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Plasmablastic Lymphoma Of Nasal Cavity : An Unusual Site Of Presentation

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Abstract

Plasmablastic lymphoma (PBL) is a rare mature B cell neoplasm arising from plasmablasts whose phenotype is switched to that of a plasma cell. PBL is seen frequently in males and commonly associated with human immunodeficiency virus (HIV) infection.[1] Oral cavity and jaw are the common sites, but it is rarely seen involving lungs, gastrointestinal tract(GIT), nasal cavity, paranasal sinuses, eyelids, orbit, CNS, skin and testicles.[1,2,3] A 35-year-old, HIV positive male presented with a nasal cavity mass which on histopathology revealed to be PBL but with overlapping histomorphological and immunophenotypic features between Diffuse large B cell lymphoma(DLBCL) and plasma cell neoplasm. Recognizing plasmablastic morphology, in an HIV positive patient, should alert one to perform other B cell markers particularly that will detect plasma cell differentiation. Due to its rarity and overlapping morphologic and immunophenotypic characteristics, PBL represents a diagnostic challenge.[2]

Keywords: CD138, EBV, HIV positive, Plasma blast.

INTRODUCTION

Plasmablastic Lymphoma (PBL) is a rare neoplasm with overlapping morphologic and immunophenotypic characteristics between aggressive large B-cell lymphomas and with plasma cell neoplasms. It was first described in 1997 to occur in the oral cavity in the setting of HIV infection.[4] A strong association with (Epstein-Barr Virus) EBV was also observed. But later PBL was identified in patients with other immunodeficiencies like post-transplant patients. The neoplastic cells lack expression of pan–B-cell antigens and CD45, but positive for plasma cell markers like CD38, CD 138, MUM1.[1] Now it is added as a specific subtype of immunodeficiency-associated lymphoproliferative disorder by the World Health Organization.[5] Other lymphomas recognized among HIV / AIDS are Burkitt's lymphoma, DLBCL, anaplastic large-cell lymphoma and primary effusion lymphoma.[2]

PBL has an aggressive and relapsing course, high rates of progression and fatality causing difficulties in management.[4] Recently, MYC gene rearrangements have been identified in 40% to 50% of patients with PBL which confer a worse prognosis. Chemotherapy/ radiotherapy remains the mainstay of treatment. Autologous hematopoietic cell transplantation showing better results than chemotherapy are

documented, but definitive data are sparse.[3] Also literatures on allogeneic hematopoietic stem cell transplantation for PBL are rare.[6]

CASE REPORT: A 35 year old, male presented with blockage of right side of the nose. There was no systemic symptoms. On examination a friable small polypoidal tissue was identified in the right nasal cavity. Serology was positive for HIV. Endoscopy assisted excision biopsy of the tissue was performed. Grossly, a friable grey white tissue bit measuring 1x1x0.5 cm was received in 10% buffered formalin and was routinely processed. H & E sections were examined.

Histopathological sections of the tumour tissue showed sheets of undifferentiated large round tumour cells. These cells contain a large vesicular nuclei, prominent 1-2 nucleoli and abundant basophilic cytoplasm (Fig1 A, B). Also seen were good number of tumour cells with plasmacytoid differentiation with paranuclear holf, eccentric nucleus and cart wheel chromatin (Fig 1 C). Frequent mitosis, many tingible body macrophages imparting starry sky appearance(Fig1A) and large areas of tumor necrosis were also noted. On light microscopy a provisional diagnosis of poorly differentiated carcinoma/ lymphoma/ plasma cell neoplasm was suggested. Formalin-fixed paraffin-embedded tissue was used to perform immunohistochemistry (IHC) panel. IHC markers: LCA / CD 45 (Fig 2D), PAN CK (Fig 2E) were negative. CD 20 (Fig 2F), CD 3 (Fig 2G) which are B and T cell markers respectively, were negative. CD 138 (Fig 3H), CD10 (Fig 3I) were positive showing plasma cell differentiation. High Ki 67 index (Fig 3J) of more than 90% was noted. ALK was negative (Fig 3K). Final diagnosis of PBL was made considering both histomorphological and IHC findings. The patient was referred to higher oncology centre for further management and further follow up were not possible.

DISCUSSION: Non Hodgkins Lymphoma is the second commonest malignancy occurring in HIV positive patients of which PBL are a characteristic type of DLBCL.[7] They were first identified in the jaws, oral mucosa, but documented in extra-oral sites as well.[1,2,3] The present case an HIV positive individual presented with mass in the nasal cavity. PBL shows strong association with HIV infection and other immunodeficiencies.[8] PBL shows a male predominance, with median age at diagnosis of approximately 50 years. Among HIV positive patients an earlier onset is noted with median age at presentation of 38 years.[9] Approximately 75% of cases occur in the setting of AIDS are positive for EBV.[1] The morphology is similar to DLBCL with plasmacytoid differentiation but tumour cells are negative for LCA and routine B-cell markers (CD20, CD45, PAX 5). [2,10] But, the tumour cells are positive for plasma cell differentiation markers CD79a, CD138, CD38 and IRF4/MUM1 which means these cells have the blastoid morphology of immunoblasts but antigenic profile of plasma cells.[1,2] Expression of CD4, CD10, CD30, and/or CD56 are reported in a subset of cases.[1] The cell of origin in PBL is thought to be the plasmablast, an activated B cell which has undergone somatic hypermutation and class switching recombination and is in the process of becoming a plasma cell.[4] Molecular and genetic studies suggest that it arises from activated B cells in transition from immunoblast to plasma cell that fail to undergo apoptosis in the post germinal center.[11] There is evidence of translocation /amplification of MYC gene which plays an important role in the pathogenesis. [10,12]

