Case Report

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A CASE REPORT OF NON-HODGKIN'S LYMPHOMA: A CLUE TO UNDIAGNOSED HIV

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ABSTRACT:

Non Hodgkin's Lymphoma (NHL) rarely manifests as a primary malignancy in head & neck region accounting for less than 1%, may give an important clue for undiagnosed HIV infection, which accounts for 2% of oral neoplasms in patients with AIDS.[1] The close association of NHL with HIV is formally recognized by fact that NHL is designated as an AIDS Defining Condition.

We present a case report of primary extranodal NHL presented in form of gingival growth as the first and only manifestation of HIV in an otherwise healthy appearing individual not aware of being HIV positive. The lesion was clinically diagnosed as pyogenic granuloma, histopathologically was Extranodal NHL, and raised suspicion of HIV which subsequently was confirmed by appropriate investigations. The present case also emphasizes importance of accurate diagnostic procedures to avoid delayed diagnosis or inappropriate treatment strategies while dealing with unspecific oral lesions which subsequently worsen the prognosis.

Key words: Plasmablastic NHL, primary malignancy, extranodal, HIV, HHV 8.

INTRODUCTION:

Many a times, NHL present in oral cavity as the first identifiable evidence of underlying HIV disease.[1-2] NHL exhibit greater predilection for dissemination to extra nodal tissues.[3,4] as it has propensity to affect non-lymphoid tissues including oral tissues.[5]

We present a case of Primary Extranodal NHL of the maxillary gingiva which was the only manifestation of HIV in apparently healthy appearing patient.

CASE DETAIL:

A 54 year male patient with no medical background of immediate interest, reported with chief complaint of gradually increasing gingival growth over the left maxillary posterior region since one month.

Physical examination revealed healthy looking male with vital signs stable and within normal limits. Extraorally, no obvious facial asymmetry and no regional lymphadenopathy.

Intraorally, there was a firm, reddish pink gingival growth approximately 3 × 2 cm with respect to

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24, 25, 26 and 27, with grade I mobility, generalized periodontitis. Orthopantomogram (OPG) revealed haziness in left maxillary sinus and generalized bone loss. Lesion was diagnosed as Pyogenic granuloma. After routine blood investigations, excisional biopsy was done along with extraction of 24, 25, 26 and 27.(Fig1).

Histopatholgic report revealed diffuse infiltration of highly anaplastic lymphoid cells in scanty connective tissue stroma. These cells exhibited highly malignant features like cellular–nuclear pleomorphism ,vesicular nuclei, patchy cellular areas with hyperchromatic nuclei , numerous bizarre mitotic figures. Plasmablastic differentiation was evident at places.(Fig 2).

An immunohistochemical study was done for confirmation. The tumor cells expressed MUM1, CD 138(focal), & CD 38(focal), positive for plasma cells.(Fig3).

The diagnosis of NHL of Plasmablastic type (PBL) of maxillary gingiva was established. In Situ hybridisation for EBV-RNA was positive.

Histologic picture raised strong suspicion of underlying immunocompromised status for which HIV test was advised. Patient was reactive for HIV 1 with CD4 count of 200.

Further investigations were done. CBCT showed obliteration of left maxillary sinus. CT scan revealed mild heterogenous enhancing soft tissue mass involving left sinus with destruction of inferio-lateral wall, no suspicious nodes on CT scan imaging of neck. MRI excluded visceral/nodal involvement. Lesion was categorized as stage IAE, primary maxillary NHL according to Ann Arbor classification.

Radiotherapy in the range of 40Gy was delivered in 25 fractions of 180cGy daily for treating NHL along with ART medication.(Fig4).

DISCUSSION:

Plasmablastic lymphoma is a rare subcategory of NHL, considered to be neoplasm arising predominantly in oral cavity of HIV patients.[6] Indeed extra-nodal tissue involvement is a rule rather than exception and often site sampled for diagnosis.[2]

The exact etiology is unknown. Genetic predisposition, immunodeficiency state like HIV are implicated.[7] The viruses commonly associated are EBV, HTLV-1, HHV-8.[3,8,9,10] It has been suggested that chronic antigenic stimulation due to these viruses act on multiple B-cell clones, which may synchronously or metachronously undergo neoplastic transformation.[11,12] The medical evaluation of patient revealed HIV and EBV infection with CD4 count of 200.

