Localized, Splenic, Diffuse Large B-Cell Lymphoma Presenting with Hypersplenism: Risk and Benefit of Splenectomy

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Abstract

Herein we report a case of a localized, massive, diffuse large B-cell splenic lymphoma diagnosed by splenectomy. A 57-year-old man complaining of weight loss and abdominal pain, was admitted to our hospital. Enhanced computed tomography scanning showed an irregularly enhanced effect in the spleen suggesting a diffuse splenic tumor. Splenectomy was done and the operation progressed without severe complications. The resected spleen weighed 3,500 g. After the operation, the patient recovered from the pancytopenia and pathology diagnosed diffuse large B-cell lymphoma. Standard CHOP plus rituximab chemotherapy was given. Complete remission has continued for 30 months.

Key words: splenic diffuse large B-cell lymphoma, splenectomy, hypersplenism, high mortality

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Introduction

Malignant lymphoma occurs in systemic organs everywhere. Therefore, it is important to carefully determine the appropriate diagnostic measures and treatment. Splenectomy is useful as a diagnostic or a therapeutic method but it is associated with high morbidity and mortality in patients with massive splenomegaly. Here, we report a splenic lymphoma with huge splenomegaly. The risk and benefit of splenectomy are described.

Case Report

A 57-year-old man, complaining of weight loss and abdominal pain, was admitted to Nihon University Nerima-Hikarigaoka Hospital. He was afebrile, and physical examination revealed no palpable peripheral lymphadenopathy. Head and neck examination revealed moderate anemia, no jaundice, and a clear oral pharynx. Heart and lung examinations were normal. He had a protuberant abdomen with a firmly palpable spleen that extended below the navel. There was no ascites or hepatomegaly. His performance status as measured by the Eastern Cooperative Oncology Group score was 1. Abdominal ultrasonography (US) and plain computed tomography (CT) scanning showed massive splenomegaly without a mass lesion (Fig. 1a). Blood 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), CRP 1.79 mg/dL (normal range: less than 0.3 mg/dL), and soluble interleukin-2 receptor (sIL-2R) 4,780 U/mL (normal range: 145-519 U/mL). Air-dried peripheral blood smear showed no abnormal lymphocyte including hairy cell or villous lymphocyte. Liver and renal functions were within normal limits. Bone marrow aspiration and biopsy revealed normocellular bone marrow without abnormal cell involvement, fibrosis, dysplasia or haeomophagocytosis and showed normal karyotype. Flow cytometric analysis also did not identify monoclonal population. Liver cirrhosis and idiopathic portal hypertension were ruled out because of lack of portal hypertension on 3D-CT.

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Figure 1. Plain CT showed massive splenomegaly without a particular finding suggesting malignancy (A), however, enhanced CT scanning did an irregularly enhanced effect in spleen (B). Gallium scintigraphy demonstrated abnormal accumulation in the huge spleen (C). The resected massive spleen weighed 3,500g; the longitudinal diameter was 29cm (D).

Discussion

Here, we reported a case of a solitary, huge, splenic DLBCL presenting with hypersplenism. Lymphoma is the most common tumor presenting with an enlarged spleen or mass lesion in the spleen. In contrast, solitary splenic non-Hodgkin Lymphoma (NHL) is rare; the incidence is less than 1% (1). Some solitary lymphomas cannot be pointed out by plain CT scanning alone or US alone. Consequently, enhanced CT scanning and Gallium scintigraphy are absolutely required to identify splenic tumor. Splenectomy is often chosen to diagnose solitary splenic lymphoma.

However, several previous reports discussed the risk of splenectomy for massive (greater than 1,500 g) splenomegaly. Splenectomy for massive splenomegaly showed a high rate of peri-operative mortality-about 20% (2, 3). In some cases with a poor general status, a bleeding tendency, complications due to infection, or organ failure, one should be hesitant to use an invasive diagnostic method. On the other hand, splenic needle biopsy may provide an adequate diagnosis without severe complications. Tam et al. reported percutaneous image-guided splenic needle biopsy in 156 consecutive cases and concluded that
splenic needle biopsy in the evaluation of new or recurrent neoplasm is a minimally invasive procedure with low complication rates and a high diagnostic yield. If it is institutionally and technically possible, splenic needle biopsy should be taken into consideration for high risk patients (4). Recently, laparoscopic splenectomy has often been used for splenic masses because of fewer complications and because it is rather appropriate for moderate splenomegaly (5).

The present patient underwent splenectomy because he had no severe complication and the following advantages of splenectomy have been proposed: 1) establishing the correct pathological diagnosis; 2) reducing the hypersplenism; 3) reducing the radiation field; 4) relieving symptoms; and 5) preventing spleen rupture.

Pathology-based diagnosis via spleen biopsy in splenic lymphoma has been reported for DLBCL, marginal zone B-cell lymphoma, Hodgkin lymphoma, follicular lymphoma, gamma/delta T-cell lymphoma, and for intravascular lymphoma as a rare case (1-3, 6-12). Moreover, other benign diseases such as tuberculosis, Castleman disease, sarcoidosis and adult onset Still disease have also been diagnosed by splenectomy (2, 3, 7, 8).

Localized indolent lymphoma is expected to be a good prognosis despite the absence of further treatment with chemotherapy. On the other hand, most cases of aggressive lymphoma such as DLBCL show disease expansion and progression, resulting in a requirement for immediate chemotherapy. It is speculated that the high peri-operative mortality rate in massive splenomegaly might include rapid progression of the disease, and such patients should be given a less invasive diagnostic method and treated immediately.

Splenic lymphoma, splenomegaly underlying lymphoma or other hematological malignancies are often reported as a cause of hypersplenism, and cytopenias were resolved after splenectomy in most cases (2, 7, 9-12). Therefore, splenectomy is useful not only for diagnosis but also for treatment underlying hematologic malignancy.

Here, we reported a rare case of localized huge splenic lymphoma diagnosed by splenectomy and the risk and benefit of splenectomy were described. In conclusion, we recommend splenectomy for patients with huge splenomegaly in order to determine the appropriate diagnosis and therapeutic efficacy, unless severe complications are present.

References

4. Tam A, Krishnamurthy S, Pillsbury EP, et al. Percutaneous image-