GI Lymphomas

성균관대학교 의과대학 내과 이준행
모든 사람은 죽는다.
모든 사람은 세금을 내야 한다.
림프종 분류는 항상 변한다.
Biologically rational classification
- morphology
- immunophenotype
- genetic features
- clinical features

Clinically useful classification
- clinical features
- natural history
- prognosis
- treatment
Immunohistochemical markers

- B-cell: **CD20**, CD79a
- T-cell: **CD3**, CD45RO
- NK-cell: **CD56**
- Ki-67: Burkitt lymphoma > DLBL > others
- Mantle cell lymphoma: **cyclin D1**
- HTLV-1 serology
- TdT, CD4, CD5, CD8, CD10, CD30, Bcl-2, cytokeratin AE1/AE3, etc
Table 1. WHO 2008: the mature B-cell neoplasms.

<table>
<thead>
<tr>
<th>Neoplasm</th>
</tr>
</thead>
<tbody>
<tr>
<td>Chronic lymphocytic leukemia/small lymphocytic lymphoma</td>
</tr>
<tr>
<td>B-cell prolymphocytic leukemia</td>
</tr>
<tr>
<td>Splenic marginal zone lymphoma</td>
</tr>
<tr>
<td>Hairy cell leukemia</td>
</tr>
<tr>
<td>Splenic lymphoma/leukemia, unclassifiable</td>
</tr>
<tr>
<td>- Splenic diffuse red pulp small B-cell lymphoma*</td>
</tr>
<tr>
<td>- Hairy cell leukemia-variant*</td>
</tr>
<tr>
<td>Lymphoplasmacytic lymphoma</td>
</tr>
<tr>
<td>Waldenström macrogobulinemia</td>
</tr>
<tr>
<td>Heavy chain diseases</td>
</tr>
<tr>
<td>- Alpha heavy chain disease</td>
</tr>
<tr>
<td>- Gamma heavy chain disease</td>
</tr>
<tr>
<td>- Mu heavy chain disease</td>
</tr>
<tr>
<td>Plasma cell myeloma</td>
</tr>
<tr>
<td>Solitary plasmacytoma of bone</td>
</tr>
<tr>
<td>Extramedullary plasmacytoma</td>
</tr>
<tr>
<td>Extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT lymphoma)</td>
</tr>
<tr>
<td>Nodal marginal zone B-cell lymphoma (MZL)</td>
</tr>
<tr>
<td>- Pediatric type nodal MZL</td>
</tr>
<tr>
<td>Follicular lymphoma</td>
</tr>
<tr>
<td>- Pediatric type follicular lymphoma</td>
</tr>
<tr>
<td>Primary cutaneous follicle center lymphoma</td>
</tr>
<tr>
<td>Mantle cell lymphoma</td>
</tr>
<tr>
<td>Diffuse large B-cell lymphoma (DLBCL), not otherwise specified</td>
</tr>
<tr>
<td>- T cell/histiocyte rich large B-cell lymphoma</td>
</tr>
<tr>
<td>- DLBCL associated with chronic inflammation</td>
</tr>
<tr>
<td>- Epstein-Barr virus (EBV)+ DLBCL of the elderly</td>
</tr>
<tr>
<td>Lymphomatosoid granulomatosis</td>
</tr>
<tr>
<td>Primary mediastinal (thymic) large B-cell lymphoma</td>
</tr>
<tr>
<td>Intravascular large B-cell lymphoma</td>
</tr>
<tr>
<td>Primary cutaneous DLBCL, leg type</td>
</tr>
<tr>
<td>ALK+ large B-cell lymphoma</td>
</tr>
<tr>
<td>Plasmablastic lymphoma</td>
</tr>
<tr>
<td>Primary effusion lymphoma</td>
</tr>
<tr>
<td>Large B-cell lymphoma arising in HHV8-associated multicentric Castleman disease</td>
</tr>
<tr>
<td>Burkitt lymphoma</td>
</tr>
<tr>
<td>B-cell lymphoma, unclassifiable, with features intermediate between diffuse large B-cell lymphoma and Burkitt lymphoma</td>
</tr>
</tbody>
</table>

Table 2. WHO 2008: the mature T-cell and NK-cell neoplasms.

