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Case Report

Plasmablastic lymphoma of transverse colon: a rare case report

Pinal Shah, R. N. Hathila, Jahnavi Vyas*, Rishikesh Balvalli

Department of Pathology, Government Medical College, Surat, Gujarat, India

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***Correspondence:**
Dr. Jahnavi Vyas,
E-mail: vyasjahnavi05@gmail.com

ABSTRACT

Plasmablastic lymphoma (PBL) is a rare aggressive subtype of non-Hodgkin's lymphoma with large neoplastic cells. It is usually associated with human immunodeficiency virus (HIV) infection but also identified in patients with solid organ transplantation and in immunocompetent patients. It frequently presents as a mass in oral cavity, but has also been described in other extra-oral sites like gastrointestinal track, skin, genitourinary track, nasal cavity, paranasal sinuses, etc. It is characterized by plasmablastic features and an immunoprofile close to plasma cells, Epstein–Barr virus (EBV) positivity and MYC gene dysregulation. We report a case of a 40 year old HIV positive male who presented with intestinal obstruction having mass in transverse colon. Histopathological examination of the excised mass revealed submucosa and muscularis propria infiltrated by monotonous population of medium to large sized lymphoid cells with plasmacytic differentiation. The tumour cells were immunoreactive for EMA, CD138 and Vimentin and immunonegative for LCA, CK, S-100, Chromogranin, CD20, CD30, CD3. Thus the final diagnosis of Non-Hodgkins Lymphoma – Consistent with Plasmablastic Lymphoma was made. PBL should be carefully differentiated from Plasmablastic Plasma cell myeloma, other CD20 negative B-cell neoplasma i.e. primary effusion lymphoma, anaplastic lymphoma Kinase (ALK)-positive large B-cell lymphoma, large B-cell lymphoma arising in human herpesvirus 8 (HHV-8)-associated multicentric Castleman disease.

Keywords: Extraoral site, HIV positive, Plasmablastic lymphoma

INTRODUCTION

Plasmablastic lymphoma (PBL) is a very rare and aggressive lymphoma with a diffuse proliferation of large neoplastic cells, most of which resemble B-immunoblasts or plasmablasts, that have a CD20-negative plasmacytic phenotype. PBL remains a diagnostic challenge due to its characteristic morphology and an immunohistochemical profile similar to plasma cell myeloma.[1] PBL is a very infrequent entity, thought to account for approximately 2.6% of all AIDS-related lymphomas.[2]

The diagnosis of PBL is challenging and difficult because it has overlapping features with those of myeloma and with lymphomas that have plasmablastic morphology. This complexity declares that PBL cannot be readily classified as a B-cell lymphoma or a plasma-cell neoplasm. The difficulty of diagnosing this disease is compounded by its aggressive, relapsing clinical course, which poses management and therapeutic challenges, and also by high rates of disease progression and fatality despite the use of state-of-the-art treatment modalities. Given its rarity, no standard of care has been established for PBL. However, with a better understanding of the biology of PBL, there is promise for improved outcomes.[3]

CASE REPORT

Authors report a case of 40 years old HIV positive male who presented with abdominal pain, abdominal distension and not passing stool since 2 days. He had a past history of tuberculosis 6 years ago, along with history of alcohol intake since 10 years and tobacco chewing since many years.

Per abdomen examination revealed distended abdomen with umbilicus-centrally placed, guarding and rigidity-present and bowel loops palpable in right iliac fossa. On auscultation, bowel sounds were sluggish. Ultrasonography showed intestinal obstruction, dilated bowel loops and possibility of mass lesion in intestine along with mild hepatomegaly. Exploratory Laparotomy with resection of ascending colon and transverse colon with mass lesion with ileo-transverse anastomosis with diverting loop ileostomy was done.

Well-preserved specimen of caecum with appendix, ascending and transverse colon with mass was received and processed routinely. Grossly, one exophytic circumferential growth measuring 10x4x1.5 cm^3 in size was identified in transverse colon, greyish white in colour and firm in consistency (Figure 1).

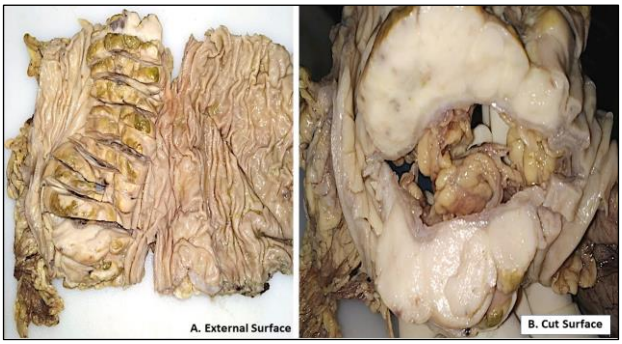

Figure 1: External and cut surface of the tumour in transverse colon.

