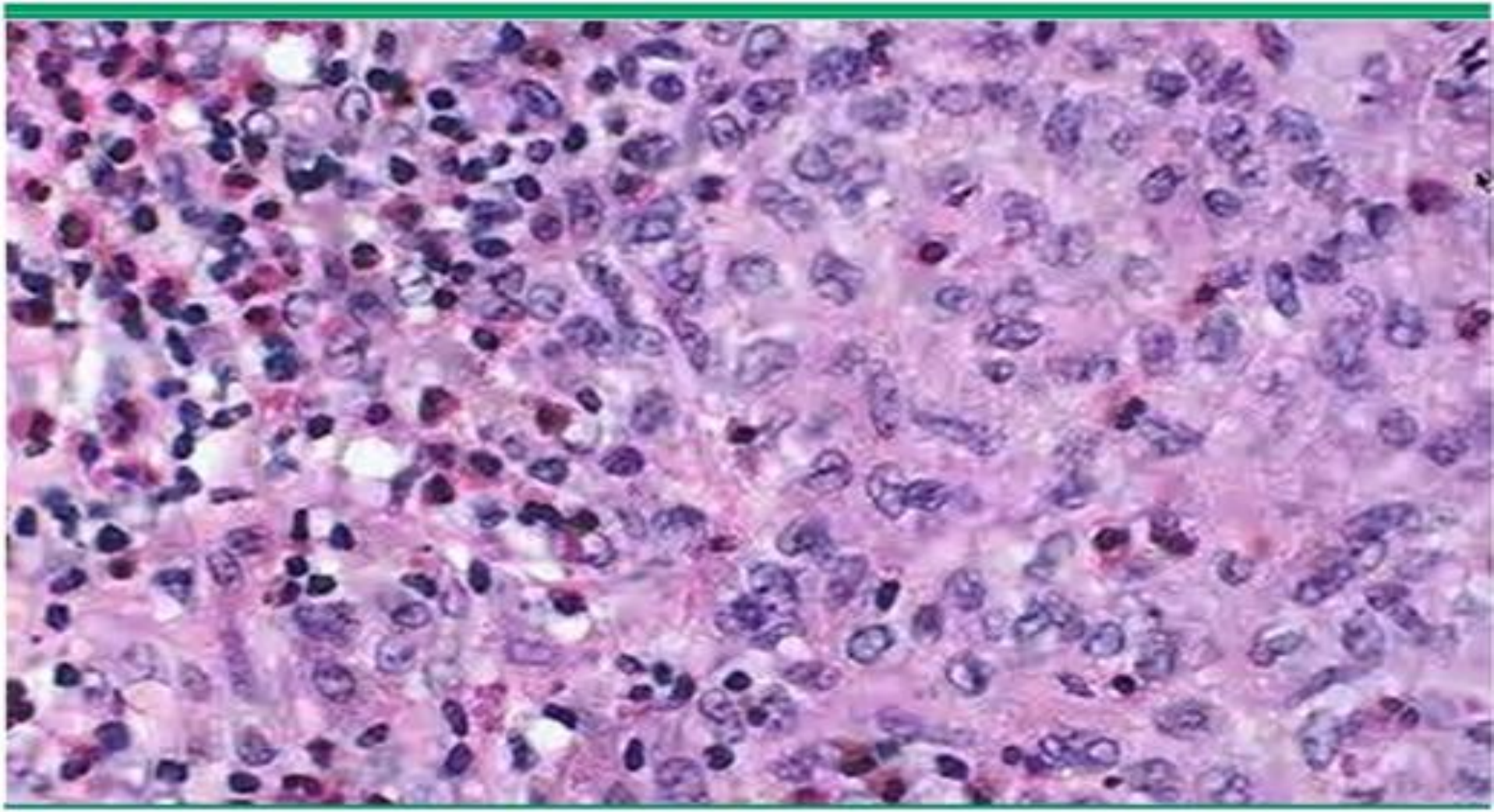
Langerhans-cell Histiocytosis
كَثْرَةُ المُنْسِجات