<table>
<thead>
<tr>
<th>Neoplasm</th>
</tr>
</thead>
<tbody>
<tr>
<td>T-cell prolymphocytic leukemia</td>
</tr>
<tr>
<td>T-cell large granular lymphocytic leukemia</td>
</tr>
<tr>
<td>Chronic lymphoproliferative disorder of NK-cells*</td>
</tr>
<tr>
<td>Aggressive NK cell leukemia</td>
</tr>
<tr>
<td>Systemic EBV+ T-cell lymphoproliferative disease of childhood</td>
</tr>
<tr>
<td>(associated with chronic active EBV infection)</td>
</tr>
<tr>
<td>Hydroa vacciniforme-like lymphoma</td>
</tr>
<tr>
<td>Adult T-cell leukemia/ lymphoma</td>
</tr>
<tr>
<td>Extranodal NK/T cell lymphoma, nasal type</td>
</tr>
<tr>
<td>Enteropathy-associated T-cell lymphoma</td>
</tr>
<tr>
<td>Hepatosplenic T-cell lymphoma</td>
</tr>
<tr>
<td>Subcutaneous panniculitis-like T-cell lymphoma</td>
</tr>
<tr>
<td>Mycosis fungoides</td>
</tr>
<tr>
<td>Sézary syndrome</td>
</tr>
<tr>
<td>Primary cutaneous CD30+ T-cell lymphoproliferative disorder</td>
</tr>
<tr>
<td>Lymphomatoid papulosis</td>
</tr>
<tr>
<td>Primary cutaneous anaplastic large-cell lymphoma</td>
</tr>
<tr>
<td>Primary cutaneous aggressive epidermotropic CD8+ cytotoxic T-cell lymphoma*</td>
</tr>
<tr>
<td>Primary cutaneous gamma-delta T-cell lymphoma</td>
</tr>
<tr>
<td>Primary cutaneous small/medium CD4+ T-cell lymphoma*</td>
</tr>
<tr>
<td>Peripheral T-cell lymphoma, not otherwise specified</td>
</tr>
<tr>
<td>Angioimmunoblastic T-cell lymphoma</td>
</tr>
<tr>
<td>Anaplastic large cell lymphoma (ALCL), ALK+</td>
</tr>
<tr>
<td>Anaplastic large cell lymphoma (ALCL), ALK-</td>
</tr>
</tbody>
</table>
**B-cell neoplasms of GI tract**

- MALT lymphoma
- Diffuse large B-cell lymphoma (DLBCL)
- Follicular lymphoma
- Mantle cell lymphoma
- Burkitt lymphoma
T-cell neoplasms of GI tract

- Adult T-cell leukemia/lymphoma (HTLV-1+)
- Enteropathy-type intestinal T-cell lymphoma
- Anaplastic large cell lymphoma, T- or null cell type
- NK/T-cell (angiocentric) lymphoma
- Peripheral T-cell lymphoma, unspecified
위장관 림프종의 분포

위장관

55-65%

20-35%

7-20%
Non-Hodgkin’s lymphoma of the GI tract
- Danish Lymphoma Study Group

Fig 1. Mean annual age-specific IRs of gastric (□) and intestinal (○) NHL in western Denmark.
최근에는 건진에서 발견되는 예가 많다.
- 서울대 강남센터 (Yang HJ. JGH 2016.)

이준행 해설: 10만 5천여명의 검진 내시경 중 429명(0.41%)에서 위암이 발견되었습니. 250명 검진내시경에서 한 명의 위암이 발견된 셈이니 우리나라 대형병원의 평균적인 수치였습니다. 그 중 12.1%(52/429)가 위림프종이었으며 DLBCL (diffuse large B cell lymphoma) 한 명 빼고 모두 저도 MALT 림프종이었습니다.
내시경 진단이 항상 쉬운 것은 아니다.

