RESEARCH PAPER

A Rare Case Report of Plasmablastic lymphoma

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Abstract:
Plasmablastic Lymphoma is a rare large B-cell lymphoma with unique immunohistochemical and morphological features. It is most commonly associated with HIV infection; however, it's now seen in other immunosuppressed states like autoimmune conditions, post-transplant settings, and even in elderly immunocompetent individuals. Although rare, it is an aggressive lymphoma with unfavorable outcomes. The aim of this manuscript is to have an in-depth review of the current knowledge of epidemiology, pathophysiology, prognostic markers, and treatment approaches currently in use and in clinical trials for this challenging disease.

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Introduction

Plasmablastic lymphoma (PBL) was initially recognized in 1971 as a large B cell lymphoma with unique immunohistochemical and morphological features most associated with HIV infection. Subsequently, it was also found in other immunosuppressed states like autoimmune conditions, post-transplant setting and even in elderly immunocompetent individuals. In 2017 World Health Organization classification, it belongs to a distinct subgroup of B cell lymphomas termed lymphoid neoplasms with plasmablastic differentiation accounting for 2% of acquired immunodeficiency syndrome (AIDS) related lymphoma. Although rare, it is an aggressive lymphoma with unfavorable outcome.

Plasmablastic lymphoma (PBL) is an aggressive subtype of non-Hodgkin's lymphoma (NHL), which frequently arises in the oral cavity of human immunodeficiency virus (HIV) infected patients. PBL shows diffuse proliferation of large neoplastic cells resembling B-immunoblasts/plasmablasts, or with plasmacytic features and an immunophenotype of plasma cells. PBL remains a diagnostic challenge due to its peculiar morphology and an immunohistochemical profile similar to plasma cell myeloma (PCM). PBL is also a therapeutic challenge with a clinical course characterized by a high rate of relapse and death. There is no standard chemotherapy protocol for treatment of PBL. Cyclophosphamide, doxorubicin, vincristine, and prednisone have been the backbone while more intensive regimens such as cyclophosphamide, vincristine, doxorubicin, high-dose methotrexate/ifosfamide, etoposide, high-dose cytarabine (CODOX-M/IVAC), or dose-adjusted etoposide, prednisone, vincristine, cyclophosphamide, and doxorubicin (DA-EPOCH) are possible options. Recently, a few studies have reported the potential value of the proteasome inhibitor bortezomib and thalidomide in PBL patients. The introduction of genes encoding artificial receptors called chimeric antigen receptors (CARs) and CAR-modified T cells targeted to the B cell-specific CD19 antigen have demonstrated promising results in multiple early clinical trials.

The present case report describes a case of plasmablastic lymphoma.

II Case report

A 55 years,old female patient (Figure 1) reported to the department of oral medicine and Radiology with the chief complaint of pain and swelling in the left upper back tooth region since 1 month. Swelling was small initially and in 4 months has increased to the present size. Extraorally asymmetry of face seen in the left middle third of the face, measuring approximately 6x7 cms extending superiorly outer canthus of eye and lower eyelids, inferiorly below the lower border of mandible, posteriorly tragal and ramus of mandible, anteriorly ala of nose and corner of the mouth. The skin over the swelling appeared stretched,and firm on palpation with tenderness. (Figure 2). Intraorally there was swelling in the left upper back region, which is approximately 5x6 cms in its greatest dimensions, extending anteriorly till 23 tooth region, posteriorly crossing beyond maxillary tuberosity, medially from midline of palate and laterally buccal vestibule. The swelling was firm in consistency and non tender on palpation. The tooth are displaced in relation to 24,25 27. (Figure 3). Left submandibular lymphnodes was palpable ,two in numbers , approximately 1mm, firm in consistency and tender.

Based on the clinical findings a provisional diagnosis of fibroosseous lesion was made.

Due to extensive growth and displaced teeth, a differential diagnosis of squamous cell carcinoma of alveolar ridge and malignant tumour of maxilla was made.

