Angioimmunoblastic T-cell lymphoma

Definition

-A subtype of peripheral T-cell lymphoma
-Systemic disease
-Polymorphous infiltrate involving LN
-Prominent proliferation of high endothelial venules
-Prominent proliferation of follicular dendritic cells

Synonyms

Lukes-Collins: immunoblastic lymphadenopathy

Working Formulation: various categories
diffuse mixed small and large cell,
diffuse large cell,
immunoblastic,
atypical hyperplasia

Kiel: AILD-Type (lymphogranulomatosis X) T-cell lymphoma

REAL: angioimmunoblastic T-cell lymphoma

Epidemiology

-Middle aged and elderly
-M=W
-15-20% of total PTCL
-1-2% of NHL

Sites of involvement

-Generalized peripheral LNs
-Hepatosplenomegaly
-Skin rash
-BM involvement is common

Clinical features

-Presents with advanced stage
-Systemic symptoms:
  Skin rash, pruritus
  Edema, pleural effusion, arthritis, ascites
-Association with drug hypersensitivity reactions (early series)
-Lab: polyclonal hypergammaglobulinemia,
  immune complexes, cold agglutinins, hemolytic anemia,
  rheumatoid factor, anti-smooth muscle Ab

Etiology

-Immunodeficiency appears to be secondary to AITCL, rather than preceding it
-EBV(75% of cases) in B-cells

Precursor lesions of AITCL

-Angioimmunoblastic lymphadenopathy
(previous thinking)
-de novo PTCL(current thinking)

Histopathology

-LN architecture: partially –completely effaced

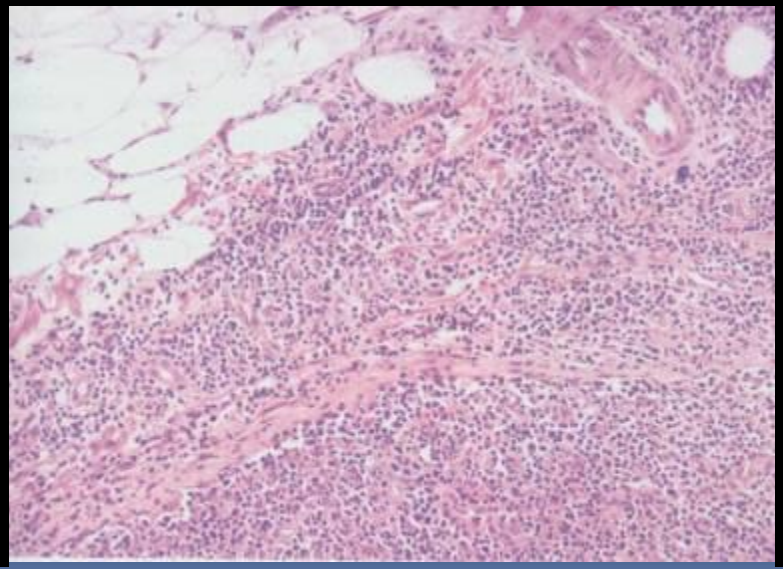
-Regressed follicles

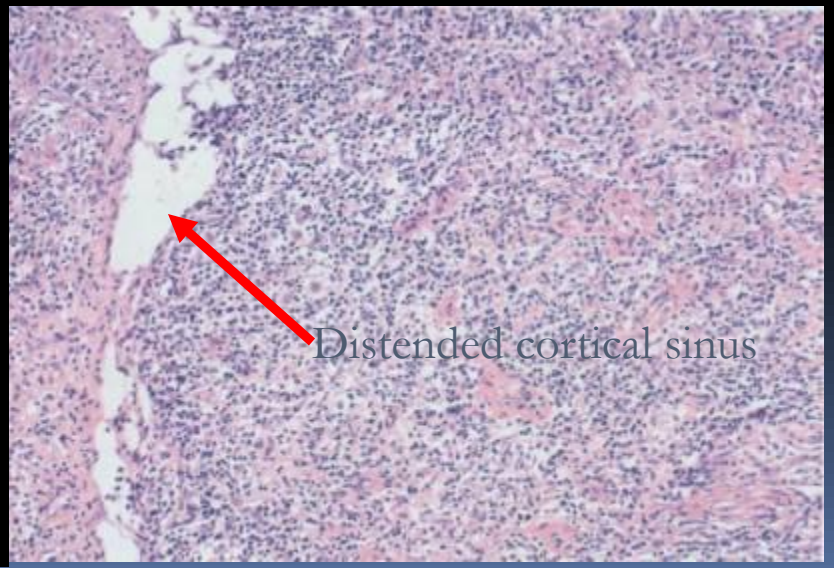
-Paracotex: polymorphous infiltrates

Lymhoma cells: medium to large, pale cytoplasm, distinct membrane, minimal atypia