성균관대학교 의과대학 내과 이준행
궤양형 병소의 감별진단은 늘 어렵습니다.
일반적으로 림프종은 잘 퍼집니다.
- DLBCL
- This stomach with DLBCL did not expand well by air infusion.
대부분은 공기 확장을 볼 필요도 없습니다.
- 모두 DLBCL
보만 4형 진행성 위암과 유사한 DLBCL

사진 제공: 부산대학교 김광하 교수님
Follicular lymphoma
임신 16주. Diffuse large B cell lymphoma → Therapeutic abortion 후 항암치료

EGD after referral
- DLBCL. Rapid progression during 3 months
AGC + follicular lymphoma

AGC B-II, tubular adenocarcinoma, M/D, 3.1x2.9 cm, subserosa (pT2b), LN + (2/49)
Follicular lymphoma, follicular, grade I, involving all regional lymph nodes
림프종 진단에 대한 개인적 의견

1. 위장관 림프종은 점막하병소처럼 보이는 경우도 있으나 점막병소인 경우가 더 많습니다.

2. 아래와 같은 경우에는 림프종을 의심하는 것이 좋습니다.
   - 암은 암 같은데 adenocarcinoma의 일반적인 모양이 아닌 경우
   - 종양 크기에 비해 obstruction이 가벼울 때 혹은 잘 퍼질 때
   - 조직검사에서 "dense lymphoid infiltration 이나 림프종으로 진단하기 어려다 (not sufficient for the diagnosis of lymphoma)"와 같은 결과일 때
   - 짧은 기간에 병소가 많이 커졌을 때
   - 다양하고 bizzare한 모양의 병소가 여러개 있을 때
   - IBD나 기타 염증성 질환 환자가 일반적인 치료에도 호전이 없을 때
병리 검사 해석에 주의하자.

성균관대학교 의과대학 내과 이준행
Make friends with a pathologist

• There is lymphoid hyperplasia with mild to moderate cellular atypism, but these findings are not sufficient for the pathologic diagnosis of GI lymphoma.
DLBCL
- 암 의심이었는데 조직검사에서 암으로 나오지 않아서 의뢰됨
진단이 지연될 수 있다.

성균관대학교 의과대학 내과 이준행
2014. 3. (외부)
Bx: gastritis

2014. 3. (전원 후 재검)
Bx: gastritis with ulcer

2014. 5. (2개월 후)
Bx: gastritis with lymphoid follicles

Bx #3: diffuse large B cell lymphoma
MALT 림프종 환자가 급작스런 복통
→ ileoceleal intussusception was diagnosed
Primary Colon Lymphoma in Korea: A KASID (Korean Association for the Study of Intestinal Diseases) Study

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H. S. KIM, MD,** S. J. MYUNG, MD,† W. H. KIM, MD,** J. C. RHEE, MD,* K. Y. CHOI, MD,** I. S. SONG, MD,**
J. H. HYUN, MD,**§§ and Y. I. MIN, MD†

Although almost all primary colorectal lymphomas are of B-cell lineage in Western countries, primary colorectal T-cell lymphomas are not uncommon in the East. The aim of this study was to review the clinical characteristics and treatment outcomes of primary colorectal lymphomas, with special emphasis on the differences between T-cell and B-cell lymphomas. Ninety-five cases of primary colorectal lymphomas that satisfied Dawson’s criteria were identified from the clinical databases of 13 university hospitals in Korea. The mean age at the time of presentation was 51.1 years and the male:female ratio was 64:31. The clinical information, including endoscopic and histological characteristics, was retrospectively analyzed. Of the primary colorectal lymphomas, 78 cases (82.1%) were of B-lineage and 17 cases (17.9%) were of T-cell lineage. Patients with T-cell lymphomas presented at a younger age than patients with B-cell lymphomas (42.8 vs 52.9 years, respectively; P =
## Endoscopic findings

<table>
<thead>
<tr>
<th>Classification</th>
<th>B cell (n=63)</th>
<th>T cell (n=15)</th>
<th>All (n=78)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Fungating</td>
<td>34 (54.0%)</td>
<td>2 (13.3%)</td>
<td>36 (46.2%)</td>
</tr>
<tr>
<td>Ulcerative</td>
<td>3 (4.8%)</td>
<td>7 (46.7%)</td>
<td>10 (12.8%)</td>
</tr>
<tr>
<td>Infiltrative</td>
<td>5 (7.9%)</td>
<td>0</td>
<td>5 (6.4%)</td>
</tr>
<tr>
<td>Ulcerofungating</td>
<td>17 (27.0%)</td>
<td>1 (6.7%)</td>
<td>18 (23.1%)</td>
</tr>
<tr>
<td>Ulceroinfiltrative</td>
<td>4 (6.3%)</td>
<td>5 (33.3%)</td>
<td>9 (11.5%)</td>
</tr>
</tbody>
</table>

