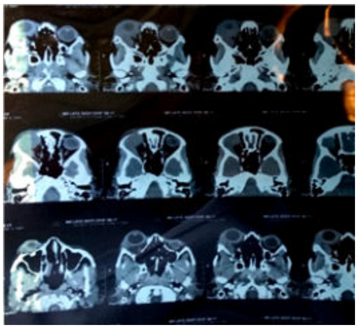

Figure 5: axial non-contrast ct scan orbit showing lobulated isodense soft tissue density mass in extraconal spaces of right orbit.

On FNAC, there were sheets and clumps of monomorphic round cells predominantly centroblasts, centrocytes & immunoblasts with high hyperchromatic nuclei which was suggestive of Non-Hodgkin's Lymphoma [figure 6].

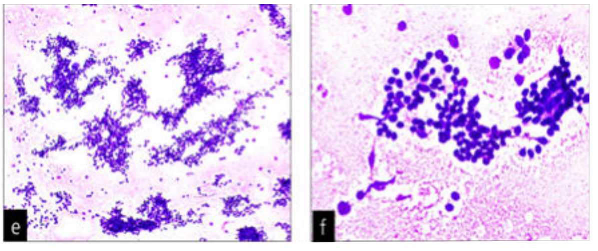

Figure 6: showing FNAC of the mass suggestive of Non-hodgkin's Lymphoma of right orbit.

Figure 7: Follow up after 2nd cycle of chemotherapy showing reduction of Proptosis of right eyeball.

A complete systemic examination was done to rule out metastatic lymphoma to the orbit included CT of the chest, abdomen and pelvis as well as renal and liver function tests. A complete haematological examination with blood count and bone marrow aspiration was also done. There was no evidence of any lymphoproliferative disease or abnormality elsewhere. Once the diagnosis was confirmed the patient was staged as Stage IB, according to Ann Arbor classification.[5]

The patient was treated with 6-8 cycles of Chemotherapy (RCHOP regime) every 3 weekly. Artificial tear substitute, topical antibiotic was given and eyelid tapping was done for exposure keratopathy. Follow-up after the 2rd cycle of Chemotherapy showed reduction of swelling and conjunctival chemosis [figure 7]. The tumour showed recurrence following the 5th cycle of chemotherapy. The patient was advised to undergo surgery but she refused to undergo surgery.

DISCUSSION

Primary NHL of orbit is a rare presentation, representing 8-10% of Extra-nodal NHL & only 1% of all NHL.[1] Usually NHL occurs in the age group of 50- 70 years however it has occurred at an unusually young age in our patient.[4] CT orbit and FNAC of the mass helps to differentiate it from other orbital tumours. Essadi et al.[4] have also showed successful management with RCHOP regimen in a similar case of orbital lymphoma. Periodic follow-up of the patient is mandatory as there is still 30-40% chance exists of developing systemic lymphoma within 5 -10 years. However, there were certain challenges encountered in management of the above case due to lack of awareness, knowledge and social taboos and the patient was lost to further follow up.

CONCLUSION

Even though rare, primary orbital NHL should be kept in consideration in a young female patient with slow growing unilateral Proptosis. After imaging & diagnostic biopsy, patient should be immediately staged & treated for better prognosis & rate of survival.

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