



# Massive Splenomegaly Successfully Treated with Splenic Irradiation and Rituximab

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## Case report

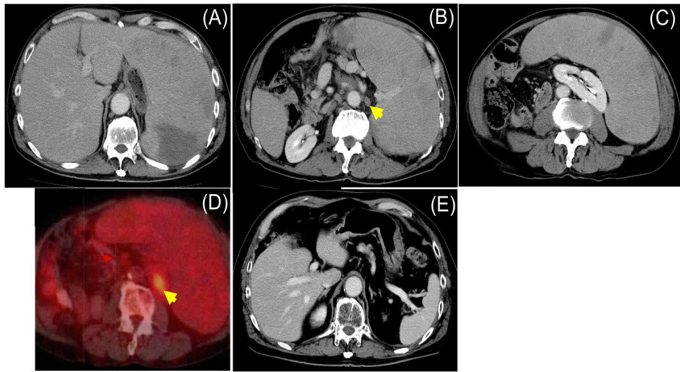
We report an elderly patient with massive splenomegaly in whom treatment with splenic irradiation and rituximab was successful, although the diagnosis was difficult to confirm. A 73-year-old male patient who had been followed for 6 years and who remained untreated for massive splenomegaly and cytopenia was hospitalized with severe abdominal pain. Abdominal Computed Tomography (CT) revealed worsening splenomegaly (maximum major axis diameter 38 cm) with multiple low-density areas suggestive of necrosis (Figure 1A-C). Splenic hilar lymph nodes were slightly swollen (Figure 1B, arrow). Due to splenic enlargement, the left kidney was shifted caudally (Figure 1C). In addition, 18F-2-Fluoro-2-Deoxy-D-glucose Positron Emission Tomography (FDG-PET)/CT revealed no significant uptake in

the spleen, except in the splenic hilar lymph nodes (maximum standardized uptake value (SUVmax) 2.0) (Figure 1D). Pancytopenia persisted (white blood cells 2,300/ $\mu$ L, neutrophils 75%, lymphocytes 20%, hemoglobin 11.7 g/dL, platelets  $5.0 \times 10^4$ / $\mu$ L), and CRP, which remained negative, surged to 9.27 mg / dL. In addition, the soluble interleukin-2 receptor (IL-2R: normal range 124-466 U/mL) level increased from approximately 600 U/mL to 2,150 U/mL. No obvious morphological abnormalities were observed in peripheral blood lymphocytes. Bone marrow aspiration and biopsy revealed some small lymphocytes, and chronic myeloproliferative neoplasms were excluded. The results of flow cytometry, chromosomal analysis, and immunohistochemically evaluation were unremarkable. Our patient was



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considered primary splenic indolent lymphoma; however, not only splenectomy but also biopsy of the spleen was not possible due to the risk of bleeding. Although the diagnosis remained unconfirmed, we initiated treatment with splenic irradiation (52 Gy) after informed consent from the patient and his family. Massive splenomegaly was regressing, and we subsequently administered rituximab (375 mg/m<sup>2</sup>) once a week for 4 consecutive weeks. The size of the spleen gradually decreased and eventually returned to its normal dimensions. The peripheral blood cell count and IL-2R level returned to normal ranges. Currently, the patient (80 years old) has been living splenomegaly free for more than 6 years without additional therapy (Figure 1E).



**Figure 1:** Abdominal images (A-C, E: CT, D: PET-CT) Massive splenomegaly and mild swollen lymph nodes (B and D-arrow) were seen. More than 6 years after treatment, normalized spleen was observed (E).

### Discussion

Our patient was suspected to have slow-growing, primary splenic indolent lymphomas, such as follicular lymphoma, lymphoplasmacytic lymphoma, leukemic non-nodal mantle cell lymphoma and splenic diffuse red pulp small B-cell lymphoma. In particular, we strongly suspected Splenic Marginal Zone Lymphoma (SMZL). SMZL is characterized by splenomegaly, cytopenia (anemia and/or autoimmune thrombocytopenia), and an indolent clinical course [1].

This diagnosis can be differentiated by the immunophenotypic and molecular/cytogenetic findings of resected spleen and/or the bone marrow. However, splenectomy is not feasible in elderly patients with massive splenomegaly, such as our patient [2]. It is more difficult to diagnose in the case of no lymphoma cells in the bone marrow, however, we must not stick to the definitive diagnosis and delay treatment. Core biopsy / Fine Needle Aspiration Cytology (FNAC) was previously not recommended in view of high fragility of splenic tissue leading to hemorrhagic complications, but currently it can be a useful test to confirm the diagnosis [3]. However, we thought that neither reliable diagnosis nor safety can be guaranteed on FNAC, in our patient with diffuse massive splenomegaly that does not form a mass, with low platelet counts.

In our patient, splenic irradiation was chosen to reduce massive splenomegaly. The spleen began to shrink steadily, further suggesting that this patient had primary splenic lymphoma. Patients with SMZL generally exhibit hematological responses to splenectomy and/or rituximab [4,5]. We also believe that it was meaningful to reduce the volume of the spleen by irradiation before administering rituximab to avoid tumor lysis. It is important that hematologists and radiologists work closely together to manage such cases. In the future, a method for treating such patients with massive splenomegaly and low platelets should be established.

### Conclusion

We reported a case of an elderly patient with massive splenomegaly whose platelet count was reduced to 50,000 /  $\mu\text{l}$ , who had been significantly effective for more than 6 years by irradiation and rituximab administration. Because the abnormal lymphocytes in the peripheral blood and bone marrow was observed, it was difficult to diagnose. This time, we did not stick to the diagnosis and intervened in the treatment, but it is hoped that medical treatment methods will be established in the future.

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**Conflict of interest statement:** I declare that I have no conflict of interest in this work.

**Informed consent statement:** Written Informed consent was obtained from the patient for publication of this article.

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