



## RESEARCH ARTICLE

### PRIMARY ORBITAL LYMPHOMA - A CASE REPORT FROM MAURITIUS

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#### Abstract

We report the case of a fifty-two year old lady with a primary large B-cell Non-Hodgkin lymphoma, NHL, of the orbit which is a rare presentation of NHL. The tumour was amenable to chemotherapy and radiotherapy. Lymphomas are malignant neoplasm arising from the lymphoid cells (Kumar *et al* 2005). They are broadly classified into two groups namely the Hodgkin Lymphoma, HL and NHL. Lymphomas are not rare in occurrence; they are considered as the eighth and ninth leading cause of death due to cancers in women and men respectively in the United State in 2017 (Longo *et al* 2012). NHL occurs 10 times more frequently than the HL and in general has a poorer prognosis as compared to the latter (Cancer treatment center of America, 2017).

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#### Introduction:-

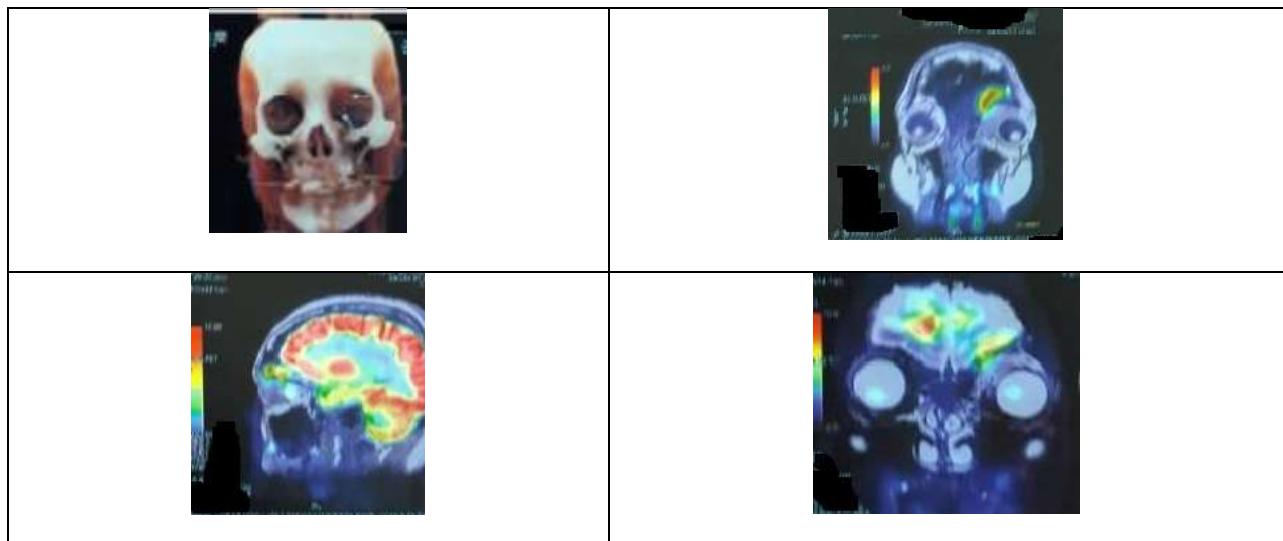
Primary Orbital lymphomas are rare tumours constituting of about only 1 % of all Non Hodgkin Lymphomas (Eckardt *et al*, 2013; Mallick *et al*, 2015; Schalenbourg and I. Mantel. 2015). However, orbital lymphomas are considered to be the most common type of malignant tumours of the orbit and the orbital adenexa (Margo & Mulla 1998). Such tumours have a slight higher female preponderance, affecting particularly individuals in the age group 50-70 years more often (Stefanovic A & Lossos IS. 2009). Generally, they are painless and slow growing B-cell lymphomas such that they are usually detected accidentally or detected only when the patient presents with pressure symptoms which occur when the tumour has become of a significant size. Primary orbital lymphomas are usually confined to the orbit. However, in certain instances, when left untreated, they have been found to spread to the adjacent organs by eroding the bony barrier. Radiotherapy is considered as curative treatment for the low grade tumours while a combination of radiotherapy and chemotherapy are preferred for high grade tumours (Mallick *et al*, 2015).

