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Primary Splenic Diffuse Large B Cell Lymphoma after Splenectomy: A Rare Case with Literatur Review

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Abstract Primary splenic diffuse large B cell lymphoma is rarely clinical entity and comprises 1% of all malignant lymphomas. The spleen is also involved in immune defense against blood-borne antigens. Though the haematological malignancies, spleen is usually affected as a part of multisystemic involvement rather than isolated involvement. More than half of patients affected by Hodgkin's disease and about a third of those with non-Hodgkin lymphoma have splenic involvement. The involved spleen may be complicated with rupture due to massive splenomegaly, which may need urgent intervention. In this report, we present a 47-year-old female patient with massive splenomegaly, who was diagnosed with primary splenic diffuse large B cell lymphoma after splenectomy.

Keywords: *diffuse large B-cell lymphoma, splenomegaly, splenic neoplasms*

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1. Introduction

Non-Hodgkin lymphoma (NHL) originates from lymphatic hematopoietic system and can be classified as B cell lymphomas (about 90% of all NHL) and T cell lymphomas (10% of all NHL) [1]. Since the spleen plays a major role in the filtration of blood, primary splenic tumors are generally haematological malignancies, which are mostly lymphomas [2]. Hodgkin and Non Hodgkin lymphomas may affect spleen as a part of systemic involvement, whereas primary splenic lymphoma is rather rare and has an incidence of less than 1% [3]. In this report, we present a case evaluated for surrenal malignant mass, who was later diagnosed with primary splenic diffuse B cell lymphoma.

2. Case Report

A 47-year-old female patient was admitted to the outpatient clinic with a 6 months history of abdominal pain which was precipitated with breathing, fatigue, and weight lost. The patient lost 12 kilograms weight in one year. Her past medical history revealed 6 years history of antidepressant drug use and she has a 17 pack-year history of smoking. Her family history revealed nothing abnormal. On physical examination, arterial blood pressure was 120/80 mmHg, pulse was 82/min and body temperature was 36.6°C. There was no palpable lymphadenopathy on the neck, axillary and inguinal region. On abdominal examination, a hard, painful and rough surfaced mass lesion reaching to splenic area, which was mobile on respiration was palpated on the left upper quadrant, about 6 centimetres below left costal archus. Examinations of other organ systems were normal. All laboratory examinations were normal except high LDH value of 766 U/L (normal: 125-220). The patient underwent abdominal ultrasonography examination, which showed hepatomegaly, measuring 19 centimetres in long axis. There was a 15x10 mm sized isohyperechogenic solid nodular lesion on segment 3 of left liver lobe, which was compatible with haemangioma or metastasis. Size of the spleen was also increased (16.5 centimetres) with expansile, 16x8 cm sized, heterogeneous mass lesion in middle-lower part, which had necrotic center and showed hypervascular character on doppler ultrasonography. For the exclusion of angiosarcoma, contrast enhancing abdominal MRI was performed and lesion was reported as angiosarcoma (Figure 1). Since 15x10 mm sized isohyperechogenic solid nodular lesion of the liver was consistent with metastasis, 18-fluorodeoxyglucose positron emission tomography/computed tomography (^{18}F-FDG PET/CT) examination was performed and showed increased FDG

uptake of 14x10 cm sized splenic lesion (SUVmax=13.5) which was consistent with splenic malignancy. Considering splenic rupture, the patient was vaccinated against pneumococcus and haemophilus influenzae and undergone diagnostic splenectomy. Grossly, the weight and the size of the spleen was increased (1089 gr in weight and 17x13x9 cm in size) and spleen had a lobulated appearance (Figure 2a). Cut surface of the spleen showed diffuse tumoral infiltration containing patchy and yellowish areas (Figure 2b). Histologically, nodular and diffuse patterned neoplastic infiltration of atypical lymphocytes some of which exhibiting plasmacytoid cytomorphology was seen (Figure 3a-3f). Deposition of eosinophilic material accompanying the neoplastic infiltration was noted (Figure 3a-3c). Histochemical studies such as Kongo Red and Crystal Violet revealed that this material was not amyloid Immunohistochemical studies revealed diffuse large B cell lymphoma with plasma cell differentiation via positive immunoreaction of neoplastic cells with the antibodies such as CD20 and CD79a. (Figure 4a-4b). Eosinophilic material was positive with Kappa and Lambda was negative (Figure 4c-4d). Ki-67 proliferating index was 85-90% (Figure 4e). The patient was discharged on 4th postoperative day uneventfully and referred to haematology department for adjuvant oncological therapy.

