



# **Enteropathy-Type T-Cell Lymphoma**

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# Enteropathy-Type T-Cell Lymphoma

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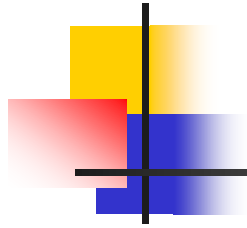
## Definition

- Intraepithelial T lymphocytes
- Varying degrees of transformation
- Usually presenting as a tumor with large lymphoid cells

## Synonyms

Intestinal T-cell lymphoma (REAL)

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## 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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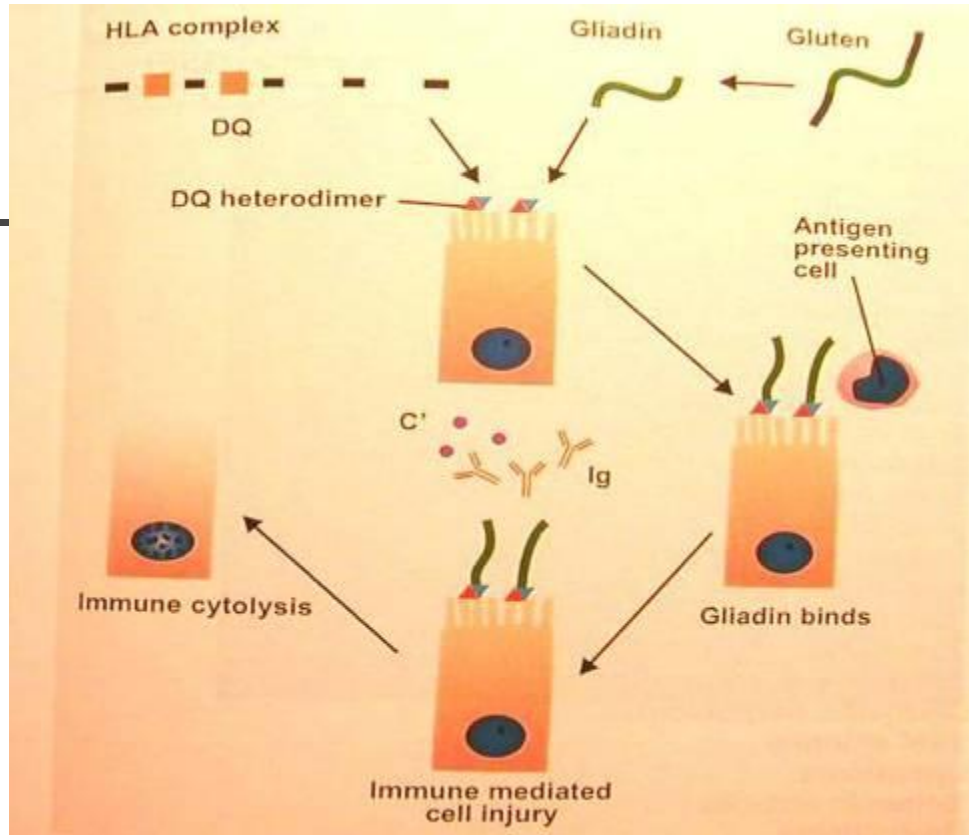
## Celiac disease

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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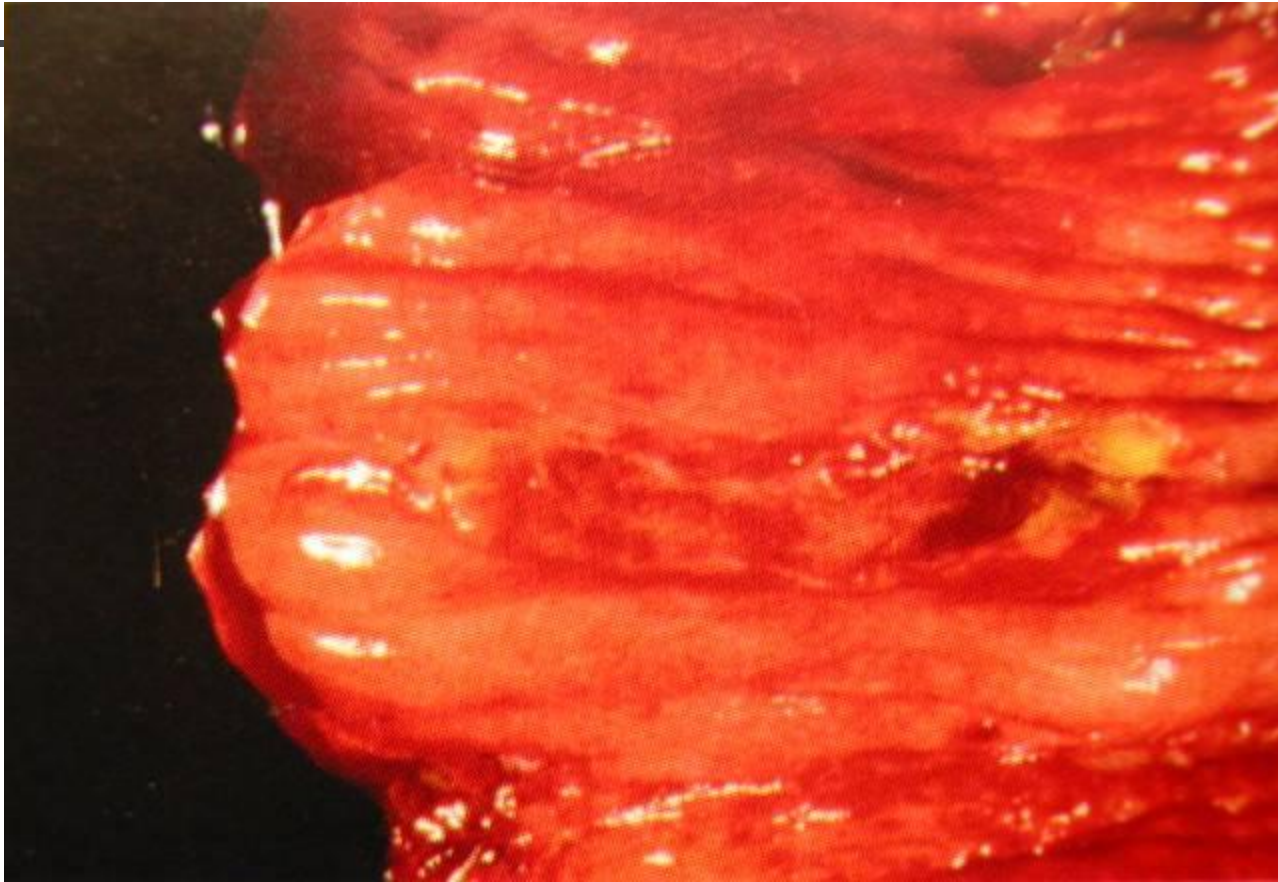


