National Comprehensive Cancer Network®

NCCN Clinical Practice Guidelines in Oncology (NCCN Guidelines®)

Non-Hodgkin's Lymphomas

Version 2.2015

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Lymphoblastic Lymphoma

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Lymphoblastic Lymphoma[a]

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Discussion

DIAGNOSIS[b]

ESSENTIAL:

- **Hematopathology review of all slides with at least one paraffin block representative of the tumor. Rebiopsy if consult material is nondiagnostic.**
- **An FNA or core needle biopsy alone is not generally suitable for the initial diagnosis of lymphoma. In certain circumstances, when a lymph node is not easily accessible for excisional or incisional biopsy, a combination of core biopsy and FNA biopsies in conjunction with appropriate ancillary techniques for the differential diagnosis (immunohistochemistry, flow cytometry, PCR for IgH and TCR gene rearrangements, and FISH for major translocations) may be sufficient for diagnosis.**
- **Adequate immunophenotyping to establish diagnosis[c]**
  - **IHC panel: CD45 (LCA), CD19, CD20, CD79a, CD3, CD2, CD5, CD7, TdT, CD1a, CD10, cyclin D1**
    or
  - **Cell surface marker analysis by flow cytometry: kappa/lambda, CD45, CD3, CD5, CD4, CD7, CD8, CD19, CD20, CD10, TdT, CD13, CD33, CD1a, cytoplasmic CD3, CD22, myeloperoxidase**
- **Cytogenetics ± FISH: *MYC*; t(9;22); t(8;14), and variants or PCR for *BCR-ABL***

USEFUL UNDER CERTAIN CIRCUMSTANCES:

- **Additional immunohistochemical studies to establish lymphoma subtype**
  - **Paraffin panel: CD22, CD4, CD8, cyclin D1**
- **Molecular analysis to detect: antigen receptor gene rearrangements**

WORKUP

ESSENTIAL:

- **Physical exam: attention to node-bearing areas, including Waldeyer's ring, and to size of liver and spleen**
- **Performance status**
- **B symptoms**
- **CBC, differential, platelets**
- **LDH**
- **Comprehensive metabolic panel**
- **Uric acid, phosphate**
- **Chest/abdominal/pelvic CT with contrast of diagnostic quality**
- **Lumbar puncture**
- **Flow cytometry of cerebrospinal fluid**
- **Bilateral or unilateral bone marrow biopsy ± aspirate with flow and cytogenetics**
- **Hepatitis B testing[d]**
- **MUGA scan/echocardiogram if anthracycline or anthracenedione-based regimen is indicated**
- **Pregnancy testing in women of child-bearing age (if chemotherapy planned)**

USEFUL IN SELECTED CASES:

- **Head MRI**
- **Discussion of fertility issues and sperm banking**
- **Beta-2-microglobulin**
- **PET-CT scan[e]**

See NCCN Guidelines for Acute Lymphoblastic Leukemia

[a] The lymphoblastic lymphoma (LL) category comprises two diseases, T-cell LL (LL-T;90%) and B-cell LL (LL-B;10%), which corresponds to T-ALL and B-ALL, respectively, with presentations in extramedullary sites.

[b] This disease is complex and curable; it is preferred that treatment occur at centers with expertise in the management of the disease.

[c] Typical immunophenotype: *LL-B*: sIg-, CD10+/-, CD19+, CD20-/+, TdT+. *LL-T*: sIg-, CD10-, CD19/20-, CD3-/+, CD4/8+/+, CD1a+/-, TdT+, CD2+, CD7+ cytoplasmic CD3+, sCD3-/+.

[d] Hepatitis B testing is indicated because of the risk of reactivation with immunotherapy + chemotherapy. Tests include hepatitis B surface antigen and core antibody for a patient with no risk factors. For patients with risk factors or previous history of hepatitis B, add e-antigen. If positive, check viral load and consult with gastroenterologist.

[e] Initiation of therapy should not be delayed in order to obtain a PET-CT scan.

Note: All recommendations are category 2A unless otherwise indicated.
Clinical Trials: NCCN believes that the best management of any cancer patient is in a clinical trial. Participation in clinical trials is especially encouraged.