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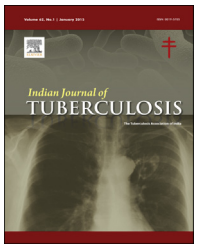

Case Report

Rare interstitial lung disease: Pulmonary Langerhans Cell Histiocytosis in a young non smoking Indian female

Lalita Fernandes [a,*], *Rohit Vadala* [b], *Anthony Menezes Mesquita* [c], *Pradeep Vaideeswar* [d]

[a] *Professor, Department of Pulmonary Medicine, Goa Medical College, India*
[b] *Junior Resident, Department of Pulmonary Medicine, Goa Medical College, India*
[c] *Director-Professor & HOD, Department of Pulmonary Medicine, Goa Medical College, India*
[d] *Professor (Additional), Department of Pathology, Seth GS Medical College, Mumbai, India*

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ABSTRACT

Adult Pulmonary Langerhans Cell Histiocytosis (PLCH) is a rare interstitial lung disease which occurs almost exclusively in smokers. A marked male predominance was initially reported, but recent studies show both men and women are equally affected due to the increasing smoking habits in women. The natural history is variable with 25% of patients having asymptomatic disease while 10–20% progress rapidly to respiratory insufficiency and death. The diagnosis is not easily recognized by clinicians or pathologists. Awareness of the clinical presentation and classical HRCT findings helps in early diagnosis and management of this disease. We report a rare case of severe PLCH in a young non smoking female with a short history who progressed rapidly to respiratory failure and died.

1. Introduction

Pulmonary Langerhans Cell Histiocytosis is an isolated form of LCH and is characterized by peribronchiolar nodular proliferation of Langerhans' cells, which gradually progresses to fibrosis with formation of irregular cysts. This results in classical High Resolution Computed Tomography (HRCT) findings that differentiates it from other interstitial lung diseases. Smoking is a risk factor. Management includes smoking cessation, corticosteroid therapy or immunosuppressive therapy. Outcome is variable with some showing spontaneous remission while a subgroup of patients develop severe disease leading to respiratory failure and death. Severe PLCH is rare[1] and hence this case is being reported.

2. Case report

A 26 years old married female with two children and housewife by occupation presented with history of occasional dry

* *Corresponding author*. TB and Chest Diseases Hospital, St. Inez, P.O. Caranzalem, Goa 403002, India. Tel.: +91 (0)9822102177 (mobile), +91 (0)8322225088 (office); fax: +91 (0)8322425007.
E-mail address: drlalitafernandes@gmail.com (L. Fernandes).
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cough, low grade fever, breathlessness, easy fatigability and loss of appetite for two weeks. There was no associated hemoptysis, chest pain, joint pain, or skin rash. There was neither a past history of tuberculosis or contact with tuberculosis nor any other relevant medical history. She was a non - smoker and non - alcoholic and she was not exposed to environmental tobacco smoke. She gave a history of using biomass fuel for cooking (chulha smoke exposure) for the last eight years with 40 hour years of exposure.

Chest X-ray showed bilateral reticulo-nodular shadows (Fig. 1). Sputum for acid fast bacilli (AFB) and Tuberculin Test were negative. She was started on DOTS Category 1 (Isoniazid, Ethambutol, Pyrazinamide and Rifampicin) as a case of Miliary Tuberculosis at her primary health centre.

Despite seven doses of anti-tubercular therapy, patient continued to be breathless with development of swelling of feet. Chest X-ray taken during this episode showed increase in diffuse reticulo-nodular shadows in the lung and was admitted for further investigations.

On physical examination, she was thinly built, afebrile with pulse rate of 108/minute, respiratory rate of 28/minute and blood pressure of 108/70 mmHg. She had facial puffiness, increased jugular venous pressure, cyanosis and bilateral edema of feet. She had no evidence of lymphadenopathy or clubbing. On systemic examination of respiratory system, crackles were heard in bilateral bases. Investigations done at admission showed hemoglobin of 10.9 gm%, total leukocyte count of 12400/cmm with neutrophil predominance, normal coagulation profile and normal renal and liver functions tests. Anti-nuclear antibodies and rheumatoid factor were negative. C-reactive protein was positive. Assays for human immunodeficiency virus1 and 2 antibodies were negative. Venous Doppler study of both lower limbs was negative for deep vein thrombosis.

