**TABLE 134-10 CLINICAL CHARACTERISTICS OF PATIENTS WITH COMMON TYPES OF NON-HODGKIN'S LYMPHOMA (NHL)**

| Disease | Median Age, Years | Frequency in Children | % Male | Stage I/II vs III/IV, % | B Symptoms, % | Bone Marrow Involvement, % | Gastrointestinal Tract Involvement, % | % Surviving 5 Years |
|---|---|---|---|---|---|---|---|---|
| B-cell chronic lymphocytic leukemia/small lymphocytic lymphoma | 65 | Rare | 53 | 9 vs 91 | 33 | 72 | 3 | 51 |
| Mantle cell lymphoma | 63 | Rare | 74 | 20 vs 80 | 28 | 64 | 9 | 27 |
| Extranodal marginal zone B-cell lymphoma of MALT type | 60 | Rare | 48 | 67 vs 33 | 19 | 14 | 50 | 74 |
| Follicular lymphoma | 59 | Rare | 42 | 33 vs 67 | 28 | 42 | 4 | 72 |
| Diffuse large B-cell lymphoma | 64 | ~25% of childhood NHL | 55 | 54 vs 46 | 33 | 16 | 18 | 46 |
| Burkitt's lymphoma | 31 | ~30% of childhood NHL | 89 | 62 vs 38 | 22 | 33 | 11 | 45 |
| Precursor T-cell lymphoblastic lymphoma | 28 | ~40% of childhood NHL | 64 | 11 vs 89 | 21 | 50 | 4 | 26 |
| Anaplastic large T/null-cell lymphoma | 34 | Common | 69 | 51 vs 49 | 53 | 13 | 9 | 77 |
| Peripheral T-cell NHL | 61 | ~5% of childhood NHL | 55 | 20 vs 80 | 50 | 36 | 15 | 25 |

***Abbreviation:*** MALT, mucosa-associated lymphoid tissue.



$>10 \times 10^9$/L) is found (Fig. 134-6) that are monoclonal B cells expressing the CD5 antigen. Finding bone marrow infiltration by the same cells confirms the diagnosis. The peripheral blood smear in such patients typically shows many "smudge" or "basket" cells, nuclear remnants of cells damaged by the physical shear stress of making the blood smear. If cytogenetic studies are performed, trisomy 12 is found in 25–30% of patients. Abnormalities in chromosome 13 are also seen.

If the primary presentation is lymphadenopathy and a lymph node biopsy is performed, pathologists usually have little difficulty in making the diagnosis of small lymphocytic lymphoma based on morphologic findings and immunophenotype. However, even in these patients, 70–75% will be found to have bone marrow involvement and circulating monoclonal B lymphocytes are often present.

The differential diagnosis of typical B-cell CLL is extensive (Table 134-1). Immunophenotyping will eliminate the T-cell disorders and can often help sort out other B-cell malignancies. For example, only mantle cell lymphoma and typical B-cell CLL are usually CD5 positive. Typical B-cell small lymphocytic lymphoma can be confused with other B-cell disorders, including lymphoplasmacytic lymphoma (i.e., the tissue manifestation of Waldenström's macroglobulinemia), nodal marginal zone B-cell lymphoma, and mantle cell lymphoma. In addition, some small lymphocytic lymphomas have areas of large cells that can lead to confusion with diffuse large B-cell lymphoma. An expert hematopathologist is vital for making this distinction.

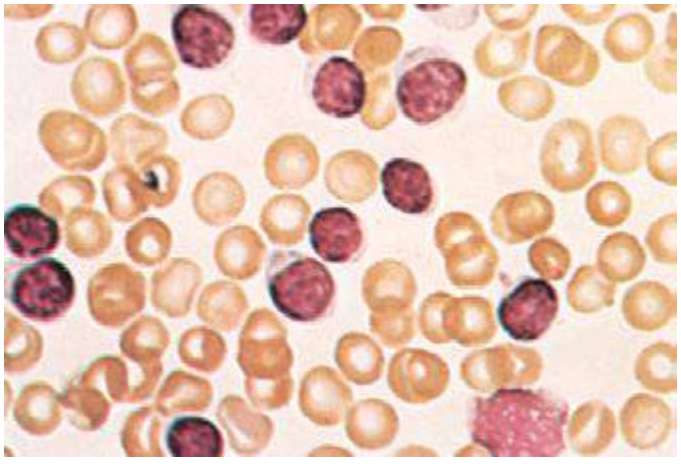

**FIGURE 134-6 Chronic lymphocytic leukemia.** The peripheral white blood cell count is high due to increased numbers of small, well-differentiated, normal-appearing lymphocytes. The leukemia lymphocytes are fragile, and substantial numbers of broken, smudged cells are usually also present on the blood smear.

Typical B-cell CLL is often found incidentally when a complete blood count is done for another reason. However, complaints that might lead to the diagnosis include fatigue, frequent infections, and new lymphadenopathy. The diagnosis of typical B-cell CLL should be considered in a patient presenting with an autoimmune hemolytic anemia or autoimmune thrombocytopenia. B-cell CLL has also been associated with red cell aplasia. When this disorder presents as lymphoma, the most common abnormality is asymptomatic lymphadenopathy, with or without splenomegaly. The staging systems predict prognosis in patients with typical B-cell CLL (Table 134-7). The evaluation of a new patient with typical B-cell CLL/small lymphocytic lymphoma will include many of the studies (Table 134-11) that are used in patients with other non-Hodgkin's lymphomas. In addition, particular attention needs to be given to detecting immune abnormalities such as autoimmune hemolytic anemia, autoimmune thrombocytopenia, hypogammaglobulinemia, and red cell aplasia. Molecular analysis of immunoglobulin gene sequences in CLL has demonstrated that about half the patients have tumors expressing mutated immunoglobulin genes and half have tumors expressing unmutated or germline immunoglobulin sequences. Patients with unmutated immunoglobulins tend to have a more aggressive clinical course and are less responsive to therapy. Unfortunately, immunoglobulin gene sequencing is not routinely available. CD38

**TABLE 134-11 STAGING EVALUATION FOR NON-HODGKIN'S LYMPHOMA**

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| Physical examination |
| Documentation of B symptoms |
| Laboratory evaluation |
| Complete blood counts |
| Liver function tests |
| Uric acid |
| Calcium |
| Serum protein electrophoresis |
| Serum $\beta_2$-microglobulin |
| Chest radiograph |
| CT scan of abdomen, pelvis, and usually chest |
| Bone marrow biopsy |
| Lumbar puncture in lymphoblastic, Burkitt's, and diffuse large B-cell lymphoma with positive marrow biopsy |
| Gallium scan (SPECT) or PET scan in large cell lymphoma |

***Abbreviations:*** CT, computed tomography; PET, positron emission tomography; SPECT, single-photon emission computed tomography.