BURKITT LYMPHOMA AND LYMPHOBLASTIC LEUKEMIA/LYMPHOMA

Burkitt lymphoma

Highly aggressive but curable B-cell lymphoma with a proliferation of medium sized lymphocytes. May be solid (lymphoma) or liquid (leukemia) or both

Occurs in all age groups, more common in children and young adults (30-50% childhood lymphoma), male:female: 2-3:1

Three epidemiological variants: Endemic (central Africa), sporadic (throughout the world), HIV-associated (in HIV+ patients)

Association with EBV: >95% EBV+ in endemic cases, ~30% EBV+ in sporadic and HIV-associated cases

Tumor sites: Endemic: Jaws and facial bones, Sporadic: Cecum most common, but can involve other extranodal and nodal sites, HIV-associated: extranodal and nodal sites, bone marrow common

Morphology: Diffuse monotonous proliferation of medium-sized nuclei, round nuclear contour, finely clump chromatin, several (2-5) small nucleoli, and a small rim of pink cytoplasm. “Square off” unclear morphology on histology. Very high proliferation rate with many mitotic figures and increased tingible body macrophages (“starry sky” pattern). Deep blue cytoplasm with vacuoles on cytomorphology

Phenotype: Express surface B-cell markers CD19+, CD20+, CD22+, Pax-5 and GC markers CD10+ (consistently) and BCL-6+. CD38+ (bright), clonal surface IG light chain+

Strong MYC+, 100% Ki-67+

Consistently negative for BCL-2, and negative for CD5, CD23, CD34, TdT

Genetics: All cases have MYC translocation: 80% t(8;14), 15% t(2;8), 5% t(8;22). No BCL-2 translocation

Prognosis: Better prognosis, 70-90% long term survival

Variant: Burkitt-like lymphoma with 11q aberration: Morphologically and phenotypically resembles Burkitt lymphoma but lacks MYC translocation, gain of 11q23 and loss of 11q24-ter

B-lymphoblastic leukemia/lymphoma (B-ALL)

A precursor B-cell neoplasm

More common than T-lymphoblastic leukemia, more common in children (75%) May present as leukemia (B-ALL), lymphoma (B-LBL), or both

Morphology: Lymphoblasts in peripheral blood and bone marrow (morphologically indistinguishable from myeloblasts, but typically have more scant cytoplasm)

Phenotype: Expresses one or several surface B-cell markers, consistently expresses cytoplasmic CD22, cytoplasmic CD79a, subset expresses CD10, usually expresses either one or both CD34 and TdT, may express aberrant myeloid markers (CD33 most common)

Prognosis: Good prognosis in children, less favorable in adults

Subtype:

B-ALL with t(9;22)BCL/ABL1: More common in adults, usually presented with high white count, similar morphology and phenotype to other B-ALL, but usually CD10+, p190 protein in children, p190 or p210 in adults, response to TKI, favorable prognosis
**B-ALL BCR/ABL1-like (or Ph-like):** Gene signature similar to B-ALL with BCR/ABL1, but lack this translocation. CRLF2 rearrangement in 50%, remaining cases with variable translocations or mutations in tyrosine kinase genes, response to TKI

**B-ALL with t(v;11q23):** Translocation of KMT2A with various partners, most common t(4;11), more frequent in infants, high white count, CNS involvement, similar morphology and phenotype to other B-ALL (except for CD10- and CD24-), worse prognosis

**B-ALL with t(12;22):** Translocation involving ETV6-RUNX1. Nearly all cases are in children, The cryptic translocation can only be detected by FISH. Better prognosis

**B-ALL with hyperdiploidy:** More common in children, better prognosis

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**T-lymphoblastic leukemia/lymphoma (T-ALL)**

A precursor T-cell neoplasm

Affects children (adolescents) and adults, male>female

T-LBL: mediastinum (thymic) most common, other sites include testes, CNS, LN, liver, spleen

**Morphology:** similar to B-ALL

**Phenotype:** variable pan T-cell antigen expression, CD1a+/-, CD4+/CD8+ or CD4-/CD8-, CD99+ (also+ in Ewing sarcoma), TdT+, CD34+, cCD3+

**Genetics:** TCRαβ (14q11) rearrangement in ~50% cases

**Prognosis:** Children: worse than B-ALL. Adults: better than B-ALL

**Subtype: Early T-cell precursor T-ALL (ETP-ALL):** CD7+, cCD3+, CD1a-, CD8-, Outcome similar to regular T-ALL