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## Macroscopy

Multiple ulcerating raised mucosal masses

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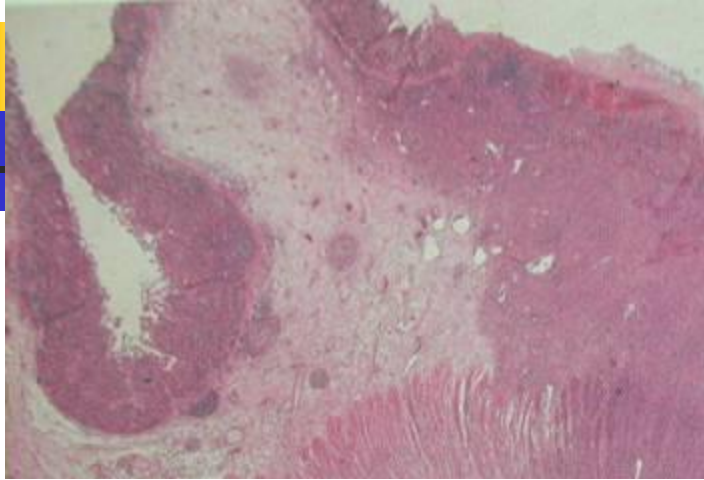


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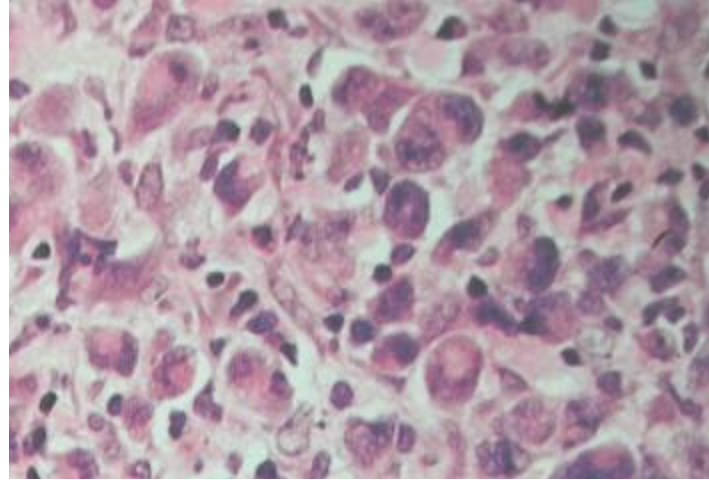
## Histopathology

- 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

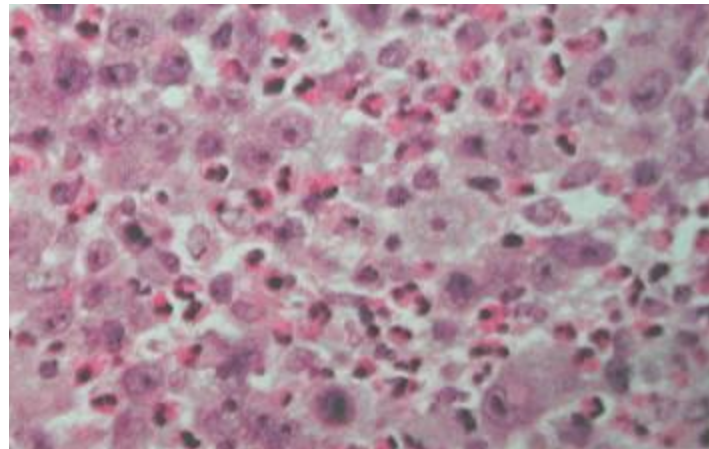
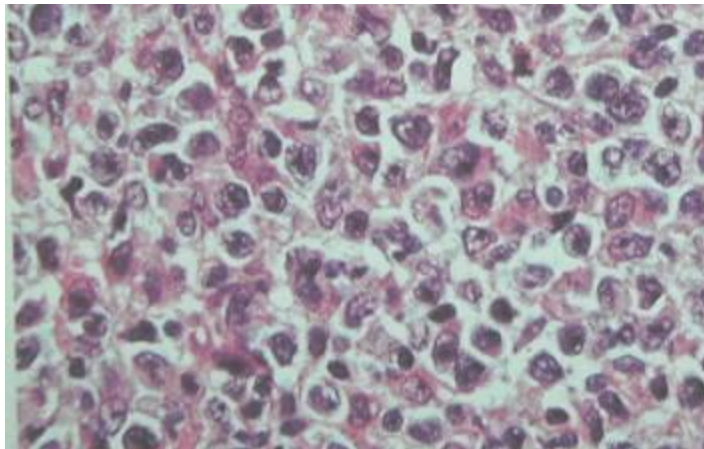
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Deeply-infiltrating tumor