Histological examination revealed submucosa and muscularis propria infiltrated by monotonous population of medium to large sized lymphoid cells (Figure 2) which were round / plasmacytoid having round / oval nuclei, coarse chromatin, prominent nucleoli and moderate amount of cytoplasm (Figure 3) along with binucleated and multinucleated cells and frequent mitosis. At places, apoptotic bodies and tingible body macrophages were also seen (Figure 4).

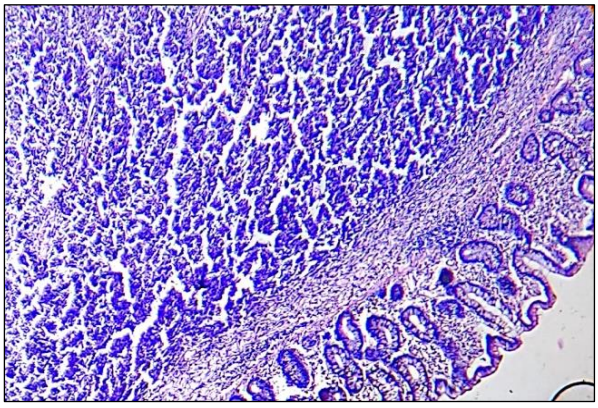

Figure 2: 4x view of tumour (H and E stain) showing diffuse infiltration of lymphoid cells in the submucosa and muscularis.

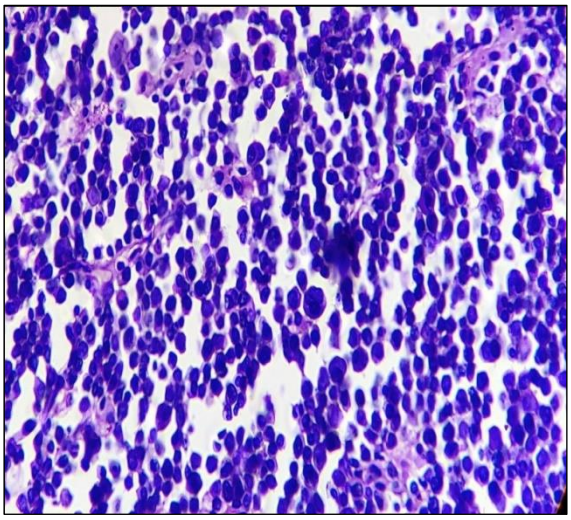

Figure 3: 40x view of tumour cells (H and E stain) showing medium to large lymphoid cells.

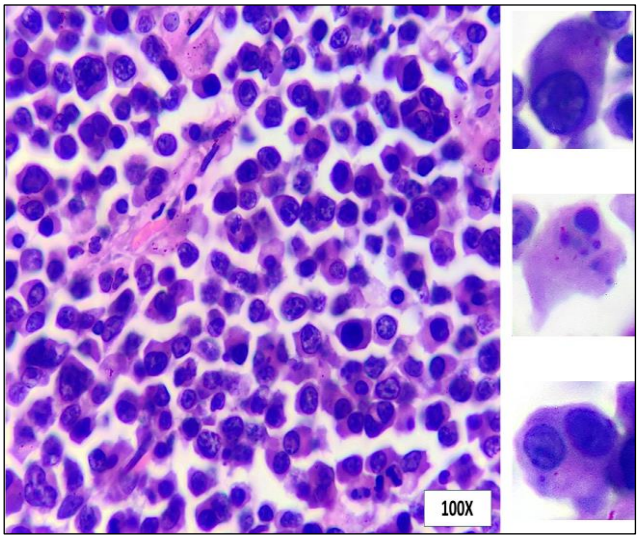

Figure 4: 100x view of tumour cells (H and E stain).

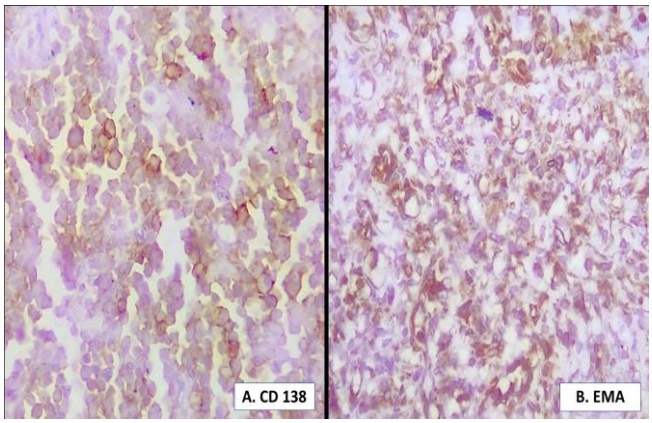

Figure 5: Immunohistochemical staining (A) The lymphoid cells were immunopositive for CD138. (B) The lymphoid cells showed immunopositivity for EMA.

Immunohistochemistry showed that the lymphoid cells were immunoreactive for EMA, CD138 and Vimentin and immunonegative for LCA, CK, S-100, Chromogranin, CD20, CD30, CD3. LCA and CD3 highlighted reactive lymphocytes (Figure 5).

