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Muhammad Irfan
Aga Khan University

Javed Hussain
Aga Khan University

Riaz Lakdawala
Aga Khan University

Fayyaz Hussain

Javaid Khan
Aga Khan University

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PULMONARY LANGERHANS CELL HISTIOCYTOSIS

*Muhammad Irfan, **Javed Hussain , ***Riaz Lakdawala,
****S. Fayyaz Hussain , ***** Javaid Khan

Introduction:

Langerhans cell histiocytosis (LCH), also known as Langerhans cell granulomatosis, eosinophilic granuloma and histiocytosis X, is a disease characterized by an abnormal proliferation of Langerhans cells in various tissues.⁽¹⁾ Depending on the extent of organ involvement, LCHs are classified as LCH with single-organ involvement (eg, PLCH), LCH with multiorgan involvement (as in Hand-Schüller-Christian disease), or multisystem disease (formerly Letterer-Siwe disease, seen rarely in adults).⁽¹⁾ Most patients are cigarette smokers in their third and fourth decades of life. Men are more commonly affected than women, by a ratio of 1.5:1. Cystic bone lesions are present in 4 to 20 percent of patients.

Here we describe a case of Pulmonary langerhans cell histiocytosis (PLCH) along with bone involvement

Case Report:

A 30 years old engineer, heavy smoker, presented in a surgical clinic with 1 month history of pain in right hip. He was a known case of diabetes mellitus. X-ray pelvis showed a bony lesion at upper right femoral shaft. Excision curettage of the lesion was done and histopathology showed cellular infiltrates composing of cells with grooved nuclei along with multinucleated cells. Immunohistochemical stains were positive for S100 and CD68 stains and negative for leucocyte markers. The features were consistent with LCH. Three months later he again came in orthopedic clinic with 2 months history of increasing backache. MRI of thoracic spine showed compression collapse of D8 vertebral body (fig 1). Thoracotomy, D8 compectomy and tricortical reconstruction was done. Four months later he presented at a pulmonology clinic with dry cough and exertional dyspnea. He was hemodynamically stable at that point and his general physical examination was unremarkable. Chest examination was also normal.

Chest radiograph showed diffuse reticulonodular shadowing in both lung fields (Fig 2). C.T chest revealed diffuse, ill defined nodular infiltrations and cystic lesions in both lung fields with sparing of

costophrenic angles (Fig 3). There was no hilar or mediastinal lymphadenopathy. The CT scan findings were very much suggestive of PLCH. Spirometry revealed moderate restrictive impairment. He was diagnosed as a case of PLCH and advised to quit smoking. On follow up after 4 months he showed significant improvement in his symptoms.

Discussion:

LC histiocytoses (LCH) are disorders characterized by disease manifestations that are explained by excess of activated Langerhan cells (LCs) in various tissues. Between 4% and 20% of patients with PLCH display cystic bone lesions, and a number of case reports demonstrate involvement of other sites, including mediastinal lymph nodes, pituitary gland, skin, gut, heart, and brain.⁽²⁾ Bony lesions may present before the respiratory symptoms, as in our case.

The diagnostic feature of PLCH is proliferation of LCs. The LCs is a specialized immune cell belonging to the family of dendritic cells that form a network of antigen-presenting and migratory cells in lymphoid and nonlymphoid organs such as the skin, heart, and lung.⁽²⁾ These modified macrophages are immunoreactive for S-100 protein and CD1a and contain Birbeck granules at an ultrastructural level.⁽³⁾

The etiology and pathogenesis of Langerhans cell histiocytosis are poorly understood. The majority of patients with LCH are smokers who commonly acquire disease in the third or fourth decade of life.^(4,5) It has given rise to the hypothesis that LCs accumulation in PLCH occurs as a response to cigarette smoke. The finding of increased numbers of LCs in the BAL fluid of smokers,⁽⁶⁾ along with abnormal T-cell proliferative responses to tobacco glycoprotein in patients with PLCH,⁽⁷⁾ lends support to this hypothesis. Our patient was also a middle aged heavy smoker.

The clinical presentation of PLCH varies; dyspnea, cough, and chest pain are predominant symptoms.⁽⁴⁾ Constitutional symptoms and hemoptysis can occur. The physical examination is quite notably unremarkable as in our patient. An abnormal chest radiographic finding may be the only clue to the disease in the 25% of asymptomatic patients with PLCH.⁽⁸⁾ Pneumothorax with chest pain is the initial clinical manifestation in 15% of patients with PLCH.⁽⁹⁾ Pneumothoraces may be recurrent, requiring thorocotomy and chest tube placement for relief of symptoms. The reticulonodular changes predominate in the mid-lung zones with some radiographic series reporting more upper-lobe involvement. Significant lower-lobe involvement has been reported as well, but is less common.⁽⁹⁾ The costophrenic angles are generally spared^(8,9)

Chest CT findings vary depending on the stage of the disease. Cysts (80%) and/or nodules (60 to 80%) are present in the majority of patients.⁽¹⁰⁾ Cystic changes appear later and vary in shape, size, and wall thickness. The CT appearance of cysts and nodules in an adult heavy smoker is virtually diagnostic of PLCH.⁽¹¹⁾ The high diagnostic accuracy of HRCT in PLCH has resulted in fewer invasive pathologic studies, especially since there is no current specific treatment modality for disease.⁽¹²⁾

Smoking cessation is recommended because of a potential pathogenetic association, and documented resolution of disease occurs following cessation of smoking.⁽¹³⁾ Corticosteroids and other immunosuppressive therapies have not been evaluated in well-designed clinical trials. Lung transplantation can be performed in patients with advanced progressive disease. Recurrence of the condition in the transplanted lung may occur.

Poor outcome in PLCH has been associated with an older age at diagnosis, increasingly severe airway obstruction (lower FEV₁/FVC ratio and higher residual volume/total lung capacity ratio), reduced carbon monoxide diffusion capacity, and use of steroid therapy during follow-up.⁽¹⁴⁾

In conclusion PLCH is rare interstitial lung disease of young to middle aged smokers. Avoidance of smoking may help in prevention of this rare disease.

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