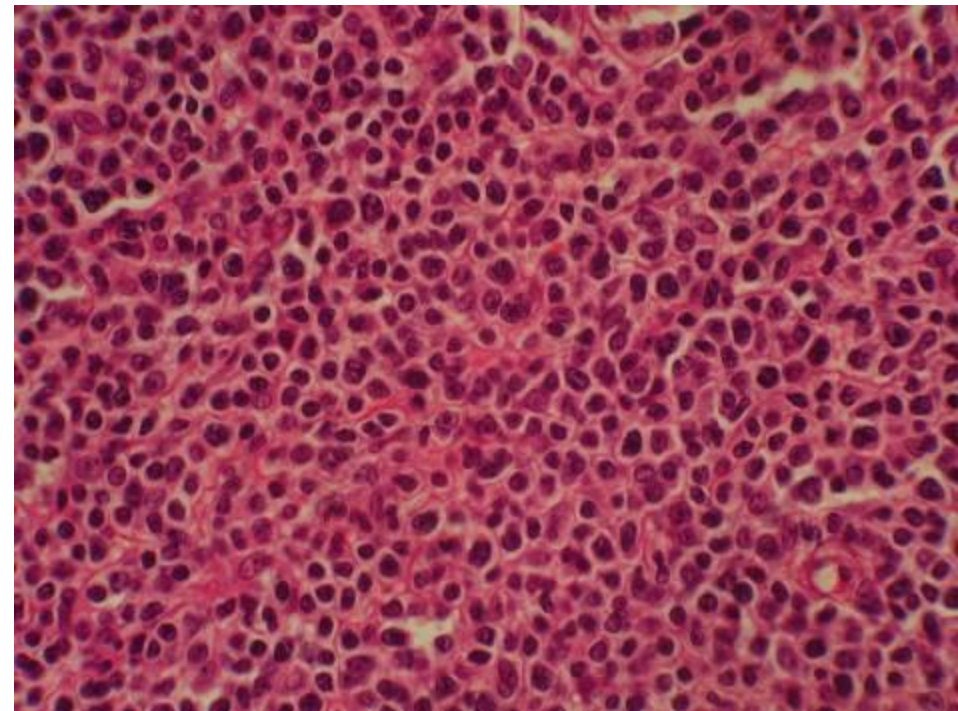
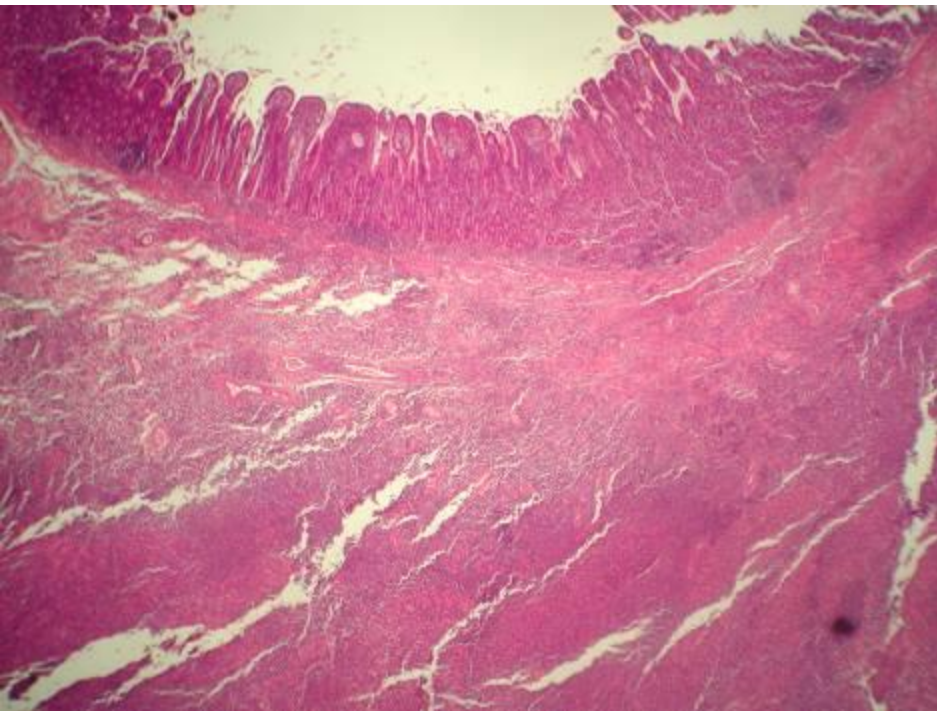