Primary intraoral NHL are rarely initial manifestation,[2] usually misdiagnosed.[7] Oral NHL may appear as swelling, exophytic mass, delayed

healing of extraction site. Recognition of distinctive type of lymphoma confined to gingiva is important as PBL may mimic reactive gingival enlargements like pyogenic granuloma , peripheral giant cell granuloma.[13,14]

A painless gingival enlargement being highly suspicious of NHL, must be considered in differential diagnosis.[13] In our case, lesion appeared primarily as firm maxillary gingival growth,without evidence of pain, paraesthesia or regional lymph node involvement.

Plasmablasts are lymphoid cells that morphologically resemble B-cell immunoblasts but have acquired a plasma cell immunophenotype (i.e., loss of B-cell markers andsurface immunoglobulin with the acquisition of plasma cell surface markers). Thus, unlike immunoblasts, plasmablasts fail to express CD45 (leukocyte common antigen) as well as the B-cell marker CD20 and are only variably immunoreactivefor CD79a—a broader-spectrum B-cell marker. They are also negative for pan-T-cell markers. Positive staining for plasma cell markers such as VS38c, CD38, MUM-1, and CD138 indicates a phenotype akin to plasma cells.[13,10]

The diagnosis was based on histopathological findings coupled with Immunohistochemistry.

Radiotherapy in the range of 35-40Gy for treating the NHL along with the ART treatment has proven successful in this case.

In 2 years of follow up, patient is receiving ART with no evidence of relapse.(Fig5).

CONCLUSION:

NonHodgkins Lymphoma (NHL) often involves the extranodal site of the head and neck but intraoral locations are much less frequent, especially when they are the only manifestation of this disease. Oral NHL are now considered as first indicator of HIV.Thus, with rising incidence of extra-nodal lymphomas it has become very important for present age dentists not to take any orofacial swellings at face value but to properly investigate its pathology and treat it judiciously.

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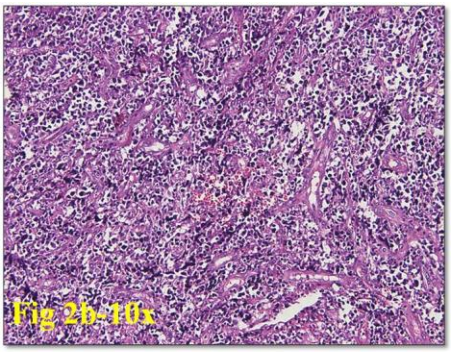
FIGURES:

Fig1a-Post-excisional clinical photograph.

Fig 1b : OPG showing–Haziness in left maxillary sinus, generalised horizontal bone loss, complete interdental alveolar bone loss between 26 and 27.



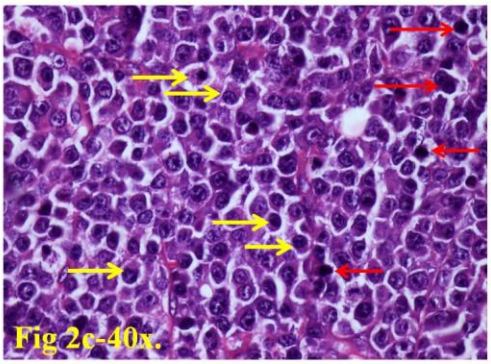
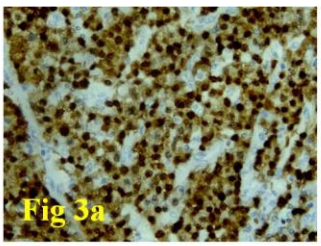
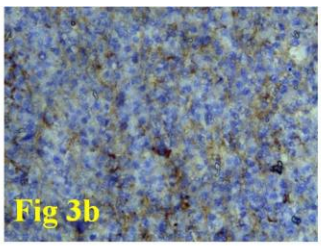
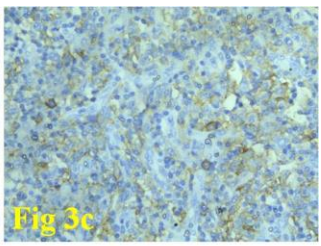
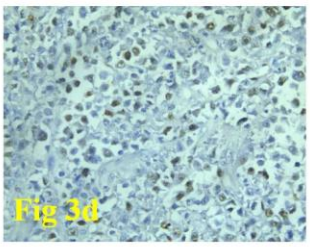


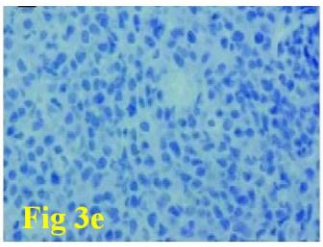
Fig 2- Histopathological images of H&E stained section
Fig 2a- Lesional tissue separated from overlying surface epithelium.
Fig 2b- Diffuse infiltration of lymphoid cells in a scanty CT stroma.
Fig 2c- Presence of large , highly pleomorphic and hyperchromatic lymphoid cells with numerous plasmacytoid differentiation (yellow arrows) numerous bizarre mitotic figures(red arrows) evident.