Histiocytosis
كَثْرَةُ المُنْسِجات

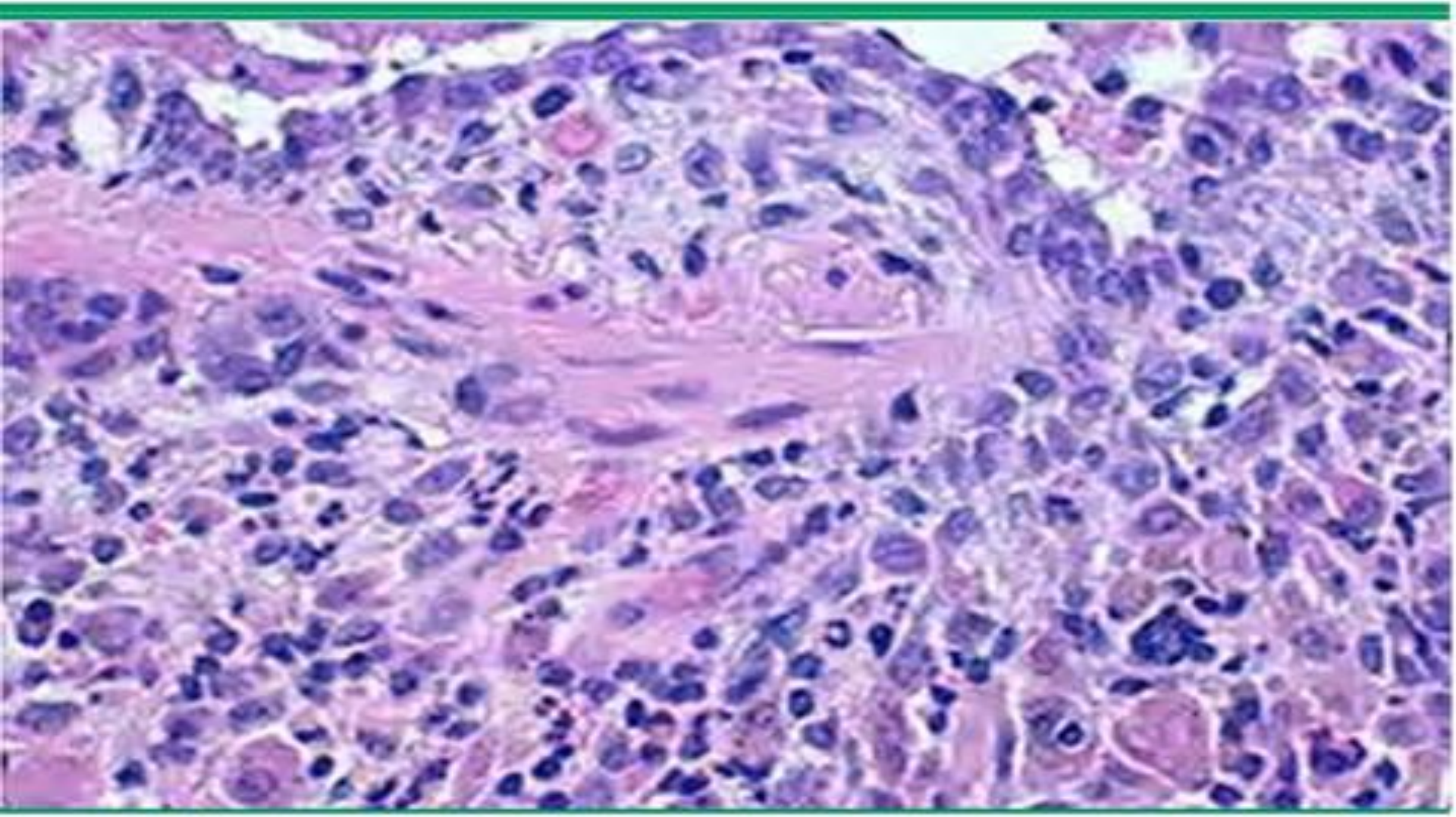
- eosinophilic granuloma, histiocytosis X
- clonal proliferation of dendritic cells
- cells have X-bodies

Pulmonary Langerhans cell histiocytosis



The cellular infiltrate of pulmonary Langerhans cell histiocytosis is characterized by Langerhans cells, eosinophils, lymphocytes, neutrophils, and alveolar macrophages.

