AN UNUSUAL CASE OF SEVERE BREATHLESSNESS IN A NINE-YEAR-OLD PATIENT PRESENTING TO A TERTIARY CARE CENTRE

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ABSTRACT

BACKGROUND
A 9-year-old boy from rural area in Andhra Pradesh came to the tertiary care centre with symptoms of predominant breathlessness since childhood which aggravated during the last 10 days. It was associated with cough with expectoration and high grade fever. Patient was given antituberculosis therapy with no benefit for two months before coming to the tertiary care centre. Patient was acutely breathless with tachypnoea and tachycardia. There was chest wall retraction of the anterior chest with each respiration. There was no wheeze on examination. Sputum for AFB was negative. Patient was negative for HIV and HBsAg. CT scan chest was done and was suggestive of interstitial lung disease with lower zone sparing. Open lung biopsy was done and was suggestive of Langerhans’-cell histiocytosis and biopsy specimen was positive for S100 protein.

KEYWORDS
Langerhans’-cell histiocytosis, Sputum for AFB, Interstitial Lung disease, Open Lung Biopsy, Bronchoalveolar Lavage.


BACKGROUND
Case Report
A 9-year-old male patient from a rural background came with a chief complaint of fever since 10 days, breathlessness since 10 days, cough with expectoration since 4 days, tightness of chest since 4 days. Started as a fever since 10 days, high grade, associated with chills and rigors. Breathlessness since 4 days, initially grade 3 progressed to grade 4. Cough with expectoration since 4 days, sputum was yellowish in colour. Moderate in quantity and not foul smelling. Patient had no history of contact with Tuberculosis. This boy was the eldest child of two and other sibling was normal. There was no history of consanguineous marriage of parents. Frequent absence from school was reported by parents because of sickness. Patient was on a mixed diet. Bowel and bladder habits were regular.

On examination, patient was thin built, anaemic, clubbing+, no cyanosis, no peripheral lymphadenopathy and no dependent oedema. PR: 145/min. BP: 100/60 mmHg. Temp: 101 degrees F. SpO2: 70%. RR: 52/min.

Examination of Respiratory System

Upper Respiratory Tract
ENT: Ears- There was no discharge. Nose- Normal oral cavity, poor oral hygiene. Oropharyngeal lumen- Class I Mallampati.

Patient was extremely breathless with inward movement of anterior part of the chest with each inspiration.

Lower Respiratory Tract
Patient had tachypnoea and tachycardia, tracheal deviation to right. Respiratory movements were equal on both sides. Bilateral rhonchi and coarse crepitations were present on both sides.

Cardiovascular system, central nervous system and abdomen were within normal limits.

Chest X-Ray PA suggestive of bilateral pulmonary infiltrates and cavitary and fibrotic lesions.

Hb 7 g%. Total Count 8900. Differential count: Polymorphs 52%, Lymphocytes 47%, Eosinophils 1%.

Biochemistry
Blood Sugar 92 mg%, Urea 33 mg%, serum creatinine 1 mg%, serum bilirubin 0.8 mg%.

HIV test negative, HBsAg Negative, Sputum for AFB was negative. Widal test and Peripheral smear for Malarial Parasites were negative.

Sputum culture and sensitivity: Streptococcus pneumonia sensitive to Clavulanate Amoxicillin, Azithromycin, Cephalexin.

Figure 1. X-ray of the Patient at the time of Admission

Echocardiography

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Normal cardiac chambers and valves. No LV regional wall motion abnormality. Mild tricuspid regurgitation with moderate pulmonary arterial hypertension. No mitral regurgitation or aortic regurgitation. Good LV and RV Function. Septae were intact.

ABG: Showed pH 7.43, pCO2 32 mmHg, pO2 45 mmHg, HC03 26 mEq/L.

