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Aggressive plasmablastic lymphoma presenting as nasal mass: an unusual Presentation

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ABSTRACT

Background: Plasmablastic lymphoma is an aggressive non Hodgkin lymphoma. It was first described in the jaws and oral cavity of HIV infected patients.

Case presentation: A 30 yr. old male presented with left sided nasal bleed from 6 months and left nasal obstruction since 3 months. Anterior rhinoscopy showed a firm, non-tender fleshy mass occupying the entire nasal cavity. Biopsy revealed plasmablastic lymphoma with the support of immunohistochemistry.

Conclusion: Unusual clinical behaviour of tumor in terms of site, immunocompetency and young age all these features are infrequent in the case of plasmablastic lymphoma that lead to present the case.

Keywords: Immunocompetent, nasal cavity, plasmablastic lymphoma

INTRODUCTION

Unilateral sinonasal mass or polyps are common presentation in otorhinolaryngological department. The majority of the pathology in nasal cavity are inflammatory with neoplasia accounting for 3% of head and neck malignancy. Neoplasticism lesion of the nose are seen in patients between 5 to 7th decade of life with a male to female ratio of 2:1.[1] Malignant tumors of sinonasal cavity comprises 1% of all head and neck tumors. Epithelial carcinomas are most common in comparison to mesenchymal tumors and lymphomas.[2] Lymphomas in the nasal cavity presents as a diffusely infiltrating tumor with homogenous soft tissue density resulting in remodeling or erosion of bones [2].Plasmablastic lymphoma(PBL) is a most common HIV related non Hodgkin lymphoma located usually in oral cavity and jaws. It is a very aggressive tumor. In recent times PBL have been reported in lungs, stomach, cervical lymph nodes, jejunum in a immunocompetent individual. Here we report the first case of PBL presenting as obstructive nasal mass in a immunocompetent patient.[3] Here we report second case of plasmablastic lymphoma arising from the nasal cavity in a 30 year old immunocompromised man. First case was reported by Nguyen et al in 2003.[4]

This case report is relevant in the context of primary health care of patient as clinician must have knowledge of unusual clinical presentation of tumor to pick up at the earliest.

presentation

A 30 yr. old heterosexual man presented with left sided nasal bleed from 6 months and left nasal obstruction since 3 months. Patient has a history of beedi smoking,10-12/day for 10 years. No history of unexplained fever, drenching sweats or weight loss seen. Anterior rhinoscopy showed a swelling over left dorsum of nose with septum deviated to right, firm, non-tender fleshy mass occupying the entire

nasal cavity on the left side reaching up to the vestibule with my mucoid secretions along the floor of nose. It bled on touch. No other palpable swelling or lymph nodes identified all over the body. Laboratory investigation showed complete blood count, electrolytes, liver function test and renal function test within normal range. A viral marker screen for HIV, hepatitis B and C were negative. Chest X-ray and ECG was normal, Contrast Enhanced Computed Tomography(CECT) nose and paranasal sinus showed a homogeneously enhancing soft tissue mass measuring 3.2x2.4x4.2 cm seen in the left nasal cavity anteriorly involving inferior and middle turbinate, superiorly abutting the cribriform plate, inferior surface of hard plate and medially involving the cartilaginous part of nasal septum, bony erosion also noted.[Figure-1] Patient underwent excision of the nasal mass. Histopathological examination demonstrated diffuse sheets of tumor cells exhibiting marked nuclear pleomorphism, with bizarre cells showing round to oval nucleus, an occasional prominent nucleoli and abundant eosinophilic cytoplasm. Few of the tumor cells have eccentrically placed nucleus. Mitotic figures and apoptotic bodies are seen exhibiting morphological features of a poorly differentiated malignancy.[Figure-2] The tumor cells were negative on immunohistochemistry(IHC) for Pan Cytokeratin(Pan-CK), Chromogranin, synaptophysin and S-100 immunomarkers. Cells were positive for CD 10, CD 79a, BCL2 with a strong nuclear positivity with MUM1 and membranous positivity for CD 138 and negative for CD20, BCL 6, Pax 5, cMYC and CD 56 on immunohistochemistry.[Figure-3] The results were consistent with a diagnosis of plasmablastic lymphoma. However the patient was lost for follow up.