Pulmonary Langerhans cell histiocytosis

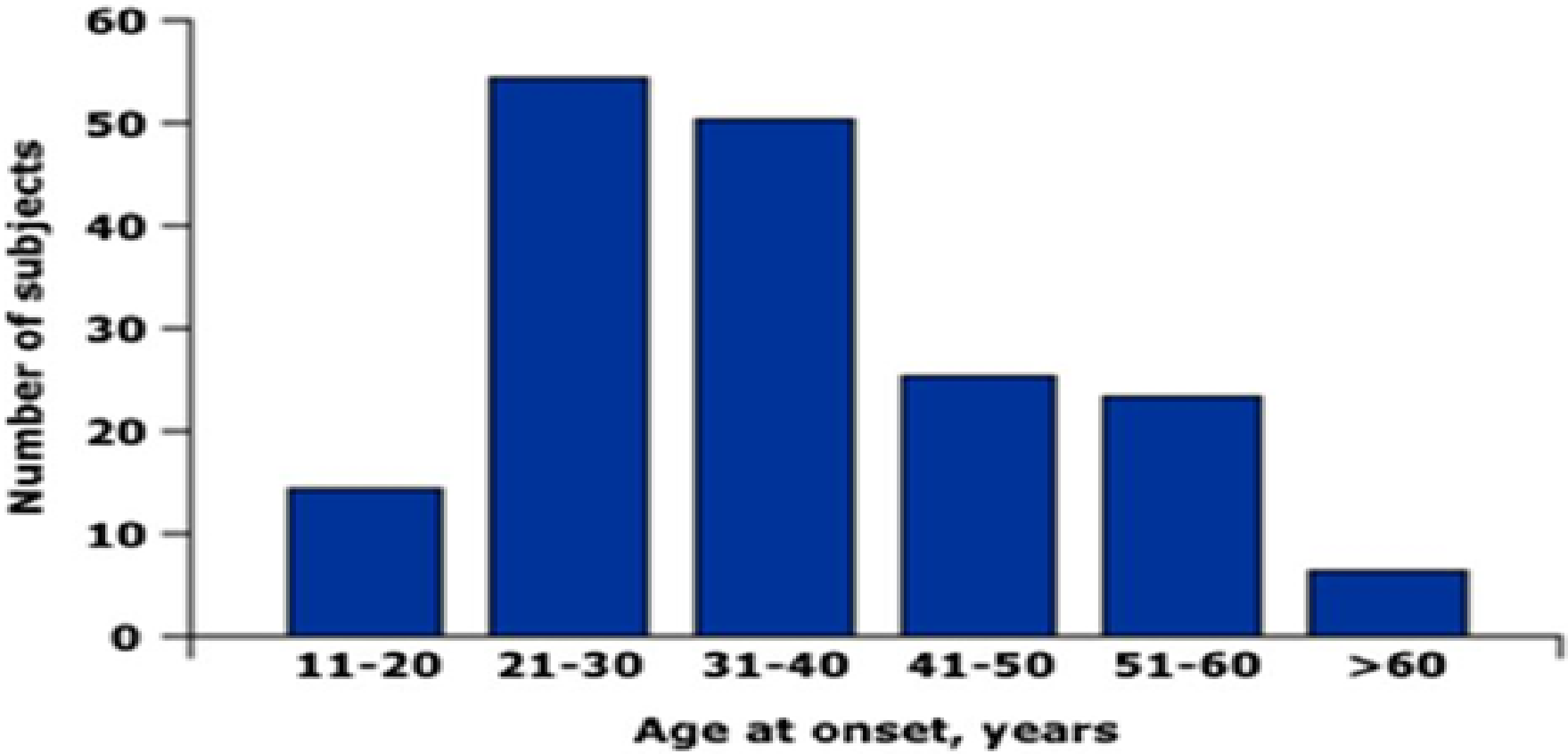


Early lesions of pulmonary Langerhans cell histiocytosis appear as cellular infiltrates along small airways.

Clinical presentation

- typically affects young-middle aged smokers
- presentation: dyspnea, cough or both
  - spontaneous pneumothorax