Investigations

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An



orthopantomograph , Paranasal sinus waters view were taken ,which revealed radiolucency involving the sinus.(Figure 4). And also Computed tomography was taken which revealed Large expansile lobulated heterogenous soft tissue density expansile mass lesion measuring 7.2 (AP) x 6.4 (RL) x 6.5 (CC) cms with epicenter and completely opacifying the left maxillary sinus eroding the adjacent maxillary sinus walls as well as the left superior alveolus and partially eroding the left pterygoid plate extending into the left half of the nasal cavity, left half of the oral cavity, left infratemporal fossa, left premaxillary soft tissue space and into the left sphenoid sinus consistent with neoplastic etiology. Mucosal thickening in the left ethmoidal and left frontal sinuses. (Figure 5). CBCT imaging was also done which showed an aggressive ,ill defined osteolytic bone defect with extensive destruction of the jawbone. (Figure 6).

Hematological investigations were done which revealed Hemoglobin 9.3 gms% ,increased lymphocytes of 49% & increased value of MCHC 38.9g/d.

Serologic values for HIV 1 and 2, HBsAg, were negative .

HbA1C being 5%.Fasting sugar levels and postprandial values were 79mg/dl &103mg/dl.

Incisional biopsy was taken and microscopic examination (Figure 7) of H & E stained section revealed parakeratinized stratified squamous epithelium and underlying connective tissue . The epithelium is 3-4 layers thick and shows flat to short rete ridges.The connective tissue shows diffuse proliferation of monomorphic sheets of round cells.The cells are large with central round to oval vesicular nuclei, prominent,multiple nucleoli and basophilic cytoplasm resembling plasmablasts. Numerous mitotic figures are evident.The surrounding stroma is eosinophilic,owing to the extruded immunoglobulins from the neoplastic cells. Blood vessels engorged with RBCs and mild amount of mixed inflammatory infiltrate predominantly of plasma cells, lymphocytes and few eosinophils are seen in the background of connective tissue stroma.

Immuno histochemical findings reveals CD 45 and CD 138 - positive for neoplastic cells.

CD20 and ALK 1 - negative for neoplastic cells.



Figure 1 Extra oral picture of patient aged 55 years old Figure 2 lateral view of swelling



