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NON-HODGKINS LYMPHOMA OF ORBIT WITH SYSTEMIC SPREAD- A CASE SERIES

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Abstract : Primary Non Hodgkins lymphoma of orbit is rare, representing 8-10 percent of extranodal NHLs and 1 percent of all NHLs. Of these, High grade lymphomas have an aggressive course and early propensity for systemic spread despite early aggressive treatment. We present 2 cases of Orbital NHL who presented initially with Unilateral upper lid mass, and bilateral proptosis which on further evaluation were diagnosed to have Orbital NHL with tissue biopsy showing Diffuse Large B cell lymphoma in one and mixed variety in other respectively. We started them on chemotherapy. Our first patient defaulted after 2 cycles and came later with inguinal and axillary lymphadenopathy and painful arm swellings. Second patient received 10 cycles of chemotherapy but he developed diffuse Lymphadenopathy and hepatosplenomegaly. Orbital lymphomas, if diagnosed and treated early, have an excellent prognosis. Stringent follow up is mandatory to check any systemic spread and hence improve survival in these patients.

Keyword : Lymphoma, Proptosis, chemotherapy, hepatosplenomegaly, biopsy

INTRODUCTION

Orbit is an uncommon primary site for Non Hodgkin's lymphoma, accounting for less than 1% of all sites of primary presentations.[1] However, Lymphoid tumors are the most common primary orbital malignancies in adults. Majority of them are low grade lymphomas(80%) and respond well to radiotherapy.[2] High grade lymphomas are rare(16%) and are known to have a higher incidence of systemic spread. Hereby, we report two interesting cases of primary orbital non hodgkin's lymphomas with systemic spread.

CASE 1

A 54 year old male came with complaints of swelling in Right Upper eyelid for 1 month duration. The swelling was insidious in onset,painless and non progressive .On examination, A firm, non tender mass noted in the medial canthal region with eccentric proptosis. CT Orbit showed Sino-ethmoidal mass encroaching medial quadrant causing eccentric proptosis (fig 1). We did a biopsy from the mass and Histopathological examination showed Diffuse Large B-cell lymphoma (fig 2).We started him on CHOP chemotherapy (Cyclophosphamide 800 mg/ sq. metre body surface area as i.v infusion over 2 hours, Adriamycin 70 mg/sq.m as i.v infusion over 1 hour, Vincristine 1.4 mg/sq.m i.v bolus over 1 to 2 minutes and Prednisolone 40mg orally twice daily for day 1 to 5), this cycle repeated every 21 days. But, the patient lost to follow up after 4 cycles.

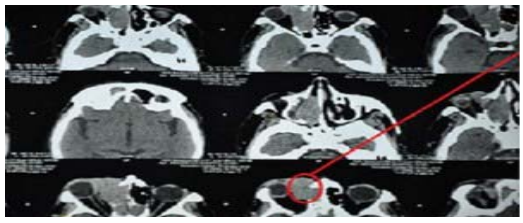

fig 1: CT orbit axial section showing sino ethmoidal mass encroaching medial orbital wall

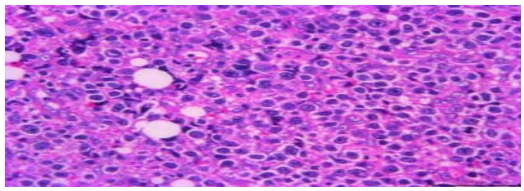

fig 2: Histopathology showing Diffuse large lymphoma cells

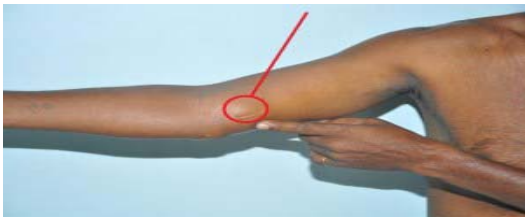

fig 3: Mass in the arm

He presented after 1 year with bilateral focal painful arm swellings (fig 3) and bilateral axillary and inguinal lymphadenopathy which on biopsy showed Diffuse Large B cell Lymphoma. Bone marrow biopsy revealed Mild hyper cellular bone marrow which was suggestive of systemic spread. This time we offered Palliative radiotherapy for arm masses.

