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Primary Diffuse Large B-Cell Lymphoma of spleen

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ABSTRACT

Splenic lymphoma is often a manifestation of the diffuse dissemination characteristic of Hodgkins and non-Hodgkins lymphoma, the diagnosis of which is generally made via biopsy of peripheral lymph nodes or bone marrow. However, primary splenic lymphoma (PSL) is quite rare, with a reported incidence of less than 1%. In this report, we present the case of LARGE B - CELL LYMPHOMA(LBCL) with prominent splenic involvement as PRIMARY SPLENIC LYMPHOMA in which the original diagnosis was made at splenectomy.

KEYWORDS : Spleen, Primary, Large B cell lymphoma, Splenectomy.

INTRODUCTION;

Non-Hodgkin lymphomas of various histologic types can occur as primary splenic lymphomas. The great majority of cases are of B-cell lineage, with diffuse large B-cell lymphoma being the most common.[1,2,3]However, initial presentation in the spleen occurs infrequently.To qualify for a diagnosis of primary splenic lymphoma, the lymphoma should be confined to the spleen and/or splenic hilar lymph nodes, although some authors also accept the presence of bone marrow involvement.[4] The patients present with splenomegaly (abdominal pain or mass), with or without fever, systemic upset, or Thrombocytopenia.[4]

Typically when diffuse LBCL involves the spleen it forms discrete tumoral masses within the splenic parenchyma.[5]

A recent report detailed other less common patterns of splenic involvement by LBCL including diffuse infiltration of the splenic red pulp.[6]Because different studies have used different criteria for inclusion of cases, it is difficult to obtain an accurate view of the clinico-pathologic features.[1] Here we report a case of unusual primary splenic LBCL.

CASE HISTORY :

A 71 yr old male tribal Patient came with complaints of lump in left hypochondrium since 3 months. O/E: A swelling in left hypochondrium and extending into left lumbar region. Margins and borders of swelling are ill defined, surface of the swelling is smooth. No lymphadenopathy. Peripheral smear – shows normal study. On USG of abdomen – Gross splenomegaly with a differential diagnosis of ? Splenic granuloma / metastasis / lymphoma. On CECT – Splenomegaly with hypoechoic lesion - splenic infarct. Splenectomy was done. The indication for splenectomy was mass per abdomen. Post-operative history is uneventful. We could follow the patient for 8 months.

PATHOLOGICAL FINDINGS

GROSS EXAMINATION – Splenectomy specimen measuring 21x15x7cm of weight 1.3 kg. Cut section –shows multiple, circumscribed gray white to gray tan nodules of varying size measuring 0.5cm to

Figure :1 a- Spleen is enlarged and is nodular. 1b- cut section shows multiple circumscribed gray white to gray tan nodules of varying sizes. 3cm in diameter. (Figure 1a & 1b)

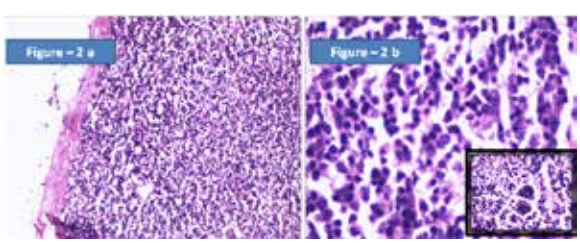

Figure-2: Histopathology shows (2a) Splenic capsule with atypical lymphocytes, seen as (2b) monotonous sheets of round to oval cells with hyper chromatic nuclei showing coarse chromatin and cleaving of nuclei. Inset shows multi nucleated giant cells interspersed in-between the tumor cells.

HISTOPATHOLOGY – Multiple sections studied show thickened capsule with loss of normal architecture of spleen and infiltrated with diffuse sheets of large lymphoid cells with wide range of cell types including centroblasts, immunoblasts and occasional multinucleated giant cells and focal cleaving of nuclei. There are areas of hemorrhage, necrosis & extramedullary hematopoiesis. Considering clinical and histopathological findings we offered a diagnosis of Primary Non-Hodgkin's lymphoma of spleen. **(Figure-2)** Immunohistochemistry was advised.