* P < 0.001

## Delayed diagnosis of colon lymphoma was seen in T cell lymphomas (n=6, 35.3%)

<table>
<thead>
<tr>
<th>Age/Sex</th>
<th>Symptoms</th>
<th>Initial impression</th>
<th>Initial management</th>
<th>Time to final diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td>24/M</td>
<td>Abdominal pain</td>
<td>UC with perforation</td>
<td>Persistent Sx after operation</td>
<td>3 months</td>
</tr>
<tr>
<td>66/F</td>
<td>Frequent loose stool</td>
<td>UC</td>
<td>Persistent Sx after steroid</td>
<td>13 months</td>
</tr>
<tr>
<td>45/M</td>
<td>Hematochezia</td>
<td>Intestinal tuberculosis</td>
<td>Weight gain after anti-tbc</td>
<td>12 months</td>
</tr>
<tr>
<td>67/M</td>
<td>Frequent loose stool</td>
<td>Intestinal tuberculosis</td>
<td>Weight gain after anti-tbc</td>
<td>6 months</td>
</tr>
<tr>
<td>33/M</td>
<td>Abdominal pain</td>
<td>Crohn’s Disease</td>
<td>Medication</td>
<td>15 months</td>
</tr>
<tr>
<td>30/M</td>
<td>Diarrhea</td>
<td>r/o amebiasis</td>
<td>Medication</td>
<td>6 months</td>
</tr>
</tbody>
</table>

Chronic diarrhea, loss of weight (15 kg) and night sweat (F/43)

Final diagnosis: peripheral T cell lymphoma
다양한 위장관 림프종 진단별 접근법

성균관대학교 의과대학 내과 이준행
림프종은 종류가 많아서...

• B 세포인지 T 세포인지 구분한다.
  → T 세포면 대부분 항암치료

• B 세포 중 MALT 림프종인지 아닌지 구분하다.
  → MALT 림프종이면 대부분 Helicobacter 제균 치료

• 다른 것들은 각자 개별 진단에 따라 치료한다.
stem cell

lymphoid progenitor

progenitor-B

ALL

pre-B

immature B-cell

cLL

mature naive B-cell

germinal center B-cell

D LBCL, FL, HL

memory B-cell

plasma cell

Bone marrow

Lymphoid tissue
DLBCL of the stomach
DLBCL of the duodenum
위암, 심이지장 DLBL
DLBCL of the small bowel
DLBCL, colon
Gastroduodenal involvement of diffuse large B cell lymphoma
Lymphoepithelial lesions

Monocytoid B-cells
MALT 림프종의 다양한 내시경 소견

Gastritis-like  Multifocal atrophic  Multinodular
Ulcerative  Polypoid  EGC-like
High grade MALToma ???

- MALT lymphoma without high grade component
- MALT lymphoma with high grade component
- Extranodal marginal zone B cell lymphoma of MALT
- DLBCL with a MALT lymphoma component
- DLBCL without a MALT lymphoma component
## The criteria is analogue.

