

## Primary Splenic Lymphoma Presenting as Large Left Upper Quadrant Mass: Case Report with Literature Review

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### Abstract

Primary splenic lymphoma (PSL) is a rare clinical entity with diffuse large B cell lymphoma type (DLBCL) being an even rarer diagnosis. We present a case of primary splenic DLBCL presenting with fever, weight loss and a palpable mass in the left upper quadrant. We discuss the clinical picture, imaging features, hematological investigations and the treatment.

**Keywords:** Primary Splenic Lymphoma; Diffuse Large B Cell Lymphoma; PET/CT Scan

### Case Report

Our patient is a 63 -year-old female who presented with a diffuse pruritic nodular rash and later developed severe fatigue, night sweats, loss of appetite and weight loss. She also had swelling in her left upper quadrant of the abdomen due to splenomegaly. Blood work was suggestive of iron deficiency anemia. CT scan demonstrated a 7.5 cm large hypodense heterogeneously enhancing mass in the spleen with an additional mass at splenic hilum invading the left adrenal and stomach. Core biopsy of the splenic mass was performed which revealed CD20 positive diffuse large B cell lymphoma with large nuclei and prominent nucleoli. The Ki- 67 index in the large neoplastic cells was brisk at approximately 60%. The Ki-67 index is tumor proliferative index and higher value > 60% is suggestive of decreased time to progression and poor overall survival [1]. Approximately 30% of the neoplastic cells expressed MUM-1 (Multiple Myeloma 1) and MYC protein (Myelocytomatosis) and a majority of the neoplastic cells expressed bcl-2 (B-Cell Lymphoma 2). The overall morphology and immunophenotype were diagnostic for a DLBCL of non-germinal center origin. FISH (Fluorescence In Situ Hybridization) studies for bcl-2 and MYC rearrangements were negative.

### Introduction

Splenic lymphoma is well demonstrated by cross-sectional imaging such as CT and MRI but the presentation is usually nonspecific. Most common primary tumors of the spleen are benign such as hemangioma (most common benign primary splenic tumor), hamartoma, lymphangioma, hemangioendothelioma, hemangiopericytoma, Littoral cell angioma, lipoma, fibroma and inflammatory pseudotumor. Malignant tumors of the spleen are uncommon and include lymphomas, metastases, and hemangiosarcoma's. Primary splenic lymphoma, although a rare entity, is still the most common primary splenic malignancy.

### Discussion

Primary splenic lymphoma (PSL) is defined as a lymphoma confined to the spleen and splenic hilar lymph nodes. PSL is extremely uncommon with a reported incidence of only 1% and is mostly non-Hodgkin's type [2]. Splenic involvement is reported in 15 - 40% of all

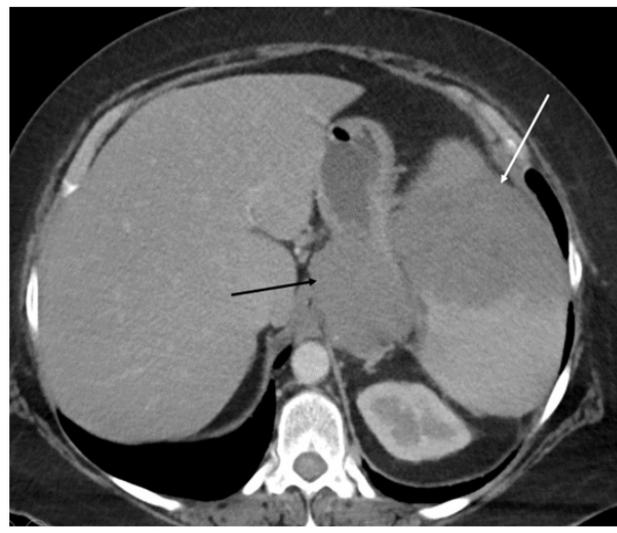
NHL and up to 50 - 80% at autopsy but PSL is only found in 1-6% of lymphoma cases. PSL is mostly small-B cell lymphoma such as marginal zone, mantle cell or follicular lymphoma and less commonly (< 1%) diffuse large B-cell lymphoma (DLBCL) [3,4]. Early diagnosis is helpful in good prognosis but is difficult as the clinical presentation is usually nonspecific with fatigue, fever, weight loss and abdominal pain being the most common initial symptoms. Some cases present with symptoms secondary to direct invasion of adjacent organs like the stomach, pancreas, omentum, and diaphragm.