IMMUNOHISTOCHEMISTRY: Tumor cells are strongly Positive for CD20 and proliferative marker KI 67; Negative for CD3,CD10, CD138, EBER, PAX5, MPO.**(Figure-3)**

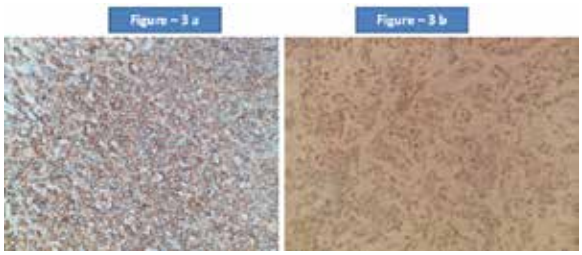

Figure-3: Immunohistochemistry showing tumor cells strongly positive for 3a- CD20 (B-cell marker) and 3b- ki-67(a proliferative marker)

Hence a final diagnosis of Primary Non-Hodgkin's lymphoma of spleen – LARGE B-CELL LYMPHOMA , DIFFUSE TYPE was made considering the clinical, histopathological and Immunohistochemical findings.

DISCUSSION:

Primary splenic lymphoma is an unusual disease, comprising less than 1% of Non-Hodgkin's lymphoma. The spleen may be the primary site of lymphoma or it may be an element of disseminated malignancy. The most common histological diagnoses are large B Cell Lymphomas, which commonly present as diffuse splenomegaly. Symptoms of Primary splenic lymphoma include fever, weight loss, generalized weakness, and left upper quadrant pain or discomfort from splenomegaly.

There are also other specific symptoms that result from direct invasion of the pancreas, stomach, diaphragm, colon, or greater omentum. The most common appearance of Primary splenic lymphoma on diagnostic imaging studies is hypodense lesions on contrast-enhanced CT scans and hypoechoic lesions on sonography. Primary splenic lymphoma also has a clinically recognizable pattern.

The definition of PSL is controversial; it can be summarized with three different definitions. Das Gupta et al. adopted a restrictive definition of PSL as a lymphoma involving only the spleen and the splenic hilar lymph nodes.[7]

Skarin et al. suggested that the diagnosis of PSL can be made if splenomegaly is a predominant feature in any lymphoma involving the spleen.[8]

On the other hand, Kraemer et al. reserved the diagnosis of PSL for patients with splenomegaly, cytopenia of at least two hematologic cell lines, and the absence of peripheral adenopathy.[9]

Because different studies have used different criteria for inclusion of cases, it is difficult to obtain an accurate view of the clinic-pathologic features.[1]

Three patterns of splenic involvement by diffuse large B-cell lymphoma have been identified:.[8]

Macronodular tumor, with predominantly stage Idisease, BCL6 positivity, and favorable clinical outcome.

Micronodular pattern, with predominantly advanced stage disease, BCL6 positivity, and poor prognosis. This pattern is often produced byT-cell/histiocyte–rich large B-cell lymphoma

The micronodules comprise numerous CD3+ T cells, CD68+ histiocytes, and scattered large CD20+ B cells with immunoglobulin light chain restriction. The large B cells arepositive for BCL6 but negative for CD10. No FDC meshworks are demonstrated in the micronodules.

Other splenic lymphomas with large B-cells are T/HRBCL -
Splenic B-cell lymphomas with >55% prolymphocytes in blood are considered to be a transformation of underlying low-grade B-cell lymphoma including SMZL or splenic diffuse red pulp small B-cell lymphoma and they are often p53+, de(7q) .[11]

Red pulp primary DLBCL is an aggressive disease,Often it is disseminated in the bone marrow (intrasinusoidal and interstitial) and liver; typically, LNs are not involved, but involvement of perisplenic LN has been reported. Spread in LN different than intravascular DLBCL.[12]

The overall survival is about 50%, irrespective of histologic type. Some studies suggest that splenectomy followed by combination chemotherapy may improve the survival.[2]

In our case, as the patient had only complaint of splenomegaly without any other system involvement, basing on clinical, pathological and immunohistochemical findings we made a final diagnosis of Primary diffuse large B-cell lymphoma of spleen.

CONCLUSION

We presented this as Primary diffuse large B-cell lymphoma of spleen is very rare and there is no data up to now concerning the best treatment for PSL. After 8 months followup the patient is doing good. Further study is required to compare the outcomes among the following modalities: splenectomy only, chemotherapy after splenectomy, radiation therapy after splenectomy, or radiation therapy with chemotherapy.

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