Anaplastic variant

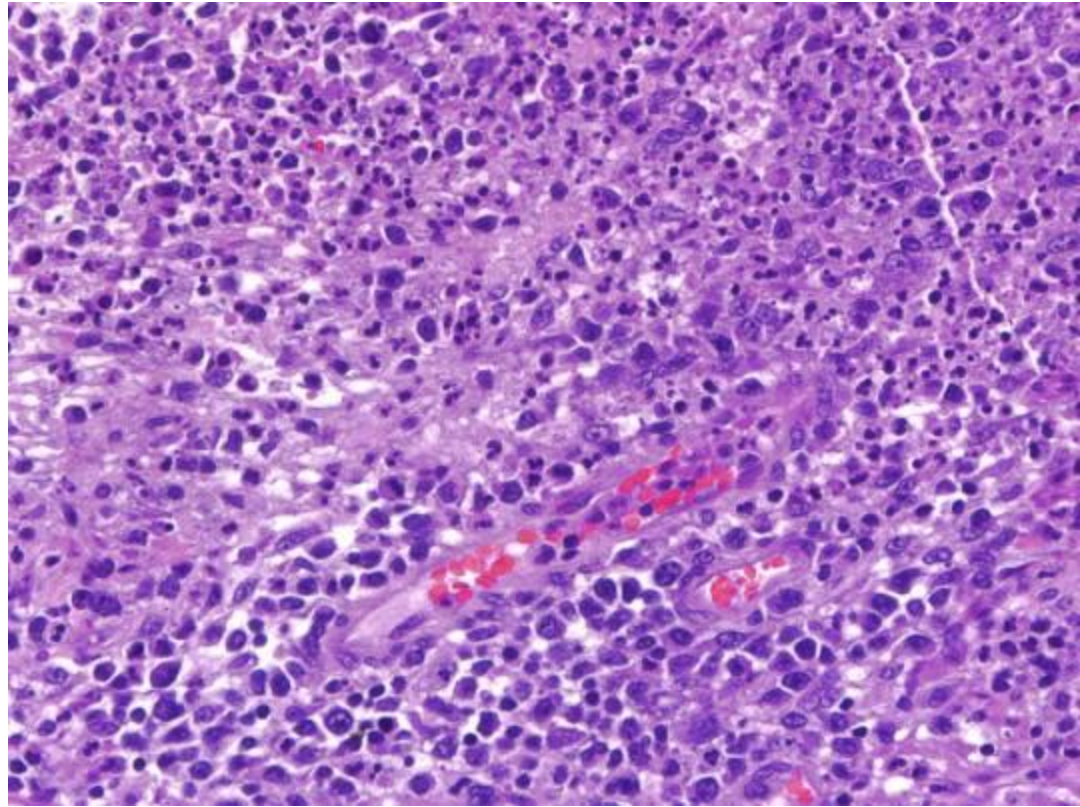
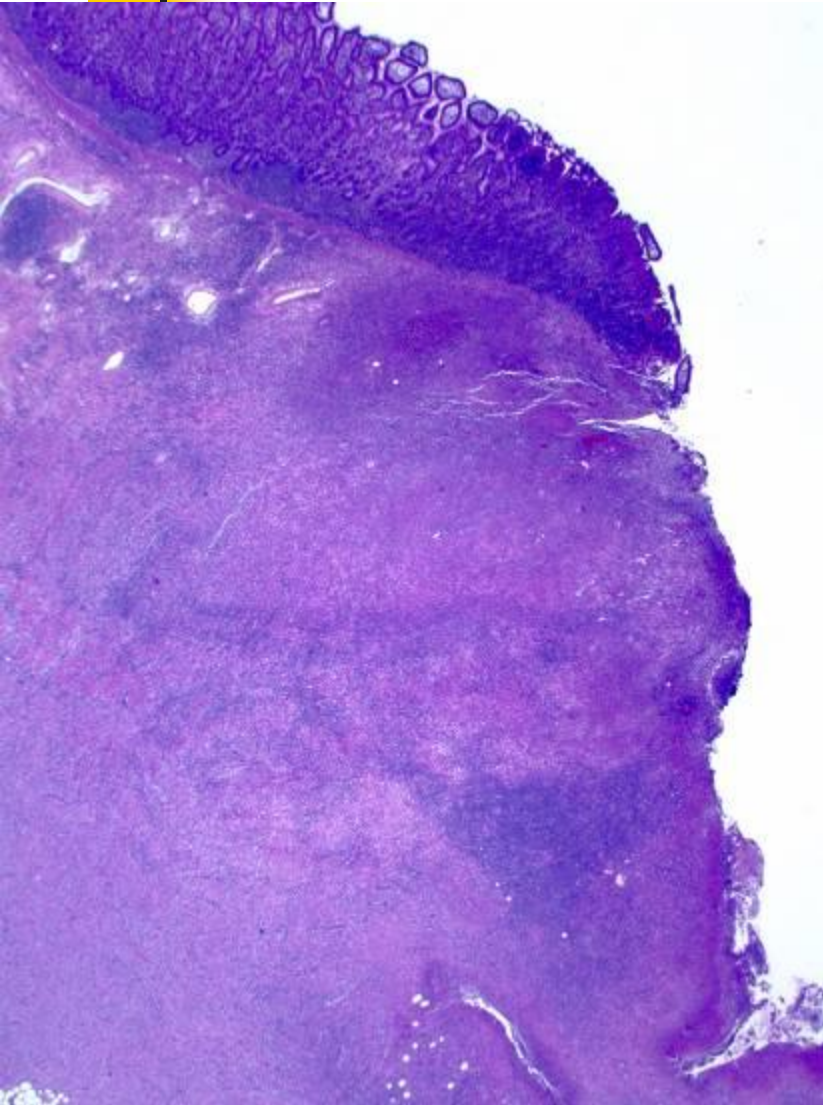