CT Chest Report
Diffuse lung disease bilaterally with ground glass pattern, mosaic pattern, cystic bronchiectasis and lung cysts predominantly in upper lobes, minimal pneumomediastinum and calcification nodes suggestive of interstitial lung disease, ?pneumocystis carinii pneumonia, ?Langerhans’-cell histiocytosis, ?cystic fibrosis, ?tuberculosis.

Bronchoscopy and Pulmonary function tests could not be done because of poor general condition of the patient.

Treatment
O2 supplementation with Venturi mask, Head end elevation.

Fever subsided with antibiotic therapy and expectoration decreased.

Open lung biopsy was done from right middle lobe by thoracic surgeon. Intercostal tube drainage was done because of pneumothorax. Inj. Cefotaxime and Inj. Metronidazole were started after biopsy.

Patient was intubated and kept on pressure control ventilation mode. Patient developed empyema on the right side of the chest. He was put on Inj. Imipenem 500 mg IV q.i.d. Inj. Linezolid 600 mg IV b.i.d. and Amikacin. He was extubated after 4 days. ICT was removed. Pleural pus for c/s was sent. Pseudomonas species was isolated after 24 hours of aerobic incubation.

Open Lung Biopsy Report: Wet preparation with toluidine blue staining revealed plenty of monomorphic cells with abundant granular cytoplasm and round to oval nuclei. Some resembled plasma cells. Occasional multinucleated giant cells were seen.

Histopathological sections from different areas of the biopsy specimen reveal immature lung parenchyma with lobulations. There was dense fibrocollagenous tissue separating the lobules. The alveoli showed variable patency. Some areas were totally atelectatic with monomorphic cellular exudates filling the narrow air spaces while others were partially open with prominent type I and type II pneumocytes. A few foci showed dilated air spaces with flattened lining epithelium. Interstitium was prominent with increased fibrocollagenous tissue and inflammatory infiltrates. Most of the inflammatory cells were lymphocytes. Lymphoid aggregates were present along with one focus of non-caseating granuloma (sarcoid like) seen in the interstitium. There was diffuse histiocytic infiltration in the interstitium and also in the alveolar spaces.

The histological features were suggestive of interstitial lung disease. It was chronic because of the nature of inflammatory cell infiltrate and fibrosis of alveolar walls.

The possible diagnoses considered were: 1. Congenital cystic adenomatoid malformation of the lung. 2. Cystic fibrosis. 3. Langerhans’-cell histiocytosis or sarcoidosis cannot be excluded with the histopathology. Biopsy specimen was positive for S-100 protein suggestive of Langerhans’-cell histiocytosis.
Differential Diagnoses

- Pulmonary Langerhans’-cell histiocytosis.
- Interstitial lung disease.
- Granulomatous lung disease.
- Congenital cystic adenomatoid malformations.
- Diagnosis of Langerhans’-cell histiocytosis was arrived at after consultation with the pathologist.

Course

The patient developed pneumothorax and empyema but responded to antibiotic therapy and was discharged with expansion of lung. But breathlessness continued unabated. There was no fever or constitutional symptoms of infection at the time of discharge. Patient was given oxygen support at home but breathlessness continued and the patient expired one month after discharge from the hospital.

Pulmonary Langerhans’-cell histiocytosis is a rare interstitial disease of the lung. It usually occurs in 20-40 years of age group among smokers. Pulmonary Langerhans’-cell histiocytosis is also called eosinophilic granuloma of the lung or pulmonary Langerhans’-cell granulomatosis. Letterer-Siwe disease and Hand-Schuller-Christian disease and PLCH are together grouped under Histiocytosis X- characterised by abnormal organ infiltration by Langerhans’ cells. Langerhans’ cells are highly differentiated cells in the monocyte-macrophage line that are also found in the dermis of the skin, the reticuloendothelial system, the pleura, and the lung. Letterer-Siwe disease is an acute, fulminant disease of children less than 2 years of age that is characterised by widespread infiltration of the reticuloendothelial system, bones, and lungs. Hand-Schuller-Christian disease is a more indolent disorder of children and young adults that involves bones and the lungs. Diabetes insipidus, exophthalmos, and osteolytic skull lesions form the classic clinical triad associated with this disorder.

