Lymphomas continue surprising us: typical and atypical presentations

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Learning objectives

1. Describe typical and atypical presentation forms of lymphomas according to type, location and radiological imaging.
2. Illustrate cases in which we should be suspicious of lymphoma as first option.
3. Show atypical cases of lymphomas that simulate other diseases.

Background

Lymphomas have a large variety of imaging appearances and they can be characteristic or simulate other diseases.

We will show the following typical and atypical lymphomas according to location, type and imaging findings. (Fig. 1)

a) LOCATION:

a.1) Typical: retroperitoneum, anterior mediastinum, spleen.

a.2) Atypical: ureter, ovarium, urinary bladder, frontal bone, large bowel.

b) TYPE:

b.1) Typical: gastric MALT lymphoma.

b.2) Atypical: gastric and pulmonary T-cell lymphoma.

c) IMAGING FINDINGS:

c.1) Typical: "butterfly pattern", "sandwich sign", "floating aorta sign", perirrenal involvement, sacral foramina involvement.
c.2) **Atypical:** extraaxial primary central nervous system (CNS) lymphoma, small bowel obstruction, peritoneal lymphomatosis, heart infiltration.

**Images for this section:**

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<td><strong>LOCATION</strong></td>
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<td><strong>TYPE</strong></td>
<td>- gastric MALT lymphoma</td>
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<td><strong>IMAGING FINDING</strong></td>
<td>- “butterfly pattern”</td>
<td>- extraaxial primary CNS lymphoma</td>
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<td>- “sandwich sign”</td>
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<td>- perirenal involvement</td>
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**Table 1:** Typical and atypical cases of lymphoma illustrated in the following section, according to the location, type and imaging finding.
Imaging findings OR Procedure details

a) **LOCATION**:

a.1) Typical:

- **Retroperitoneum**: Lymphoma may result in lymphadenopathy almost anywhere in the body, but one of the locations where it more frequently results in lymphadenopathy is the retroperitoneum and the mesentery. In fact, the most common malignancy resulting in mesenteric lymphadenopathy is lymphoma. (Fig. 1)

- **Anterior mediastinum**: The mediastinum is commonly involved in lymphoma, either as part of disseminated disease or less commonly as the site of primary involvement. Mediastinal lymphomas usually arise from either the thymus or lymph nodes, thus accounting for their predilection for the anterior and middle mediastinum. (Fig. 2)

- **Spleen**: Secondary involvement of the spleen is common in both non-Hodgkin lymphomas and Hodgkin disease. Primary splenic lymphoma is rare. Diffuse uniform infiltration is the most common form of splenic lymphoma. Other imaging patterns diffuse infiltration with miliary lesions, multiple focal nodular lesions and large solitary mass. (Fig. 3).

a.2) Atypical:

- **Ureter**: Ureteric involvement in lymphoma is rare and has largely been reported in patients with aggressive non-Hodgkin lymphomas. When it does occur, it is usually secondary to renal involvement or retroperitoneal lymphadenopathy. Direct ureteric involvement in lymphoma is extremely rare. (Fig. 4)

- **Ovarium**: Primary ovarian lymphoma is a rare manifestation of non-Hodgkin lymphomas, even though secondary involvement is well recognized. Primary ovarian lymphoma accounts for 0.5% of non-Hodgkin lymphomas and 1.5% of ovarian tumours. This is partly because of the fact that there is no lymphoid tissue within the ovary. So the diagnosis of ovarian lymphoma is almost invariably unsuspected until the tumor is examined histologically. (Fig. 5)
• **Urinary bladder**: Ninety-five percent of bladder neoplasms arise from the epithelium and the most common subtype is urothelial carcinoma, which accounts for 90% of cases. Primary bladder lymphoma is rare, as there is no lymphoid tissue in the bladder, but secondary involvement of the bladder may be present in 10%-25% of patients with lymphoma and leukemia. The diagnosis of lymphoma is further strengthened if there is involvement of other organs. (Fig. 6)

• **Frontal bone**: The primary lymphoma of the skull is extremely rare and the imaging findings are inespecific. Primary lymphoma of bone is a rare malignant condition that accounts for less than 5% of all primary bone tumors. The lytic-destructive pattern is the most common radiographic appearance and the diagnosis is further strengthened if the soft tissue mass is associated with surprisingly little cortical destruction. Secondary or metastatic lymphoma is indistinguishable from primary bone lymphoma without whole-body surveillance to diagnose the source. (Fig. 7)

• **Large bowel**: Primary lymphoma of the large bowel accounts for 0.4% of all tumors of the colon, and colorectal lymphomas constitute 6%-12% of gastrointestinal lymphomas. More often affects the cecum and rectum than other parts of the large bowel. (Fig. 8)

b) **TYPE**:

Hodgkin disease is usually supradiaphragmatic, with mediastinum involvement and contiguous spread from one nodal group to the next along the lymphatic pathways. It is usually almost entirely confined to the lymph nodes. Extranodal involvement is much less common in Hodgkin disease than in non-Hodgkin lymphoma.

