Case Report

Primary malignant lymphoma of the spleen

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Splenic lymphoma or primary malignant lymphoma of the spleen is rare with reported incidence less than 1% of all Non Hodgkin's lymphomas. Some authors consider PMLS is an entity limited to the spleen and splenic hilum[1].

Most PMLS are of B cell origin, most common histological picture being low grade or intermittent grade lymphomas[2].

Clinical picture of anemia and pan cytopenia which are usually refractory to medical therapy may be due to PMLS.

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Key words **: PMLS, lymphoma of spleen, splenectomy**

Primary malignant lymphoma of the spleen (PMLS) is an unusual malignancy. It comprises less than 1% of all Non Hodgkin's lymphomas. Pan Cytopenia and anemia secondary to primary lymphoma of the spleen is refractory to medical therapy and responds well to splenectomy (Fig 1).

We report a case hypersplenism needed splenectomy which turned out to be PMLS.

Case Report

60 years old woman presented with mass per abdomen, generalized weakness, on and off breathlessness of over 2 years duration. She was a known diabetic and ischemic heart disease patient on medications.

Examination — Her examination was significant for conjunctival pallor, absence of generalized lymphadenopathy, breathless at rest, and hepatomegaly of 2 cm below the right costal margin and massive spleenomegaly of 4 cm below the left costal margin crossing the mid line, and there was no ascites.

Laboratory data included hemoglobin of 5.5 g/dl, haematocrit of 25%, total leukocyte count of 4200/µl (differential: neutrophil-60, lymphocytes- 40). A peripheral smear showed pan cytopenia with dimorphic anemia, platelet count of 80,000/ µl with reticular count of 1.4% and other investigation such as LFT, Urine and Serum chemistry were within normal limits.

Bone marrow revealed hyper plastic megaloblastic picture with mild dyserythropoiesis, and meta myelocytosis .

Chest X-ray showed cardiomegaly, and ECG - sinus tachycardia with ischemic changes.

USG revealed, hepatomegaly of 15cm with normal architecture, and massively enlarged spleen of 25cm, homogenous structure with pre and para aortic lymph adenopathy.

The patient underwent splenectomy through left sub costal incision under G.A, along with excision of lymph nodes at the hilum.

Grossly, the spleen weighed 2500gms, measured 28cm x13cm x 7cm in dimensions.

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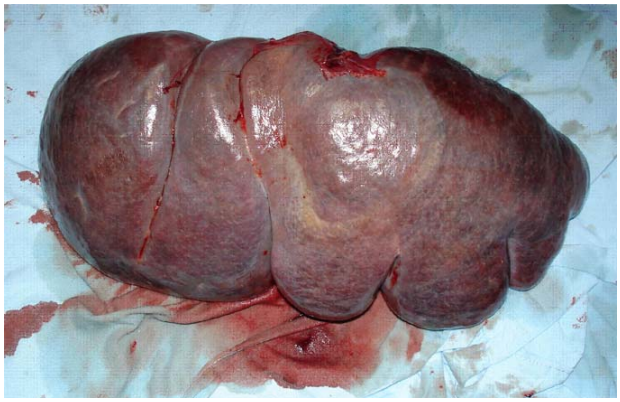

Fig 1 — Splenectomy - Specimen (28x13x7 cm weight 2500 gms)

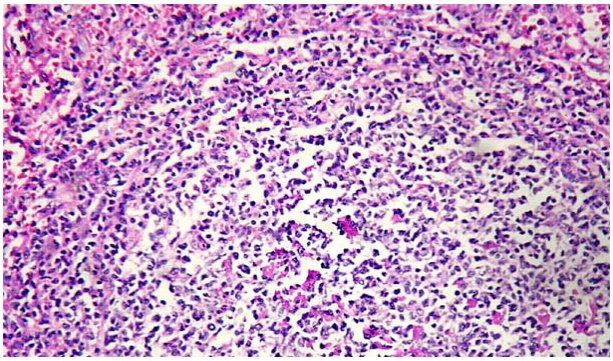

Fig 2 — Small lymphocytic type, Follicular arrangement Malignant lymphoma – Spleen (Microscopy 20x H &E)

Cut section showed multiple small grayish white miliary nodules.

Microscopy showed large nodular areas of lymphoid aggregates both in red and white pulp (Fig 2).

Nodules varying in size, follicles composed of monotonous population of cells.

Hilar lymph node show similar cell pattern, adherent pancreatic tissue is free.

IMPRESSION : MALIGNANT LYMPHOMA SPLEEN

IMMUNO- HISTO CHEMISTRY WAS CONSISTENT WITH

SPLENIC MARGINAL ZONE LYMPHOMA, CD 20 + Ve.

LIVER BIOPSY SHOWED NORMAL HEPATIC ARCHITECTURE WITH NO TUMOR INFILTRATION.

The patient doing well at 6 months follow-up.

Discussion

Spleen is commonly involved in disseminated lymphoma, to qualify for primary splenic lymphomas, the lymphoma should be confined to the spleen and /or splenic hilar lymph nodes[3].

Non Hodgkin's lymphomas of various histologic types are represented amongst the primary splenic lymphomas. Majority of cases are of B-cell lineage and, diffuse large cell lymphomas is the commonest type.

Malignant lymphomas of the spleen may be detected as asymptomatic spleenomegaly or result in a picture of hypersplenism.

Gross patterns are (1) Homogenous (diffuse), (2) Miliary (micro nodular), (3) Multiple masses (macro nodular), (4) Solitary masses (Macro nodular).

World Health Organization classification system, splenic marginal zone lymphoma is described as an indolent B-cell lymphomas which generally presents as spleenomegaly with involvement of the bone marrow and peripheral blood. Splenic MZL is characterized by micro nodular infiltration of the spleen with marginal zone differentiation[4].

Splenic marginal zone lymphoma (SMZL) is characterized by micro nodular infiltrate of the splenic white pulp, centered on preexisting follicles with a peripheral rim of marginal zone B-cells, always accompanied by a variable degree of red pulp infiltration. Immuno histochemistry is helpful in differentiating various types of lymphomas[5].

In large analysis of 42 cases of splenic marginal zone lymphoma in post splenectomy specimens after established World Health Organization criteria, following pattern were noted 1) Predominantly random growth. (24) 2) predominantly (11/42) or exclusively (7/4) diffuse infiltration, 21 cases showed bi-phasic appearance, 13 cases showed marginal zone morphology.

These results confirm heterogenesity regarding histologic, immuno histo-chemical and molecular variety of splenic marginal zone lymphomas and indicate origin from the diverse resident B-cell population of the normal splenic marginal zone.6

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