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Primary Splenic Diffuse Large B-Cell Lymphoma with Massive Splenomegaly

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Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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Case Study

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ABSTRACT

Primary malignancies of the spleen are very rare, diffuse large B-cell lymphoma of the spleen is a rare primary neoplasm. Primary splenic diffuse large B-cell lymphoma is a type of non-Hodgkin's lymphoma that constitute, less than 1% of non -Hodgkin's lymphoma and less than 2% of all lymphomas. Primary splenic lymphoma has been associated with the hepatitis C and HIV virus. It affects men more than women and present in late middle age.

Patient with diffuse large B cell lymphoma typically present with fever, fatigue, weight loss, palpable splenomegaly and left upper quadrant pain. Diffuse large B cell lymphoma stage I being limited to the spleen, whereas stage II spleen and hilar Lymph-node involvement and stage III is distant metastasis including liver and surrounding organs involvements, stomach, pancreas, kidney and colon. Stage I disease, a splenectomy is the standard treatment for DLBSL. Radiation and chemotherapy can be used. When the cancer is confined to the spleen, the prognosis is quite favourable with a median survival of 7.48 years.

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We are reporting an extremely rare case of primary splenic lymphoma in a 45 years old female patient, presented with abdominal pain and huge splenomegaly. The CT revealed a bulky, low density, irregular mass in the spleen and clinically disease limited to spleen only. So we performed open total splenectomy.

Keywords: Diffuse large B. cell lymphoma; primary splenic lymphoma; splenomegaly.

1. INTRODUCTION

"Lymphoma is a type of blood cancer that develops from white blood cells called lymphocytes grow out of control. There are two types of lymphocytes. B-lymphocytes (B-cell) and T-lymphocytes (T-cell). Lymphoma can be grouped as Hodgkin's lymphoma or Non-Hodgkin's lymphoma. Primary splenic diffuse large B-cell lymphoma DLBCL is a rare type of Non-Hodgkin's lymphoma. It commonly present with abdominal pain, splenic mass and an elevated level of lactate dehydrogenase level. Primary splenic lymphoma are again subtypes" [1-4].

1. Marginal zone lymphoma
2. Diffuse large B-cell lymphoma
3. T-cell lymphoma

"Diagnosis is bases on imaging findings of splenomegaly with hypoechoic masses on ultrasonography and CT scan. Treatment option includes total splenectomy, chemotherapy and splenic radiation" [2-4].

2. CASE PRESENTATION

A 45 years old female was admitted to our centre on 10/02/2011, with complaints of weight loss, abdominal pain and distention of abdomen. Physical examination revealed, she had a protuberant abdomen with firm, huge palpable splenomegaly extending from xiphisternum to below the umbilicus. There was no ascites or hepatomegaly or lymphadenopathy. Abdominal ultrasonography and CT showed massive splenomegaly size 27x15x10 cm with a huge heterogeneous hypoechoic masses in the spleen. Haematological examination revealed pancytopenia. WBC 1.9 × 109 /L (neutrophils 48.0%, eosinophils 0%, basophils 0%, monocytes 4.0%, lymphocytes 48.0%), Hb 10.3 g/dL, platelet count 99 × 109 /L, LDH 127 U/L (normal range 100-220 U/L). Liver and renal functions were within normal limits. Bone marrow test was normal. A final diagnosis of a primary splenic malignant tumour was reached and patient was prepared for surgery.

The patient underwent total splenectomy, through a mid-line surgical approach. During the operation, Patient had huge splenomegaly with multiple nodular lesions on the surface of the spleen. The liver, mesenteric or retro peritoneal lymph nodes were not involved. The spleen was adherent to stomach, omentum and sub-diaphragmatic area and was separated carefully. Total splenectomy done, achieving good haemostasis, drainage tube kept and abdomen closed in layers.