Besides, the following are more common in non-Hodgkin lymphoma that in Hodgkin disease: extranodal involvement, mesenteric adenopathies, hepatic involvement without spleen disease and bone marrow disease.

b.1) Typical:

• **Gastric MALT lymphoma**: Gastrointestinal lymphoma is an uncommon disease but is the most frequently occurring extranodal lymphoma and is almost exclusively of non-Hodgkin type. The stomach is the most frequently involved, accounting for 50%-70% of all primary gastrointestinal lymphomas and the majority are mucosa-associated lymphoid tissue (MALT) lymphomas. (Fig. 9)
b) **TYPE:**

b.2) **Atypical:**

- **Gastric T lymphoma:** peripheral T-cell lymphoma represents a relatively small proportion of lymphomas and has a lower prevalence in Western countries, with an aggressive course. According to gastrointestinal system it most frequently involves the small intestine; however, it could involve the colon and stomach. At gross examination, multiple intestinal ulcers are present, often with perforation. Pathologic differentiation from inflammatory bowel disease is difficult as atypical lymphoma cells and inflammatory lymphocytes mix at the site of mucosal ulceration. (Fig. 10)

c) **IMAGING FINDINGS:**

c.1) **Typical:**

- **"Butterfly pattern" primary CNS lymphoma:** Lymphomas are 16% of all primary CNS tumors. In patients with normal immunity, classically presents as a solitary homogeneously enhancing mass. Unenhanced CT typically shows a high-density (70%) lesion in a central hemispheric location, which often reaches or crosses the midline giving the "butterfly pattern". (Fig. 11)

- **"Sandwich sign":** the mesenteric lymph nodes involved by lymphoma often coalesce, forming a conglomerate soft-tissue mass. It tends to grow around and displace normal anatomic structures such as vessels or bowel, without destroying them, giving an appearance of a hamburger, called as "sandwich sign". This sign is specific for mesenteric lymphoma. (Fig. 12)

- **"Floating aorta sign":** lymphoma tends to surround adjacent vessels and in retroperitoneal involvement sometimes the aorta seems to be immersed in the tumor, giving the "floating aorta" or "CT angiogram" sign. (Fig. 13)

- **Perirenal involvement:** Perirenal involvement by lymphoma is usually the result of direct extension from retroperitoneal disease or transcapsular spread of renal parenchymal disease. Less commonly, perirenal disease is isolated from the renal parenchyma, in which case the disease can completely surround the kidney without parenchymal compression or functional impairment. Although this finding in renal lymphoma occurs infrequently, it is virtually pathognomonic. (Fig. 14)
• **Sacral foramina involvement**: Lymphoma is a common tumor of the paravertebral region. It usually originates in the para-aortic lymph nodes and extends to involve adjacent structures. The typical route of dissemination to the spine is by direct, contiguous spread from the retroperitoneum to the paraspinous and epidural spaces through the neural foramina. (Fig. 15)

c) **IMAGING FINDINGS**:

c.2) Atypical:

• **Leptomeningeal lymphoma**: Intraaxial lymphoma is usually primary and extraaxial lymphoma is usually secondary. In one-thirds of the cases secondary CNS lymphomas present as leptomeningeal metastases, while only one-third do it as parenchymal metastases. Thus, leptomeningeal primary lymphoma is rare (8% of all primary CNS lymphomas). It is often undetectable or inespecific. Imaging techniques can show meningeal calcification, enhancement or masses. (Fig. 16)

• **Small bowel obstruction**: Lymphoma is the most common malignancy of the small bowel. As in other sites of lymphomatous involvement, obstruction is uncommon in the small bowel, since the tumor does not elicit a desmoplastic response. Although less commonly the radiologic appearance of lymphoma may mimic that of adenocarcinoma with bowel obstruction. (Fig. 17)

• **Peritoneal lymphomatosis**: Peritoneal lymphomatosis is a very rare complication of non-Hodgkin lymphoma (most commonly diffuse large B-cell lymphoma) most often arising in patients with known disease. Patterns of tumor involvement of the mesentery, omentum, and peritoneum are indistinguishable from those seen in peritoneal carcinomatosis. Looking for other lymphomatous disease signs is almost necessary to suspect the diagnosis. (Fig. 18)

• **Mediastinal aggressive lymphoma**: as everywhere, lymphoma is a soft tumor that tends to surround and displace other structures. A mediastinal lymphoma invading adjacent structures such as heart is very uncommon. (Fig. 19)