<table>
<thead>
<tr>
<th>Score</th>
<th>Diagnosis</th>
<th>Histological features</th>
</tr>
</thead>
<tbody>
<tr>
<td>0</td>
<td>Normal</td>
<td>Scattered plasma cells in lamina propria. No lymphoid follicles.</td>
</tr>
<tr>
<td>1</td>
<td>Chronic active gastritis</td>
<td>Small clusters of lymphocytes in lamina propria. No lymphoid follicle. No lymphoepithelial lesions.</td>
</tr>
<tr>
<td>2</td>
<td>Chronic active gastritis with florid lymphoid follicle formation</td>
<td>Prominent lymphoid follicles with surrounding mantle zone and plasma cells. No lymphoepithelial lesions.</td>
</tr>
<tr>
<td>3</td>
<td>Suspicious lymphoid infiltrate, probably reactive</td>
<td>Lymphoid follicles surrounded by small lymphocytes that infiltrate diffusely in lamina propria and occasionally into epithelium.</td>
</tr>
<tr>
<td>4</td>
<td>Suspicious lymphoid infiltrate, probably lymphoma</td>
<td>Lymphoid follicles surrounded by marginal zone cells that infiltrate diffusely in lamina propria and into epithelium in small groups.</td>
</tr>
<tr>
<td>5</td>
<td><strong>MALT lymphoma</strong></td>
<td>Presence of dense infiltrate of marginal zone cells in lamina propria with prominent lymphoepithelial lesions.</td>
</tr>
</tbody>
</table>
What for Wotherspoon 3 and 4 ???

First routine gastric biopsies

Wotherspoon 1,2
Gastritis

Wotherspoon 3,4
PCR
Clonal
Lymphoma*

Wotherspoon 5
Lymphoma*

Not clonal
Gastritis
Personal protocol for MALToma

- Low-grade MALT lymphoma in the first endoscopic biopsy
- Staging work-up including EUS, CT, BM
- Stage E-I₂, II, III, IV or *H. pylori* (-) or high-grade
- *H. pylori* eradication (PCA 2 weeks) + 2nd eradication, if necessary
- UBT 4-6 wks after completing antibiotic treatment
- Endoscopy, 3 months after completing eradication
Bone marrow involvement
- lymphoid aggregate, CD20(+)

• Bone marrow, left, biopsy:
• Normocellular marrow (cellularity 50 %) with three lineage hematopoiesis and involvement of malignant lymphoma (tumor volume: less than 5 %)
Bone marrow involvement
- lymphoid aggregate
Bone marrow involvement
- lymphoid aggregate, CD20 (+)
Subtle CD20 positivity in the bone marrow of a patient who has a mucosa-associated lymphoid tissue lymphoma should not be regarded as evidence of involvement in the bone marrow.

Figure 2. Progression-free survival according to bone marrow involvement in patients with mucosa-associated lymphoid tissue lymphoma.
Management of MALToma at SNUH

FIGURE 1. Schematic study design for the treatment approach and follow-up of patients with gastric MALT lymphoma. Bx indicates biopsy; CR, complete remission; CTx, chemotherapy; Hp, Helicobacter pylori; MALT, mucosa-associated lymphoid tissue; mo, month; NC, no change; Op, operation; PR, partial response; q, every; RTx, radiation therapy.
2004. 11. 29
하부 영역 CT 재검
chest PA

2004. 11. 29

p) 비정상
chest PA

s-EGD

2007. t=18

saddle area of fundus

white atrophic scar

2004. 1. 03

CT.

10.

2004. 1. 03

2004. 3. 17

CT.

10.

2004. 3. 17

2004. 1. 03

CT.

white atrophy

10.

2004. 1. 03

CT.

white atrophy

10.

2004. 1. 03

CT.

white atrophy

10.

2004. 1. 03

CT.

white atrophy

10.

2004. 1. 03

CT.

white atrophy

10.

2004. 1. 03

CT.

white atrophy

10.

2004. 1. 03

CT.

white atrophy

10.
From HPE to complete remission

Figure 1 Interval to CR after eradication. The median interval was 3 months. Six patients reached CR after 12 months.
Initial EGC  
2 months later  
6 months later
# MALToma at SMC