PLCH patients are diagnosed incidentally with a screening chest radiograph, after pneumothorax, respiratory or constitutional symptoms. Symptoms usually include non-productive cough, dyspnoea, chest pain, fatigue, weight loss and fever. Pulmonary hypertension and cor pulmonale can occur. Peripheral eosinophilia may not be present.

Radiological Presentation: Nodular lesions, reticular opacities, upper-zone cysts or honeycombing, preservation of lung volume, and costophrenic angle sparing are highly specific for this disorder. Reticular or nodular opacities are seen in the middle to upper zone with relative sparing of lower zones. Normal, obstructive or restrictive pattern can be seen in pulmonary functions.

The pathological cell type of pulmonary Langerhans’-cell histiocytosis is the Langerhans’ cell, a differentiated cell of the monocyte-macrophage line. They are distinguished by a pale-staining cytoplasm and large nucleus and nucleoli. Electron microscopy can demonstrate the classic pentilaminar cytoplasmic inclusion or Birbeck granule (X-body). Cigarette smoking is causative in stimulating the production of monocyte macrophage cells through GM-CSF and development of Langerhans’ cells, resulting in formation of inflammatory nodules and fibrosis.

Diagnosis is by identification of Langerhans’ cell in bronchoalveolar lavage fluid characteristic staining for S-100 protein or peanut agglutination antigen. Treatment and Prognosis: Patient can improve spontaneously or can develop end stage lung fibrosis. Smoking cessation, corticosteroid therapy or cytotoxic therapy, Interleukin-2 and anti–tumour necrosis factor-α, radiotherapy, lung transplantation are tried.

DISCUSSION

LCH usually occurs in smokers in 20-40 years age group. Our patient is a 9-year-old non-smoker. Interstitial lung disorders are heterogeneous and present diagnostic dilemma. LCH can involve lung, bone and other organs. Histiocytosis X of bone can involve skull or can be polyostotic. Can be associated with Diabetes insipidus, thrombocytopenia or hepatosplenomegaly. Can occur in patients of 2 months to 71 years of age. Summation of clinical, radiologic, and pathologic findings are necessary for children’s interstitial Lung disorders.

Dayananda et al noted LCH in a 14-year-old, similar to our case. LCH must be distinguished from lymphangioleiomatosis which usually occurs in females and pleural effusions are more common. Pavan Kumar et al presented an LCH case with a mandibular swelling. Isolated pulmonary lesions were presented by authors with pulmonary nodular lesions. Proper diagnosis is possible only with demonstration of Langerhans’ cells by biopsy or S-100 proteins.

LCH is usually underdiagnosed. Sometimes all the modalities of radiology, BAL and immunohistochernistry are needed. Uday Bande reported it in a 27-year-old male smoker, diagnosed by histopathological examination.

Wudthichai Suttithawil reported an 18-year-old non-smoker with predominant LCH of lung diagnosed by pulmonary wedge biopsy and Langerhans’ cells were diagnosed by histopathology and presentation of CD1a in lung tissue. Several studies showed LCH presentations in extrapulmonary bone involvement among smokers. Sneha Varkki presented a similar case like ours with an isolated pulmonary involvement in a six-year-old male. Pulmonary LCH can present with bilateral pneumothorax as observed in Aleric et al presentation. But in our study pneumothorax occurred after lung biopsy. LCH nodules can resemble isolated pulmonary metastases.

CONCLUSION

Pulmonary Langerhans’-cell histiocytosis is a rare interstitial lung disease presenting with pulmonary nodules. High degree of suspicion is needed in diagnosis. Prognosis is variable. Presentation can be isolated pulmonary to multiorgan involvement. Langerhans’-cell histiocytosis involving bone is common. Diagnosis is by demonstrating the Langerhans’ cells in the biopsy specimen or bronchoalveolar lavage specimen. No satisfactory treatment is available. Prognosis is variable.

REFERENCES