Increased eos

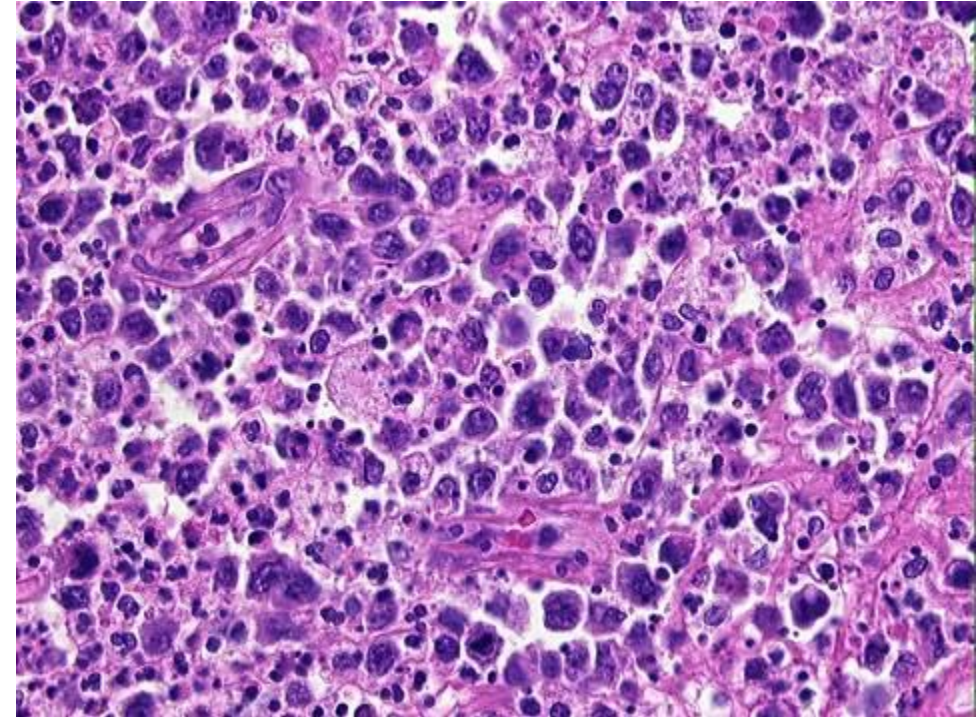
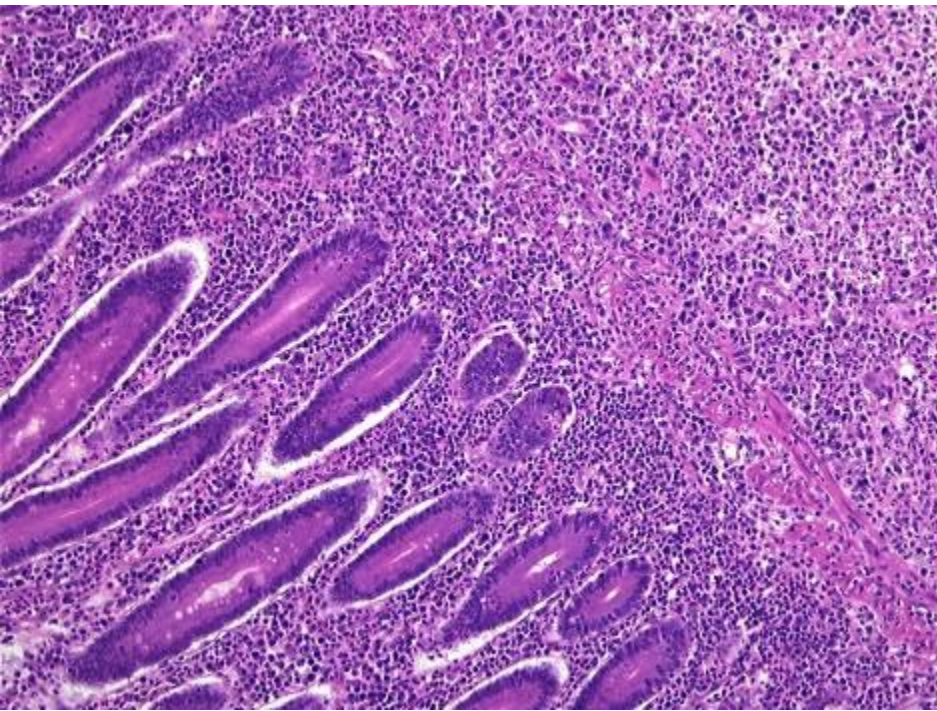
# Intestinal/Enteropathy T-cell Lymphoma



