Is it Primary Myelofibrosis or Chronic Lymphocytic Leukemia Related Secondary Myelofibrosis?

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Dear Editor,

I read with great interest the short communication by Palta et al. [1] titled, “Simultaneous Presence of Two Hematological Malignancies: Chronic Lymphocytic Leukemia and Myelofibrosis in a Patient,” in which the authors report the unusual coexistence of these two disorders. Focus is given in the central issue and challenge in distinguishing between primary and secondary myelofibrosis due to chronic lymphocytic leukemia (CLL). In this case, there are several arguments against primary myelofibrosis (PMF). Furthermore, reactive myelofibrosis is reversible if the underlying cause can be eradicated so, in this case myelofibrosis cannot be considered a hematological malignancy per se.

Firstly, secondary myelofibrosis is not an uncommon event in CLL, occurring in 20–30% of cases and structural and functional changes of bone marrow stroma including the pattern of fibrosis in CLL have been described before [2].

Secondly, a “sine qua non” prerequisite for the diagnosis of PMF which the authors fail to demonstrate, is the presence of marked megakaryocyte proliferation with atypia in bone marrow. These megakaryocytes show aberrant nucleo/cytoplasmic ratio and hyperchromatic and irregularly folded nuclei with dense clustering, accompanied by

reticulin and/or collagen fibrosis. The demonstration of reticulin fibrosis is not a required criterion for PMF diagnosis. In its absence, in addition to the aforementioned megakaryocyte changes, a hypercellular marrow with granulocyte proliferation and decrease in erythroid precursors is essential [3].

Thirdly, in this case report there was no evidence of demonstration of a clonal marker, i.e. JAK2V617F mutation. Besides, the MPLW515K/L status is unknown [4]. Hereupon, the marrow fibrosis cannot be attributable to a clonal process. Instead, strong evidence points out to a nonclonal nature and the most likely culprit in this case is a lymphoid neoplasm.

Finally, the distinction between PMF and secondary myelofibrosis due to CLL has important clinical and therapeutic implications since the latter can be reversed by controlling the underlying disease while the former has a completely different treatment strategy. Besides, during the clinical follow up of PMF the clinician must stay alert for the small, but real risk of progression from the chronic phase to acute leukemia [5].

Conflict of interest Nothing to declare.

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