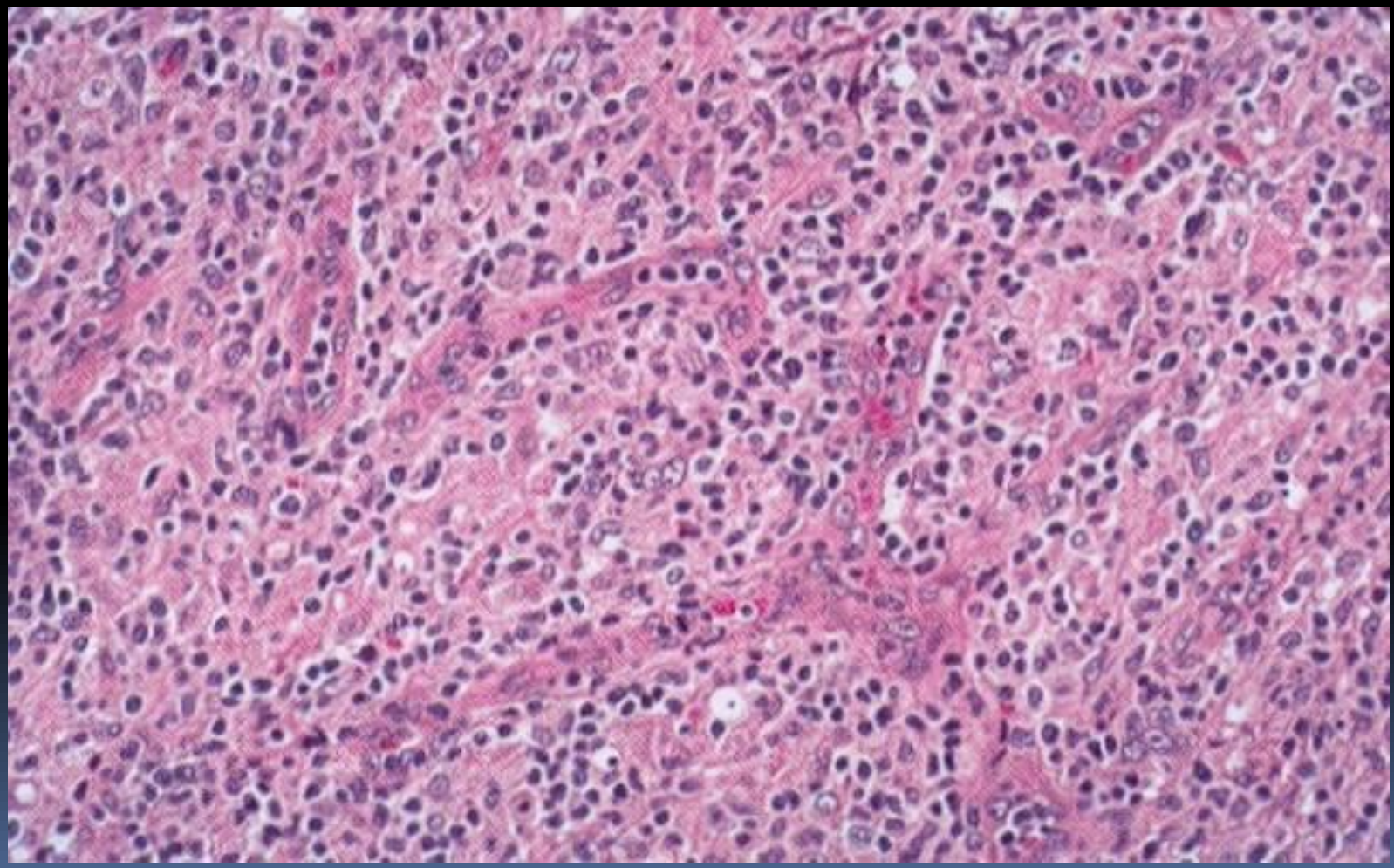
Others: small lymphocytes, plasma cells, eos, histiocytes, follicular dendritic cells, rare HRS-like cells