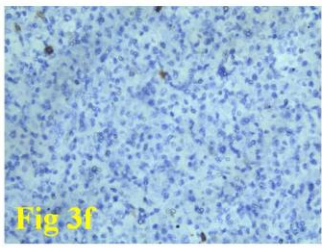


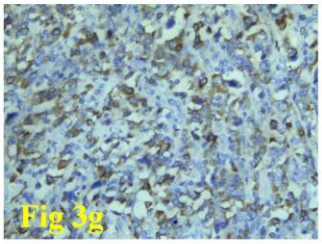


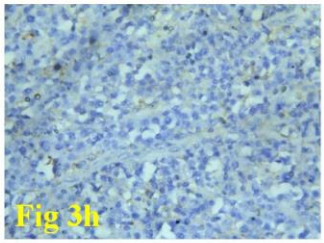




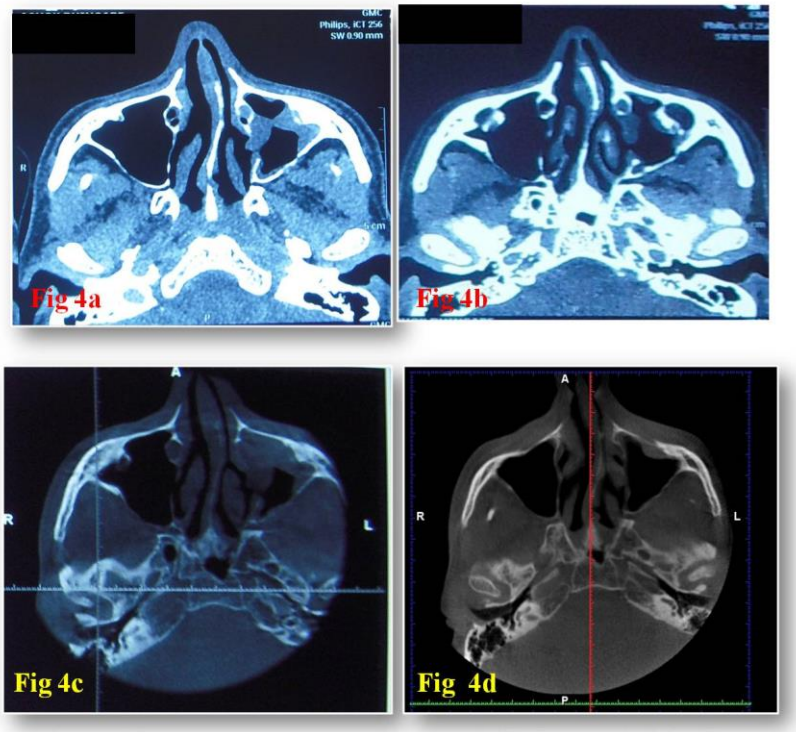








Immunohistochemical profile
Fig 3a-MUM 1 positive Fig 3b-CD 138 Focal positive
Fig 3c-CD38 Focal positive
Fig 3d-In Situ hybridisation for EBV RNA is positive
Fig 3e- CD20 negative Fig 3f- CD79a negative
Fig 3g and Fig 3h-There is predominance of lambda expressing chains over kappa expressing cells.



Preoperative and Posttreatment comparison of CT Scan and CBCT

Fig 4a -Pre treatment CT Scan showing soft tissue growth causing destruction of left maxillary sinus walls.

Fig 4b- Post treatment CTScan showing considerable reduction in the soft tissue growth.

Fig 4c -Preop CBCT showing soft tissue growth obliterating the left maxillary sinus.

Fig 4d-Post treatment CBCT showing appearance of radiolucency with decreasing size of the growth.

Fig 5- Postradiotherapy follow up clinical photograph.