## Table 1. Baseline Characteristics and Risk Factor Analysis of Failure of MALT-lymphoma Remission

<table>
<thead>
<tr>
<th></th>
<th>Remission (n=88)</th>
<th>Failure (n=7)</th>
<th>p-value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age, mean (range), years</td>
<td>50±12</td>
<td>48±9</td>
<td>0.465</td>
</tr>
<tr>
<td>Sex (M:F) (%)</td>
<td>48:40 (55:45)</td>
<td>3:4 (43:57)</td>
<td>0.083</td>
</tr>
<tr>
<td>Follow up duration, (mean±SD, month)</td>
<td>40±25</td>
<td>35±15</td>
<td>0.513</td>
</tr>
<tr>
<td>Endoscopic location (%)</td>
<td></td>
<td></td>
<td>0.001</td>
</tr>
<tr>
<td>Distal</td>
<td>65 (73.9%)</td>
<td>1 (14.3%)</td>
<td></td>
</tr>
<tr>
<td>Both</td>
<td>14 (15.9%)</td>
<td>2 (28.6%)</td>
<td></td>
</tr>
<tr>
<td>Proximal</td>
<td>9 (10.2%)</td>
<td>4 (57.1%)</td>
<td></td>
</tr>
<tr>
<td>Endoscopic appearance (%)</td>
<td></td>
<td></td>
<td>0.344</td>
</tr>
<tr>
<td>Ulcer</td>
<td>33 (37.5%)</td>
<td>4 (57.1%)</td>
<td></td>
</tr>
<tr>
<td>Erosion</td>
<td>20 (22.7%)</td>
<td>2 (28.6%)</td>
<td></td>
</tr>
<tr>
<td>Erythema</td>
<td>29 (33.0%)</td>
<td>0 (0%)</td>
<td></td>
</tr>
<tr>
<td>Thick folds or tumor</td>
<td>6 (6.8%)</td>
<td>1 (14.3%)</td>
<td></td>
</tr>
<tr>
<td>Presence of large cell component (+) (%)</td>
<td>1 (1.1%)</td>
<td>1 (14.3%)</td>
<td>0.067</td>
</tr>
</tbody>
</table>
Post-therapy grading system

Table 1  GELA histological grading system for post-treatment evaluation of gastric MALT lymphoma

<table>
<thead>
<tr>
<th>Score</th>
<th>Lymphoid infiltrate</th>
<th>LEL</th>
<th>Stromal changes</th>
</tr>
</thead>
<tbody>
<tr>
<td>CR (complete histological remission)</td>
<td>Absent or scattered plasma cells and small lymphoid cells in the LP</td>
<td>Absent</td>
<td>Normal or empty LP and/or fibrosis</td>
</tr>
<tr>
<td>pMRD (probable minimal residual disease)</td>
<td>Aggregates of lymphoid cells or lymphoid nodules in the LP/MM and/or SM</td>
<td>Absent</td>
<td>Empty LP and/or fibrosis</td>
</tr>
<tr>
<td>rRD (responding residual disease)</td>
<td>Dense, diffuse, or nodular extending around glands in the LP</td>
<td>Focal LEL or absent</td>
<td>Focal empty LP and/or fibrosis</td>
</tr>
<tr>
<td>NC (no change)</td>
<td>Dense, diffuse, or nodular</td>
<td>Present, “may be absent”</td>
<td>No changes</td>
</tr>
</tbody>
</table>

MM, muscularis mucosa; LP, lamina propria; SM, submucosa; LEL, lymphoepithelial lesions.
**Probable minimal residual = CR ???**

