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PRIMARY ORBITAL LYMPHOMA WITH PROPTOSIS : A CASE REPORT

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ABSTRACT Primary orbital lymphoma constitute for around 10 % of the orbital lymphomas. Among the ocular adnexal lymphomas, around 2/3 cases are of orbital lymphomas. Clinical presentation can vary from proptosis, reduced vision, restricted motility, diplopia to periorbital edema that is seen rarely. Diagnosis is confirmed after histopathological confirmation and the treatment constitutes of surgery, radiotherapy, chemotherapy or a combination of both. We report a case of a 58 year old male patient with primary orbital non Hodgkin's lymphoma who presented with proptosis and its management.

KEYWORDS : Primary Orbital Non Hodgkin's Lymphoma, Proptosis

INTRODUCTION

Lymphoid tumours of the orbit are the most common primary tumours of the orbit. Primary orbital non Hodgkin's lymphoma(NHL) constitute around 4% of all tumours of orbit. Primary orbital NHL is a type of extranodal lymphoma. It is quite rare.(Tranfa et al., 2001) Clinically, it can present as a firm, rubbery mass in the orbit, proptosis, reduced vision, restricted motility, diplopia etc. The management of the disease is multidisciplinary and consists of surgery, radiotherapy or chemotherapy or a combination of both. (Galieni et al., 1997) We report a case of a male patient with primary orbital NHL and its management.

CASE REPORT

A 58 year old male presented to our Ophthalmology department with complaints of prominence of left eye since 6 months. The proptosis was painless and significant. There was restriction of the lateral movement of the left eye . Patient didn't give any history of trauma or any thyroid disorder. Ophthalmological examination revealed the Best corrected visual acuity(BCVA) in both eyes was 6/6 with normal colour vision and Intraocular pressure(IOP) was 18 mmHg in both eyes, measured with applanation tonometry . There was an axial proptosis of 6 mm, noted in the left eye(Figure 1 and 2). The proptosis was non- pulsatile and no ocular bruit was noted. There was a significant restriction in the outward lateral movement of the left eye. Dilated fundus examination was normal, there was no evidence of Optic disc pallor or papilledema. On systemic examination, no organomegaly was noted and no lymphadenopathy was seen.

Figure 1 – Picture showing proptosis in the left eye

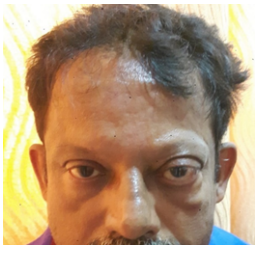


Figure 2 Picture taken from above (Naffziger's method) showing axial proptosis of left eye.

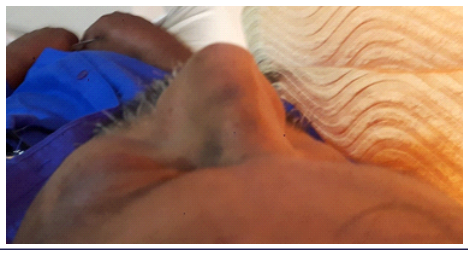
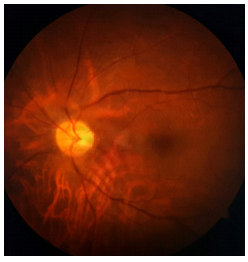
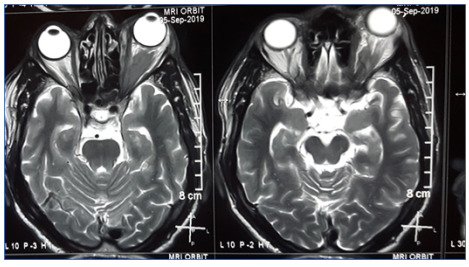


Figure 3 : Fundus picture of the left eye of the patient showing no significant findings



CT scan after i.v. contrast showed evidence of peripheral enhancement of thickened isodense optic nerve. Magnetic resonance imaging(MRI) of the orbit showed the lesion encircling the left optic nerve that was isointense to hypointense in both T1 and T2 weighted image with fairly definite margins. The lesion showed moderate enhancement and was closely abutting the extraocular muscles.

Figure 4 T2 weighted axial images showing iso to hypointense mass lesion encircling the left optic nerve



The patient was taken up for left orbito-zygomatic craniotomy and excision of the space occupying lesion. Histopathology demonstrated sheets of predominantly small to medium sized neoplastic lymphoid cells with scanty cytoplasm, round uniform nucleus with clumped chromatin. A diagnosis of low grade non Hodgkin's lymphoma was established. The patient then received chemotherapy consisting of Cyclophosphamide, Vincristine , Doxorubicin and Prednisolone with Rituximab Immunotherapy followed by radiotherapy.

DISCUSSION

Orbital lymphomas account for around 10 % of the orbital tumours. These lymphoid tumours constitute around 2% of nodal and extranodal lymphomas. (Tranfa et al., 2001) Among the lymphomas of ocular adnexa , around 2/3 cases are of orbital lymphoma. These orbital lymphomas comprise mostly of low grade B cell type. (Galieni et al., 1997) Orbital lymphomas most commonly occur in patients of age 50-70 years and are more common in females in comparison to males. In this case, the patient was a 58 years old male. Commonly, proptosis is the presenting complaint and other clinical features such as

reduced visual acuity, restricted eye movements, double vision and rarely periorbital edema can also be seen. This patient had progressive proptosis with restriction of abduction of left eye.(Eckardt et al., 2013; Priego et al., 2012)

Complete clinical examination, CT scan ,MRI and histopathological examination are required to confirm the diagnosis. (Khmamouche et al., 2017) A differential diagnosis of Optic nerve sheath meningioma was also considered as per the presentation of axial proptosis, history and initial CT scan reports. Since, the mass was abutting the extraocular muscles , there was a restriction of ocular movements noted .For confirmation by histopathology and for decompression, craniotomy was done.

The treatment of orbital lymphomas are multidisciplinary including surgery, chemotherapy, radiotherapy or their combination. Surgery is used as a tool to aid in diagnosis .Surgery as the only treatment modality isn't sufficient and is linked with increased chances of relapse.(Borkar et al., 2018) This patient after the craniotomy and histopathological confirmation ,received chemotherapy including Cyclophosphamide, Vincristine, Doxorubicin, Prednisolone and Rituximab monotherapy followed by radiotherapy.

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