Age at onset of pulmonary Langerhans cell histiocytosis

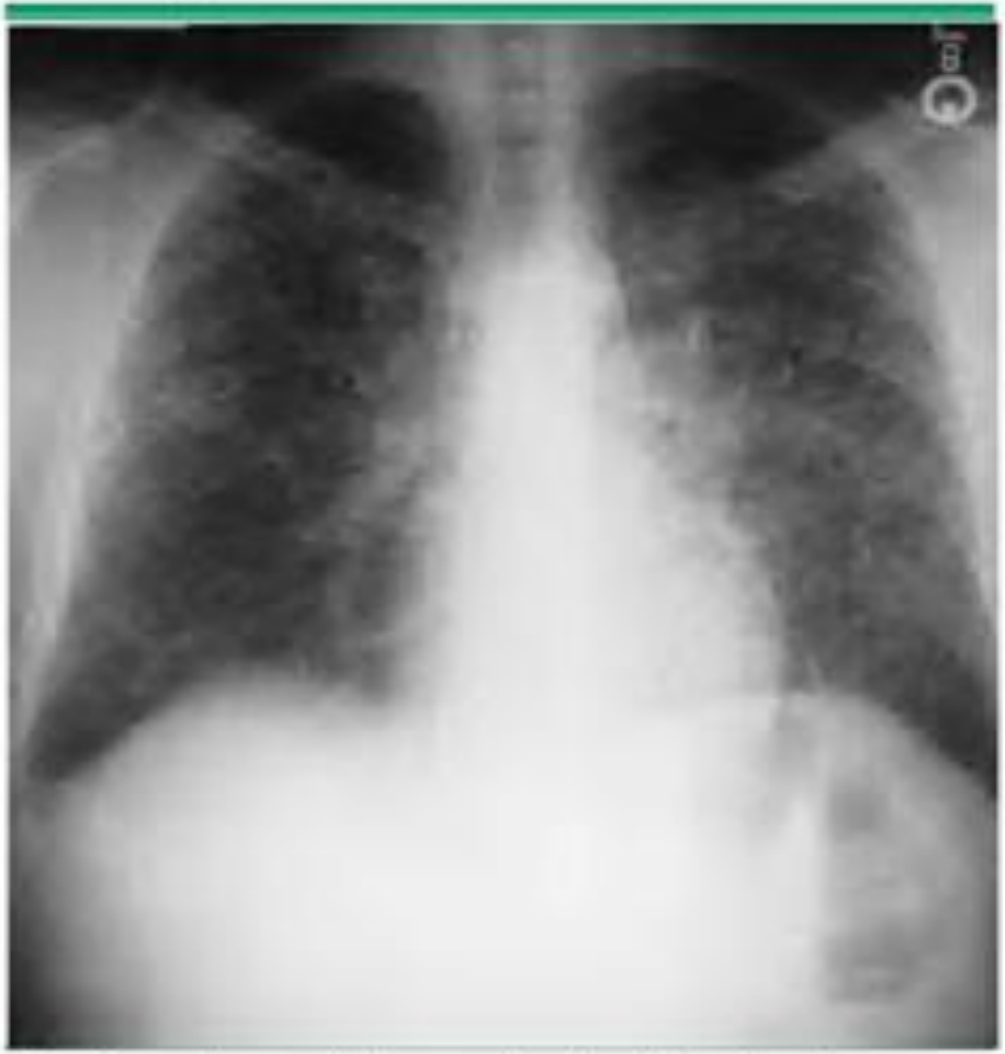


The data shown were obtained from published papers in which the age at onset of symptoms or diagnosis could be determined (total number of subjects = 168). The peak age at onset is between 20 and 40.

CXR

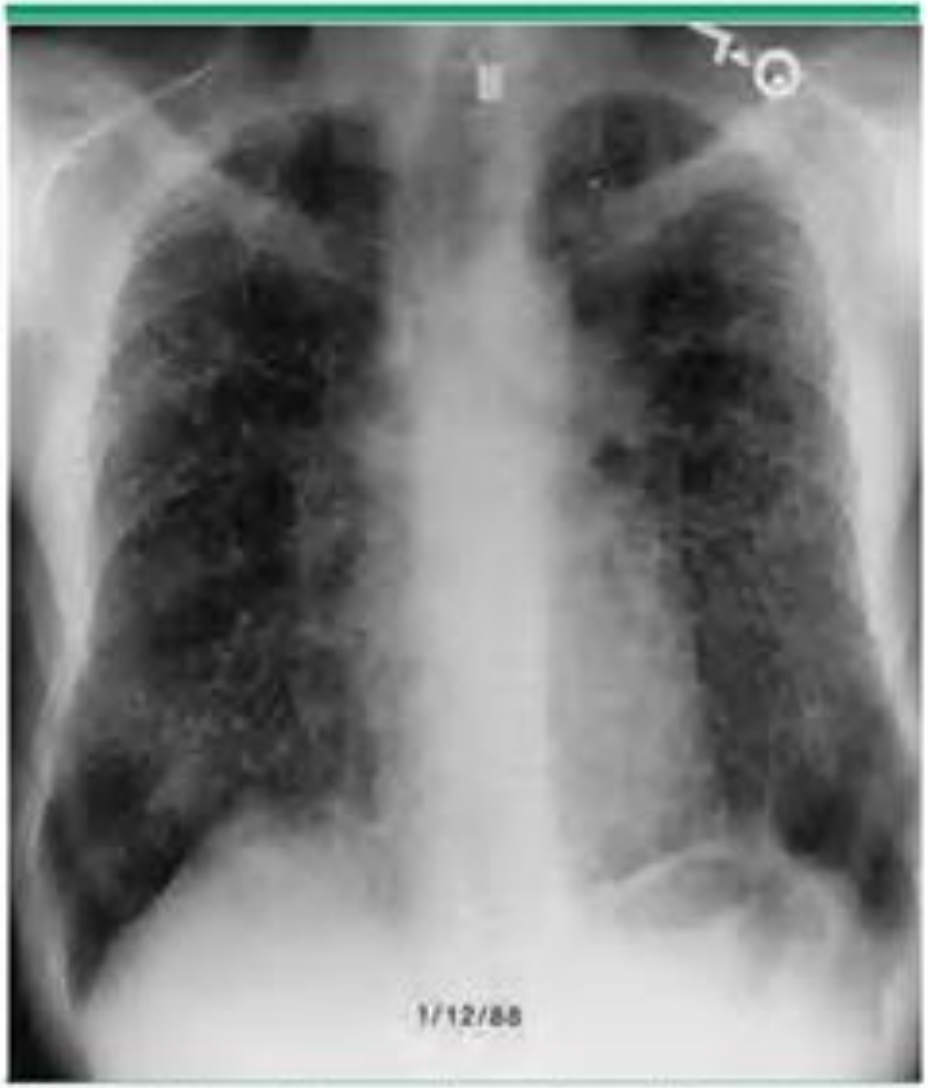
- primarily in upper lung zones :
  - cystic
  - nodular and/or reticulonodular chnges that progress to honeycombing