-Infiltrate bridges capsule; distended cortical sinus; vascular proliferation





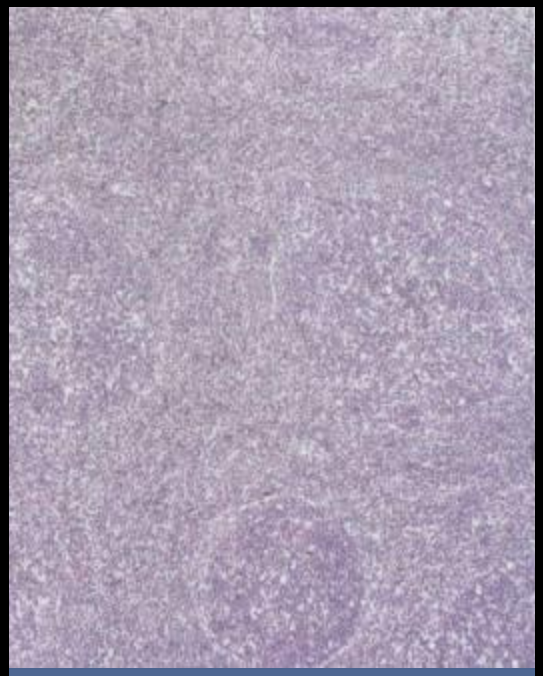
Angioimmunoblastic T-Cell Lymphoma



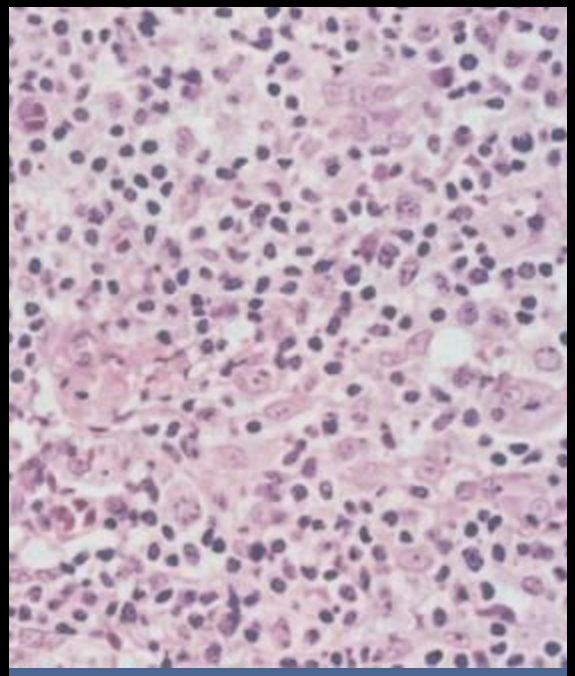
Histopathology(continued)