Differential diagnosis include –

A) Anaplastic plasma cell myeloma: Clinical, laboratory and imaging findings are helpful in distinction between the two. Renal dysfunction, monoclonal paraproteinemia, osteolytic lesions, hypercalcemia, bone marrow involvement favour plasmablastic plasma cell myeloma. But HIV association and a high Ki-67 index favours diagnosis of PBL.[1, 4]

B) ALK-positive large B-cell lymphoma: the large immunoblasts express plasma cell antigens, lack pan B-cell markers and show weak CD45 expression but on IHC show ALK positivity in a granular, cytoplasmic staining pattern, which helps in easy distinction from PBL.[1]

C) Immunoblastic DLBCL: express pan–B cell antigens.[1]

D) Poorly differentiated carcinoma: shows immunoreactivity for cytokeratins and ⁄ or epithelial membrane antigen.[5]

Volume 4, Issue 5; September-October 2021; Page No 946-949

CONCLUSION:

PBL is a rare, aggressive neoplasm carrying a poor prognosis. Many times, it is considered as undifferentiated malignant tumour due to lack of expression of CD45 and B-cell or T-cell markers. However, recognizing plasmablastic morphology, in an HIV positive patient, should alert one to perform CD 138 marker particularly to detect plasma cell differentiation. But the diagnosis is challenging in a HIV negative set up, among immunocompetent and in rare sites, during which correlation with clinical, morphological, phenotypic and molecular features would become crucial. [5]

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Volume 4, Issue 5; September-October 2021; Page No 946-949

FIGURES

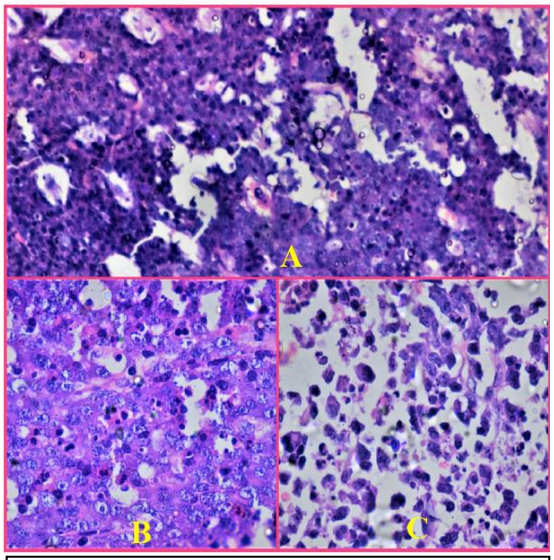

Fig1: A(sheets of tumour cells with starry sky appearance), B(tumour cells with large vesicular nuclei and prominent nucleoli), C(Plasmacytoid cells)

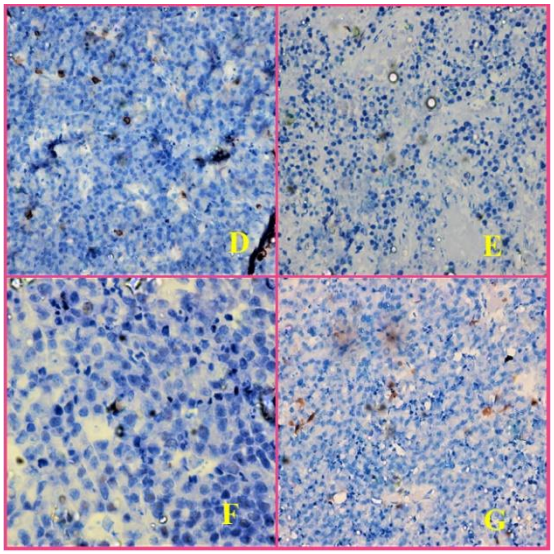

Fig 2: D (LCA Negative), E (PAN CK Negative), F (CD 20 Negative),

G (CD 3 Negative)

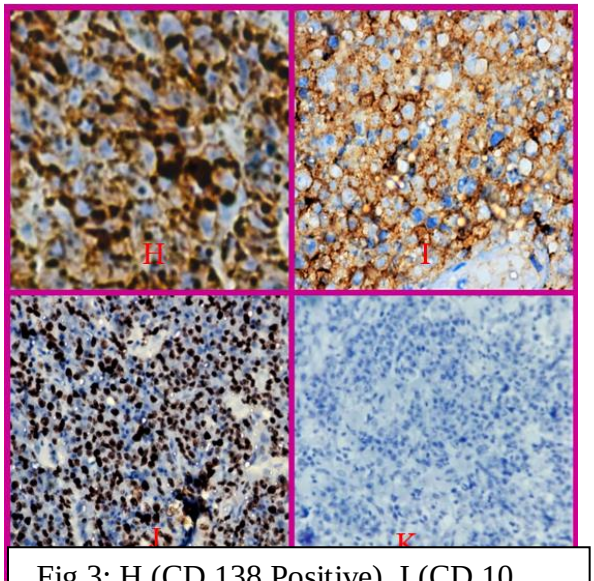

Fig 3: H (CD 138 Positive), I (CD 10 Positive), J (Ki 67 more than 90%),

K (ALK Negative)

Volume 4, Issue 5; September-October 2021; Page No 946-949