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
  Lymphoma 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
Histopathology (continued)

-A: Arborizing blood vessels
-B: Dendritic cells abut and extend from venues (CD21)
-C: medium-sized lymphocytes 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)
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