



Lymphoproliferative and Subsequent Myeloproliferative Disorder – A One Way Street?

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To the Editor,

Chronic lymphocytic leukemia (CLL) is a malignancy of immunologically incompetent but morphologically mature B lymphocytes. Essential thrombocythemia is a myeloproliferative neoplasm characterized by excessive clonal platelet production. When lymphoproliferative and myeloproliferative disorders are present in the same individual, they are usually discovered at the same time.^{1,2} When separated by time, a new lymphoproliferative neoplasm is usually diagnosed in individuals with established myeloproliferative neoplasm.³ We discuss a case of stable, Stage 0 CLL, who was confirmed to have new Essential thrombocythemia, 7 years after the diagnosis of CLL.

A 63-year-old male with a 14-year history of a treated malignant melanoma presented with left lower limb numbness and tingling. Laboratory results showed elevated IgM of 248 with normal total protein and serum IgG/IgA levels. Serum protein electrophoresis showed no M spike. In the index hematology visit, all blood cell counts were normal which were monitored annually. Subsequent WBCs showed persistent lymphocytosis of $> 5 \times 10^9/L$, with normal hemoglobin (16.1 g/dL) and platelet count ($421 \times 10^9/L$). Peripheral blood flow cytometry confirmed CLL/SLL cells that consisted of 13% of the total nucleated cells which were CD19+, CD20+, CD5+, CD23+, ZAP 70+, surface Lambda-light chain+, and CD38-. (Figure 1a). Peripheral blood fluorescent *in situ* hybridization showed 13q deletion. A diagnosis of Stage 0 CLL/SLL and low-risk disease was made, and the patient is on a watch-and-wait approach to date.

Three years after the diagnosis of CLL/SLL, he developed a superficial venous thrombosis in the left lower extremity. Laboratory results revealed thrombocytosis of $494 \times 10^9/L$, $13.8 \times 10^9/L$ WBCs, 16.2 g/L hemoglobin, and hematocrit of 50.6%. The patient had persistent thrombocytosis with further workup showing positive JAK2 V617F mutation. Break-point cluster region protein/Abelson protooncogene and bone marrow aspiration with biopsy were negative, ruling out Chronic Myelogenous Leukemia. This

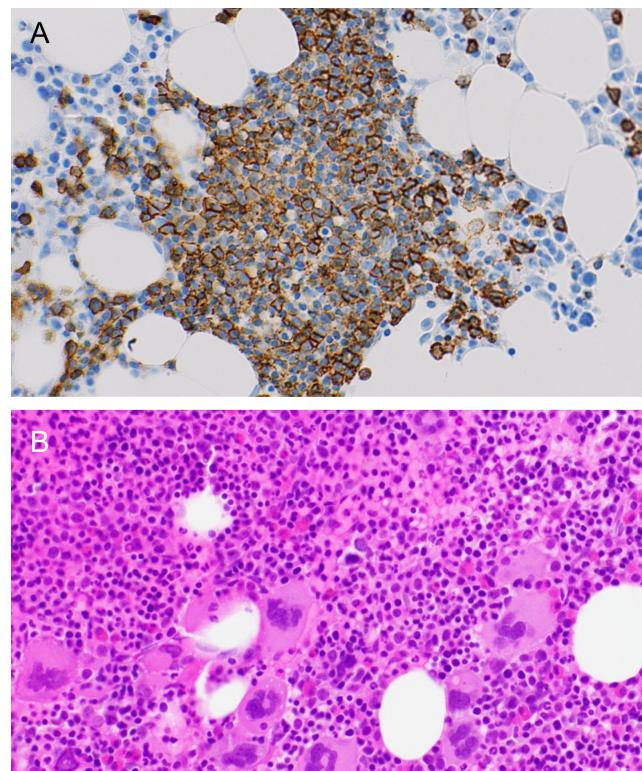


FIG. 1. A, B. (a) Immunohistochemical (IHC) stained CD20+ Silelim lymphoid aggregates in the bone marrow under 400× magnification and (b) hematoxylin and eosin (H & E) stained slide with increased number of megakaryocytes in the bone marrow under 400× magnification.

confirmed the diagnosis of a myeloproliferative neoplasm, most consistent with hemorrhagic Essential thrombocythemia (Figure 1b), a high-risk disease with age > 60 years with leukocytosis, neutrophilia, and a history of superficial venous thrombosis. The patient was then started on hydroxyurea and informed consent was taken.

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Unlike solid tumors, the co-existence of hematologic neoplasms, lymphoproliferative neoplasm, and myeloproliferative neoplasms are not common. Possible pathogenetic mechanisms for their occurrence might include the independent proliferation of 2 distinct cell lines, bi-lineage development of a common pluripotent stem cell proliferation, or an accidental situation.⁴ Some hypothesize that initial “trigger hits” occur in pre-JAK2 progenitor pluripotent hematopoietic stem cells, which makes them genetically labile, and they differentiate into myeloid and lymphoid lineages, with the myeloid cells gaining JAK2 mutation while lymphoid cells undergo malignant transformation prior to the loss of Zap 70 mutation.⁵

We report this case to highlight the co-existence of a myeloproliferative disorder with an established lymphoproliferative disorder, even in patients who have not received chemotherapy.

Patient Consent for Publication: Informed consent was obtained from the patient.

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