Definition

Lymphoma limited to the chest and lungs, with or without mediastinal lymphadenopathy • No extrathoracic manifestation for at least 3 months.

- **Epidemiology**
  Rare compared with secondary lymphoma arising via hematogenous dissemination or by direct extension from hilar or mediastinal lymphomas.
- **Etiology, pathophysiology, pathogenesis**
  Forms:
  - In combination with intrathoracic lymphadenopathy • 10–15% of lymphomas • More common in Hodgkin disease than in non-Hodgkin lymphoma.
  - Primary pulmonary lymphoma (at most with minimal lymph node involvement) • Rare, < 1% of all malignant lymphomas • Either Hodgkin or non-Hodgkin lymphoma • In primary pulmonary non-Hodgkin lymphoma, a distinction is made between low-grade MALT B-cell lymphoma, high-grade non-Hodgkin lymphoma of B-cell type (about two-thirds of cases, usually associated with Epstein–Barr virus; risk groups—HIV-infected patients and organ transplant recipients), and the angioimmunoblastic lymphomas of T-cell type.

Imaging Signs

- **Modality of choice**
  CT is preferable to plain radiography.
- **Radiographic findings**
  Broad spectrum of findings ranging from miliary foci to nodules, pneumonia-like infiltrates (with or without air bronchogram), and interstitial and even ground-glass changes.
- **CT findings**
  Broad spectrum of findings (in two-thirds of cases there are bilateral and/or multiple foci)—one or more nodules with or without cavitation • Round or segmental infiltrates (with or without an air bronchogram) • Up to 50% of high-grade lymphomas include liquefaction that may be rapidly progressive • Reticulonodular changes.
- **Pathognomonic findings**
  Rapidly progressive consolidations with an air bronchogram and elongated bronchovascular structures (CT angiogram sign).

Clinical Aspects

- **Typical presentation**
  Low-grade lymphoma: Asymptomatic in > 50% of cases, otherwise mild nonspecific symptoms (cough, slight dyspnea, chest pain) • *High-grade lymphoma:* Generally symptomatic (symptoms of hepatitis B infection).
- **Confirmation of the diagnosis**
  Biopsy.

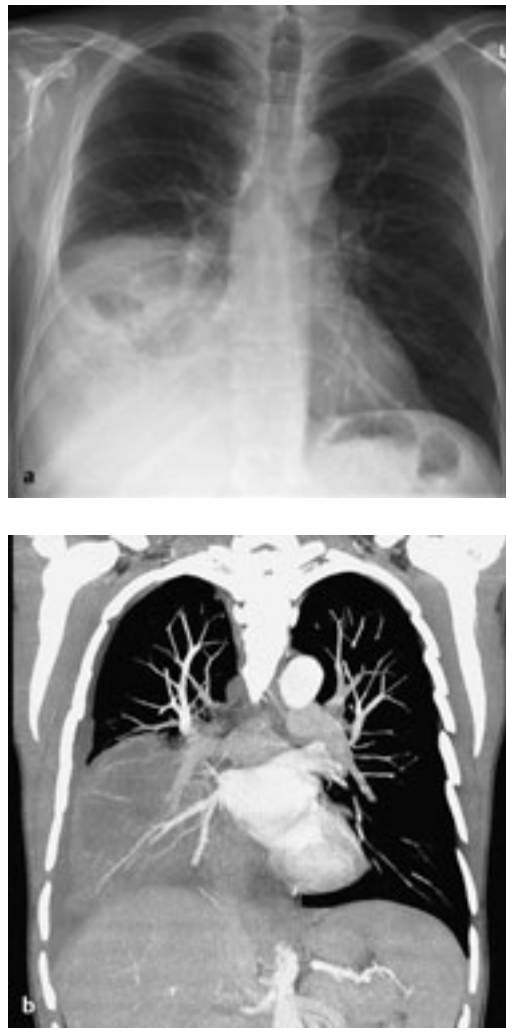
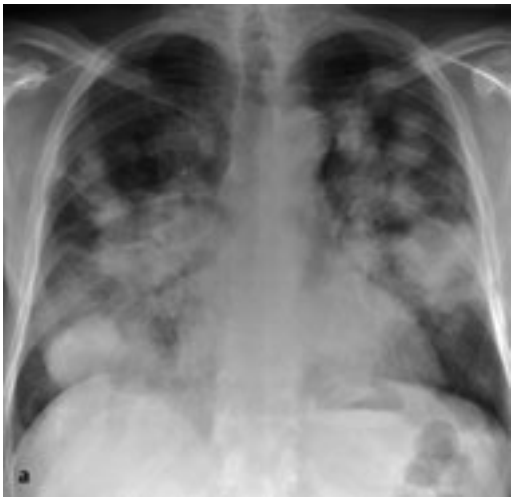


Fig. 8.9 Highly malignant Epstein–Barr virus-associated B-cell non-Hodgkin lymphoma in a 39-year-old man with HIV infection. Two-week history of fever, nonproductive cough, and rapid deterioration of general health.

a The plain chest radiograph shows an extensive, relatively homogeneous infiltration of the right lower lobe, enclosing a small radiolucency consistent with liquefaction.

b On CT (coronal MIP slices) the finding also appears relatively homogeneous. The major vessels and bronchi are intact (CT angiogram sign and air bronchogram).



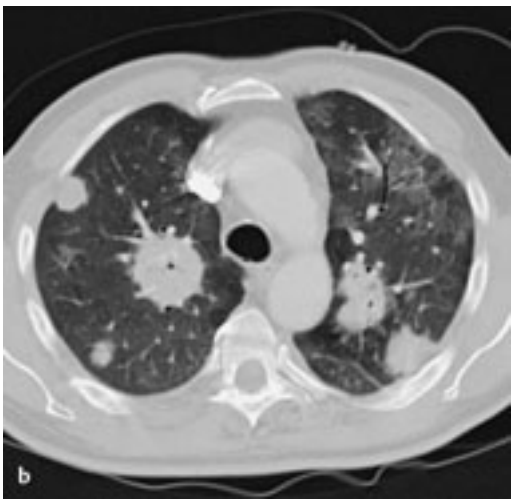


Fig. 8.10 MALT lymphoma in a 40-year-old man with HIV infection and weight loss and limited exercise tolerance. Both lungs show large infiltrates resembling round focal lesions that are partially confluent in the basal segments, forming pneumonia-like areas of consolidation. The bronchovascular structures coursing through these areas appear intact; they are not significantly shifted, compressed, or obstructed. These findings suggest lymphoma infiltrates. There may be hilar lymphadenopathy on the right side.

- **Therapeutic options**
  Low-grade lymphoma: Watch and wait, resection, or single-modality therapy • *High-grade lymphoma:* Treatment depends on the underlying disorder, chemotherapy, modulation of immunosuppression.
- **Course and prognosis**
  Low-grade lymphoma has a good prognosis (5-year survival rate is over 80%) • High-grade lymphoma has a poor prognosis, depending on the initial situation (HIV infection, organ transplantation).
- **What does the clinician want to know?**
  Staging after diagnosis by biopsy.

Differential Diagnosis

Nodular lesions	– Bronchial neoplasm – Metastases
Areas of consolidation	– Pneumonia – Distinguished by history, clinical findings, and course
Interstitial changes	– Pulmonary interstitial disorder
Kaposi sarcoma	– Radiographically indistinguishable

Tips and Pitfalls

Because pulmonary lymphomas are rare, radiographic findings may variously be misinterpreted as pneumonia, malignancy (lung carcinoma, metastases), or pulmonary interstitial disease.

Selected References

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