Imaging workup includes ultrasound, CT scan, MRI and FDG PET/CT. On ultrasound, single or multiple hypoechoic lesions are typically seen in the spleen. CT usually shows hypodense lesions with or without hilar lymphadenopathy and better characterizes adjacent organ invasion. FDG PET/CT has high sensitivity in the detection of splenic lesions and is used in pretreatment evaluation [5]. Splenic hypermetabolism more than liver, splenic nodules and splenic index (length x height x thickness) more than 725 cm<sup>2</sup> have a sensitivity of 100% and specificity of 95% to suggest splenic involvement [6]. On MRI the lesions are isointense to splenic parenchyma on T1 and T2 weighted images or hypointense on T2 images. Upon gadolinium administration hypointense lesions relative to enhancing splenic parenchyma are seen; which later become isointense [7]. This feature helps in differentiating these lesions from metastases which are rarely T2 hypointense and typically show necrosis and hemorrhage [8]. Diffusion-weighted imaging with background body signal suppression (DWIBS) has been shown to provide results similar to PET/CT [9]. DWIBS shows tissue structure and cellularity and may be complementary to PET/CT for detecting aggressiveness of the disease and is able to stage lymphoma according to Ann Arbor staging [10]. The biochemical testing includes routine blood tests, lactate dehydrogenase (LDH), beta-2 macroglobulin, hepatitis testing, uric acid levels and HIV testing. Bone marrow analysis is done to stage the disease. Additional immunohistochemical studies establish the lymphoma subtype. Histologically PSL is classified as red pulp pattern and white pulp pattern; with red pulp pattern having a poorer prognosis [4].

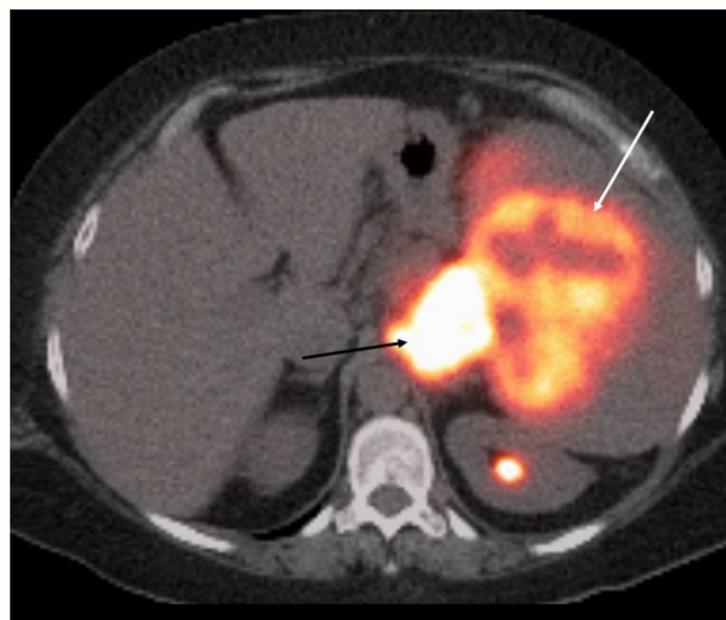
The treatment options include splenectomy, chemotherapy, and immunotherapy. Immunotherapy with rituximab has led to decreased rates of splenectomy in cases of PSL without affecting overall survival [3]. On establishing the diagnosis, primary treatment is with R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) along with involved field radiation therapy (IFRT) [1,10]. Primary splenic DLBCL has more bone marrow infiltration, B symptoms and progression-free survival in comparison to DLBCL. However, there is no difference in overall survival [4]. Our patient was treated with R-CHOP. About a year later she developed relapse and was then treated with RICE (Rituximab, Isocyanide, Carboplatin, and Etoposide). Repeat PET/CT showed remission. She was then offered salvage treatment with R-DHAP (Rituximab, Dexamethasone, High dose Cytarabine and Cisplatin). Later she received a high dose BEAM (Carmustine, Etoposide, Cytarabine, and Melphalan) therapy and autologous stem cell rescue.



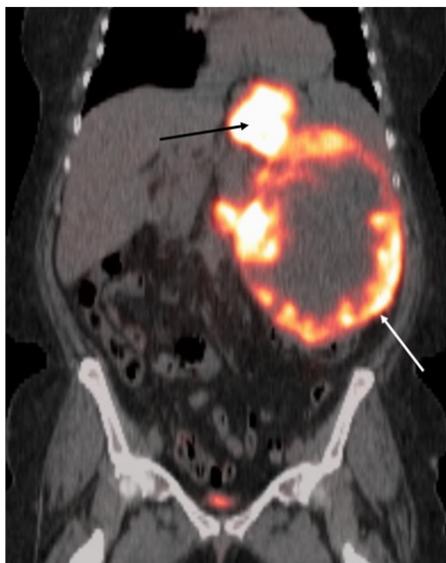
**Figure 1a:** Axial CT scan through upper abdomen showing heterogeneous mass in spleen (White Arrow) and along the posterior wall of stomach (Black Arrow).



**Figure 1b:** Coronal CT scan through upper abdomen showing heterogeneous mass in spleen (White Arrow) and along the posterior wall of stomach (Black Arrow).



**Figure 2a:** Axial PET/CT scan through upper abdomen showing hypermetabolism in spleen (White Arrow) and along the posterior wall of stomach (Black Arrow).



**Figure 2b:** Coronal PET/CT scan through upper abdomen showing hypermetabolism in spleen (White Arrow) and along the posterior wall of stomach (Black Arrow).

## Conclusion

The PSL usually presents as solitary or multiple splenic masse rather than splenomegaly alone and clinical presentation is often vague. The correctional imaging is a helpful initial step in evaluating such cases. DLBCL limited to spleen has a favorable prognosis as compared to other stages. MRI, DWIBS and PET/CT are helpful in evaluating such cases. With newer chemotherapeutic regimens the need for splenectomy for treating this entity has reduced.

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