Systemic disease with pulmonary manifestation- rare presentation

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- Master X, 3 yr old male child born of non-consanguinous marriage was brought with

  c/o breathlessness – 1 month

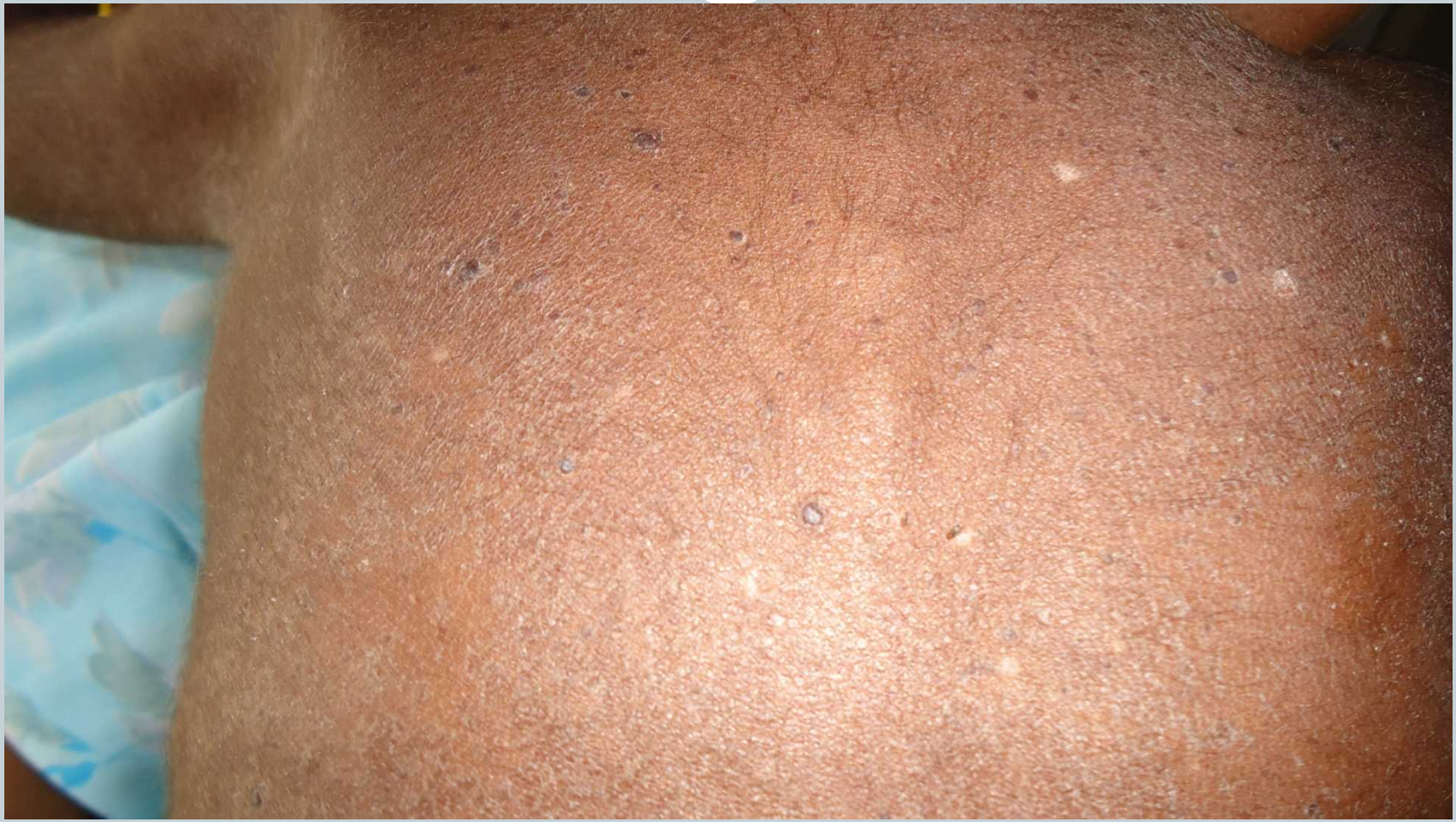
  no cough,fever.

  not improving with nebulisation

- No similar past history
- No H/O contact with TB
- No family H/O asthma
- Perinatal history - insignificant

