

## Pulmonary Langerhans cell Histiocytosis

Pulmonary Langerhans' cell histiocytosis (pulmonary LCH) is an uncommon but important cause of interstitial lung disease, and it occurs predominantly in adult cigarette smokers [1]. Pulmonary LCH belongs to the spectrum of Langerhans' cell histiocytosis (LCH), diseases characterized by uncontrolled proliferation and infiltration of various organs by specialized dendritic cells known as Langerhans' cells [2]. Other clinical entities within this spectrum of LCH are seen in adults and children, and vary in severity from mild disease that requires limited or no therapy, to severe disseminated forms with extensive organ involvement and high mortality.

Virtually any organ system may be involved by LCH, including skin, bone, pituitary gland, lymph nodes, and lungs [1]. Although LCH is more common in children than adults, pulmonary involvement is much more common in adults with LCH, in whom it frequently occurs as the sole organ involved with disease. More than 95% of patients with pulmonary LCH smoke cigarettes, suggesting a role for smoking as a trigger that induces this disease in susceptible individuals [1].

The clinical presentation is often with non-specific symptoms, particularly cough and dyspnea [1]. Up to a third of patients may be asymptomatic, while around 20% will have constitutional symptoms (weight loss, low-grade fevers, sweats, and fatigue) [1]. Definitive diagnosis requires lung biopsy, but a confident provisional diagnosis of pulmonary LCH may be made based on high resolution chest CT (HRCT) and bronchoscopic findings. The differential diagnosis includes other causes of cystic lung disease (including lymphangiomyomatosis), other diseases that preferentially infiltrate the upper lung fields (including sarcoidosis, hypersensitivity pneumonitis, silicosis, and histoplasmosis), and other diseases that cause nodular or diffuse lung infiltrates (including malignancy, vasculitic conditions like Wegner's granulomatosis, etc). PET may be helpful to define the extent and activity of disease in some patients, particularly if there are significant constitutional symptoms [3].

In every patient with pulmonary LCH, a careful physical examination and appropriate diagnostic studies should be undertaken to rule out other organ involvement. The treatment of pulmonary LCH should include absolute smoking cessation for all smokers. All patients should be screened for the presence of pulmonary hypertension with echocardiography, as a significant proportion will develop pulmonary hypertension, the severity of which does not necessarily correlate with lung volumes or spirometry [4]. Patients with pulmonary hypertension may respond to vasodilator therapy (sildenafil and bosentan). Additional complications that occur throughout the disease course include recurrent pneumothoraces and a probable increase in secondary malignancies [1]. In some patients with progressive disease, therapy with corticosteroids or chemotherapy may be needed to induce disease remission or stability. In selected patients, chemotherapy with cladribine may induce complete remission of disease [5, 6]. Pneumothoraces may require treatment with surgical pleurodesis to prevent recurrence. Lung transplantation is indicated in patients with progressive disease and substantial impairment in lung function.

### References:

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