CASE 2

A 50 year old male came with c/o bilateral protrusion of eyes for 1 year. He was a known coronary artery disease patient for 8 years. On examination, he had bilateral proptosis with bilateral Submandibular lymphadenopathy(fig.4).Right Incisional biopsy of submandibular nodes showed Small to medium sized B-cell lymphoma(fig.5). Echocardiography showed Left Ventricle Ejection Fraction-35%. So, Adriamycin was withheld for this patient due to its cardiotoxicity. We gave him 10 cycles of COP chemotherapy (Vincristine 2mg as i.v bolus injection, Cyclophosphamide 1gm as i.v infusion over 2 hours and Prednisolone orally 40 mg BD for day1 to 5, each cycle repeated every 21 days) and 1 cycle of Iphosphamide (3000mg/sq.m) with Etoposide (100mg/sq.m) and Carboplatin (635mg/sq.m), but he lost for follow up after a single cycle.

fig 4: Bilateral proptosis

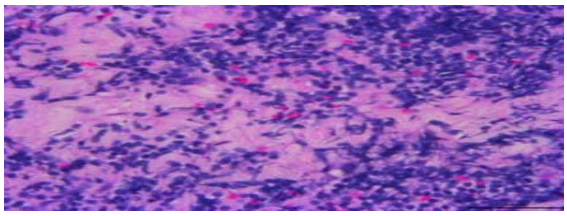

fig 5 : Histopathology showing small to medium sized lymphoma cells

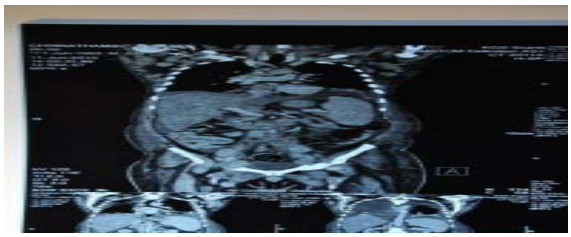

fig 6: CT chest and abdomen showing pleural effusion and hepatosplenomegaly.

The patient came after 6 months with Bilateral increasing proptosis. Systemic evaluation included Echocardiogram which showed Pericardial effusion ,CT chest and abdomen showed Diffuse lymphadenopathy, Hepatosplenomegaly and pleural effusion suggestive of systemic spread. Since the patient's general condition was poor, palliative care given. (fig 6)

DISCUSSION

Lymphomas of the orbit are uncommon and may involve any site in the orbit. They are common in the age group of 50 to 70 years. The clinical presentation of Orbital lymphomas may vary from palpable mass to ptosis, proptosis, excess tearing etc. Staging of Non Hodgkin's lymphoma is done by Biopsy and Histopathological examination of the lesion. Systemic workup includes complete blood counts, liver and kidney function tests, peripheral blood film, bone marrow biopsy, chest X-ray, computerized tomogram (CT) of the orbit, chest and abdomen. Treatment modalities of Primary Orbital lymphomas include Radiotherapy, Chemotherapy and Surgery. Excision with primary radiotherapy is very effective in localised lymphomas in early stages. In advanced cases, chemotherapy followed by Palliative radiotherapy is very effective than isolated chemotherapy. [4] Surgery alone is not useful except in conjunctival masses. [5] Major prognostic criteria for orbital adnexal lymphomas include anatomic location of the tumor; stage of disease at first presentation, lymphoma subtype as determined using the revised European American lymphoma (REAL) classification [6], immunohistochemical markers and the serum lactate dehydrogenase level (LDH). These cases are reported to emphasize the need for full staging work up for early and appropriate management of these patients. In high grade histologies, follow up is mandatory at closer intervals to recognise and treat early systemic spread.

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