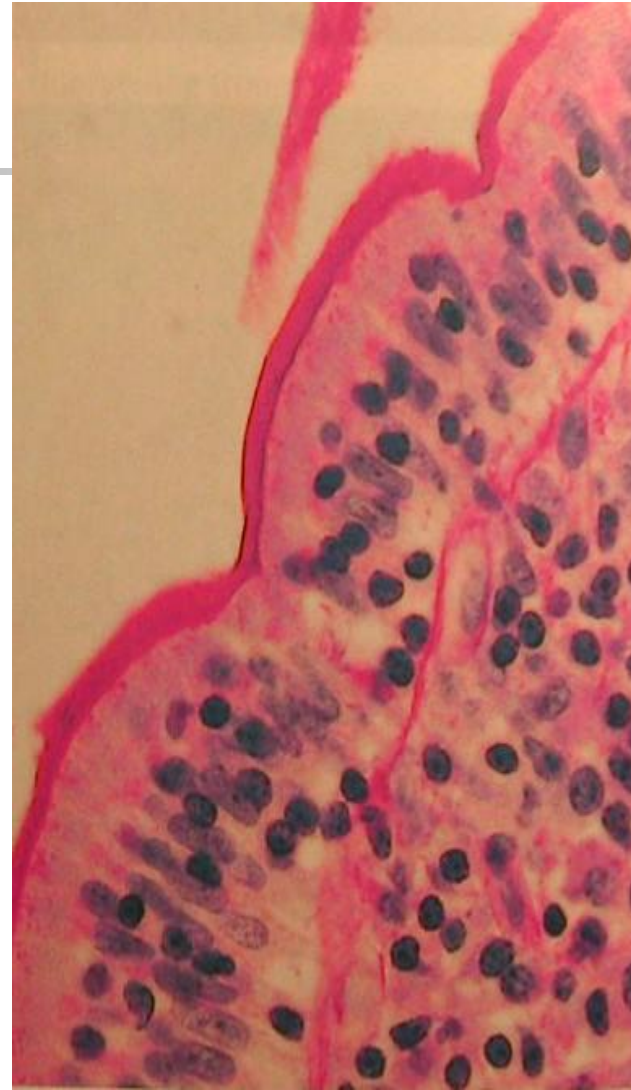
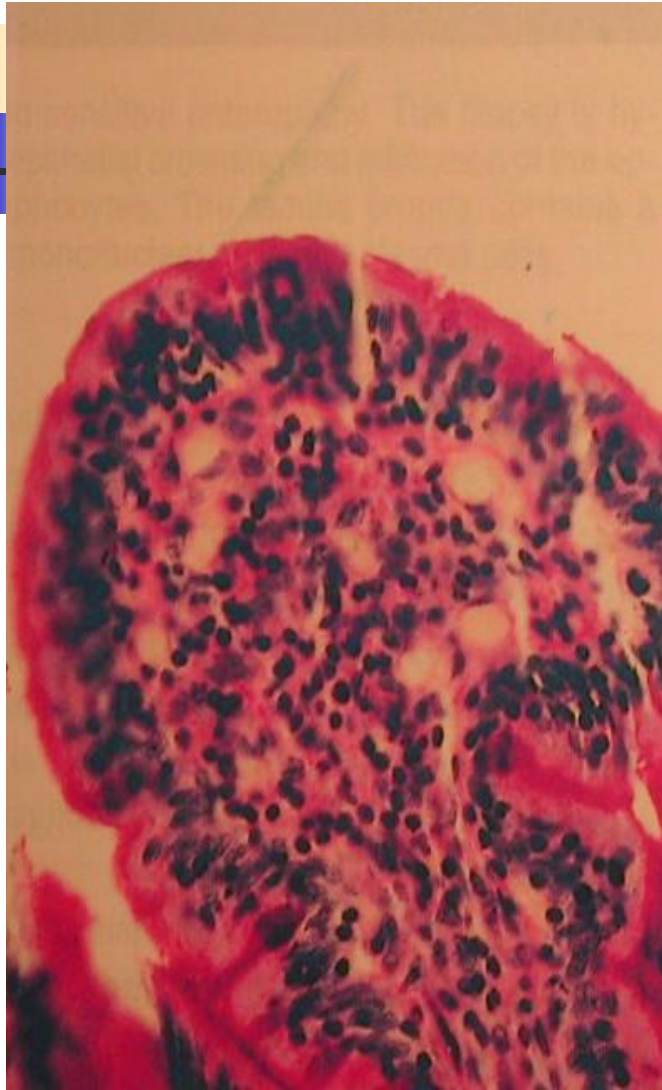
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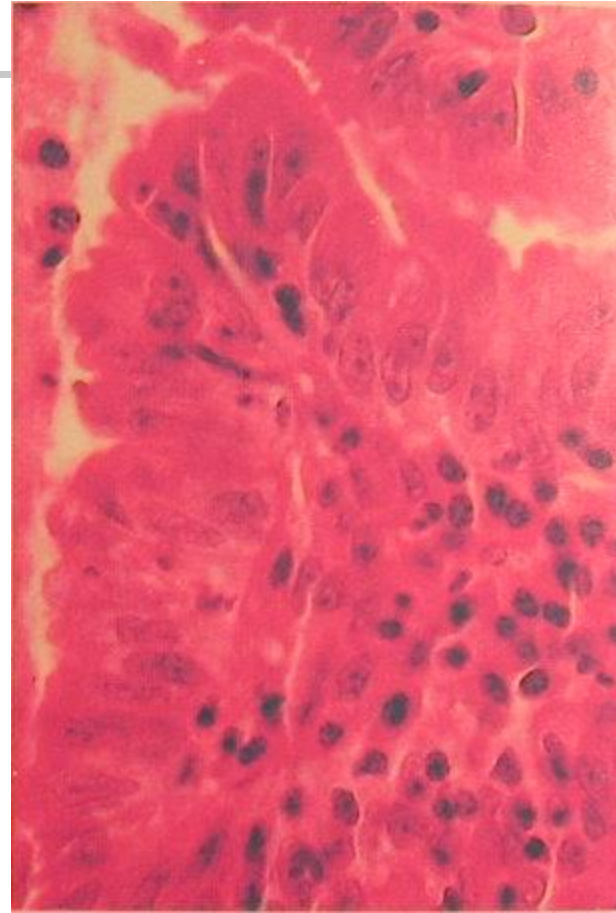
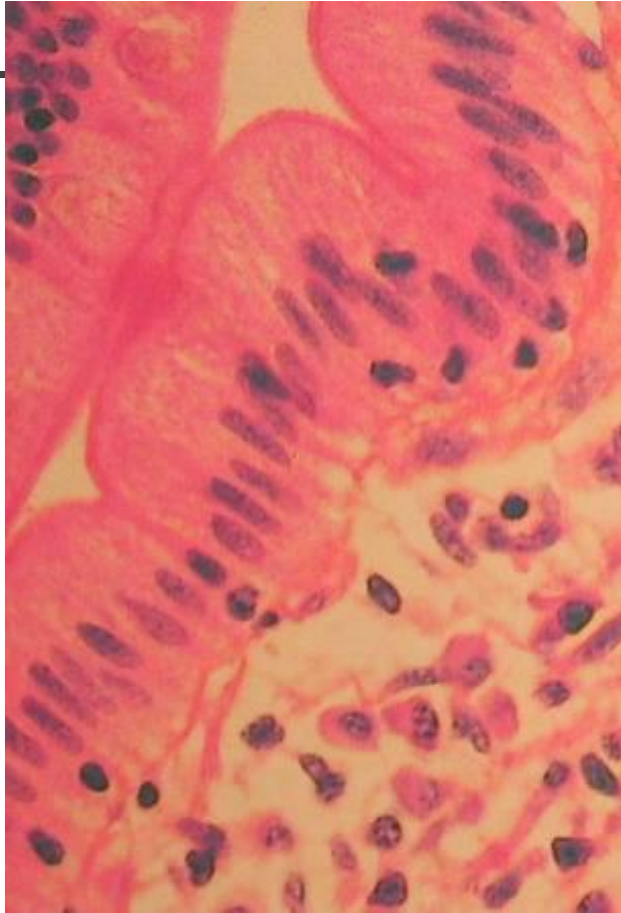


