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PARANASAL SINUS NON-HODGKIN LYMPHOMA - A RARE CASE REPORT

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Abstract : Objective - To report a rare case of paranasal sinus lymphoma in a young adult. Case report - A 23 years old male patient, known case of NHL of right maxillary sinus, treated outside with chemotherapy (CHOP with intrathecal methotrexate) and defaulted for follow up after complete resolution of symptoms, presented to our department with swelling of the right eye for about 1 month. Then on clinical examination the patient had chemosis and proptosis of the right eye and swelling of the right maxillary region. Biopsy from the lesion showed DLBCL. With imaging it was found that the lesion was localized to right maxillary sinus and adjoining structures and bilateral neck nodes only, and the case was labelled as relapse of extranodal NHL stage II AE. Since the patient was a known seropositive case of HBsAg, he was treated with palliative radiotherapy alone.
The patient achieved a very good response clinically. **Discussion** - Most extranodal NHL occurs in fifth to sixth decade, but in our case it was in third. In seropositive cases of HBsAg, chemotherapy may be avoided to prevent reactivation of hepatitis B viral infection. Lymphomas should always be kept in mind as one of the differential diagnosis when considering the tumors of sinonasal region. Radiotherapy played the main role in the management of lymphoma in seropositive cases of HBsAg, especially in the relapse or recurrent setting. The importance of looking for viral markers before starting chemotherapy is once again stressed.
Keyword :extra nodal NHL, paranasal sinus lymphoma, HBsAg, radiotherapy.

INTRODUCTION:

Non-Hodgkin lymphoma (NHL) is a heterogenous group of malignancy of the lymphoid system. Approximately one-thirds of NHL arises from extranodal sites. The commonest sites of extranodal NHL are GIT (stomach, intestine), followed by head & neck and skin. Among the head& neck region common sites are Waldeyer's ring, thyroid, salivary glands, nasal cavity and paranasal sinuses (PNS).[7] Among the malignant neoplasms of paranasal sinuses 14% is contributed by NHL. [4] The nasal cavity and paranasal sinus tumors which were grouped together earlier are now recognized to be different. In Asian population nasal cavity was more common site than paranasal sinus and was predominantly T-cell type, but in western population paranasal sinus is commoner and was predominantly B-cell type.[4] HIV, HHV-8, HCV, EBV and HTLV-1 are the viruses implicated in the etiopathogenesis of NHL.[7]

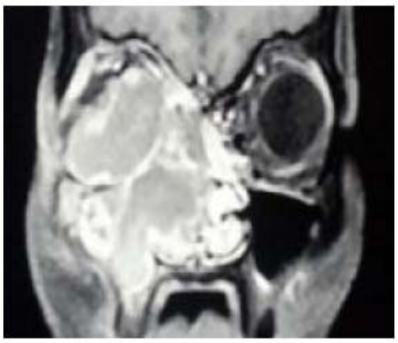
CASE REPORT:

A 23 years old male patient, known case of NHL of right maxillary sinus, treated outside with chemotherapy (CHOP + intrathecal methotrexate) and defaulted for follow up after complete resolution of symptoms, presented to our department with swelling of the right eye for about 1 month. Then on clinical examination the patient had chemosis and proptosis of the right eye and swelling of the right maxillary region.

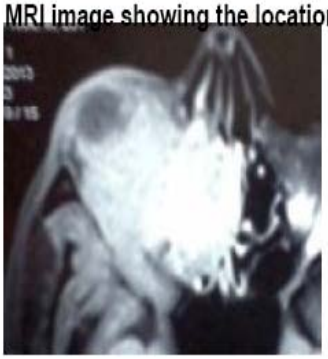
picture showing proptosis and swelling of right orbital and maxillary region

closer view of the image 1

Ophthalmologic examination showed reduced vision in right eye (1/60), minimal restriction of all extraocular movements and hyperemic disc with blurred margins in fundus examination. MRI & PET-CT report showed the disease extent and metabolically active disease in: a) Right maxillary sinus involving the right nasal cavity, ethmoid sinus, extraconal region of orbit, infratemporal region and eroding right maxilla, floor and medial wall of orbit –infiltrating the rectus and pterygoid muscle causing mass effect over the globe with proptosis, compressing and displacing the optic nerve; b) Irregular thickening in the nasopharynx with obliteration of fossa of rosenmuller and torus tubaris; c) Neck nodes involving bilateral level II and III; d) No other sites showed significant increased uptake.