Discussion

Human immunodeficiency virus(HIV) related Non Hodgkin Lymphoma(NHL) are plasmablastic lymphoma is 2.6%. The incidence of PBL occurring in HIV negative patients has not yet been estimated. In one of the literature review of 228 patients of PBL by Rafaniello et al, 69% were HIV positive, rest 31 % were HIV negative.[5] Around 1/3rd of HIV negative patients were found to have some form of iatrogenic immunosuppression (commonly solid organ transplant). Remaining cases were immunocompetent with no evidence of immunosuppression.[5] It is classified as B cell neoplasm showing proliferation of large neoplastic cells resembling B immunoblast with immunophenotyping of plasma cells. The prognosis is bad with survival between 1 to 24 months (average 6 months). [6] Most commonly affected sites irrespective of HIV status are oral cavity and gastrointestinal(GI) tract.[7] Other less common extra-oral sites included are central nervous sytem(CNS), paranasal sinus and mediastinum.[8] However oral and extra oral PBL do not show significant difference in terms of age group and sex predilection.[9] The individual cells vary in size from immature tumor cells including immunoblast or plasmablastic to a more mature plasmacytic differentiation.[10] Apoptotic bodies along with necrotic cells and tangible body macrophages are commonly seen. The IHC staining pattern of PBL is of a mature differentiated B lymphoma which expresses CD 79a along with plasma cell marker of CD 38, CD 138, multiple myeloma oncogene 1(MUM-1). Immunohistochemistry for CD10,CD56,BCL-2 marker are negative.

There are 2 closely related histological differentials for PBL . They are plasmablastic plasma cell myeloma and diffuse large B cell lymphoma with plasmacytic differentiation. Hence clinical correlation is required to distinguish PBL from plasmablastic variant of plasma cell myeloma. Plasma cell myeloma(PCM) usually shows increased serum immunoglobulin along with multiple osteolytic damages and cells are positive for CD56 hence favors PBL.[11] While a strong IHC of CD45 and CD20 goes in favors of diffuse large B cell. Ki67 of > 90 % indicating the aggressive nature of the malignancy. In our case CD20 is negative with Ki-67 score around 60% making less likely to be DLBCL. Other morphological differential diagnosis are burkitts lymphoma, poorly differentiated carcinoma, malignant melanoma that can be ruled out by IHC. Negative IHC for CD20 and BCL2 with low Ki67 exclude the Burkitt Lymphoma. While poorly differentiated carcinoma neuroendocrine tumor and melanoma are excluded by the negative immunostain for Pan- cytokeratin,chromogranin along with ,synaptophysin and S100 respectively.[Table-1] It is unclear if PBL is related to immunosuppressive in HIV negative patients. EBER is the universal accepted test for confirmation of PBL, it is detected

Volume 3, Issue 3; May-June 2020; Page No.126-130

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by fluorescence or in situ hybridization. In PBL 82% of HIV positive and 46% of HIV negative cases showed EBER expression.

Conclusion

Plasmablastic lymphoma arising as a nasal mass is rare in immunocompetent patient. The morphological and immunohistological characters overlap with other lymphoproliferative disorders, posing a diagnostic challenge for the pathologist. Since the entity is very aggressive with an early death, arriving at an early clinical diagnosis with prompt treatment is essential for the survival of such patients.

Ethical Clearance- As no data or patient intervention is done hence there is no scenario of taking ethical clearance as per institute policy.

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Volume 3, Issue 3; May-June 2020; Page No.126-130

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Legends –

Figure 1- CECT paranasal sinuses-A-Axial and B-coronal section section both show soft tissue mass in left nasal cavity with focal bony erosion of surrounding wall.

Figure-2 A Section shows a diffuse infiltration of tumor with a background of inflammatory cells.(H&E,100x). B- Section shows a large tumor cells with round to oval nuclei with vesicular chromatin and centrally placed prominent nucleoli and moderate amount of eosinophilic cytoplasm with necro-inflammatory background. Mitotic figures also noted. (H&E,400X)

Figure-3 A- Diffuse membranous positivity for CD138.(IHC,200X) B- Diffuse membranous positivity for CD 10.(IHC,200X) C-Diffuse nuclear positivity for MUM1.(IHC,200X) D-Focal membranous positivity for CD 79A.(IHC,200X) E- Tumor cells show KI-67 score around 60 %.(IHC,200X) F- Tumor cells are negative for CD56.(IHC,200X) G-Tumor cells are negative for CD20.(IHC,200X) H-Tumor cells are negative for PAX5.(IHC,200X)

