

## Case Report

# Human immunodeficiency virus associated plasmablastic lymphoma: A case report

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### Abstract

Non-Hodgkin's lymphoma (NHL) is the third common malignant lesion of the oral region. Plasmablastic lymphomas are rare, aggressive neoplasms occurring mostly in human immunodeficiency virus (HIV) infected individual which accounts for approximately 2.6% of all NHL. It usually presents as a diffuse growth and with diffuse pattern of histological presentation. It is very difficult to differentiate this lymphoma from other NHL. Immunohistochemical evaluation of various markers is an important criteria of the diagnostic protocol. Here, we describe a case of plasmablastic lymphoma in a 50-year-old female HIV-infected patient. The diagnosis was based on histopathological examination and immunophenotyping.

**Key words:** Human immunodeficiency virus infection, non-Hodgkin's lymphoma, plasmablastic lymphoma

### INTRODUCTION

Plasmablastic lymphoma is a subtype of non-Hodgkin B-cell lymphoma that has a predilection in the oral cavity and most commonly affecting human immunodeficiency virus (HIV) positive patients.<sup>[1]</sup> This type of lymphoma has cells resembling B immunoblasts and has the plasma cells immunophenotype.<sup>[2]</sup>

World Health Organization (WHO) has described plasmablastic lymphoma as a distinct entity that typically occurs in the oral cavity, and it accounts for 2.6% of all NHL.<sup>[2]</sup> There is a proliferation of lymphoid cells which has immunoblastic morphologic features with an immunophenotype denoting terminally differentiated B-cells. One-third of the plasmablastic lymphoma is related to HIV infection, but most of them are associated with Epstein virus infection.<sup>[3]</sup>

### CASE REPORT

A 50-year-old HIV-positive female patient came to our outpatient department with a chief complaint of

swelling in the upper right jaw region for the past 1½ months. Swelling is insidious with a gradual increase in size. The patient had pan chewing habit since 20 years. The Patient was on antiretroviral and antihypertensive therapy. Her blood investigation was within the normal limits except hemoglobin that was less (9.1 g/dl).

On extra-oral examination, the swelling was diffuse with the boundaries superiorly from infraorbital margin to inferiorly up to line drawn from the corner of the mouth to tragus [Figure 1]. On intra-oral examination, there was an ulceroproliferative growth, which extended in the buccal vestibule of upper second premolar to lateral incisors [Figure 2]. Radiographic examination showed evidence of mildly hyper dense, soft tissue in the right maxillary sinus extending

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through the walls into the subcutaneous plane of the oral cavity, ethmoid sinus, and frontal sinus [Figure 3].

The biopsy tissue obtained from the lesion was stained with hematoxylin and eosin staining. It showed the histological pattern of densely proliferating lymphocytes with a large nucleus and conspicuous nucleolus in the connective tissue infiltrating into the muscle tissue. The monotonous pattern of lymphocytes were also seen [Figure 4]. Immunophenotypic analysis showed negativity for B-cell lymphoma 6 protein, cluster of differentiation (CD3), multiple myeloma oncogene 1 (MUM1) and CD20 antibodies [Figures 5-8], but positivity for epithelial membrane antigen, leucocyte common antigen (LCA), and Syndecan 1 (CD138) antibodies [Figures 9-11]. Immunohistochemistry (IHC) results gave a concrete idea about the diagnosis of plasmablastic lymphoma in the biopsy.

## DISCUSSION

NHL is the third most common group of malignant lesions seen in the oral cavity.<sup>[4]</sup> The incidence of NHL is 100 times more in HIV-infected individuals than in the general population, mostly secondary to HIV-induced immunosuppression.<sup>[5]</sup> HIV-associated NHL is categorized into three broad types: Small noncleaved type (Burkitt's lymphoma), large cleaved or noncleaved type (centroblastic lymphoma) and immunoblastic lymphoma with plasmacytoid differentiation.<sup>[6]</sup>