MRI image showing the location and extent of tumor



axial cut of MRI

Mucosal biopsy of right maxilla showed diffuse large B cell lymphoma (DLBCL). IHC report was CD20 –Positive; Bcl-6 – Posiive; Ki67 was 90%; Mib was 90%. Bone marrow biopsy showed mildly h ypocellular marrow. USG abdomen showed mild hepatomegaly. Viral markers: HBsAg – Positive**,** Anti-HCV Ab – Negative, HIV –Negative. The case was labelled as relapse of non-Hodgkin lymphoma of right maxillary sinus -diffuse large B cell type –stage II AE. The patient was started on tab.Entecavir 0.5 mg o.d., as antiviral prophylaxis. Case was discussed in tumour board and since chemotherapy may cause immunosuppression and reactivation of the hepatitis B viral infection, it was decided to treat with palliative irradiation alone. Pa tient was treated with RT to a dose of 40Gy in 20 fractions to the target area of right maxillary sinus, nasal cavity, ethmoid sinus and orbit. There was a complete response to radiation clinically.

PET-CT image showing the localized uptake of tumor

post treatment: complete clinical response

DISCUSSION:

Diagnosis of NHL arising from paranasal sinus will be challenging because of the misleading symptoms.[3] Lymphomas are usually submucosal but carcinomas are usually ulcerative. Early diagnosis is the mainstay in the treatment of lymphomas. But because the lesion can spread easily through the anatomical spaces, it rarely produces symptoms in the initial stage. Jaffe E.S. has reported the common age of occurrence for NHL as 70 years.[4] Indian data stated the median age of occurrence as 55.5 years. [2] But in our case the age of occurrence was younger, in the third decade. In the literature review did by Su Zy et al., in patients with primary paranasal sinus lymphoma, maxillary sinus was the commonest site, followed by ethmoid and sphenoid sinus[4],[5]; diffuse large B cell was the common histologic subtype[2],[4] which was agreed in our case.

The common symptoms of sinonasal lymphoma are nasal obstruction, epistaxis, headache and unilateral facial or cheek swelling. Extension to orbit can lead to proptosis and visual disturbances.[4] Most of the extra nodal lymphomas present in early stages (stage I and II) [5]. In DLBCL, the tumor cells express pan B markers (eg: CD20, CD79a).[4] The management of extranodal lymphomas is not different from those of nodal lymphomas of similar stage and histopa-thology, and similar is the prognosis.

[4],[5] The prognosis is little favourable for DLBCL compared to NK/T cell type.[4] There were controversial reports of increased risk of CNS spread in PNS lymphomas, for which intrathecal chemotherapy was given.[6] Combined modality therapy (chemotherapy + IFRT) results in better outcome compared to either alone. Patients who achieve a CR (complete response) after chemotherapy should receive a consolidation RT 30 Gy. For partial responses a dose of 40 Gy is recommended. The desired target volume is Involved Field Radio Therapy (IFRT) that includes involved region with generous margins. HBV reactivation can occur in a patient with resolved or inactive HBV infection who receive chemotherapy or Rituximab; defined as an abrupt increase in serum HBV DNA and serum ALT levels. It can rarely lead to fulminant hepatitis and death, but mostly self-limiting. Hence, it can lead to premature termination of a chemotherapy regimen or delay in starting it. Few studies successfully used entecavir or lamivudine prophylaxis, with better results from entecavir in HBsAg positive patients undergoing chemotherapy.[1]

Radiotherapy played the main role in the management of lymphoma in seropositive cases of HBsAg, especially in relapse or recurrent setting, in whom rechallenging with chemotherapy or rituximab carries significant complications. [3] The importance of looking for viral markers before starting chemotherapy is once again stressed. Wide knowledge and information about lymphoma and its various uncommon presentations in the head and neck region should reach the general practitioners It should always be kept in mind as one of the differential diagnosis when considering the tumors of sinonasal region. An interdisciplinary approach is essential for the early diagnosis and prompt treatment of these patients to increase the survival rate.

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