A-Hyperplastic germinal center
B-Large basophilic B blasts
C-RS-like cells

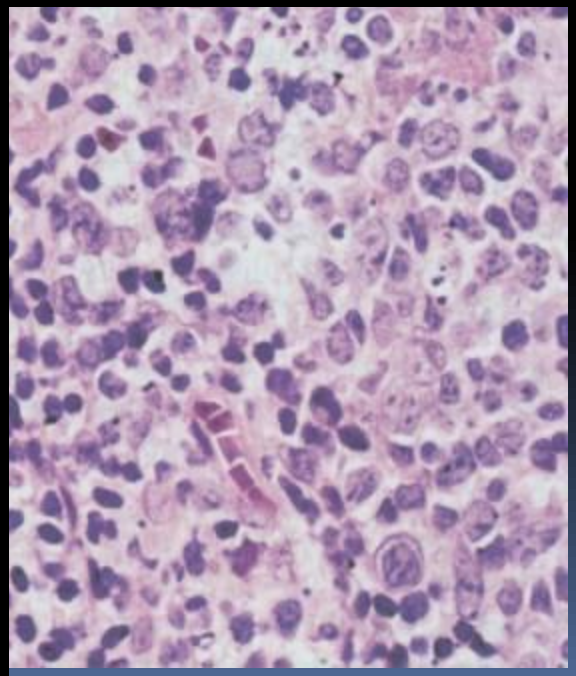
A



B

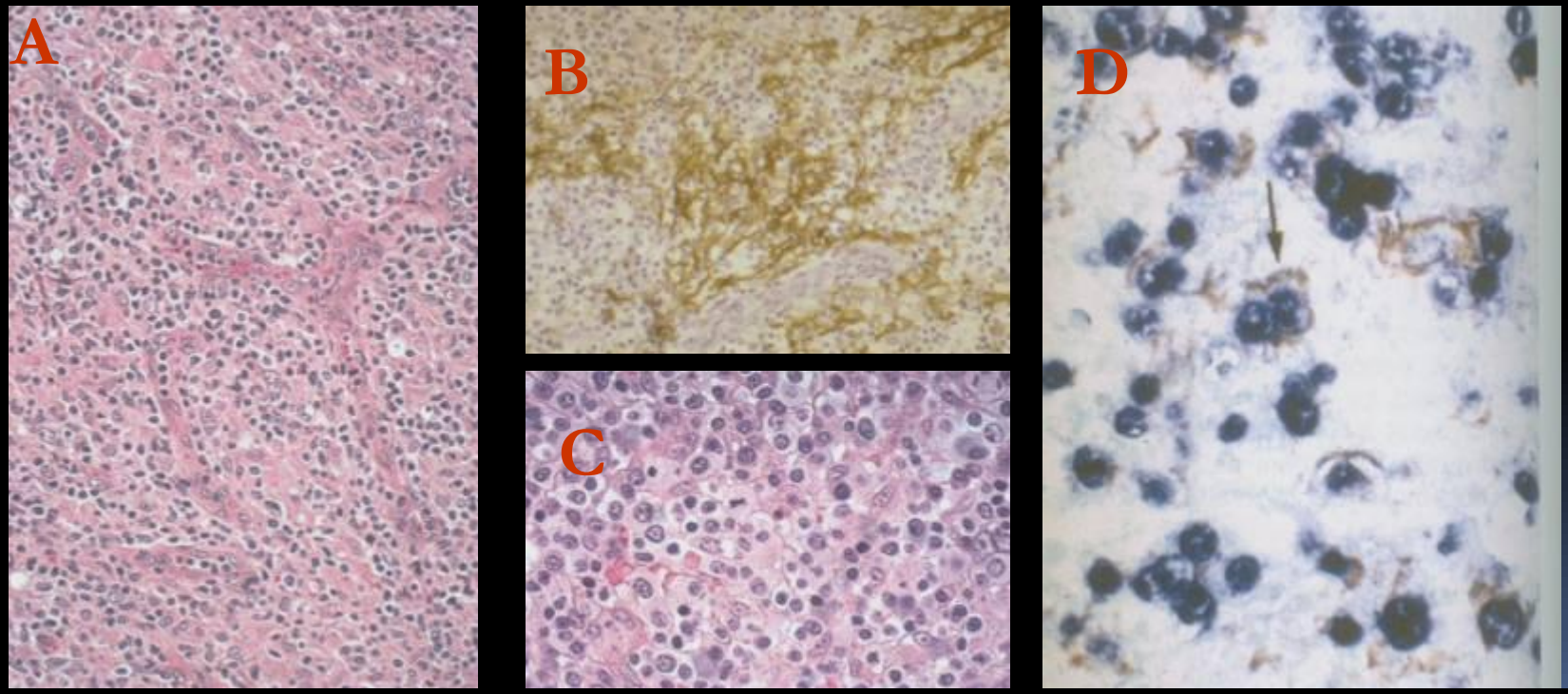


C



Histopathology(continued)

-A: Arborizing blood vessels
-B: Dentric cells abut and extend from venues(CD21)
-C: medium-sized lymphoytes with clear cytoplasm/distinct membrane
-D: Double staining: EBER/CD20