Images for this section:
Fig. 1: 53-year-old male with centrofollicular lymphoma. Contrast-enhanced CT image shows mesenteric and retroperitoneal lymphadenopathy.
Fig. 2: 23-year-old male with dysnea and neck swelling. A, B) Posteroanterior and lateral chest radiographs show an anterior mediastinal mass. C) Contrast-enhanced CT scan demonstrates an homogeneus soft tissue mass in the anterior mediastinum that compresses superior cava vein and right pulmonary artery (arrow). Diffuse large B-cell lymphoma produces superior vena cava syndrome.
Fig. 3: 67-year-old female with non-Hodgkin disease. Coronal contrast-enhanced CT image reveals homogeneous moderate splenomegaly due to linfomatous involvement.
Fig. 4: 36-year-old female with ureteral primary follicular lymphoma. A) Intravenous urography shows stenosis of the right midureter with dilatation of its collecting system. B, C) Contrast-enhanced CT image demonstrates right excretory system dilatation due to ureteral thickening.
Fig. 5: 43-year-old female with abdominal distension and flatulences. Pelvic US demonstrates ascitis and two heterogeneous pelvic masses (X) dependent from ovaries (o). The uterus (u) is normal.
Fig. 6: 80-year-old female with anemia and right abdominal pain. A) US images show right renal pelvis dilatation produced by a mass in the right lateral wall of the bladder. B) Contrast-enhanced CT confirms the irregular thickening of the urinary bladder wall.
Fig. 7: 81-year-old female with right side face swelling. CT scan of the brain shows a soft-tissue mass in right frontal region which enhance markedly after contrast administration (B) and destroys both the external and internal laminae of the skull (D). The mass extends into right orbital (A) and anterior cranial fossas (B. Frontal bone anaplastic lymphoma.
Fig. 8: 74-year-old male with lymphomatous polyposis. Double-contrast barium enema examination demonstrates multiples polypoid masses in ascending and transverse colon.
**Fig. 9:** 75-year-old female with loss of weight and epigastric pain. Contrast-enhanced CT image show hipodense and irregular antrum wall thickening produced by MALT lymphoma.
**Fig. 10:** 46-year-old male with gastric and pulmonary T-cell lymphoma. A) Contrast-enhanced CT image reveals antrum wall irregular thickening. B) Pulmonary window of CT scan shows nodular consolidation with air bronchogram (arrowhead) in the left lower lobe.

**Fig. 11:** 81-year-old female with confusion and change in mental status. A) Unenhanced CT shows a high-density mass extending across splenium of corpus callosum and crossing the midline. B) Contrast-enhanced CT and C) coronal and axial gadolinium-enhanced T1-weighted MR images demonstrate marked homogeneous enhancement of callosal tumor with extension into both occipital lobes. Diffuse large B cell lymphoma.
**Fig. 12:** 61-year-old male with non-Hodgkin disease. Contrast-enhanced CT image illustrates a conglomerate mass formed by mesenteric and retroperitoneal lymphadenopathy that surrounds but does not occlude the superior mesenteric artery, showing the "sandwich sign".
Fig. 13: 53-year-old male with low grade follicular lymphoma. Contrast-enhanced CT image reveals a soft-tissue attenuation mass in the retroperitoneum which separates the aorta from the vertebral body. This is called "floating aorta sign".
Fig. 14: 36-year-old female with perirenal lymphoma. Contrast-enhanced CT scan shows perirenal soft-tissue mass enveloping both kidneys, predominantly the left one.
Fig. 15: 50-year-old male with non-Hodgkin disease. Contrast-enhanced CT images show a soft-tissue mass in internal iliac chain region extending to left anterior sacral foramina. Look the lack of fat in the left sacral foramina comparing with the right one.
Fig. 16: 58-year-old female with sensibility disorders. Before (A, C) and after (B) contrast material administration CT images reveal an extraaxial left frontal lesion spontaneously hyperdense (arrow) and enhanced after contrast administration. A meningioma en plaque was suspected. Axial and coronal before (D) and after (E,F) gadolinium-enhanced T1-weighted MR images show an irregular and focal meningeal thickening and enhancement in the left frontoparietal convexity (arrow heads). Surgery biopsy demonstrates leptomeningeal primary lymphoma.
**Fig. 17:** 55-year-old female with abdominal distension. Axial and coronal contrast-enhanced CT images show a diffuse thickening of the ileum which produces bowel obstruction. Histological examination after surgery demonstrates non-Hodgkin lymphoma.
**Fig. 18:** 69-year-old female with abdominal non-Hodgkin lymphoma. Contrast-enhanced CT scan illustrates diffuse omental thickening in the left flank (arrowheads) and the right paracolic gutter (arrow) due to peritoneal lymphomatosis. It has soft-tissue masses in mesentery (X) and retroperitoneum according to lymph nodes conglomerates and left perirrenal space involvement (*).
Fig. 19: 22-year-old female with a left ventricular mass view in US. Thromboembolism suspect. CT pulmonary angiogram demonstrates an anterior mediastinum mass (X) which surrounds aorta and infiltrates pulmonary artery (A) and left heart (B).
Conclusion

Lymphomas have a wide variety of imaging appearances and familiarity with the spectrum of findings is essential for radiologists.

Some presentation forms are almost pathognomonic and make us think on lymphoma as the first possibility. Instead, in other cases the findings lead to a different entity and finally the diagnosis of lymphoma surprises us.

Personal Information

References