On the basis of morphology and immunohistochemical features, the pathological diagnosis of Non- Hodgkins Lymphoma consistent with plasmablastic lymphoma was made.

DISCUSSION

Plasmablastic lymphoma (PBL) probably is a heterogeneous group of aggressive large B-cell lymphomas composed of large neoplastic cells most of which resemble immunoblasts or plasmablasts.[1]

PBL was originally described as a variant of DLBCL, often presenting in the oral cavity of HIV+ patients and referred to as PBL of oral mucosa type.[4] However, PBL is now described as a new disease entity in the 4th WHO classification. PBL accounts for 1.5% of all nodal non-Hodgkin's lymphomas.[5] PBL occurs predominantly in adults with immunodeficiency, most commonly due to HIV infection, however it may be associated with other causes of immunodeficiency. The median age group is 39 years. It most frequently presents as a mass in in the oral cavity with GIT being the 2nd most common site.[1]

PBL show a morphological spectrum varying from a diffuse and cohesive proliferation of cells resembling immunoblasts to cells with plasmacytic differentiation, which may resemble cases of plasmablastic plasma cell myeloma. Cases with monomorphic plasmablastic morphology are seen in HIV infection in oral mucosal type, while cases with plasmacytic morphology are more commonly seen in extranodal sites.[1]

The neoplastic cells express a plasma cell phenotype, including positivity for CD138, CD38, VS38c, IRF4/MUM1, PRDM1 (also called BLIMP1), and XBP1. CD45, CD20, and PAX5 are either negative or sometimes weakly positive in few cells. CD79a is positive in approximately 40% of cases. EMA and CD30 are frequently expressed. The Ki-67 proliferation index is usually very high (>90%). In situ hybridization for EBER is positive in 60-75% of cases. HHV8 is consistently absent.[1]

The main differential diagnosis of PBL are poorly differentiated primary or metastatic carcinoma, malignant melanoma, gastrointestinal stromal tumor (GIST) and lymphoma.

Other differentials include plasmablastic or anaplastic multiple myeloma, Primary effusion lymphoma (PEL), anaplastic large cell lymphoma (ALCL) and diffuse large B-cell lymphoma (DLBCL).

Poorly differentiated carcinoma can be differentiated from PBL according to its consistent immunological staining for cytokeratin. Malignant melanoma can be distinguished by using S-100 protein and HMB45. GIST is mesenchymal tumor of the small intestine, usually characterized by expression of CD34 and CD117.[6]

According to morphology, the main differential diagnosis of PBL is plasmablastic or anaplastic multiple myeloma which is morphologically and immunophenotypically identical to PBL. Features that favor PBL include association with HIV infection and EBER positivity in neoplastic cells. Features favoring myeloma include the presence of monoclonal paraproteinemia, hypercalcemia, renal dysfunction, and lytic bone lesions.[3]

Among lymphomas, other differential diagnosis includes primary effusion lymphoma which usually manifests as pleural or pericardial effusion and is rarely associated with lymphadenopathy or mass, but it shows a strong association with HHV-8.[3] Primary effusion lymphoma (PEL) usually presents as a serous effusion, rare cases of PEL present as a solid tumor mass which are referred to as extracavitary PEL, and must be considered in the differential diagnosis of PBL. Like PBL, extracavitary PEL is strongly associated with HIV infection and displays immunoblastic and/or anaplastic morphology. Primary effusion lymphoma has an immunophenotype that closely resembles that of PBL, with expression of plasma cell–related markers, absence of pan–B cell antigens, and frequent positivity for EBV. However, extracavitary PEL can be readily distinguished from PBL by its universal association with human herpesvirus 8, a feature not seen in PEL.[7]

ALK-positive large B-cell lymphoma is a rare type of B-cell lymphoma that, similar to PBL, is composed of large immunoblastic cells that express plasma cell-related antigens, lack expression of pan B-cell markers and typically show weak or absent CD45 expression. The tumour cells in ALK positive large B-cell lymphoma are positive for ALK in a granular, cytoplasmic staining pattern when assessed by immunohistochemistry, a feature that allows for easy distinction from PBL.

Immunoblastic diffuse large B-cell lymphoma is also a differential diagnostic consideration, however, the expression of pan Bcell antigens (ie, CD20, CD19, and PAX-5) in immunoblastic diffuse large B-cell lymphoma makes this distinction reasonably straightforward, as PBL is usually negative for these markers.[7]

CONCLUSION

Plasmablastic Lymphoma is a rare entity, seen commonly in immunocompromised patients. It commonly occurs in oral cavity, however it may present to extraoral sites like GIT, skin, etc. Because PBL does not express common lymphoid markers, it is easily mistaken as poorly differentiated carcinoma or melanoma or sarcoma.

Awareness about this entity, as well as immunohistochemistry and association with clinical findings are crucial for establishing a correct diagnosis.

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