Extranodal lymphoma

Rising Incidence and Patterns in the Abdomen

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I HAVE NO DISCLOSURES
"THE RISING INCIDENCE OF LYMPHOMA IN THE PAST 2 DECADES HAS BEEN CHARACTERIZED BY AN INCREASE IN EXTRANODAL LYMPHOMA"

1. Immunosuppression
   • HIV
   • Solid organ transplants (esp. cyclosporine)
2. Indolent viral infections (EBV)
3. Increase in skin lymphomas may be related to sunlight
Learning Objectives

1. To recognize the increasing incidence of extranodal lymphoma

2. To review the most common patterns of involvement in the abdomen

3. To review potential pitfalls and differential diagnoses in extranodal lymphoma
Outline: Extranodal Lymphoma

• Definition
• Primary vs. Secondary
• Types of Lymphoma most likely to manifest as extranodal disease

• Most Common Involvement In The Abdomen
  – Spleen
  – Liver
  – GI tract
  – Kidneys
Extranodal Lymphoma

**DEFINITION:**
- Lymphoma located outside of lymph nodes, thymus, tonsils, Waldeyer’s ring

**SPLEEN:**
- Nodal disease in HL
- Extranodal disease in NHL

**PROGNOSIS:**
- Usually intermediate to high-grade
- Typically indicates a more advanced stage of disease
- Poor prognostic factor

**FREQUENCY:**
- More common in NHL than HL (40% vs. 5%)
- More common in recurrent disease or immunodeficiency
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<th>Primary Extranodal Lymphoma</th>
<th>Secondary Extranodal Lymphoma</th>
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<td>➢ Confined to a single organ and immediately adjacent lymph nodes</td>
<td>➢ Involves nodes other than those immediately adjacent to the primary organ</td>
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<td>• Can be Stage I or II</td>
<td>➢ OR more than one extranodal site</td>
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<td>• Stage III or IV</td>
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<td>• More common than primary</td>
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Advantages of FDG PET/CT are largely due to:

1. *Extranodal sites not identified at CT*
2. Detection of FDG-avid, normal-sized lymph nodes (usually <1 cm)
Subtypes of Lymphoma involving Extrananodal sites

- Most common subtypes:
  - Diffuse large B-cell
  - Follicular lymphoma

- Most LIKELY to affect extranodal sites:
  - Mucosa-associated lymphoid tissue (MALT) lymphoma
  - Mantle cell lymphoma
  - Lymphoblastic lymphoma
  - Burkitt's lymphoma
Splenic lymphoma
SPLENIC LYMPHOMA

- Most common splenic malignancy

- PET/CT:
  - Accuracy of almost 100% for primary splenic involvement at staging (CT accuracy = 57%)
  - Evaluation more limited after treatment due to splenic activation
Splenic lymphoma: Patterns of involvement

- Normal
- Diffuse infiltration
- Splenomegaly
- Discrete Nodules
Patterns of Splenic Lymphoma: Diffuse infiltration

- Most common pattern
- May look normal

Lymphadenopathy in the splenic hilum is suggestive of splenic disease
Splenic Lymphoma: Splenomegaly

Size is a poor indicator of splenic involvement

Normal size
• Does not exclude involvement

Moderate splenomegaly
• Reactive splenomegaly occurs in 30% of patients

Marked splenomegaly
• Almost always indicates infiltration
Patterns of Splenic Lymphoma: Discrete nodules

- **CT:** Low attenuation and hypovascular
- **MRI:** hypo/isointense on T1WI, hyperintense on T2WI, hypovascular
Patterns of Splenic Lymphoma: Discrete nodules

On Ultrasound: hypoechoic with no through transmission
Hepatic lymphoma
Hepatic lymphoma

• At presentation, the liver is involved in
  – 15% of patients with NHL
  – 10% of with HD

• Primary disease is rare:
  – Most common in immunocompromised patients
Patterns of Hepatic lymphoma
Patterns of Hepatic Lymphoma: Hepatomegaly and Diffuse Infiltration

