Case Report: Rare case of plasmablastic lymphoma at right maxillary sinus with aberrant expression of CD3 (cytoplasmic) and CD4

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Introduction

Plasmablastic lymphoma (PBL) is a rare lymphoma of large B cells (1%) with plasmablastic or immunoblastic morphology and expression of plasma cell-associated markers. PBL usually presents at the mucosa of extranodal sites, especially the nasal or oral cavity (~50%), digestive system (20%), soft tissue and bone (15%). [1] Cases typically arise de novo in immunocompromised patients who have HIV-infection (31-62%), while some develop from secondary transformation of follicular lymphoma or chronic lymphocytic leukaemia/ small lymphocytic lymphoma (CLL/SLL). PBL shows association with EBV infection and complex karyotype. MYC dysregulation through translocation or amplification is common. Treatment modality includes chemotherapy, addition of antiviral therapy and surgery. The prognosis of PBL is poor and the median overall survival with chemotherapy is 6-32 months. [1-4]

Here we present a case of PBL at the right maxillary sinus with aberrant expression of CD3 (cytoplasmic) and CD4, and showed spontaneous regression after addition of antiviral therapy.

Clinical findings

A 68-year-old gentleman presented with right upper gum swelling and dental pain for two weeks. He had medical history of Hepatitis B Virus (HBV) carrier status, Human Immunodeficiency Virus (HIV) infection/ Acquired Immunodeficiency Syndrome (AIDS), Pneumocystis pneumonia requiring mechanical ventilation, upper gastrointestinal tract cytomegalovirus (CMV) infection and unascertained history of lymphoma diagnosed 15 years ago for observation.

Physical examination showed right alveolar swelling, #18 tooth socket with slough and gum swelling. Flexible laryngoscopy showed right nasal floor yellowish purulent discharge and narrowed nasal cavity passage due to congested mucosa.

PET scan showed a right maxillary sinus mass (66 x 66 x 66 mm) with heterogeneous hypermetabolism, involving the right-sided oral cavity, right tongue, nasal cavity, paranasal sinuses and the inferior extraconal right orbit.

A biopsy was taken at the oral right maxilla and the diagnosis of plasmablastic lymphoma was made.

No treatment was given and the right facial mass spontaneously regressed after two months since symptoms onset. The clinical impression was likely due to rise in CD4 count and improved immunity on HIV medication.

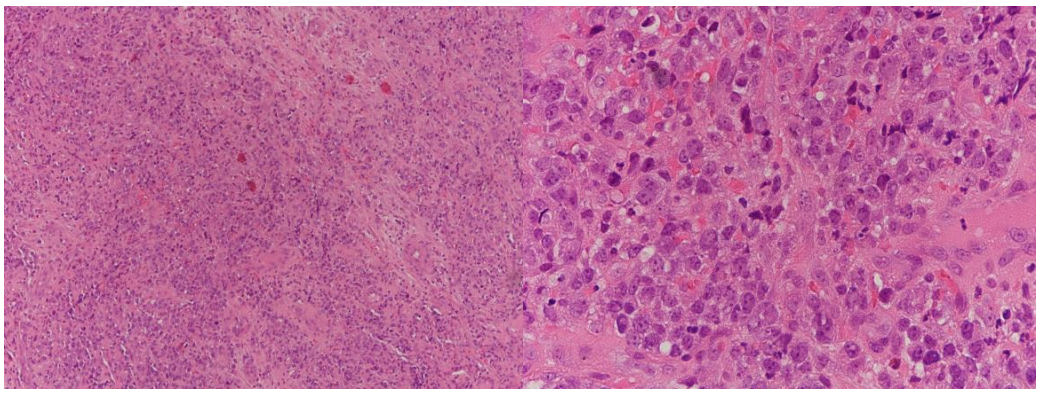

Figure 1. H&E sections of the tumour, low power 10x (left) and high power 40x (right).