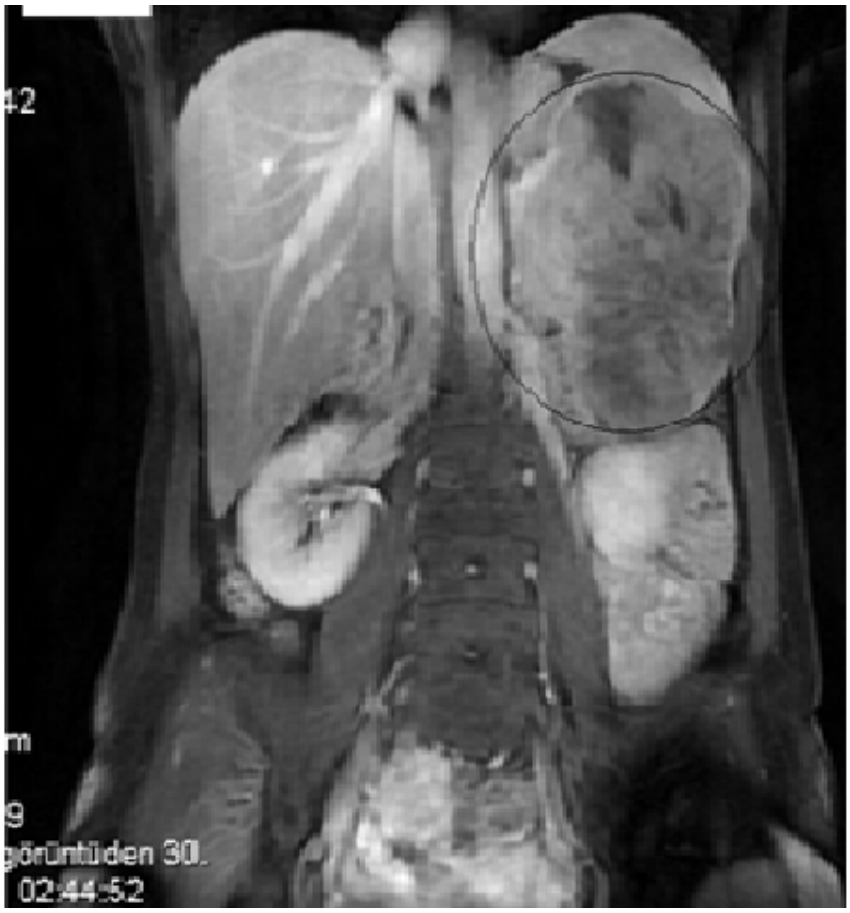

Figure 1. Contrast enhanced MRI showed a 16 cm sized heterogeneous lesion which had cystic necrotic areas