Lymphoma accounts for 3.13% of all malignancies in the female population of Mauritius in the year 2017. The incidence of Non-Hodgkin lymphoma in the same year was 1.4% while the incidence of malignancies affecting the eye, brain and CNS is 0.9%. However, the incidence of lymphomas affecting the orbit and orbital adenexa is not documented in Mauritius (National Cancer Registry, 2017). Here, we report the case of a low grade B-cell primary lymphoma of the orbit that was treated with chemotherapy as major modality of treatment. Radiotherapy was ultimately used for eradication of a small remnant mass.

#### Case Report:

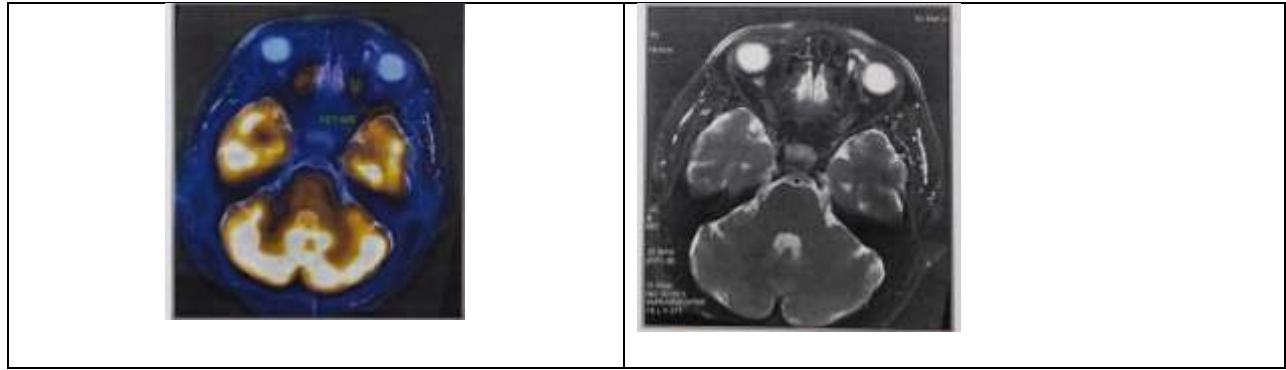
A 52-year-old lady presented to her general practitioner with complaint of a swelling in the upper part of her left orbit which she reported as has gradually been increasing in size for the past 3 months. She also complained of

progressive bulging of the left eye as well as drooping of the eyelid on the same size. The left eye was deviated downward and laterally with significant proptosis and mild restriction of medial movements of the left eye. The swelling was non-pulsatile, painless and not associated with any visual complaint in the affected (left) eye. Vision was 6/6 in the left eye and 6/6 in the right eye. There was neither history of any thyroid disorders, palpable organomegaly nor any enlarged lymph node. Furthermore, rest of the cranial nerves and neurological assessment was normal. Magnetic resonance imaging (MRI) of the left orbit and brain revealed an extraconal mass measuring  $4.8 \times 2.2 \times 1.7$  cm. The mass was isointense to muscle on T1 W1 and T2 W1 and hyperintense on STIR images. No other abnormalities were noted on the above MRI. The MRI report was suggestive of a possible Lymphoma. The patient was referred for specialised care where a positron emission tomography, PET scan and an incisional biopsy was performed. The PET scan report showed the presence of an ill-defined soft tissue mass of  $2.2 \text{ cm} \times 2.4 \text{ cm} \times 2.7 \text{ cm}$  in the medial aspect of the upper eyelid and supero-medial anterior orbit of the left side (figure 1). The lesion infiltrated the soft tissue of the upper eye-lid and encased the anterior aspect of medial rectus, superior rectus, levator palpebrae superioris and superior oblique. Moreover, the lesion appeared to be adherent to the globe near the equator and displaced the globe inferolaterally. However, no bony erosion was noted. The biopsy result confirmed the mass as being a low grade B-cell Non Hodgkin Lymphoma.