Table-1 Differential Pathological Diagnosis of PBL

Figure-1

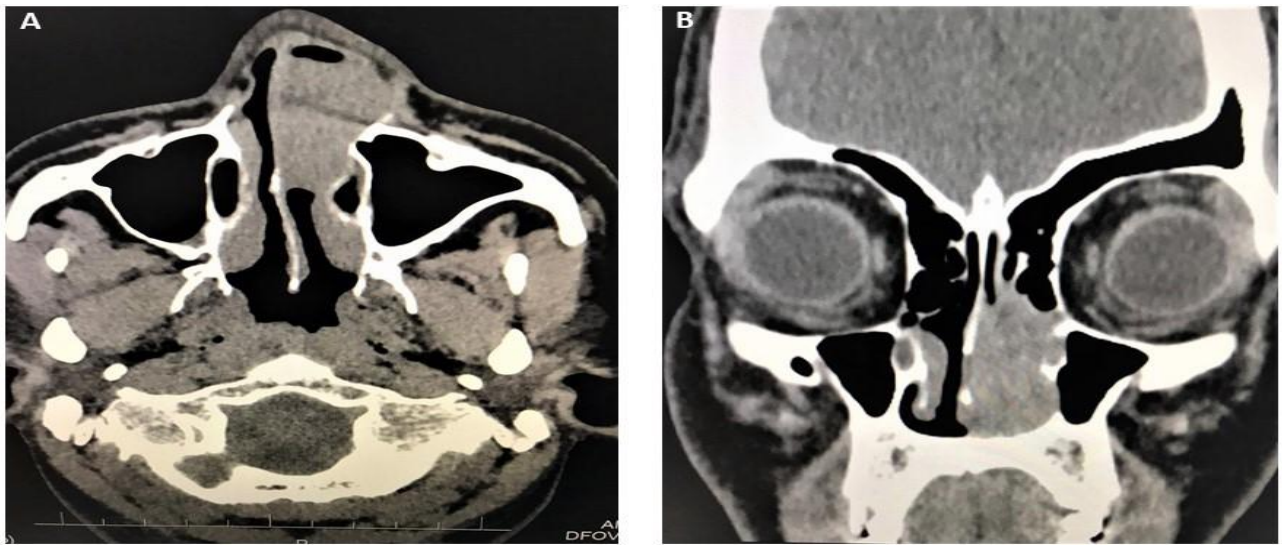
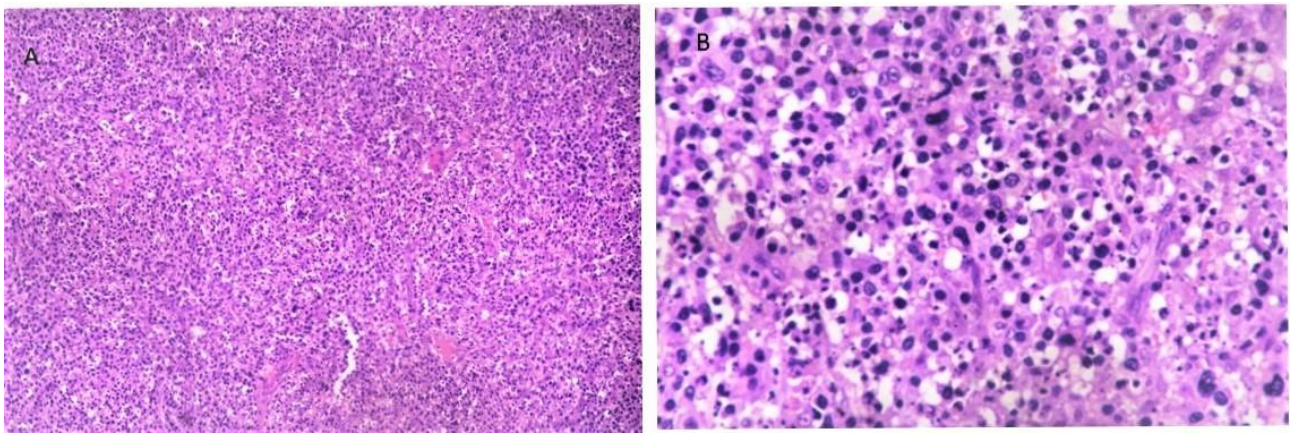