Pulmonary Langerhans cell histiocytosis



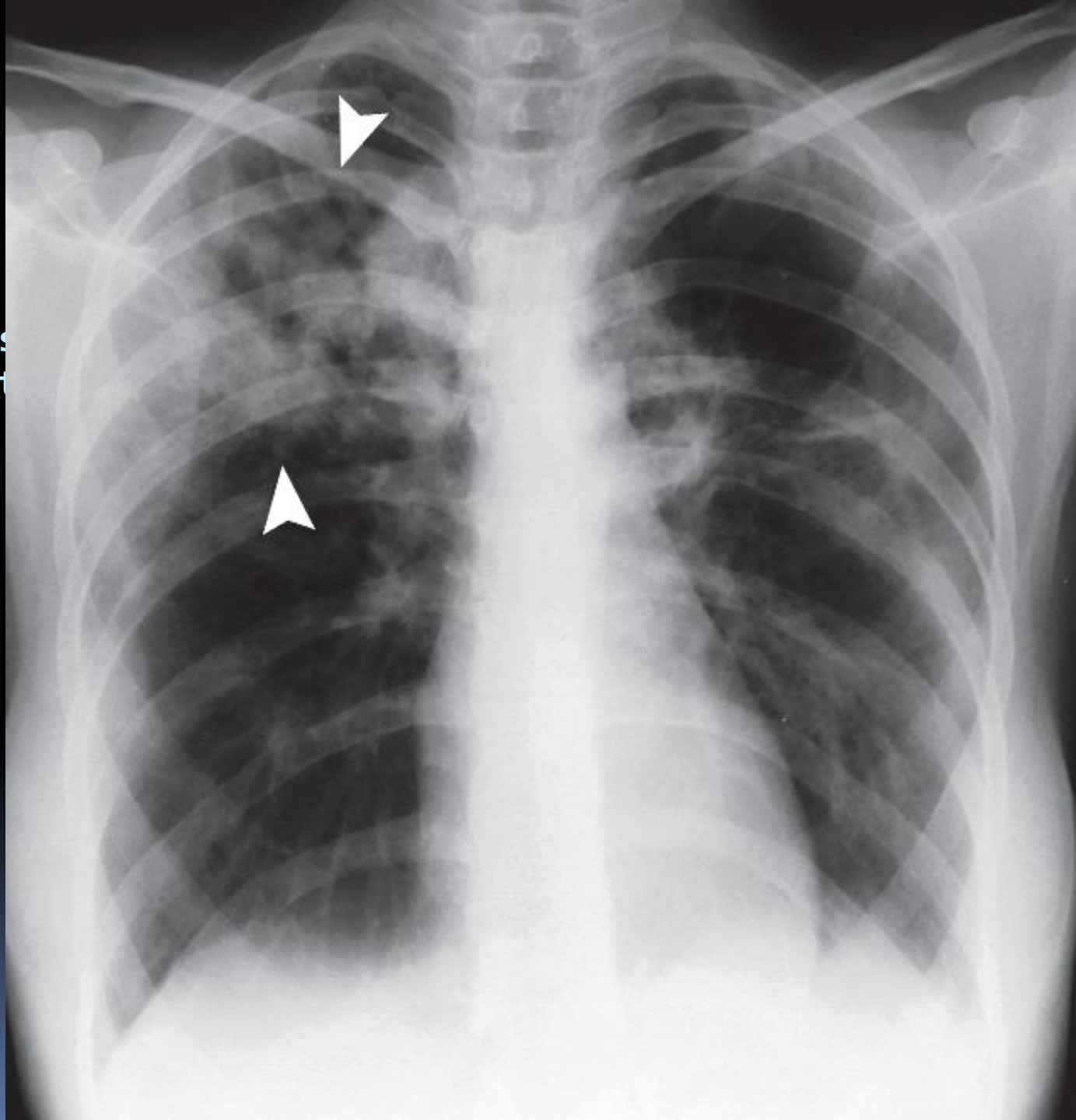
Chest radiograph demonstrates the classic features of profuse ill-defined nodules, reticulondoular opacities, cysts, and costophrenic angle sparing in pulmonary LCH. There is a mild reduction in lung volumes.