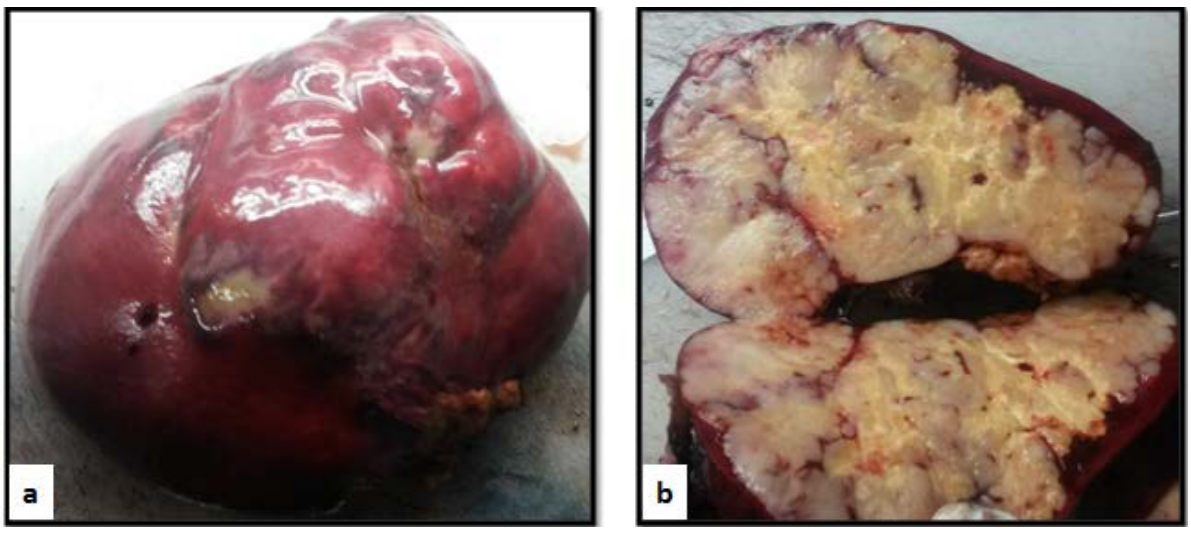

Figure 2 a,b. Grossly, spleen had a lobulated appearance. Cutsurface shows diffuse tumoral infiltration containing patchy and yellowish areas

