Journal of clinical and experimental hematopathology
Vol. 60 No.4, 122-123, 2020

Highlights: B-chronic lymphocytic leukemia

Commentary

Current diagnostic characteristics of and treatments for chronic lymphocytic leukemia in Japan

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Keywords: CLL, CD5, CD23, BTK inhibitors

Chronic lymphocytic leukemia (CLL) is a rare disease in Japan with an incidence of 1/10 that in Western countries.[1] Thus, most studies on CLL are conducted in Europe and America, and the diagnosis and treatment of CLL in Japan are based on Western data.

Recently, new and improved treatments for CLL have become available in Japan. Therefore, this commentary aims to discuss the current and future prospects of CLL treatments and provide significant information for both hematologists and hematopathologists. These findings have been reviewed by experts from the fields of both basic and clinical medicine.

Prof. Yoshino clarified the problems encountered in the diagnosis of CLL and related diseases from the viewpoint of pathology. Dr. Kikushige explained the mechanism of onset and genetic abnormalities of CLL, with reference to his own data. Prof. Kojima reviewed the latest findings regarding the treatment of CLL using targeted drugs. Dr. Uchiyama summarized the current state of minimal residual disease analysis that should be introduced for CLL treatment in the near future.

The International Workshop on Chronic Lymphocytic Leukemia guidelines are commonly followed for diagnosing and treating CLL.[2] CLL is not a general term for chronic lymphocytic proliferative disorders but is used to denote B-cell-derived leukemia with characteristic properties. Morphologically, it is characterized by the presence of mature small lymphocytes, as observed in naturally dried specimens. Immunohistochemically, positivity for CD5 and CD23 is essential for diagnosing CLL. A hematologist should diagnose CLL based on morphological and immunohistochemical findings. Additionally, pathological findings of lymph nodes, if available, can further aid diagnosis. Since there is no specific genetic marker for CLL, an exclusion diagnosis must be carried out. Particularly, mantle cell lymphoma and follicular lymphoma, which have specific genetic markers, should be ruled out. The finding of weak expression of CD20, CD22, CD79b, and surface immunoglobulin also helps in diagnosis but is not essentially required.[2]

Traditionally, CLL treatment has been aimed at improving disease symptoms, although anticancer drugs are prescribed in worse cases. Currently, there is no curative treatment for CLL; only treatments for prolonging progression-free survival and overall survival are available.[3-6] An evidence-based treatment rather than an experience-based treatment should be provided based on the patient's conditions. In particular, the efficacy of Bruton tyrosine kinase (BTK) inhibitors as the initial treatment has been established, regardless of the patient's condition and prognostic factors.[3,4] In the future, the development of new molecular targeted drugs, such as signal transduction inhibitors and antibodies, is expected in Japan. As a specialist in hematology, I think it is our responsibility to provide world-standard treatments to patients with CLL.

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Online Published: December 15, 2020
DOI:10.3960/jslrt.20039
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