Pulmonary Langerhans cell histiocytosis

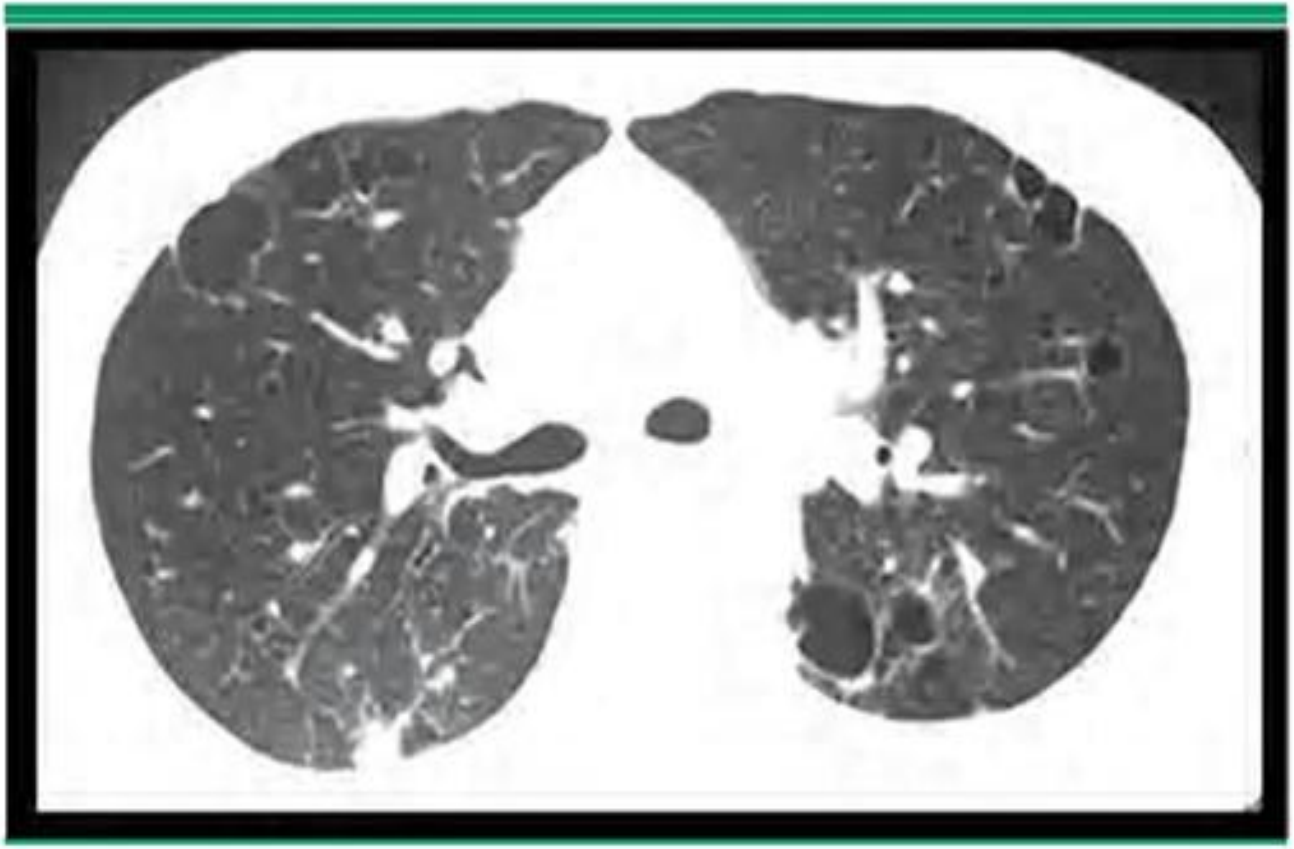


Chest radiograph reveals reticulonodular opacities in mid-lung zones, cysts, costophrenic angle sparing, and increased of lung volumes in a 58 year-old man with pulmonary LCH.

