Case Report

A Rare Case of Orbital Lymphoma

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Abstract:

Orbital lymphoma without systemic involvement is rarely seen in ophthalmological practice. We diagnosed one such case in our set-up. An old female presented with diplopia and proptosis right eye. CT-scan revealed an intraorbital mass along the superior orbital wall. MRI orbit showed space occupying lesion involving frontal sinus which was confirmed on histopathological and Immunohistochemical examinations. The histochemical studies confirmed the presence of non-Hodgkin's lymphoma. This is a rare manifestation of non-Hodgkin's lymphoma with primary involvement of orbital mass extending into frontal sinus and cranial fossa with no systemic involvement. This case was being reported for general awareness as the tumour is potentially curable.

Introduction

Primary non-Hodgkin's lymphoma (NHL) of the orbit is seen infrequently. The orbit is rare primary site for NHL, without systemic involvement, accounting for 0.5% of all presentations. However, orbital lymphomas are most common malignancy, consisting 10% of orbital tumors¹.

Case report

A 65 years old female was admitted in PNS Shifa with complaints of diplopia and painless proptosis right eye for past one and half months. There was no other co-morbidity. Corrected vision in right eye (RE) was 6/12 and left eye (LE) was 6/6 unaided. Anterior and posterior segments were normal in both eyes except early cataractous changes. Pupil was fixed nonreactive to light on A 65 years old female was admitted in PNS Shifa with complaints of diplopia and painless proptosis right eye for past one and half months. There was no other co-morbidity. Corrected vision in RE was 6/12 and LE was 6/6 unaided. Anterior and posterior segments were normal in both eyes except early cataractous changes. Pupil was fixed nonreactive to light on RE but reacting on LE. Extra-ocular motility was restricted in all direction of gaze on right side while full and unrestricted on left side. The proptosis was non-axial measuring 6mm. The eyeball was displaced 6 mm

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downwards and 10 mm outward laterally (Fig.1). Diplopia was found in lateral gaze. Intraocular pressure and visual field was normal on both eyes. Mass was firm, non-tender, non pulsatile, occupying the superior orbit and resulted in mild ptosis.

There was no cervical lymphadenopathy and systemic examination was also normal.



Fig.1 Photograph of patient.

On investigation, blood picture was normal. CT-scan orbit showed intraorbital mass 3.5x 3.2x2.5 cm along superior orbital wall with destruction of adjacent superior orbital wall and extension into frontal sinus, anterior and middle cranial fossa. MRI orbit revealed space occupying lesion involving frontal sinus. Trans-septal anterior orbitotomy was done with wedge resection for diagnostic purpose. Histopathology revealed poorly differentiated malignant growth. Diagnosis was made on immune histochemical study which was consistent with diffuse B-cell non-Hodgkin's lymphoma. Patient was referred to oncologist for further evaluation, radiotherapy and chemotherapy.

Discussion

Orbital involvement at the time of diagnosis of NHL is an uncommon presentation. Median patient age is 75 years². In older patients, the evolution of

symptoms tends to occur over a period of 6 to 12 months. In younger patients, malignant lymphoma tends to exhibit a more rapid clinical course with almost invariable evidence of systemic involvement. Orbital lymphomas are usually unilateral but may involve both orbits. It usually presents with painless proptosis, insidious onset, downward displacement of the globe, a palpable non-tender mass and ptosis³. Imaging studies usually confirm the presence of a mass, most commonly in superior and anterior orbit but less commonly deep in orbital apex. Diagnosis is accomplished with computed tomography (CT) and/or MRI. Recent valuable tool is gallium scanning. The treatment of choice is external beam irradiation. Local control of the tumor is excellent with 97% success rate. A promising frontier of treatment is proton beam radiation therapy. Non-Hodgkin's lymphomas are the most chemo-responsive of cancers⁴⁻⁵. However, few patients fail to attain complete remission. Surgery is rarely used⁶. New therapeutic modalities include monoclonal antibodies, recombinant interferons, and interleukin-2⁷.

Conclusion

Orbital involvement of non-Hodgkin's lymphoma is uncommon, but may occur at any time during the course of the disease. It should be suspected in any patient who presents with subtle ocular signs and symptoms.

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