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## Immunophenotype

-Positive: CD3,7,30,56,103

cytotoxic granules: granzyme B, TIA

-Negative: CD4,5

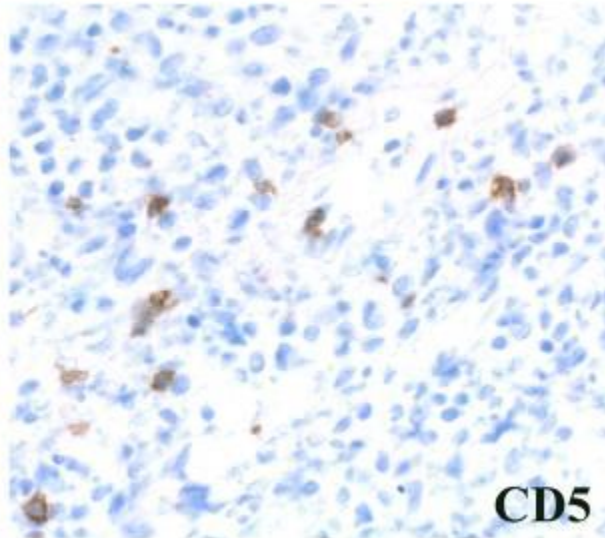
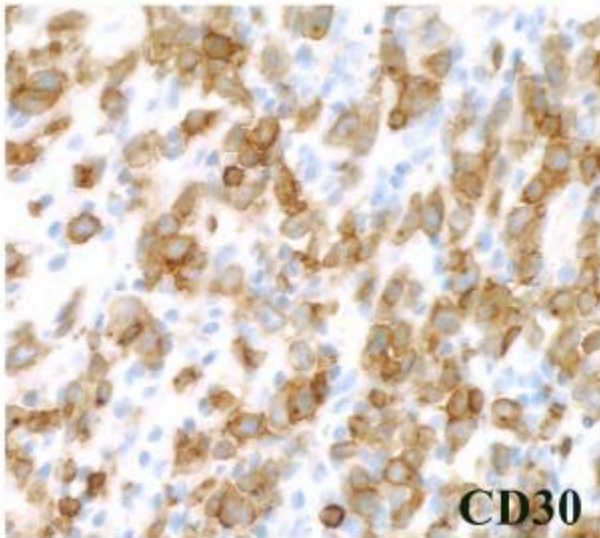
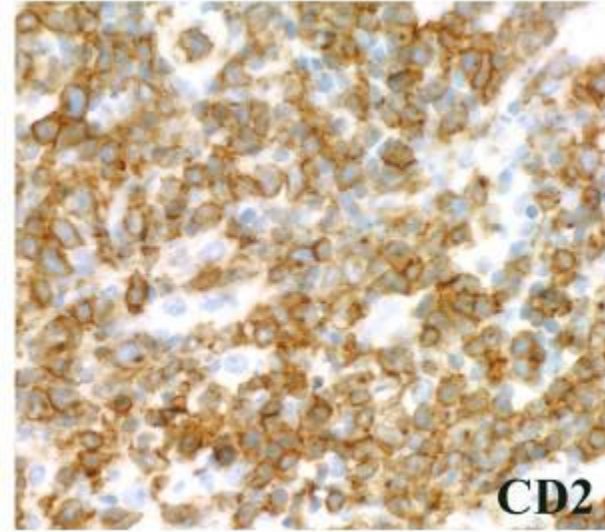
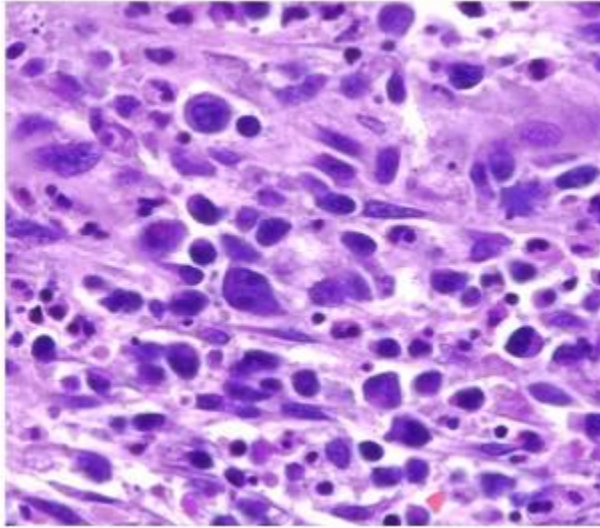
-CD8-/+, subset of small to medium sized cells, CD8/56+

Adjacent mucosa: T cell are CD8-

Refractory celiac disease: CD8-

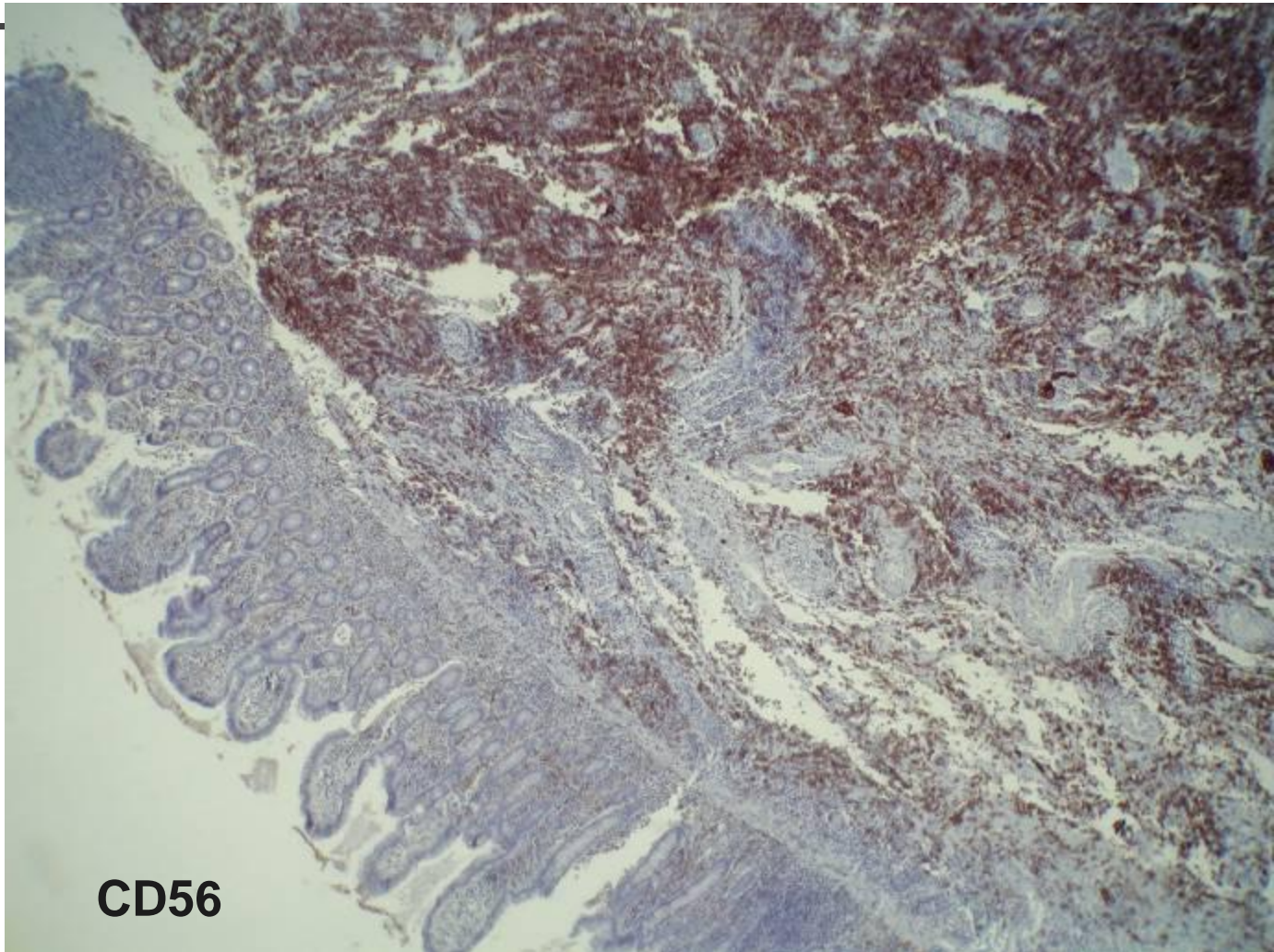
-CD103: integrin  $\alpha E$ , intestinal T cell marker