**Fig 1:-** Showing different views of the mass on PET scan prior to chemotherapy.

The patient was advised for radiotherapy but instead, she opted for chemotherapy due to fear of losing her eye-sight as a consequence of radiotherapy treatment. Four cycles of R-CHOP (Rituximab, Cyclophosphamide, Adriamycin, Vincristine and Prednisone) regimen was administered at 21 days interval each. After 21 days of the fourth cycle of chemotherapy, PET scan was repeated and the result revealed an enhancing soft tissue lesion of size  $15 \times 10 \times 8.5$  mm in the supero-medial portion of the left orbit with no metabolic activity without any intraconal extension, optic nerve involvement and adjacent bony erosions (figure 2). The Chemotherapy proved to have completely inhibited the metabolic activity of the left eye lesion although a remnant of the mass was noted. Subsequently, the patient was counselled and subjected for few sessions of radiotherapy which was found to remove the remnant mass completely without affecting the vision of the patient.



**Fig 2:-** PET scan showing size of the tumour post chemotherapy.

### Discussion:-

The occurrence of primary orbital lymphomas is considered as extremely rare, constituting of about only 1% of cases of NHL (Stefanovic&Lossos 2009). Radiotherapy, chemotherapy, surgery, immunotherapy or a combination of these modalities have been advocated in the management of orbital lymphomas (Sharma & Kamath 2015). Several studies have shown that the primary modality of treatment of primary low grade orbital lymphoma is radiotherapy which on average has shown a local control rate of 83 to 100% and a five-year disease-free survival of 62 to 100% (Yadav & Sharma 2009). However, despite radiotherapy has proved to be curative as the sole modality treatment in low grade primary orbital lymphomas, this mode of treatment is very challenging because of the presence of the radiosensitive lens, lacrimal gland, and retina which may lie in close proximity with the tumour such that any damage to these structures may lead to irreversible blindness.

The role of Chemotherapy, on the other hand, in the management of localised primary orbital lymphoma is not well defined. The Local control rate with chemotherapy was reported as 42% compared to 100% with radiotherapy (Esiket *et al.* 1996). Moreover, studies have shown that chemotherapy has not been found to have any effect on the disease-free or overall survival in localized low-grade NHL and its use for the initial management of primary orbital Non-Hodgkin's Lymphoma has been associated with a delay in the deployment of radiotherapy. Consequently, it delays the local control of the disease which in turn can promote a slow systemic progression of the disease and hence aggravate the condition (Pelloskiet *et al.* 2001).

As for the role of surgery in the management of primary orbital lymphoma, it is limited only to excisional biopsy which helps from a diagnostic point of view (Jain *et al.* 2018). Furthermore, as a sole modality of treatment, surgical excision can lead to more dreadful complications (Sharma & Kamath 2015).

With reference to the above case, we found that chemotherapy may be useful in the management of low grade primary orbital tumours which are of significant size where radiotherapy can prove to be very hazardous to the surrounding tissues. In this case, chemotherapy has shrunk the tumour remarkably such that only limited sessions of radiotherapy was needed and radiotherapy-induced local damage to surrounding structures have been avoided markedly.

### Conclusion:-

Primary non-Hodgkin's lymphoma (NHL) of the orbit is a rare entity which occurs in the orbit without any systemic manifestation. The clinical features are usually non-specific such that a biopsy coupled with adequate imaging studies is mandatory for an early diagnosis of the condition. As demonstrated in the above patient, the role of chemotherapy can be re-evaluated as a debulking agent in the management of relatively big size primary orbital lymphoma before radiotherapy is initiated. However, radiotherapy remains crucial and of paramount importance in the management of low grade B-cell primary orbital NHL.

### Ethical consideration:

An informed consent was taken from the concerned patient. She was reassured that all data will be treated with strict anonymity and confidentiality. All journals and materials used for this paper are clearly referenced.

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**Conflict of interest:**

The authors have no conflict of interest to declare.

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