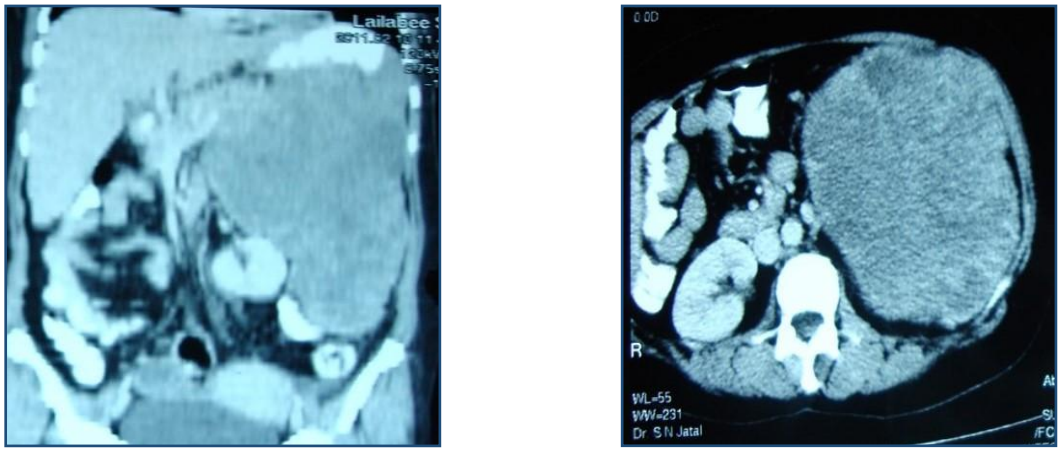

Fig. 1. CT Abdomen showing 27x15x10 cm splenomegaly with hypoechoic lesion in spleen

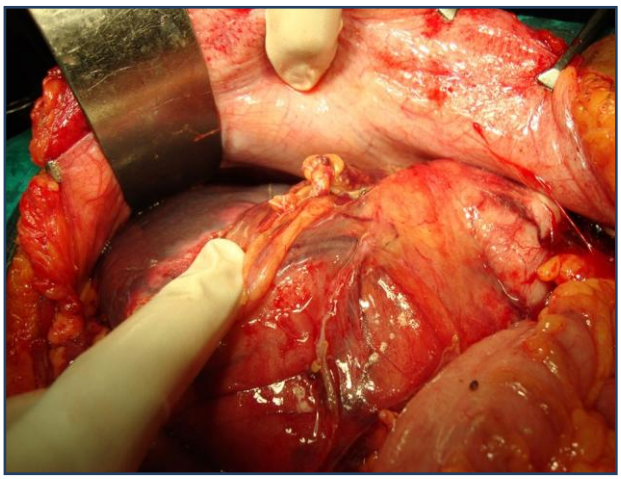

Fig. 2. Intra-operative photograph showing huge splenic mass at of lower pole of spleen