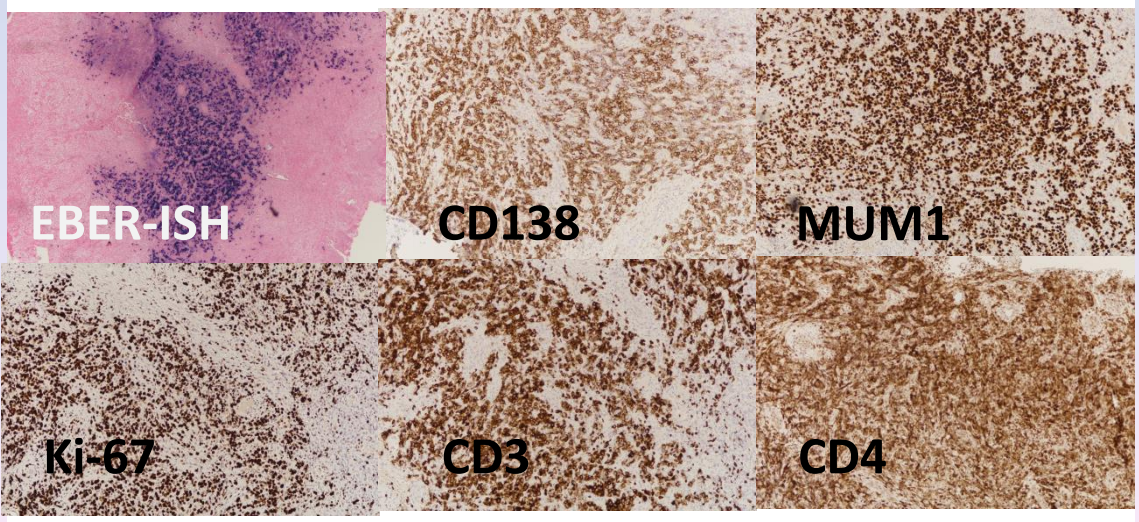

Figure 2. EBER-ISH was positive. (top left). Immunohistochemistry showed positivity for CD138 (top middle), MUM1 (top left), Ki-67 (bottom left), CD3 (bottom middle) and CD4 (bottom right)

Microscopic Findings and Ancillary Tests

The biopsy consisted of a piece of tan tissue with no orientation measuring 0.5 x 0.2 x 0.5 cm.
Histological examination showed ulcerated squamous mucosa with underlying stroma infiltrated by diffuse sheets of atypical large lymphoid cells which possessed large round nuclei, single to few prominent nucleoli, scanty amphophilic cytoplasm and brisk mitosis.

Immunohistochemically, the atypical large lymphoid cells were positive for CD138, MUM1, weakly positive for EMA (membrane staining), with aberrant expression of CD3 (cytoplasmic) and CD4. Ki67 (proliferative index) staining is approaching 100%. EBER (in situ hybridization for EBV-encoded RNA) was positive. They were negative for epithelial marker AE1/3, and were largely negative for LCA. They were negative for other B cell markers CD20, PAX5, CD79a, CD10, CD19 and CD23; other T cell markers CD2, CD5, CD7, CD8, and CD56, CD57, TIA, CD15, CD30 and ALK. Myeloid markers including myeloperoxidase, CD34, CD117 were also negative.

Discussion

Diagnosing PBL can be difficult due to the reduction/ absence of CD20, PAX5 and CD45 expression. They are typically positive for plasmacytic differentiation markers (CD138, MUM1 and CD38), EBER-ISH and show high Ki-67. CD3 and CD4-positive PBL is rare and can be misdiagnosed as peripheral T-cell lymphoma or extranodal NK/T cell lymphoma. Most reported cases show diffuse strong cytoplasmic staining of CD3, very rarely with additional CD4 positivity. Other T cell markers (e.g. CD2, CD55, CD7 and CD8) are reported to be negative. [5-7] The differentials also include plasmablastic plasma cell myeloma which requires clinical, laboratory and radiological correlation, and ALK-positive large B cell lymphoma which stains positive for ALK.

In summary, we present a rare case of PBL of the maxillary sinus in an immunocompromised patient who showed regression after addition of HIV medication. This PBL shows atypical immunophenotype and our report wishes to draw attention to this diagnostic pitfall.

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