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Simultaneous Malignancy of Primary Splenic Angiosarcoma and Low Grade B-Cell Lymphoma

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Dear editor

We present the first case of simultaneous Primary Splenic Angiosarcoma (PSA), an extremely rare and highly malignant vascular neoplasm, found under detailed examination for low-grade B-cell lymphoma. A 72-year-old male presented at our hospital with multiple lymphadenopathy of up to 15 mm, left pleural effusion, and marked splenomegaly. White blood cell count was within normal limit, but monoclonal B-cell population (1800/μL) was detected. Flow cytometry revealed CD3-, $CD5^+$, $CD10^-$, $CD19^+$, $CD20^+$, CD22+, $CD23^-$, $FMC7^+$, $SmIg\lambda^-$, and $SmIg\kappa^-$. Fluorescence in situ hybridization revealed a positive del (13q14) and trisomy 12 and negative BCL2 split and IGH/CCND1 fusion signals. Bone marrow and pleural effusion analysis detected the same monoclonal B-cell population by flow cytometry as that in peripheral blood. Bone marrow biopsy

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showed hypercellular bone marrow with $CD20^{+}bcl\text{-}2^{+}$B-cell infiltration. Cytogenetic study revealed 2 cells of a clone with trisomy 12 of 20 cells, which were analyzed in pleural effusion. A month after the first visit, he was hospitalized for hepatic bleeding and underwent trans-arterial embolization of artery no 6. Although hepatic bleeding stopped, his platelet counts kept decreasing without lymphocytosis and splenomegaly progressed. We re-examined his bone marrow analysis, which revealed normal hematopoiesis with 10% abnormal mature B-cell infiltration. We suspected his enlarged spleen as the main cause of thrombocytopenia. For histological diagnosis and treatment of thrombocytopenia, we performed splenectomy, which showed proliferation of malignant spindle cells with an irregular and dense nucleus, forming atypical vascular spaces throughout the spleen (Figures 1a-1d). Tumor cells were positive for CD34 and CD31, and negative for D2-40. Ki-67 positive index was 30%. Spleen parenchyma was not infiltrated by lymphoid cells. Based on these findings, a final diagnosis of simultaneous malignancy of PSA and Low-Grade B-Cell Lymphoma Not Otherwise Specified (LGBCL-NOS) was made. We considered that the patient's hepatic bleeding was consistent with a symptom of metastatic angiosarcoma. He received palliative care and died 2 months after the splenectomy.

PSA is an extremely rare (approximately 0.2 per million people) and highly aggressive neoplasm arising from vascular endothelial cells within the spleen [1,2]. Falk S et al. reported that thrombocytopenia was detected in 3% of cases with splenic angiosarcoma [1]. The disease is refractory to chemotherapy and radiotherapy, with a patient survival duration of approximately 4.4-14 months [1,2]. Some reports have indicated an association of PSA with previous chemotherapy for malignant lymphoma and radiation therapy for breast cancer [3].

After an extensive literature search, this was the first case of simultaneous malignancy of PSA and B cell neoplasm that we encountered. Although the patient's abnormal B-cell morphology was similar to Chronic Lymphocytic Leukemia (CLL), surface antigen expression was different to that in CLL ($CD23^{+}$, $FMC7^{-}$); hence, LGBCL-NOS was diagnosed [4]. It is well known that the most common pathological diagnosis for unexplained splenomegaly is lymphoid malignancy (57%) [5], and we considered that B-cell lymphoma might be the cause of our patient's marked splenomegaly. We believe that because of splenectomy for confirming diagnosis, the patient could avoid receiving ineffective chemotherapy.

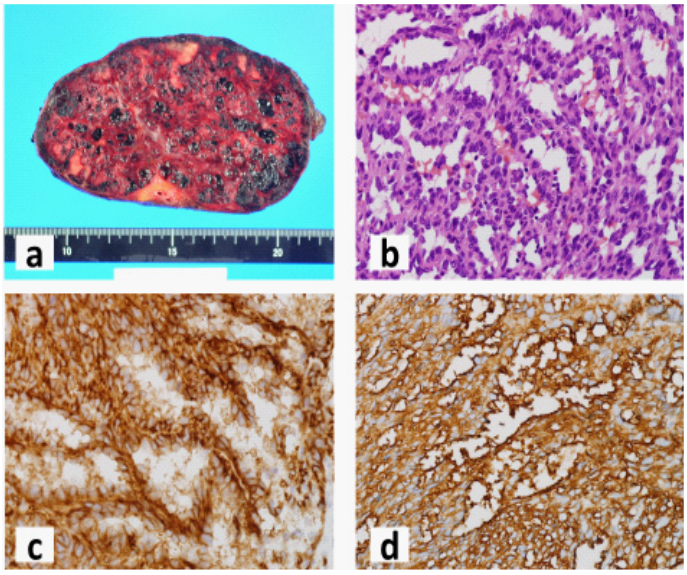

Figure 1: (a): Excised specimen of the enlarged dark brown spleen.
(b): Histopathologic examination of the spleen specimen showed a malignant spindle-cell forming irregular vascular spaces (HE stains × 400).
(c): Immunohistochemical examination of the splenic specimen showed tumor cells positive for CD31 (×400).
(d): Immunohistochemical examination of the splenic specimen showed tumor cells positive for CD34 (×400).

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