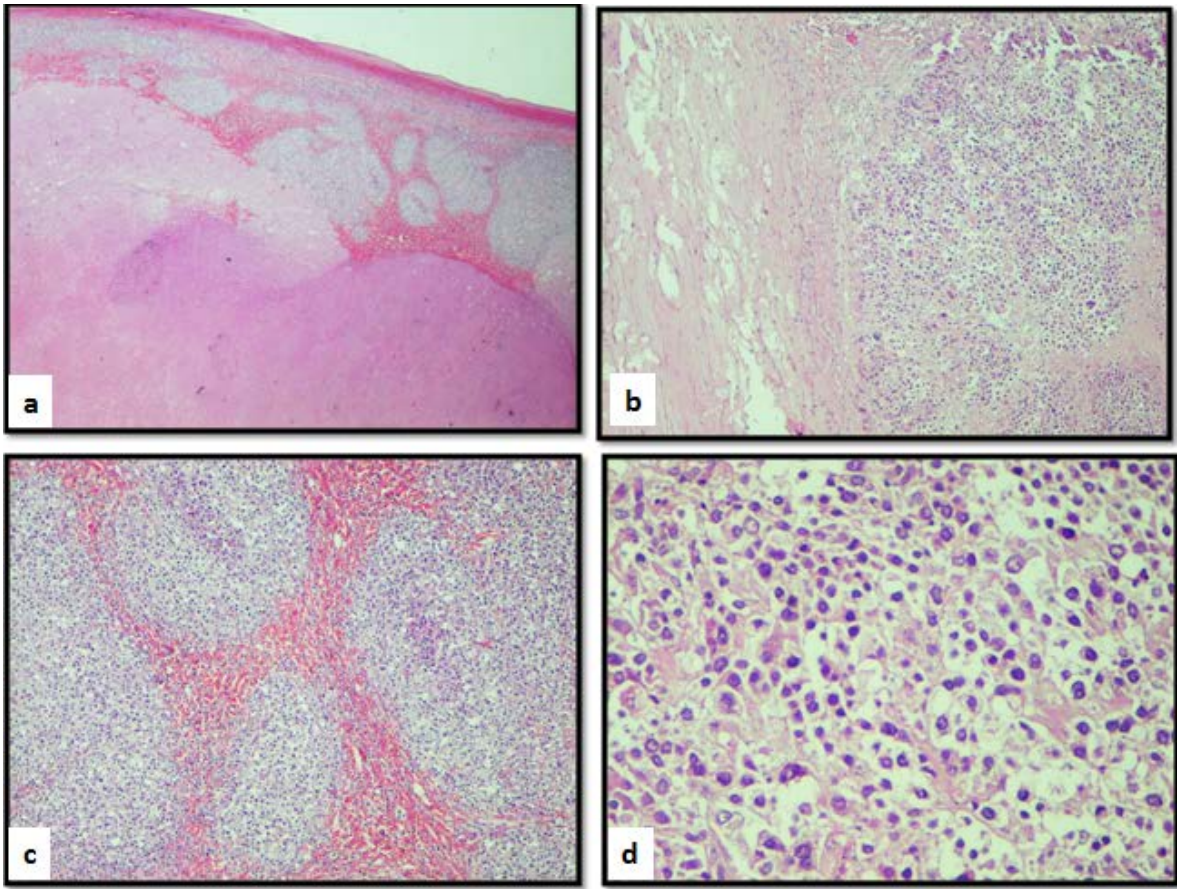

Figure 3. a-d. Histologically, neoplastic infiltration of atypical lymphocytes in nodular and diffuse pattern, some of which exhibit plasmocytoid cytomorpholgy was seen. Deposition of eosinophilic material accompanying the neoplastic infiltration was also noted

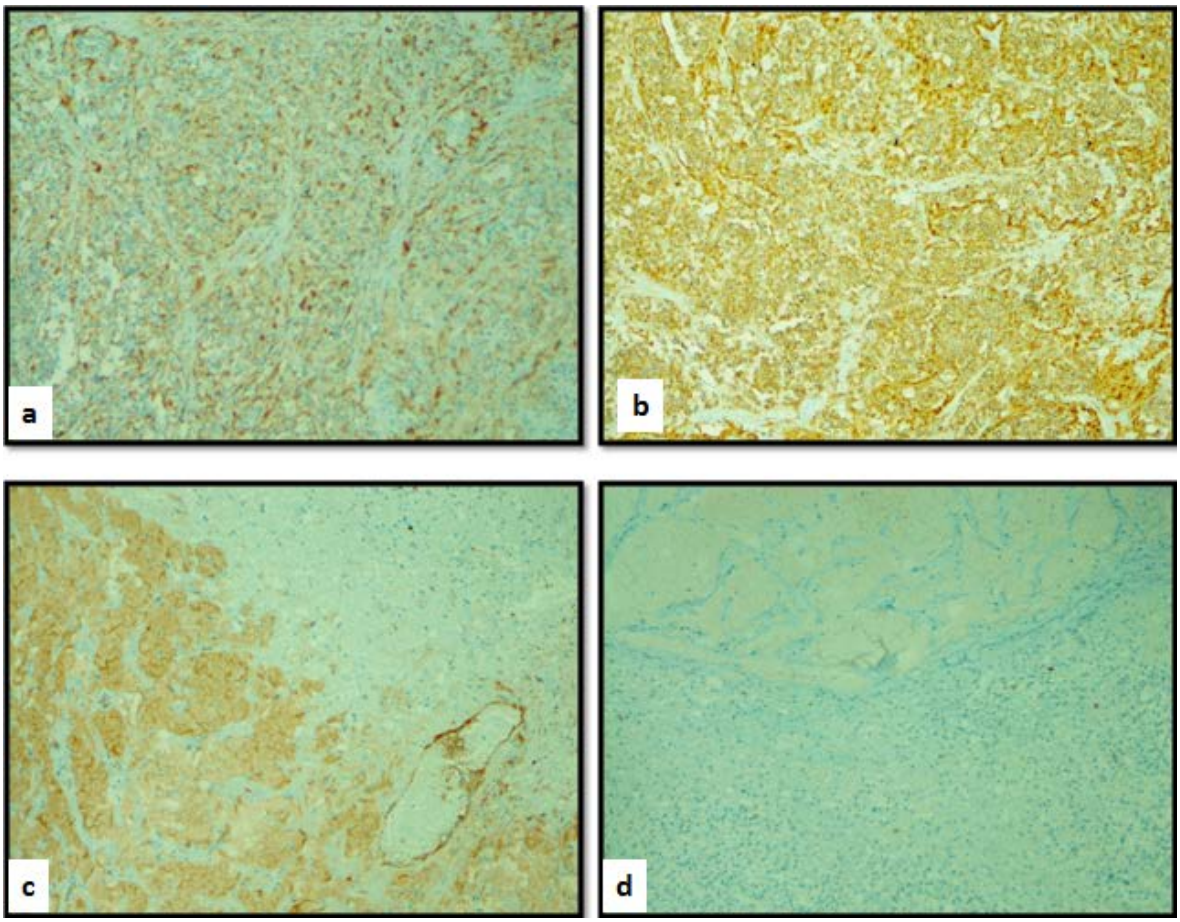

Figure 4. a-d. Neoplastic cells showed immunoreaction with CD20, CD79a and eosinophilic material was also positive for Kappa, but there was no expression of Lambda