<table>
<thead>
<tr>
<th>GELA category</th>
<th>Histology</th>
<th>Clinical significance</th>
<th>Comments</th>
</tr>
</thead>
<tbody>
<tr>
<td>Complete histological</td>
<td>Total disappearance of the lymphoid</td>
<td>Complete remission</td>
<td>Identification of CR may be subject to sampling 'artefact' and the designation of complete regression needs sustained absence of histological disease in the context of remission as assessed by all other means. No need for additional treatment.</td>
</tr>
<tr>
<td>response (CR)</td>
<td>infiltrate with only scattered small lymphocytes and plasma cells. Regressive stromal changes with fibrosis and separation of glands can be seen.</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Probable minimal residual</td>
<td>Small lymphoid aggregates present, usually at the base of the lamina propria. Associated stromal regressive changes are usually present.</td>
<td>Complete remission</td>
<td>The significance of the lymphoid aggregates is impossible to determine by morphology or immunocytochemistry, but it has been established that these nodules frequently, but not always, harbour cells with the same clonal gene rearrangement as the original lymphoma cells, consistent with the presence of a small number of residual neoplastic cells. However, no adverse prognostic significance has been demonstrated associated with this histology which is detected in early follow-up biopsies after <em>Helicobacter pylori</em> eradication of most cases undergoing subsequent complete remission. No need for additional treatment.</td>
</tr>
<tr>
<td>disease (pMRD)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Responding residual disease</td>
<td>Overt residual lymphoma with a nodular or diffuse infiltrate of neoplastic B-cells but with clear evidence of regressive stromal changes characterised by fine fibrosis and an 'empty lamina propria'.</td>
<td>Partial remission</td>
<td>Comparison with the diagnostic biopsy is helpful in this context. These features are considered to indicate a partial and ongoing response. In the absence of unfavourable endoscopic results or a clinical appearances of progression, a decision about additional treatment can be postponed until after the following endoscopic assessment. Management should be individually tailored.</td>
</tr>
<tr>
<td>(rRD)</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>No change (NC)</td>
<td>Persistence of overt lymphoma identical to that seen at diagnosis with no morphological features to suggest response to treatment (such as stromal fibrosis).</td>
<td>Stable disease or progressive disease</td>
<td>In the case of persisting macroscopic lesions or evidence of dissemination of the disease, oncological treatment should be proposed. If only microscopic infiltration is present, oncological treatment can be postponed up to 24 months after achievement of <em>Helicobacter pylori</em> eradication, after which management should be individually tailored.</td>
</tr>
</tbody>
</table>

GELA, Groupe d'Etude des Lymphomes de l'Adult; MALT, mucosa-associated lymphoid tissue.
왜 NC가 나쁜 이름인가?
- physician assistant가 정리한 의무기록

UBT : (-)
EGD : 1. Gatric Maltoma (#1-#2)
  2. CAG
* bx : 1. Stomach, antrum, anterior wall, biopsy :
   .CG, inactive, with lymphoepithelial lesion
   . GELA histologic grading system: NC (no change)
   . Hp(-)
  2. Stomach, low body, anterior wall, biopsy :
   . Chronic gastritis, inactive, with lymphoepithelial lesion
   . GELA histologic grading system: rRD (responding residual disease)
   . Hp(-)
  3. Stomach, antrum and mid body, greater curvature, biopsy : CG, Hp(-)
제균하였으나 용종 없어지지 않아서 용종절 제술 후 경과관찰함
Histology after polypectomy
MALToma mimicking plasmacytoma
Follow-up 8 years later
- MALToma mimicking plasmacytoma
MALT lymphoma simulating an extramedullary plasmacytoma of the stomach

- Approximately one third of all cases of gastric MALToma show variable degrees of plasma cell differentiation, occasionally plasma cells constitute the major population in the tumor.
- As surgical resection has been the standard treatment for gastric plasmacytoma, this case highlights the need for caution. If *H. pylori* infection is found in a patient with a gastric plasmacytoma, it should be eradicated as a first line of therapy before surgery is considered.
조직검사에서 MALToma라고 하였으나...
제균치료 후 호전 없어 RT 하였고 이후 호전
Duodenal MALToma
Cecal MALToma
Rectal MALToma
Marginal zone cells (IgM)

Mantle zone cells (IgM+D)

Follicle center cells (IgM, IgG, IgA or IgE, not IgD)
Lymphomatous polyposis (M/66)
Mantle cell lymphoma
- stomach and colon, *Cyclin D1: Positive*
GI involvement of nodal MCL
SMC experience of 19 GI MCLs

• Presenting symptoms: abdominal pain (36.8%), GI bleeding (26.3%)
• Location: colon alone (47.4%), colon and stomach (36.8%), stomach alone (10.5%)
• Endoscopy: polypoid (48.1%), infiltrative (33.3%), ulcerative (14.8%), fungating (3.7%)
Scattered tingible body-laden macrophages (macrophages containing dead body of apoptotic tumor cells)
Burkitt lymphoma at SMC, 1995-2007