1. Diffuse infiltration is the most common pattern
Patterns of Hepatic lymphoma: Discrete nodules

- Hepatic NHL lymphoma may occur in the absence of splenic disease
- Hodgkin’s Lymphoma of the liver is almost always associated with splenic disease
Primary Hepatic Lymphoma

- Single dominant mass
- Heterogeneous

Secondary Hepatic Lymphoma

- Typically multiple, smaller masses
- Homogenous
Hepatic lymphoma
Hepatic lymphoma
Hepatic lymphoma
Metastases vs. Lymphoma

1. Heterogeneous
2. Spleen usually normal

1. Homogenous
2. Bulky lymphadenopathy
3. Spleen often involved
Metastases vs. Lymphoma

Lymphoma

Metastatic Melanoma
Disseminated fungal infection in immunocompromised patients

- Typically smaller and more heterogeneous
- Peripheral enhancement
- No lymphadenopathy
- MRI most sensitive in differentiating lymphoma from fungus
Lymphoma of the GI Tract
Lymphoma of the GI tract

- Primary GI lymphoma is the most common extra-nodal manifestation of non-Hodgkin lymphoma (20% of all cases)
- Multiple sites often involved

Obstruction is uncommon ... EXCEPTION ... T cell lymphoma
Perforation is uncommon... EXCEPTION ... Post chemotherapy
Imaging appearance of GI Lymphoma

1. Nodular
2. Polypoid
3. Infiltrative
4. Aneurysmal
5. Ulcerative

CT staging has important therapeutic implications:

- Stage I and II disease may be excised
- Stage III and IV disease must be treated with radiation, chemotherapy, or both.

- GI tract lymphoma may be missed at endoscopy
Risk factors for GI Lymphoma:

1. HIV
2. *Helicobacter pylori*
3. Celiac disease (Enteropathy Type T-cell Lymphoma)
4. IBD
   - 2-3 x increase
   - Further 5x increased risk with immunosuppression
5. Immunosuppression after solid organ transplantation
Gastric lymphoma

Most common histologic subtypes:

- *H. Pylori* –associated low-grade MALT lymphoma
- High-grade diffuse large B-cell lymphoma.
Gastric MALT Lymphoma

- Chronic *H pylori* infection is associated with development of mucosa associated lymphoid tissue = MALT

  - Most low-grade primary gastric lymphomas arise from MALT
  - Low-grade MALT lymphoma diagnosed early has a good prognosis (5-year survival 75%–91%)
  - High grade MALT lymphoma: survival rate < 50%

Therefore, early diagnosis is crucial

- Detection challenging as the clinical, endoscopic, and radiologic findings can simulate gastritis and gastric carcinoma.
MALT LYMPHOMA (High Grade)

- Normal CT shown to be highly predictive of low-grade MALT lymphoma
- Greater than minimal thickening suggests transformation to a higher grade
- Lymphadenopathy less common in low-grade MALT Lymphoma
Gastric B Cell Lymphoma
Gastric Lymphoma vs. Adenocarcinoma

**Lymphoma**
- Diffuse wall thickening
- Hypovascular
- Gastric distensibility
- Lymphadenopathy below the renal pedicle

**Adenocarcinoma**
- More focal wall thickening
- Greater tumour enhancement
- Tumour infiltration beyond gastric wall
- Linitis plastica
- Gastric outlet obstruction
Gastric Lymphoma vs. Adenocarcinoma

Lymphadenopathy extending below the renal pedicle = Lymphoma
Occasionally, a GIST may mimic a gastric lymphoma
Lymphoma of the Small Bowel

- Rising incidence related to B-cell hyperactivation in HIV
- Distal ileum is the most frequently affected site
Small Bowel Lymphoma
Small Bowel Lymphoma