3. Discussion

Lymphomas are malignancies originating from lymphoid tissue and they are classified as Hodgkin and non-Hodgkin lymphomas. Most of the time, primary lymphomas occur in lymph nodes, although they may also be seen in some extranodal soft tissue and organs such as thyroid tissue, nasopharynx, tonsillar tissue, spleen and gastrointestinal system [4]. Primary splenic lymphomas (PSL) comprise approximately 1% of all malignant lymphomas [5]. Das Gupta et al. [6] described primary splenic lymphoma as 'malignancy involving only spleen and splenic hilar lymph nodes with preservation of other organ systems' PSL is generally presented as B cell non-Hodgkin lymphoma. Secondary involvement of spleen is usually seen together with the involvement of other abdominal lymph nodes. Major symptoms of PSL include fatigue, weight loss, fever, left sided abdominal pain and abdominal distension secondary to splenomegaly. Other symptoms usually develop secondary to direct invasion of pancreas, stomach, diaphragm and omentum [7]. Our patient had a 6 months history of fatigue and a 1-year history of weight loss and left sided abdominal pain. Since deep anatomical location of the disease, patients are usually asymptomatic in early phases until splenomegaly or disease progression becomes evident. Splenic weight may be up to 0.5-1 kilogram when it is diagnosed [8].

Primary splenic lymphomas have 4 distinct radiological appearances: diffuse infiltration with splenomegaly, small, focal or miliary nodules, multiple large nodular lesions and bulky solid masses [3]. On CT and ultrasound images, they are usually seen as hypoechoic lesions. Radiological images taken in early phases of the disease may be reported as angiosarcoma, metastasis or hamartoma [9]. First ultrasonography examination of our patient was reported as malignant mass originating from left surrenal region. Abdominal MR images were consistent with splenic angiosarcoma. Splenic biopsy or splenectomy may be required for definitive diagnosis of PSL [7]. Histopathologically, malignant splenic lymphomas show four different distribution pattern: diffuse infiltration of red pulp, uniform multicentric involvement of white pulp, irregular involvement of white pulp together with secondary involvement of red pulp, early localisation in periarteriolar lymphoid sheath and/or marginal zone [10]. In our case, tumoral infiltration was present in all parts of splenic tissue. Splenectomy, chemotherapy and radiation therapy without splenectomy are treatment modalities for PSL, although there is no consensus on the latter one. International Prognostic Index (IPI) is used to predict the prognosis of patients with non-Hodgkin's lymphoma. This index covers five clinical risk factors: age ≥ 60 years, elevated serum lactate dehydrogenase levels, performance status ≥2 (ECOG) or ≤ 70 (Karnofsky), Ann Arbor stage III or IV, >1 site of extranodal involvement. This is a powerful predictor of outcome in all subtypes of non-Hodgkin's lymphoma. (0-1 factor = low risk: 35% of cases; 5-year survival 73%, 2 factors = low-intermediate risk: 27% of cases; 5-year survival 51%, 3 factors = high-intermediate risk: 22% of cases; 5-year survival 43%, 4-5 factors = high risk: 16% of cases; 5-year survival 26%). In addition, patients with splenic lymphoma are divided into three groups: in group I, only spleen is involved, group II patients have splenic and hilar lymph node involvement and in group III, lymph node involvement is present beyond liver and splenic hilus. Our case is accepted as group I (only splenic involvement). Ahman et al. reported that the 5-year survival rate of all his PSL patients was 31% and 40% for those with group I and II [3].

4. Conclusion

Primary splenic diffuse large B cell lymphoma is a rare malignancy. Differential diagnosis may be difficult based on radiological images, biopsy and splenectomy may be needed for definitive diagnosis.

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