Figure 3 Intraoral view of swelling

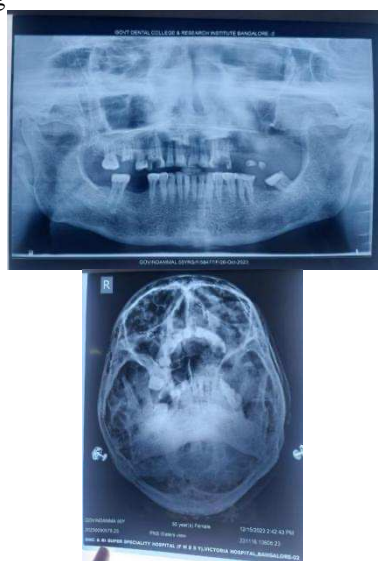


Figure 4: showing Orthopantomograph and Waters view

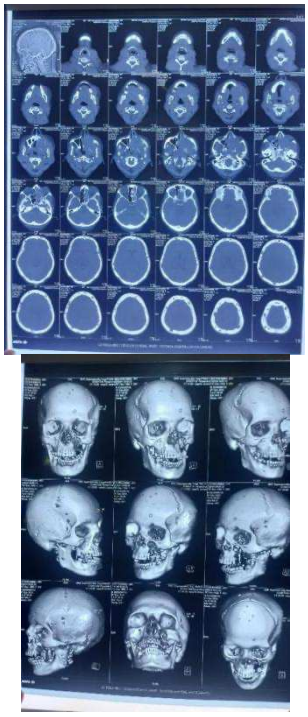


Figure 5 . Computed Tomography Axial views

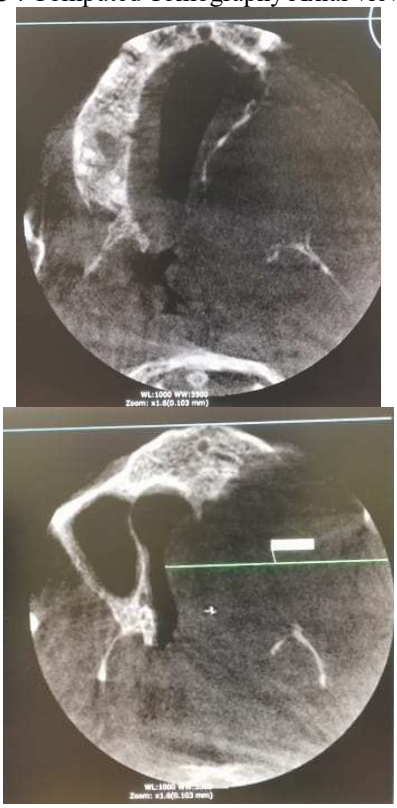


Figure 6 . Conebeam Computed Tomography

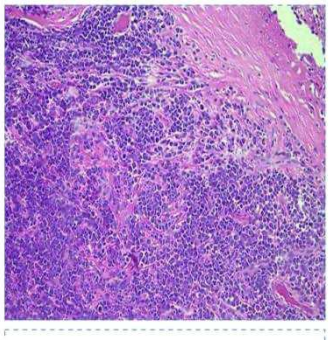


Figure 7: Histologic picture

Discussion

Plasmablastic lymphoma (PBL) is a **rare and aggressive subtype of diffuse large B-cell lymphoma** that was initially described in the oral cavity of HIV-positive patients but has since been reported in both immunocompromised and immunocompetent individuals. Its distinct morphology characterized by large cells with plasmacytoid features and its immunophenotype—typically negative for conventional B-cell markers (CD20, PAX5) but positive for plasma cell–associated markers (CD138, MUM1, BLIMP1)—make diagnosis particularly challenging clinics in oncology. This overlap with plasma cell neoplasms often leads to misclassification, underscoring the importance of comprehensive immunohistochemistry and where available molecular studies.

Clinically, PBL most commonly involves the **oral cavity**, but extraoral sites such as the gastrointestinal tract, lymph nodes, and skin have been documented. The case under discussion adds to the growing evidence that PBL should be considered in the differential diagnosis of rapidly enlarging masses in atypical locations, especially when conventional lymphoma markers are absent. Epstein–Barr virus (EBV) positivity is frequently observed, supporting its link to immunodeficiency states, although EBV-negative cases are increasingly recognized.

Therapeutic options remain limited. **CHOP-like regimens are generally ineffective**, and more intensive protocols such as EPOCH or hyper-CVAD have been employed with variable success. Given its plasma cell–like biology, novel agents such as **bortezomib, lenalidomide, and daratumumab** have been explored in small series, though evidence is still preliminary. Autologous stem cell transplantation may be considered in eligible patients, but overall prognosis remains poor, with median survival often less than two years. This highlights the urgent need for prospective studies and targeted therapies.

Our case contributes to the literature by emphasizing the **heterogeneity of PBL presentations** and the diagnostic pitfalls associated with its overlap with plasma cell neoplasms. It reinforces the necessity of a **multidisciplinary approach** involving hematopathologists, oncologists, and infectious disease specialists. Early recognition and aggressive management are crucial, but outcomes remain guarded. Reporting such cases is vital to enhance collective understanding, refine diagnostic algorithms, and inform therapeutic strategies for this uncommon lymphoma.

Conclusion

Plasmablastic lymphoma continues to test clinicians with its rarity, aggressive course, and overlapping features with other hematologic malignancies. Recent advances in the therapy of HIV-associated and aggressive lymphomas do not seem to have had a positive impact on the outcome of patients with PBL. Given the rarity and heterogeneity of PBL, it is likely that no large, randomized controlled therapeutic studies would ever be done. From our perspective, multi-institutional collaboration would be an effective mechanism by which diagnostic and therapeutic advances can come to light in rare diseases such as PBL.

Plasmablastic lymphoma remains a rare and aggressive subtype of diffuse large B-cell lymphoma, often associated with immunodeficiency but increasingly recognized in immunocompetent individuals. This case highlights the diagnostic challenges posed by its overlapping morphologic and immunophenotypic features with other hematologic malignancies, underscoring the importance of comprehensive histopathological and immunohistochemical evaluation. Early recognition is crucial, as timely initiation of intensive chemotherapy regimens may improve outcomes, though prognosis remains poor. Our report adds to the growing body of literature emphasizing the need for heightened clinical suspicion, multidisciplinary collaboration, and further research into novel therapeutic strategies to enhance survival in patients with this uncommon entity.

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