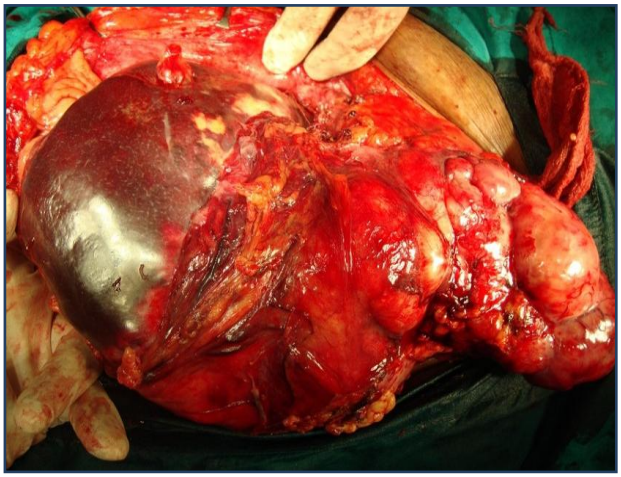

Fig. 3. Intra-operative photograph showing huge splenic mass with splenomegaly

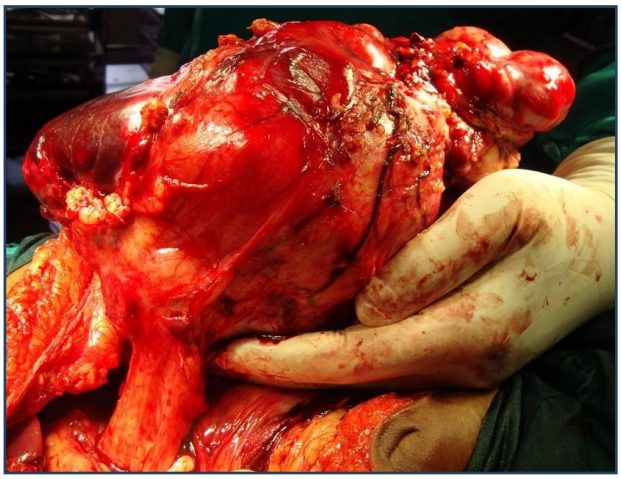

Fig. 4. Intra-operative photograph showing huge splenic mass with splenic pedicle