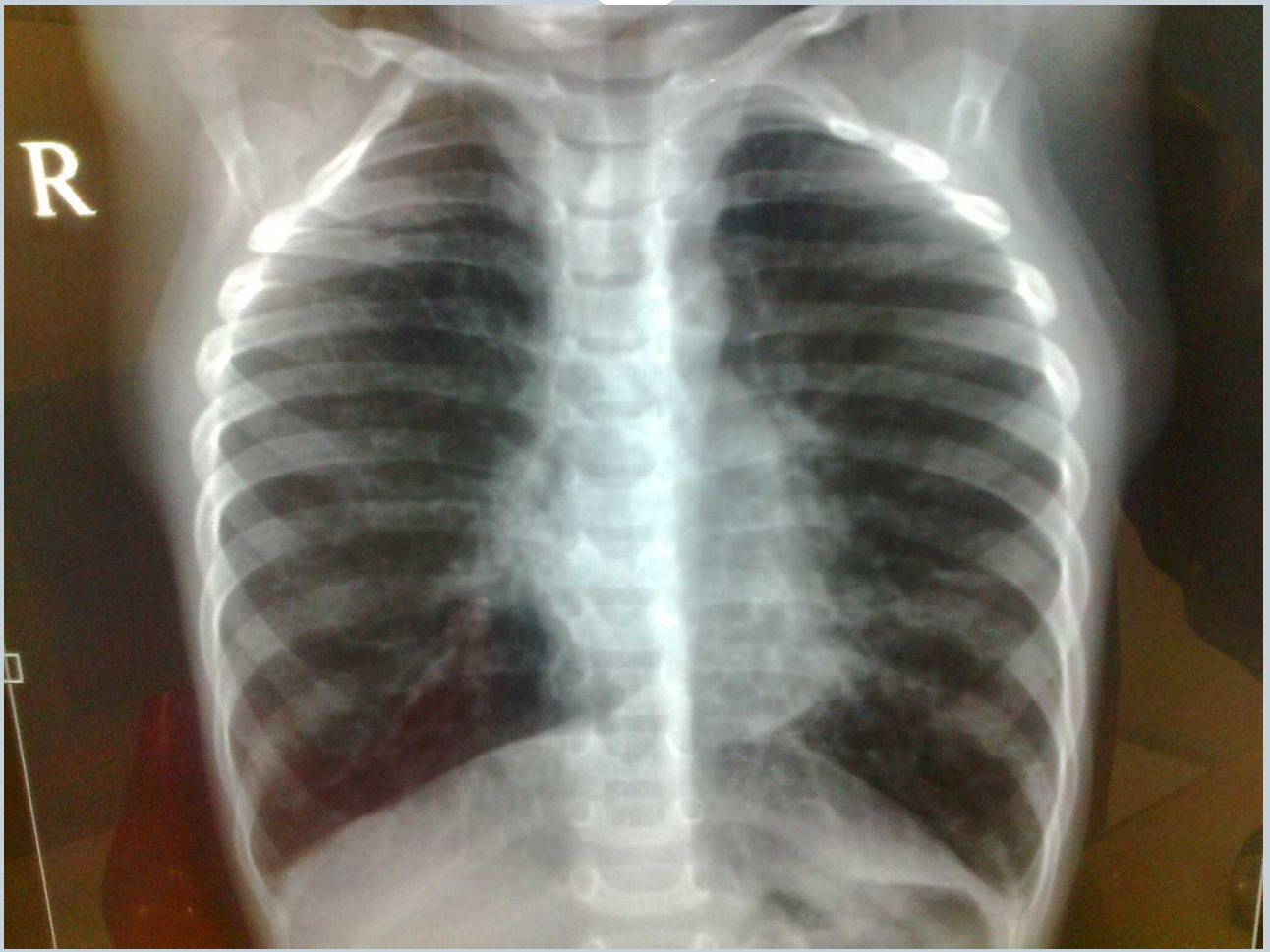
- O/E
- G/E – normal except for skin lesions
- RS – BAE equal with wheeze & crepts