Mantle cell lymphoma: Most common subtype
Enteropathy Associated T Cell Lymphoma: Obstruction
Enteropathy Associated T Cell Lymphoma: Obstruction

- 50 x increased risk with untreated Celiac Disease
- Less wall thickening than B-cell lymphomas
- Imaging features similar to IBD with extensive ulcers, refractory to treatment

***Death is usually due to perforation caused by refractory malignant ulcers.
Post Transplant Proliferative Disorder
Post Transplant Proliferative Disorder: Perforated Esophagus
SB lymphoma vs. Adenocarcinoma

- Cavitary mass
- Multifocal
- Bulky lymphadenopathy

- Solid mass
- Obstructing
Duodenal lymphoma
Lymphoma of the Large Bowel

- Rectal mass
- Bulky Lymphadenopathy

Most common in the cecum and rectum
Mantel Cell Lymphoma of the Cecum with intussusception
Mantel Cell Lymphoma of the Cecum with intussusception
Colonic lymphoma presenting with innumerable polypoidal lesions
Rectal Lymphoma
Cecal lymphoma with lymphadenopathy

Cecal Mass

Bulky Lymphadenopathy
Renal Lymphoma
Renal Lymphoma

• Usually clinically silent
• Acute renal failure is rare
• Incidence:
  – 60 % at autopsy
  – ...BUT only 8% of all patients at staging CT

Often minimal mass effect on renal contour

- Imaging findings can be subtle
- IV contrast is essential for diagnosis.
Patterns of Renal Lymphoma

- Multiple masses
- Single mass
- Invasion from retroperitoneal disease
- Perirenal disease
- Infiltrative
Patterns of Renal Lymphoma: Multiple Lesions = widespread disease

- Most common pattern
- Usually bilateral
Patterns of Renal Lymphoma: Multiple Lesions = widespread disease
Multifocal Lymphoma

Metastases (Lung Cancer)
Patterns of Renal Lymphoma: Solitary Lesion

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<th>Ultrasound</th>
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<td>• Nephrographic-phase is the most sensitive for lesion detection</td>
<td>• hypoechoic (reflects tissue homogeneity)</td>
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Lymphoma

- Typically hypovascular
- Homogeneous

Renal Cell Carcinoma

- Typically vascular (caution... papillary RCC)
- Often heterogeneous
- Homogenously hypoechoic...
PITFALL = CYST
- Hypovascular

- Varying appearances
- Typically more echogenic and heterogeneous
Diagnosis: Pyelonephritis
3 months later... Bilateral Disease
- Biopsy proven renal lymphoma

January 2008
April 2008
Transplant kidney mass

DDx: Renal cell carcinoma, PTLD
Patterns of Renal Lymphoma: 
*Renal invasion from contiguous retroperitoneal disease*
Patterns of Renal Lymphoma: Perirenal Disease

- May surround kidney without parenchymal compression
- Renal vessels remain patent despite encasement
- Can cause obstruction with hydronephrosis
Patterns of Renal Lymphoma: *Diffuse infiltration*

- Almost always bilateral
- Renal function poor, but usually sufficient to remain clinically silent.
Diffuse Infiltration

Nephromegaly and infiltrated parenchyma
Diffuse Infiltration

Nephromegaly

4 years earlier
Patient with AIDS

- Nephromegaly (17 cm)
- Echogenic Kidney

- DDx: Aids Nephropathy vs. Infiltrative Lymphoma

- Biopsy: Lymphoma
Patterns of Renal Lymphoma: 
*Focal Infiltration*

**DDx:** TCC or pyelonephritis
Infiltrating hilar mass causing hydrenephrosis
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References


9. Gurney KA, Cartwright RA. Increasing incidence and descriptive epidemiology of extranodal non-Hodgkin lymphoma in parts of England and Wales.. Leukaemia Research Fund Centre for Clinical Epidemiology at Leeds University, Institute of Epidemiology, Margaret Smith Building, 30 Hyde Terrace, LS2 9LN, UK.


Thank You

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