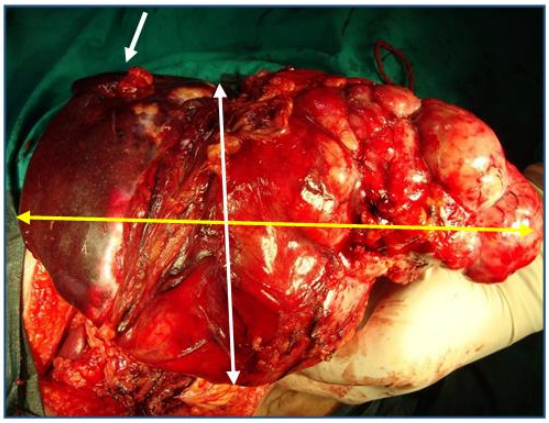

Fig. 5. Intra-operative photograph showing huge splenic mass of size 27x15x10

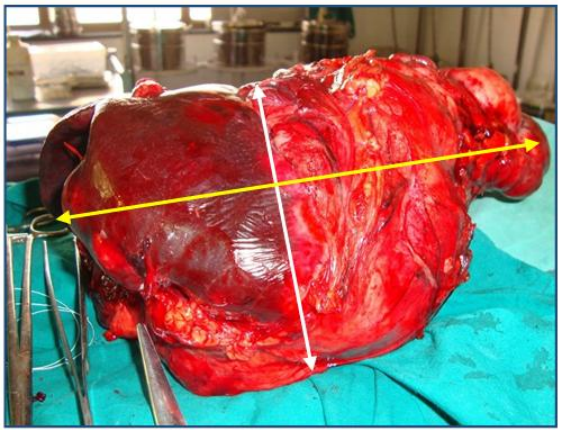

Fig. 6. Showing huge Splenomegaly with huge splenic mass of- weighing 4 kg

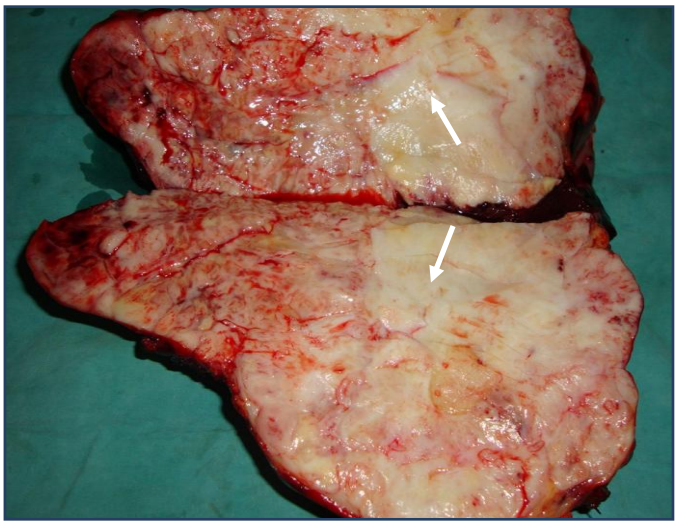

Fig. 7. Cut -section showing gray – white fleshy tumor

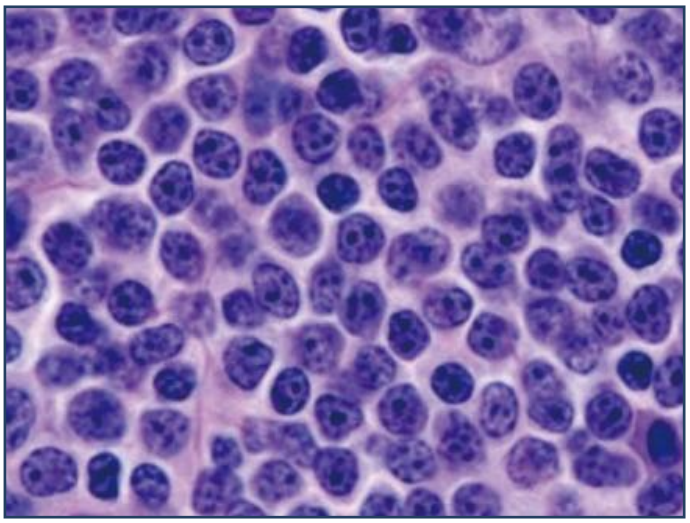

Fig. 8. Microscopic -diffuse large B-cell lymphoma

Gross specimen of spleen weighing 4 kg. Massive splenomegaly size 27x15x10 cm. The Cut surface was almost totally effaced by a huge grey-white homogenous tumour. Microscopically revealed diffuse proliferation of large neoplastic B-cell lymphoma –stage I. Splenic lymphoma was diagnosed and there were no hilar lymph-nodes. Eight days after the operation, the patient revealed from the pancytopenia. WBC of 7.6 × 109 /L, Hb 13.0 g/dL and platelet count 330 × 109 /L although CRP increased to 8.21 mg/dL. Patient recovery was uneventful and patient discharged on 10th post-operative days. The patient was advised chemotherapy with CHOP regimen and after one year follow up, patient is found to be symptom free and healthy (Figs. 1-8).

3. DISCUSSION

"Primary splenic diffuse large B Cell lymphoma is very rare as it occurs in less than 1% of non-Hodgkin's lymphoma" [2,5]. "It is most commonly found in females and older males. Symptoms can include splenomegaly, left upper quadrant pain, fever and weight loss. It can also be associated with HIV virus and can present with metastasis to hilar and retroperitoneal lymph-nodes. It also causes cystopenia" [2,5].

3.1 Ahmann's Classification

Primary splenic diffuse large B-cell lymphoma is classified in three stage.

1. Stage I – Disease confined to the spleen.
2. Stage II – Involvement of the spleen and hilar lymph-nodes.
3. Stage III - Extra splenic nodule or hepatic mesenteric or retroperitoneal lymph-nodes [2-4].

Imaging studies, ultrasonography and CT abdomen shows hypodense or hypoechoic lesion in the spleen. Primary splenic lymphomas histological subtypes including.

1. Diffuse large B-cell lymphoma
2. Marginal zone lymphoma
3. Anaplastic large cell lymphoma (ALCL)
4. Follicular lymphoma [2,3]

The most common variant is diffuse large B-cell lymphoma accounting for approximately 31% of all non-Hodgkin's lymphoma cases, anaplastic variant of diffuse large B-cell lymphoma was first defined in the 2017 [6-8]. "A rare morphological variant of DLBCL and constitutes less than 3.4% of all DLBCL cases. AV –DLBCL affect nodal and extra nodal sites. Patient present with fever, weight loss and night sweats" [2,4].

"Splenectomy is considered the first line of procedure and has shown high efficacy in both diagnosis and treatment of splenic lesions. Several studies have demonstrated the role of splenectomy in improving the prognosis of primary splenic lymphoma [9,10]. Adjutant chemotherapy including R-CHOP regimen i.e. Rituximab, cyclophosphamide, doxorubicin, vincristine and prednisolone" [2].

Bairey etal studies 87 patient with primary splenic DLBCL, Splenectomy and chemotherapy had a better free survival. Brox etal studies 9 patients with primary splenic lymphoma and median survival time of 7.48 years who underwent splenectomy with chemotherapy. "Splenic irradiation is also treatment option for inoperable cases, in order to reduce the size of the spleen" [2,5].

4. CONCLUSION

In cases of massive splenomegaly, primary splenic diffuse large B-cell lymphoma a total splenectomy is the gold standard procedure. Radiation and chemotherapy can be used. When the cancer is confined to the spleen, the prognosis is quite favourable with a median survival of 7.48 years.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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