Figure-2



Volume 3, Issue 3; May-June 2020; Page No.126-130
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Figure-3

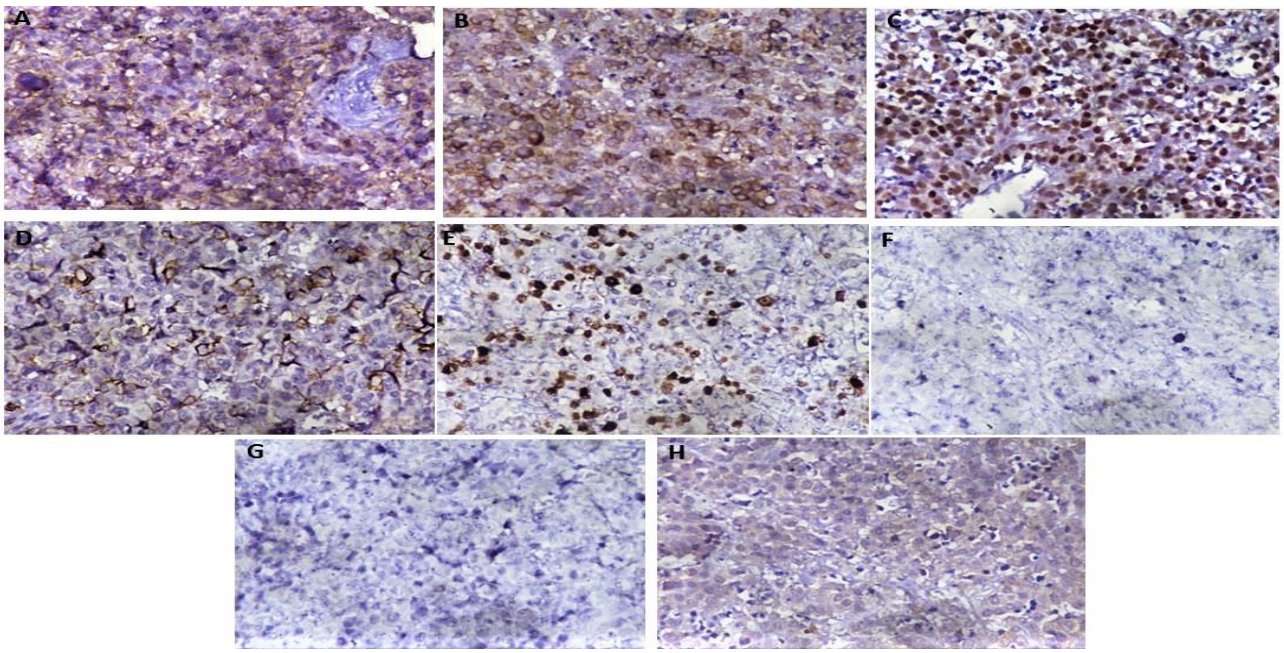

Table-1

IHC	PCM	DLBCL with PD	BL	PDCA	MM	PBL
CD138	++	+	-	-	-	++
CD56	+	-	-	-	-	-
MUM1	+	+	-/+	-	-	++
CD20	-	+	++	-	-	-
BCL2	++	+	++	-	-	-
CD79a	++	++	++	-	-	++
Ki67	Low<10%	High<90%	High>90%	High<90%	High<90%	High<90%
S100	-	-	-	-	++	-
PanCk	-	-	-	++	-	-
CRAB	+	-	-	-	-	-

Abbreviations:PCM,Plasma cell myeloma;DLBCLwith PD,Diffuse lage cell lymphoma with plasmablastic differentiation;BL,Burkitt Lymphoma;PDCA,Poorly differentiated carcinoma;MM,Malignant melanoma;PBL,Plasmablastic PanCk-Pancytokeratin Lymphoma;CRAB,Hypercalcemia,kidney disease,anemia and bone lytic lesion.: ++,Majority of cases positive;+,Few of the cases are positive;-/+ Majority of cases are negative;-,negative

Volume 3, Issue 3; May-June 2020; Page No.126-130
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