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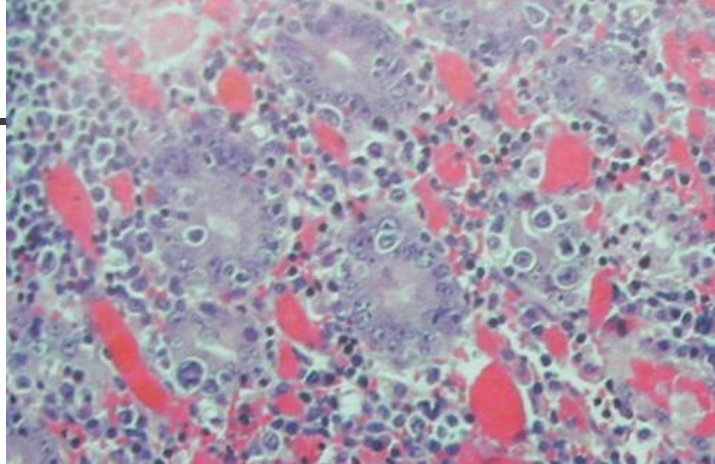
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Subset of cases with small to medium-sized cells is CD8+ and CD56+

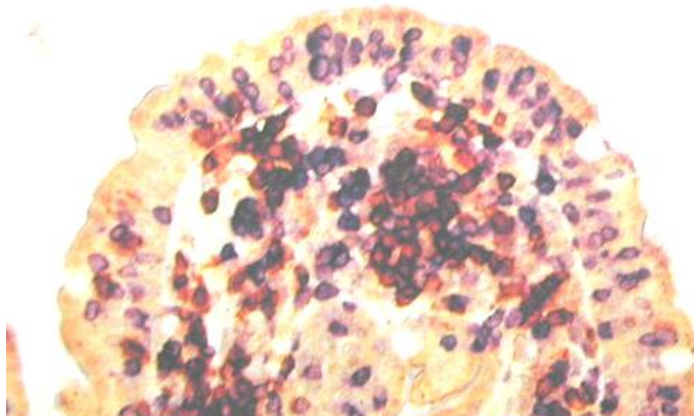
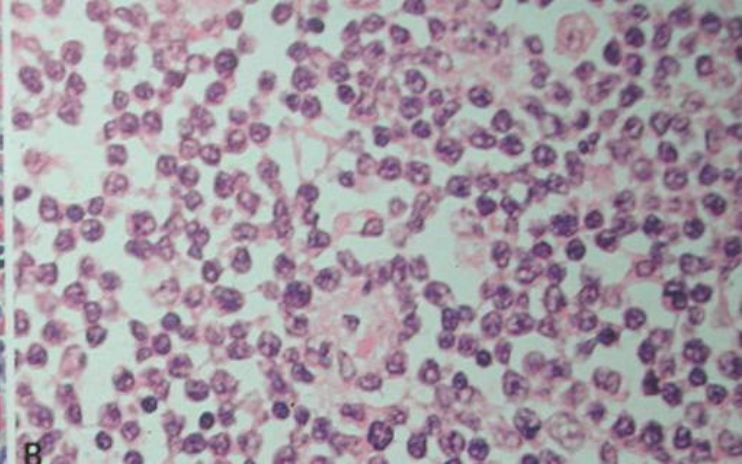


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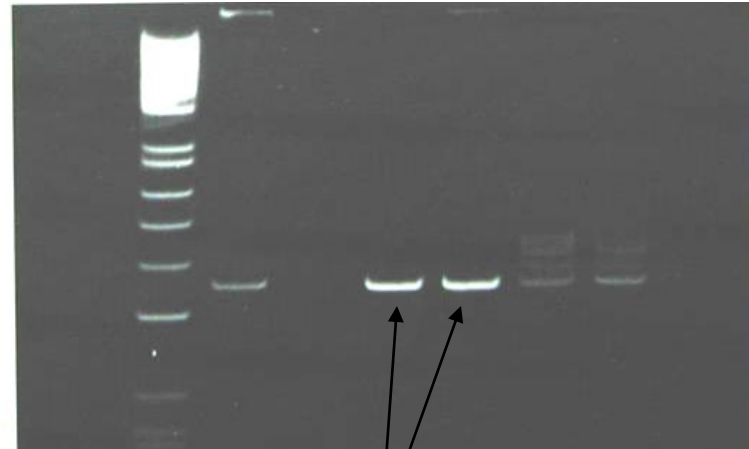
Intraepithelial invasion



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