Grading

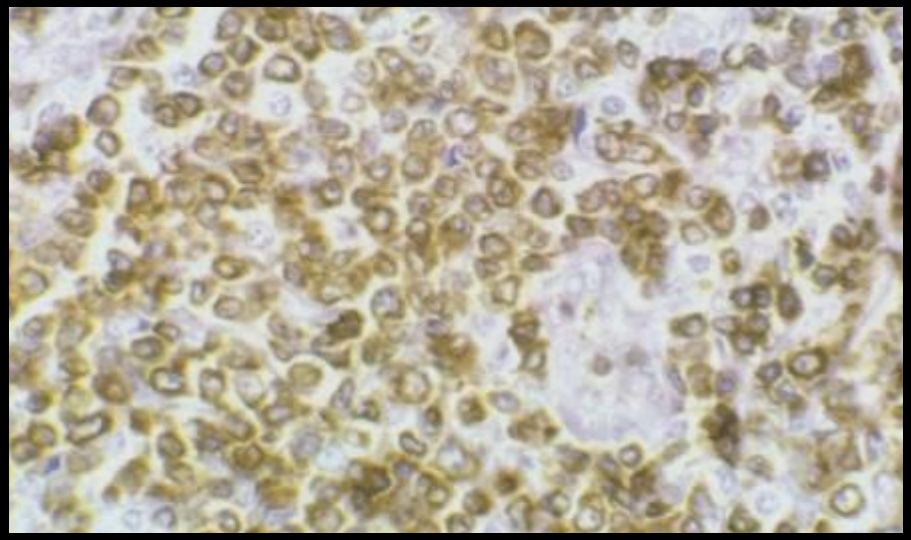
Generally not performed

Immunophenotype

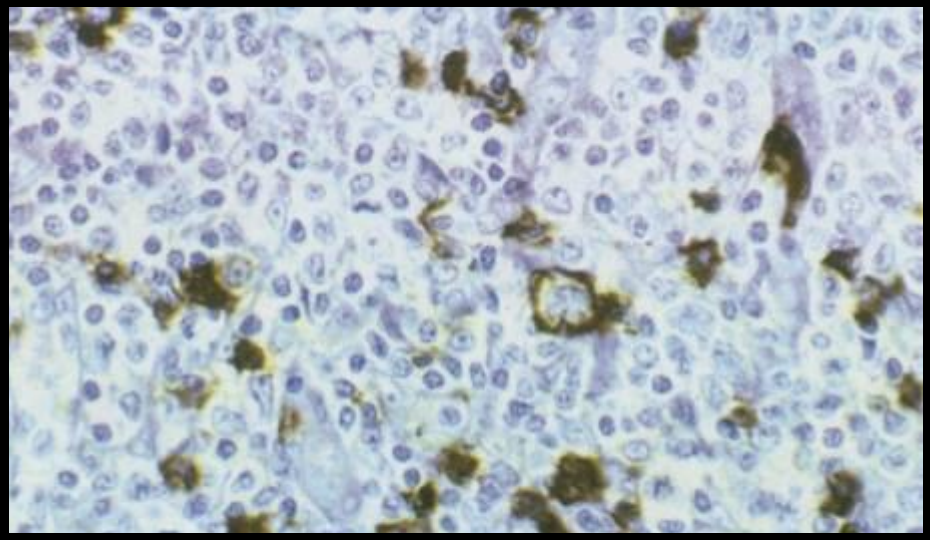
-Positive for CD4 (predominant), CD3, CD2, CD5
-Admixed with many reactive CD8+ cells
-Also positive for TFH cell markers: CD10, CXCL13, PD-1 (60-100% of cases)
-Polyclonal plasma cells
-CD21, CD21: conspicuous dendritic cells

Possible normal counterpart:
-CD4+ Follicular helper T cells (TFH) that are activated by EBV-pos B cells

Histopathology (continued)



CD3



CD20

Genetics

TCR gene rearrangement: 75-90%

Ig gene rearrangement: 30% (in expanded EBV-pos B cells)

Cytogenetics: trisomy 3, trisomy 5, additional X

Prognosis and predictive factors

-Aggressive
-Median survival: < 3 years
-Often succumb to infections
-Some patients may develop secondary EBV-pos DLBCL