PRIMARY INTRAORBITAL B-CELLS LYMPHOMA. A CHALLENGING DIAGNOSIS. CASE REPORT AND REVIEW OF THE LITERATURE

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INTRODUCTION

Orbital lymphoma is a type of non-Hodgkin lymphoma (NHL) that originates in the conjunctiva, lacrimal gland, soft tissues of the eyelid, or extraocular muscles; it is most commonly extraconal in location. Orbital lymphoma it is said to be primary when it arises spontaneously from one of these locations, and secondary when it is associated with metastatic spread from an extraorbital site. While lymphoma constitutes more than half of all orbital malignancies (55 percent), the incidence of orbital lymphoma has been reported to account for between 1 and 10 percent of NHL cases.

DIAGNOSTIC WORKUP

The following evaluations form the basis of a good diagnostic workup, undertaken in conjunction with the patient's internist: History and physical examination Dilated fundus examination, Thorough examination of opposite orbit as well as the oral cavity and oro-pharynx, Complete blood count, biochemis try profile, Fine-needle aspiration and biopsy, Liver function tests, renal function tests, Chest x-ray, Computed tomography (CT) and magnetic resonance imaging (MRI) of orbit, abdomen, thorax, and pelvis, and bone marrow aspiration also is indicated.

DISCUSSION

Orbital lymphoma may be unilateral or bilateral. Although it has been known to present in patients between 15 and 70 years of age, most cases cluster around the seventh decade. Historically, a female preponderance has been noted. Geographically, the disease is most common in Asia and Europe. Almost 80 percent of orbital and adnexal lymphomas are of low-grade variety, with B-cell lymphomas and extranodal marginal zone lymphoma of the mucosa-associated lymphoid tissue type being the most common histological diagnoses. MALT lymphomas comprise a distinct set of tumors that exhibit the unique property of "homing," which permits lymphoma cells to adhere to other epithelial and mucosal sites, thus enabling bilateral involvement B-cell lymphoma has shown 5-year relative survival rate in localized cases 73%, regional 72% and distant 55%

Fig 1 and 2: Pre operative photographs

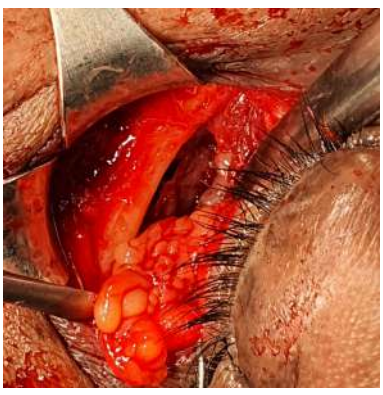
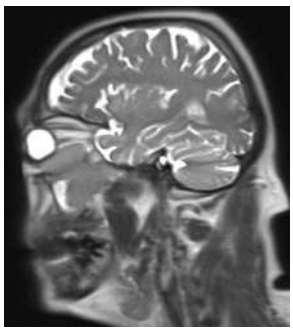


Fig 5 incisional biopsy of intraorbital lesion



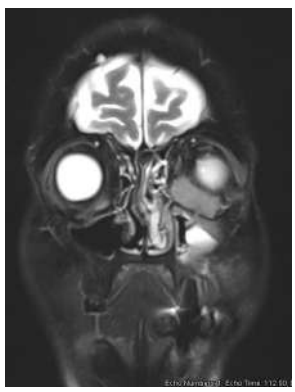


Fig 3,sagital MRI section Fig 4 coronal MRI section

Fig 6 Post treatment

CASE PRESENTATION

A 52 years old Indian male patient, known to have hypertension and type II diabetes presented to Emergency Department complaining of 4 days history of gradual onset blurring of vision, left eye pain and double vision. Clinical examination showed left eye proptosis, limitation to abduction gaze, tense and hard to digital palpation, and binocular diplopia. MRI Head and Orbit showed left intraorbital extraconal mass lesion seen along the inferior aspect of the left orbit displacing the inferior rectus muscle superiorly and pushing the eye globe forward extending to the left maxillary sinus through and the most medial part of the left nasal cavity. PET CT Scan whole body showed Left testis/epididymis uptake along with additional foci at the ductus deferens also compatible with lymphoma activity. Under general anesthesia Incisional biopsy was done using left sub ciliary incision, mass was identified in the supraperiostal plane and three main specimens incised and sent to histopathology investigations. Histopathology report showed Large B-cell lymphoma, activated B cell type. After diagnosis, case was referred to hemato-oncology and it was staged as ***Diffuse large B-cell lymphoma, NOS, activated B cell type Stage IV.*** Patient received total two cycles of Hyper-CVAD and two cycles of R-CHOP showing regression of the lesion on PET CT scan as well as regression of the symptoms.

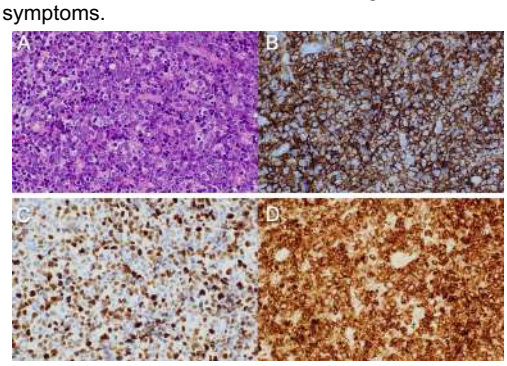


Figure 1. Diffuse large B cell lymphoma not otherwise specified (DLBCL, NOS). A. Left intra-orbital mass, incisional biopsy showing atypical lymphoid cells with mitosis (x 400). B-D. Immunohistochemistry (x 400); Tumor cells are positive for B. CD20, C. MUM-1, and D. BCL-2.

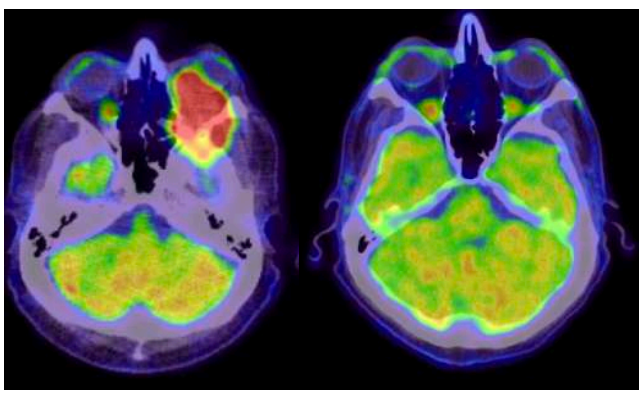


Fig 7 Comparison of PET CT Left side before Right side after

CONCLUSSION

Orbital lymphoma can be easily mistaken for another ocular disease due to the slowly progressing nonspecific complaints of the patients. If unspecific orbital symptoms are present, adequate imaging studies followed by early surgical biopsy will contribute to the early diagnosis. We should always be suspicious of this diagnosis especially in patients over sixty years of age with slowly growing mass in the orbit or proptosis non-reacting to specific treatment.

CONFLICT OF INTEREST: WE HAVE NOT CONFLICT OF INTEREST RELATED TO THIS PRESENTATION

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