HRCT thorax done showed multiple thick walled irregular cysts, multiple centrilobular nodules and peribronchial cuffing. Thick walled cysts were predominantly noted in upper and mid zones while the lower zone showed cysts and nodular distribution involving the costophrenic regions. There was no mediastinal lymphadenopathy or any pleural effusion (Fig. 2). In view of HRCT findings, a provisional diagnosis of LAM (Lymphangioleiomyomatosis) and PLCH was considered. Abdominal ultrasound was normal and renal ultrasound did not reveal any angiomyolipoma.

On the 5th day of admission, the patient developed spontaneous left-sided pneumothorax followed by right-sided pneumothorax (see Fig. 3)

CT guided lung biopsy done following left intercostal tube insertion for left spontaneous pneumothorax showed epithelioid cells, necrosis and acute inflammatory cells. She was treated with nasal oxygen and talc pleurodesis. However patient progressively deteriorated and died. Post mortem lung biopsy revealed presence of alveoli showing cystic dilatation. The interstitium showed presence of collection of lymphocytes, plasma cells, eosinophils and histiocytes. The histiocytes showed presence of moderate amount of cytoplasm and vesicular nucleus with nuclear grooving (See Fig. 4). Immunohistochemistry markers was positive for CD1a (See Fig. 5). The diagnosis of PLCH was thus confirmed.

3. Discussion

Langerhans cell was first described by Paul Langerhans in 1868. It is a bone marrow derived dendritic cell found in the skin and lung whose normal function is processing and presentation of antigens. The disease Langerhans Cell Histiocytosis results from clonal accumulation and proliferation of langerhans cell in bone, lungs, skin, lymphnodes and pituitary gland due to unknown reasons.

It is predominantly a disease of childhood and is rarely seen in adults. The Histiocyte Society founded in 1985 classified LCH into single system LCH involving one organ or system and multisystem LCH involving two or more organs or system. Acute disseminated LCH (Letterer-Siwe disease) is a severe multisystem disease affecting mostly young children with poor prognosis. Multifocal LCH seen in older children and adolescent (Hand-Schuller- Christian syndrome) almost

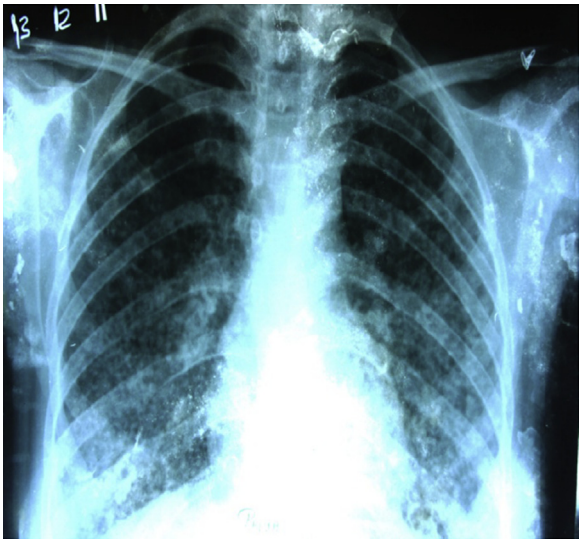

Fig. 1 – Chest X-ray showing bilateral reticulo-nodular shadows.

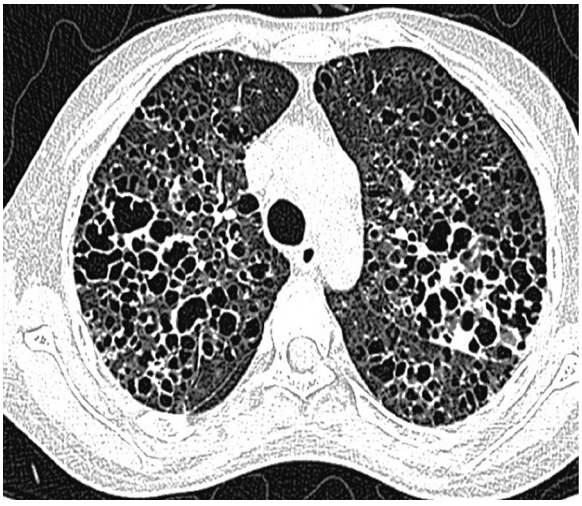

Fig. 2 – HRCT Thorax showing bilateral multiple thick walled irregular cysts.