Burkitt’s lymphoma n=80

Adult (age >18) n=47, 58.7%

Child n=33, 41.3%

GIT involve n=20, 43%

No GIT involve n=27, 57%

## Involved organs of GI Burkitt lymphoma

<table>
<thead>
<tr>
<th>Organs</th>
<th>Number (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Stomach</td>
<td>11 (55%)</td>
</tr>
<tr>
<td>Colon</td>
<td>4 (20%)</td>
</tr>
<tr>
<td>Stomach + Duodenum</td>
<td>3 (15%)</td>
</tr>
<tr>
<td>Stomach + Colon</td>
<td>1 (5%)</td>
</tr>
<tr>
<td>Stomach + Duodenum + Colon</td>
<td>1 (5%)</td>
</tr>
</tbody>
</table>

Fungiating type

Ulcerative type

Ulcerofungiating type

Ulcerinfiltrative type
Survival of GI Burkitt lymphoma

Peripheral T cell lymphoma (PTCL)
- CD3 (+), CD56(-)
PTCL detected during screening
Primary NK-/T-cell lymphoma of the gastrointestinal tract: clinical characteristics and endoscopic findings

Authors
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² Department of Internal Medicine, Konkuk University School of Medicine, Seoul, Korea
³ Department of Pathology, Samsung Medical Center, Sungkyunkwan University School of Medicine, Seoul, Korea

Background and study aims: Primary NK-/T-cell lymphoma of the gastrointestinal tract is a very rare disease with a poor prognosis. The aim of this study was to determine the clinical and endoscopic characteristics of patients with primary gastrointestinal NK-/T-cell lymphoma.

Patients and methods: The clinical features of 14 patients with primary gastrointestinal NK-/T-cell lymphoma and the endoscopic findings in 11 of these patients were reviewed. Their median age (n=8, 57%) or surgical resection (n=5, 36%). The median survival for all patients was 9 months. On endoscopy in 11 patients, the anatomic location of the primary lesion was found to be: stomach, n=3 (27%); esophagus, n=2 (18%); duodenum, n=1 (9%); and the ileocolonic area, n=5 (46%). These lesions were ulceroinfiltrative in 4 cases (36%), ulcerative in 3 cases (27%), superficial/erosive in 3 cases (27%), and infiltrative in 1 case (9%). No prominent fungating mass was
Figure 3  Overall survival of enrolled patients.
결론: 위장관 림프종

• 내시경 진단이 항상 쉬운 것은 아니다.
• 병리 검사 결과 해석에 주의하자.
• 우리나라에는 장 T-세포 림프종이 많다.
• 진단이 지연될 수 있다.
• 위장관 림프종은 매우 다양하다.
아래 종설을 참고하시기 바랍니다.

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Vol. 12, No. 3, 158-165, September 2012 • http://dx.doi.org/10.7704/khugr.2012.12.3.158

위장관 림프종

Gastrointestinal Lymphoma

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School of Medicine, Seoul, Korea

Gastrointestinal tract is the most common location of extranodal lymphoma and 95.4% of gastrointestinal lymphoma is non-Hodgkin type. Although gastrointestinal lymphoma is usually secondary to nodal lymphoma, it can present as a primary gastrointestinal lymphoma with the majority being in the stomach (74.8%). In South Korea, the most frequent histological subtype of gastric lymphoma is extranodal marginal zone B-cell lymphoma of mucosa-associated lymphoid tissue (MALT), followed by diffuse large B-cell lymphoma. Gastrointestinal lymphoma typically presents with nonspecific symptoms, and endoscopic findings are quite variable. So, the diagnosis is mainly dependent on the histopathological evaluation. Treatment of gastrointestinal lymphoma is dictated primarily by the histopathological type and the stage of the disease. This review will discuss the histopathological classification, staging systems, clinical features and treatment of gastrointestinal lymphoma. (Korean J Helicobacter Up Gastrointest Res 2012;12:158-165)

Key Words: Gastrointestinal neoplasms; Lymphoma
경청해 주셔서 감사합니다.

5. 위장관 림프종

1) DLBL (diffuse large B cell lymphoma) B세포 림프종
2) MALToma 말트림프종
3) Mantle cell lymphoma
4) Peripheral T cell lymphoma (PTCL)
5) NK T-cell lymphoma
6) Burkitt lymphoma 버킷 림프종
7) Primary gastric lymphoblastic lymphoma

경청해 주셔서 감사합니다.