Enteropathy-Type T-Cell Lymphoma
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Definition
-Intraepithelial T lymphocytes
-Varying degrees of transformation
-Usually presenting as a tumor with large lymphoid cells

Synonyms
Intestinal T-cell lymphoma (REAL)
Enteropathy-Type T-Cell Lymphoma

Epidemiology
Uncommon
Increased in areas (Scandinavia) with high prevalence of celiac disease (5-10%)
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Sites of involvement
Jejunum, ileum (most common)
Others: duodenum, stomach, colon or outside GI
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Clinical features
Hx of celiac disease, childhood or adult onset (most), Abdominal pain, intestine perforation
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Etiology and precursor lesions

-Possible EBV virus: S. and C. America
  (extranodal NK/T-cell lymphoma-nasal type, associated with EBV infection and commonly involves the gastrointestinal tract, must be considered in the differential diagnosis)

-Celiac disease: clear association, prodromal period of refractory
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Celiac disease
Gluten sensitive enteropathy
Refractory sprue
-Malabsorption due to sensitivity to food with Gluten (Gliadin)
-Association with HLA-DQA1 and DQB1
-Adenovirus infection
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HLA heterodimers on enterocytes bind Gliadin and present it to antigen-presenting cells, causing cytotoxic injury.
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Macroscopy

Multiple ulcerating raised mucosal masses
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**Histopathology**
- Monomorphic, medium to large cells
- Round or angulated nuclei
- Abundant pale cytoplasm
- Occasionally anaplastic
- Infiltration by histiocytes, eosinophils
- Epithelial infiltrate in many cases
- A subset: small monomorphic lymphocytes with scanty cytoplasm, dark nuclei.
- Villous atrophy, crypt hyperplasia, epithelial lymphocytosis
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Deeply-infiltrating tumor

Anaplastic variant

Increased eos
Intestinal/Enteropathy T-cell Lymphoma
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**Immunophenotype**
- Positive: CD3, 7, 30, 56, 103
  - Cytotoxic granules: granzyme B, TIA
- Negative: CD4, 5
- CD8-/+,
  - Subset of small to medium sized cells, CD8/56+
  - Adjacent mucosa: T cell are CD8-
- Refractory celiac disease: CD8-
- CD103: integrin αE, intestinal T cell marker
Intestinal/Enteropathy T-cell Lymphoma
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Subset of cases with small to medium-sized cells is CD8+ and CD56+
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- Intraepithelial invasion
- Monomorphic variant

CD3(+), CD8(-)  
PCR shows T cell clones
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Genetics
Genotype (most): HLA DQA1*0501, DQB1*0201
TCR clonally rearranged

Postulated cell of origin
Intraepithelial T cell of the intestine
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Prognosis
- Poor
- Abdominal complications
- Recurrences: most frequently in small intestine