Plasmablastic lymphoma is a subtype of non-Hodgkins B-cell lymphoma.<sup>[1]</sup> WHO has described plasmablastic lymphoma as a distinct entity that typically occurs in the oral cavity and it accounts for 2.6% of all NHL.<sup>[3,5,7,8]</sup> Few cases of plasmablastic affect extraoral sites such as gastro intestinal tract, lymph nodes, and skin.<sup>[7,8]</sup> The



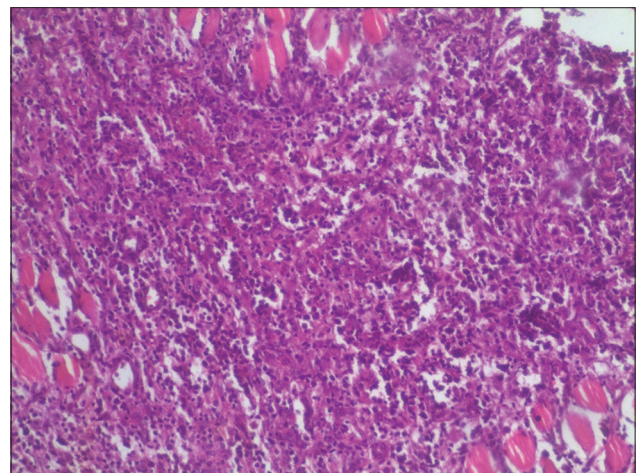
**Figure 1:** Extraoral swelling extending superiorly from infraorbital margin to inferiorly up to line drawn from corner of mouth to tragus



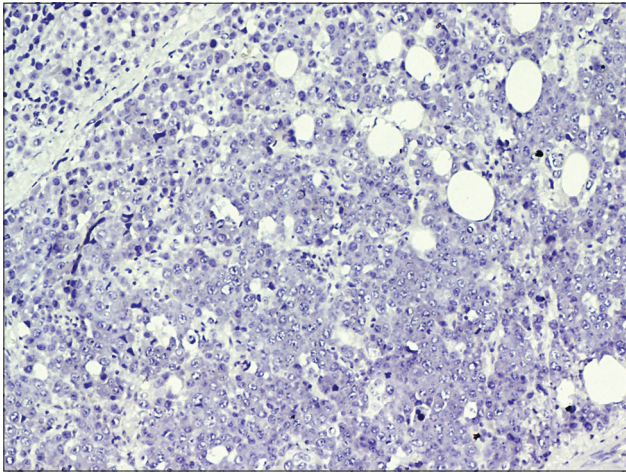
**Figure 2:** Intraoral ulceroproliferative growth seen extending in the buccal vestibule of upper right second premolar to lateral incisors



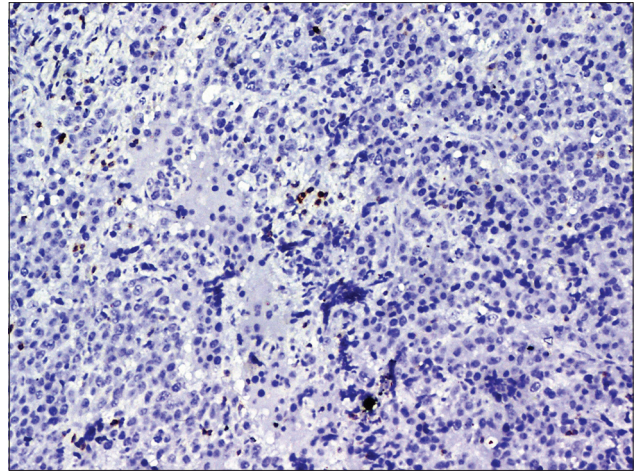
**Figure 3:** Radiography showed the presence of soft tissue mass in right maxillary sinus, extending through the walls into the subcutaneous plane, oral cavity, ethmoid sinus, and frontal sinus