adult pulmonary Langerhans' cell histocytosis (PLCH) shows multiple pulmonary nodules with different sizes located at the right upper lung zone (*arrowheads*)

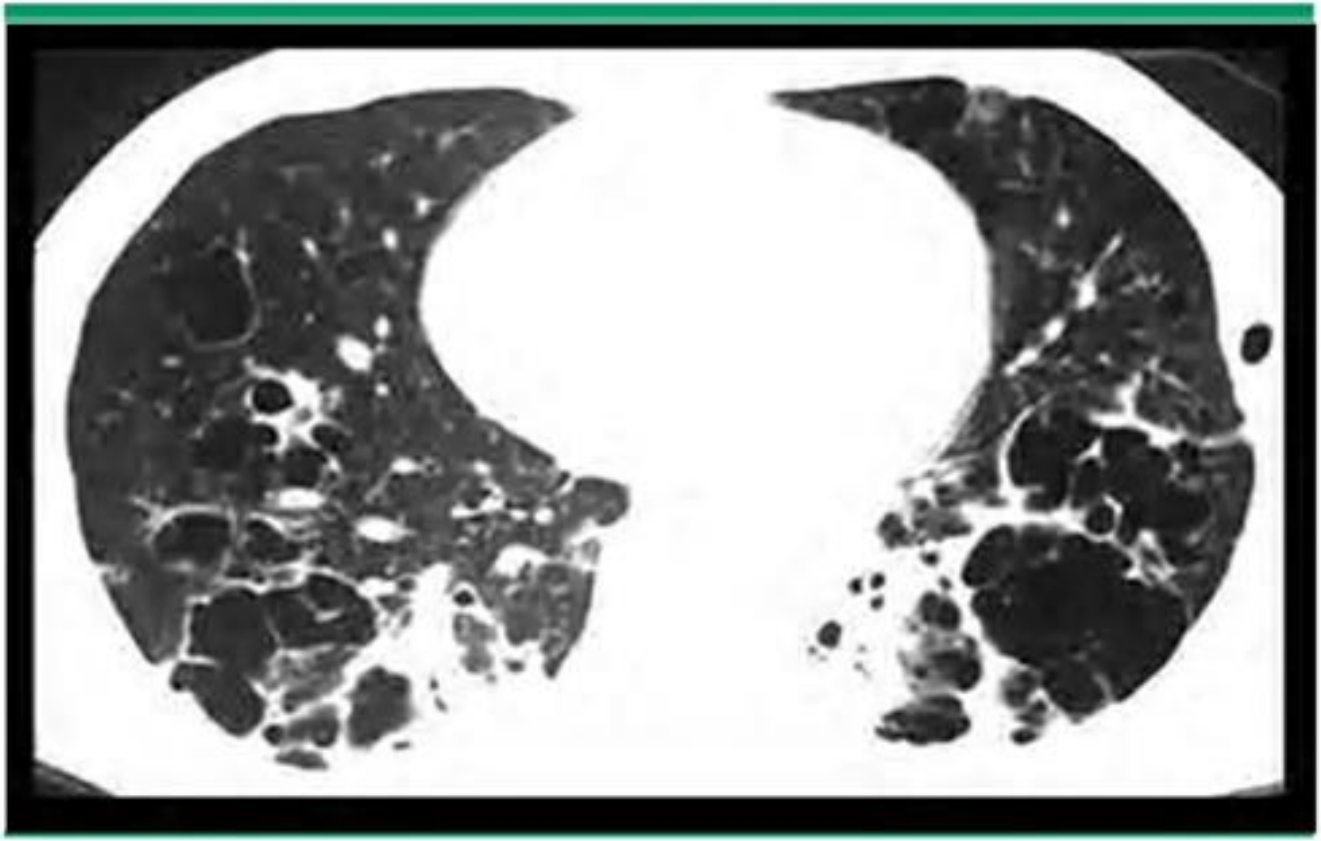


Cystic changes in Langerhans cell histiocytosis



Langerhans cell histiocytosis (eosinophilic granuloma) in an adolescent boy characterized by multiple cysts in the upper and lower lobes. The cysts are interspersed throughout the parenchyma and have a slightly thick wall. A posterior right upper lobe nodule and confluent bibasilar patchy lower lobe opacities are also part of the disease process. *Courtesy of Paul Stark, MD.*

