

# 18.14.5 Pulmonary Langerhans' cell histiocytosis 4

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### 18.14.5 Pulmonary Langerhans’ cell histiocytosis S. J. Bourke ESSENTIALS

Pulmonary Langerhans’ cell histiocytosis is characterized by a re-active monoclonal proliferation of activated histiocytes in the distal bronchioles. It presents with cough, breathlessness and (sometimes) systemic symptoms. Chest radiography and CT typically show nodules which then cavitate and may rupture, causing pneumothorax. Corticosteroids and/or cytotoxic drugs are of some benefit, and lung transplantation is an option for progressive disease.

Introduction Pulmonary Langerhans’ cell histiocytosis (LCH) is a rare disease characterized by a reactive monoclonal proliferation of activated histiocytes in the distal bronchioles, resulting in inflammatory nodules, cyst formation, and fibrosis. Langerhans’ cells are a particular type of histiocyte derived from dendritic cells in the bone marrow. They normally migrate in the blood to the squamous epithelium of the skin, lungs, gastrointestinal, and female genital tract, where they are involved in antigen presentation to T cells. Abnormal proliferation of histiocytes is also the pathological basis for acute disseminated LCH (Letterer-Siwe disease) and multifocal LCH (Hand-Schüller-Christian disease)—disorders which produce a spectrum of distinct clinical feature (see Chapter 22.3.9).

Epidemiology and aetiology LCH affects about one in 560 000 adults, with an equal male to female ratio and a peak age of onset between 20 and 40 years. There is a strong association with smoking, with more than 90% of patients having smoked tobacco. Patients with pulmonary LCH have abnormal T-cell proliferative responses to tobacco glycoproteins and an increased secretion of bombesin-like peptides from neuroendocrine cells in the lung. Recently mutations in the mitogen-activated protein kinase pathway (such as the BRAF V600 mutation) have been identified in about half of tissue samples of LCH. Clinical features Cough and breathlessness are the most common symptoms, and about one-third of patients have systemic symptoms such as fever or weight loss. Pneumothorax occurs in about 25% of patients and may be recurrent and sometimes bilateral. About 25% of patients have no symptoms and the diagnosis is made incidentally from a chest radiograph. In adult pulmonary LCH the clinical manifestations are usually confined to the lungs, but in 10–15% of cases lesions are also present in bone, skin, lymph nodes, and the posterior pituitary (sometimes causing diabetes insipidus).

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