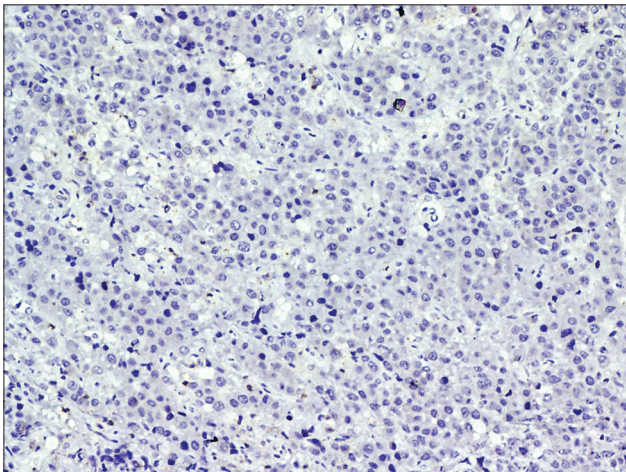
**Figure 4:** On histological examination densely proliferating lymphocytes with large nucleus and conspicuous nucleolus in the connective tissue infiltrating into the muscle. Monotonous pattern of lymphocytes are seen



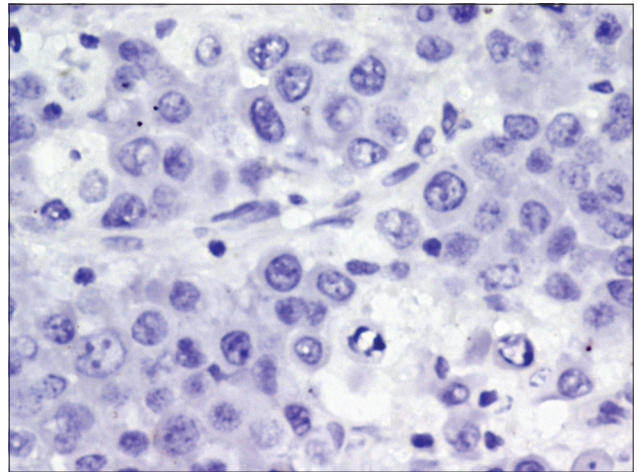
**Figure 5:** Immunohistochemistry for B-cell lymphoma 6 protein shows negative finding



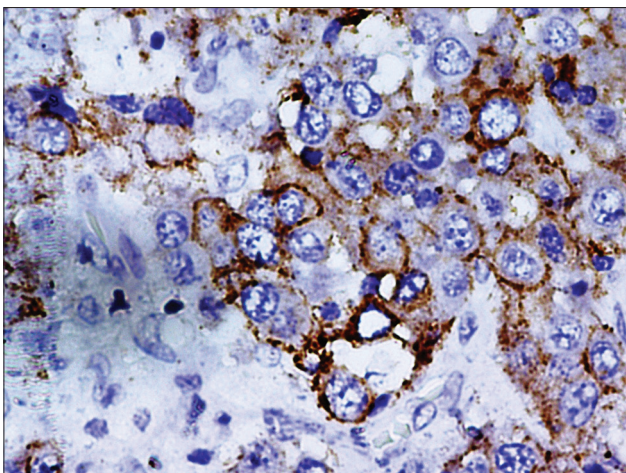
**Figure 6:** Immunohistochemistry for cluster of differentiation 3 shows negative finding



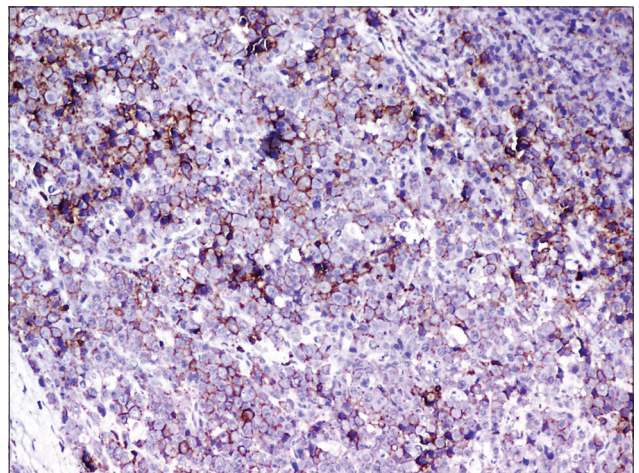
**Figure 7:** Immunohistochemistry for multiple myeloma oncogene 1 shows negative finding