Cystic changes in Langerhans cell histiocytosis



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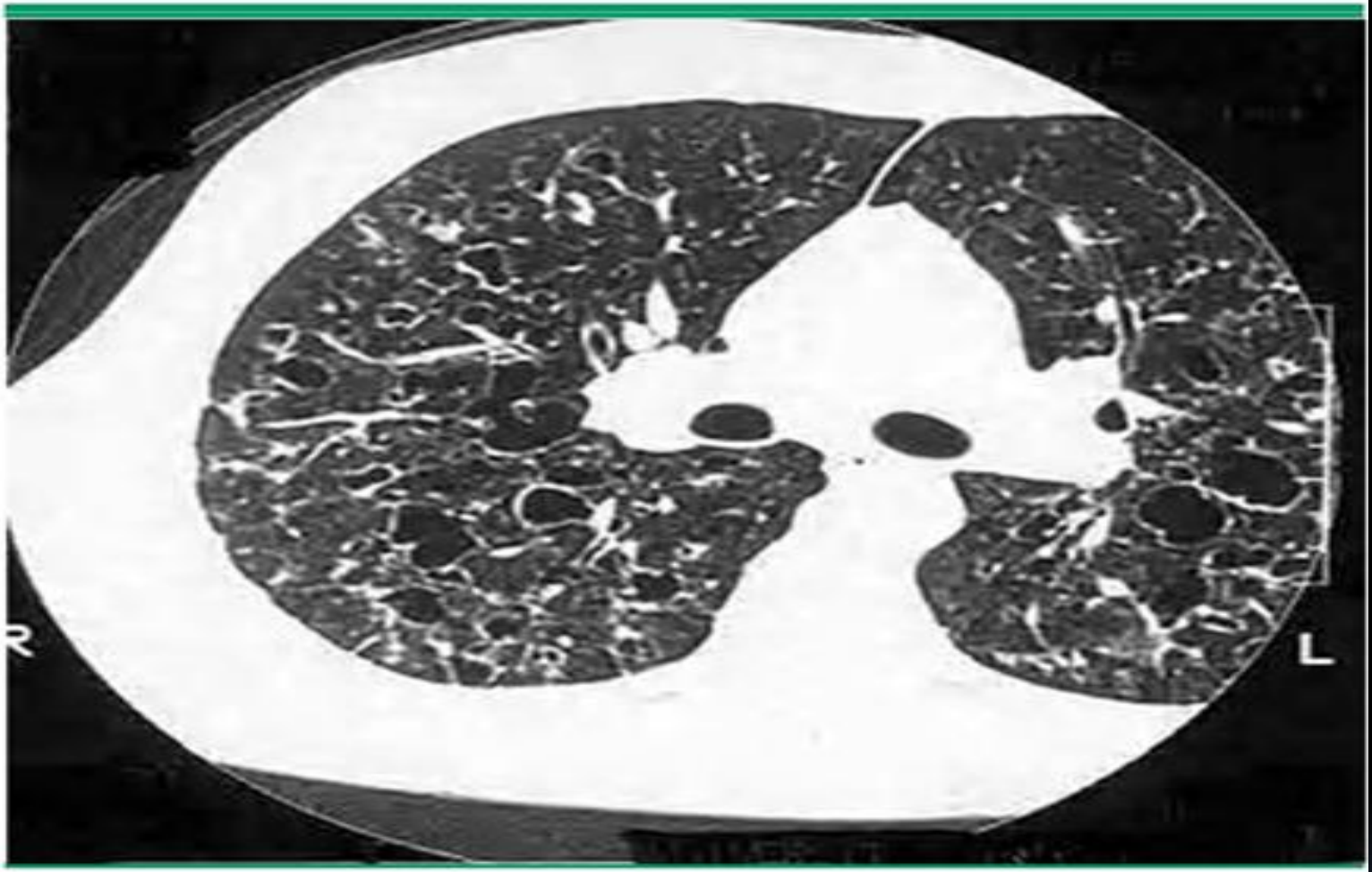
Pulmonary Langerhans cell histiocytosis



High resolution CT with thin section shows more clearly the cysts in pulmonary LCH, which vary markedly in size and may be larger than 10 mm. The cysts are bizarre in shape, many closely related to pulmonary arteries, often mimicking bronchiectasis. Few nodules are present in this case.

course

- may stabilize in some patients or may be progressive
- may progress to respiratory failure
- Smoking cessation may be followed by significant improvement
- Poor response to immunosuppressive treatment

  • no treatment