A RARE CASE OF PULMONARY HISTIOCYTOSIS.
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Abstract: Pulmonary histiocytosis is a well described entity in adults but extremely rare in children. May present with milder symptoms to severe respiratory failure. We had a 3 year old child presenting with respiratory distress, managed initially with bronchodilators and antibiotics. On further examination and investigation, he was diagnosed to have pulmonary histiocytosis. The case being reported for its rarity.

Keyword: Pulmonary histiocytosis, Langerhans cell histiocytosis, CD 1a, chemotherapy.

INTRODUCTION:
Langerhans cell histiocytosis is a multisystem disorder which has an extremely variable presentation. Pulmonary involvement is seen in only 10-15% of individuals. Very few cases are reported in literature in children.

CASE REPORT:
Master X, 3 yr old male child 1st born of non-consanguineous marriage was brought with complaints of breathlessness – 1 month, no history of cough or fever, symptoms were not improving with nebulisation. No similar past history, no H/O contact with TB, no family H/O asthma, Perinatal history – insignificant.

General Examination – normal except for skin lesions. The skin lesions are multiple, discrete, papular, scaly and greasy with seborrhic dermatitis like lesions present over the trunk. RS – Bilateral Air Entry equal with wheeze & crepts. Respiratory Rate - 40/min; retractions (+); Abdomen- firm hepatomegaly. Other systems - Normal.

Investigations:
- CBC, RFT, LFT were normal. Sepsis and TB work up were negative. Stool for fat globules were negative. CXR was normal. Arterial blood gas showed Type 2 Respiratory failure. Usg abdomen showed hepatomegaly. Sweat chloride test was normal. Provisional diagnosis of Interstitial lung disease was made. But the child did not responded to bronchodilators, antibiotics.
- Differential diagnosis for the CT Chest which showed cystic lesions in lungs - Langerhans cell histiocytosis, Lymphangiomatosis.
- Skeletal survey and Echo was normal. Bronchoscopy showed normal tracheobronchial tree. Broncho Alveolar Lavage showed sheets of 80% chronic inflammatory cells, 15% histiocytosis. Skin biopsy showed stratified squamous epithelium with focal collections of polygonal cells with eosinophilic cytoplasm and folded nucleus suggestive of langerhans cell histiocytosis. ImmunohistoChemistry showed S100 4(+), CD 1a +ve, CD 68 +ve.

A diagnosis of PULMONARY HISTIOCYTOSIS was made. Treatment: LCH International Study Protocol is followed comprising of Vinblastine, Etoposide, Prednisolone. After the first cycle of chemotherapy
the patient symptomatically improved. But the patient was
lost follow up.
Pulmonary LCH is a well described entity in adults but extremely
rare in children. Though rare, it should be considered in any child
with chronic respiratory disease when not responding to treatment.
The case is being presented for its rarity.

**DISCUSSION:** Childhood histiocytosis is classified into
A. Langerhans cell histiocytosis  B. Hemophagocytic lymphohistiocytosis  C. Malignant histiocytosis
Langerhans cell histiocytosis has 3 groups
A. Eosinophilic granuloma
B. Hand schuller christian disease
C. Letterer siwe disease
Incidence of LCH is 1 in 2 lakh. It has no sex predilection. Pulmonary Langerhans cell histiocytosis (PLCH) is rare in childhood but occurs most commonly in children with multisystem LCH. In adults, by contrast, the lung is the most common and usually the sole organ affected. Isolated PLCH is common in adults. In children it occurs with multiorgan involvement. In adults almost seen in smokers. It affects the smaller airways and blood vessels in lungs. Pathological lesion includes peribronchial inflammation with cyst formation leading on to fibrosis. Usually asymptomatic. May present with milder symptoms to severe respiratory failure. Rarely it can also present with pneumothorax. Many cases have been diagnosed following postmortem examination. HRCT is pathognomic. Lung biopsy confirms the diagnosis. Prognosis depends on the risk organ involved. In general it carries a poor prognosis.

**REFERENCES:**