**Figure 8:** Immunohistochemistry for cluster of differentiation 20 shows negative finding



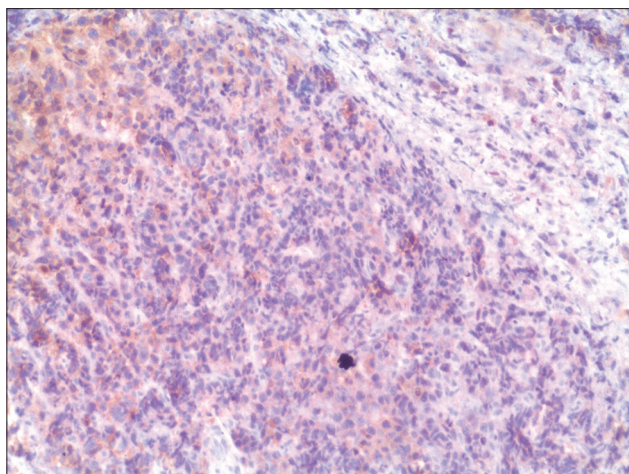
**Figure 9:** Immunohistochemistry for epithelial membrane antigen on focal membrane shows positive finding



**Figure 10:** Immunohistochemistry for leukocyte common antigen on focal membrane shows positive finding

mean age group patients affected is usually 39 years in HIV-positive patients with a male predilection.<sup>[7]</sup>

The pathogenesis of plasmablastic lymphoma is not yet understood properly. Based on the genetic,



**Figure 11:** Immunohistochemistry for cluster of differentiation 138 shows positive finding

molecular and IHC studies, plasmablastic lymphoma is considered to be derived from the postgerminal center, terminally differentiated activated B-cells during their maturation from immunoblast to a plasma cell.<sup>[7]</sup>

Considering the morphology, the differential diagnosis of plasmablastic lymphoma includes lymphoproliferative disorders such as Burkitt's lymphoma, immunoblastic lymphoma, anaplastic plasmacytoma, and anaplastic lymphoma kinase (ALK) positive diffuse large B-cell lymphoma.<sup>[7,9,10]</sup> Since anaplastic plasmacytoma is not common in immunosuppressed patient and LCA is positive, anaplastic plasmacytoma is ruled out in this case.<sup>[6,11]</sup> Burkitt's lymphoma is always positive for CD20 and since CD20 is negative in our case, ruled out Burkitt's lymphoma.<sup>[6,7,10]</sup> LCA positive gave us a clue for immunoblastic diffuse large B-cell lymphoma. Negativity for CD 20 gave a diagnosis against immunoblastic<sup>[5]</sup> and MUM1 negative gave a diagnosis against ALK-positive diffuse large cell lymphoma.<sup>[12]</sup> CD20 negative and positive expression of plasma cell marker CD138 is seen in plasmablastic lymphoma<sup>[2,5,7,8]</sup> which gave us the diagnosis.

Strong IHC expression of CD38, CD138 and weak/absent expression of CD20 and CD79a is the criteria for diagnosis of plasmablastic lymphoma.<sup>[2,6,9]</sup>

Highly active anti-retroviral therapy (HAART) along with chemotherapy is the recommended treatment given for patients with plasmablastic lymphoma.<sup>[13]</sup> Recent studies have revealed the use of proteasome inhibitor bortezomib in patients along with HAART.<sup>[6,7,14]</sup> Plasmablastic lymphoma has a poor prognosis with a survival period of 1–24 months after diagnosis.<sup>[2]</sup> Studies have shown that oral plasmablastic lymphoma has increased overall survival rate than extraoral.<sup>[7]</sup>

## CONCLUSION

Knowledge of plasmablastic lymphoma is mandatory to avoid confusion with all non-Hodgkin lymphomas because of lack of advanced lymphoid markers. Early diagnosis of this pathological lesion, management of HIV infection with more aggressive chemotherapy can account for a better prognosis in future.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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