  RR- 40/min ; retractions(+)
- Abd- firm hepatomegaly
- Other sys- Normal



Investigations

Hb	10.2
TC	16400 (P 60 L 36 E 4)
Platelet	4 lacs
P.smear	Microcytic hypochromic anemia
NEC	No growth
TB work up	negative
HIV	negative
RFT	N
LFT	N
Stool for fat globules	Negative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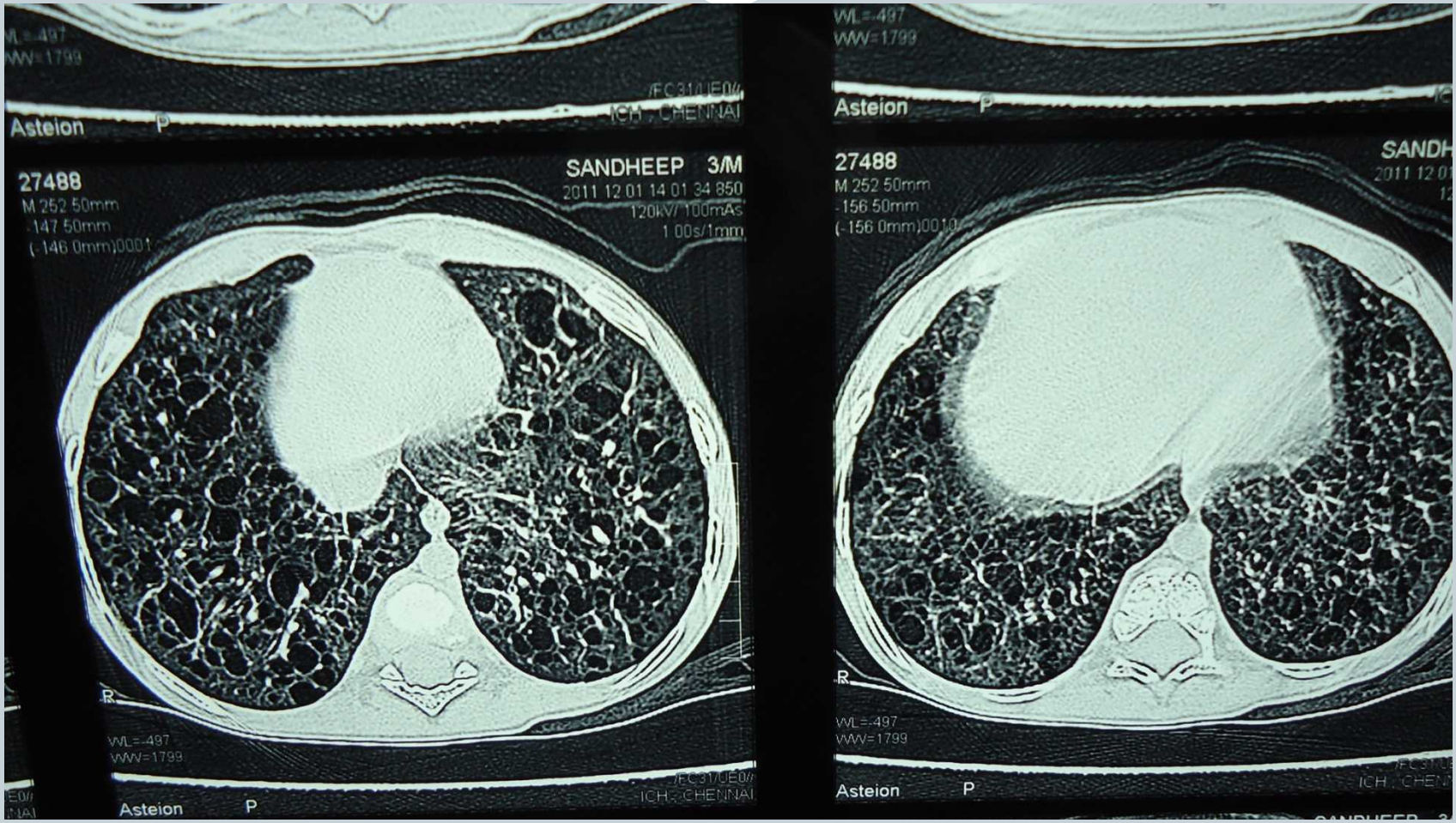
- CXR – normal
- ABG- pH-7.37, po_2-81,pco_2- 53
- Usg abdomen- hepatomegaly
- Provisional diagnosis- ? ILD
- Not responded to bronchodilators, antibiotics

CT chest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D/D

- Langerhans cell histiocytosis
- Lymphangiomyomatosis
- Pulmonary embolism

- Skeletal survey – normal
- Echo - normal
- Bronchoscopy – tracheobronchial tree normal
- BAL

  sheets of chronic inflammatory cells 80%,

  histiocytosis 15%, eosinophils 3%

- Skin biopsy-

  Stratified squamous epithelium with focal collections of polygonal cells with eosinophilic cytoplasm and folded nucleus

  S/O langerhans cell histiocytosis

- IHC- S100 4(+),

  CD 1a +ve, CD 68 +ve

  LCA – patch positivity

  Ki 67- low

Histiocytosis

- Classification of childhood histiocytosis

A. Langerhans cell histiocytosis

B. Hemophagocytic lymphohistiocytosis

C. Malignant histiocytosis

Langerhans cell histiocytosis

A. Eosinophilic granuloma

B. Hand schuller christian disease

C. Letterer siwe disease

- Incidence – 1 in 2 lakh
- No sex predilection

Histiocytosis X

Skeleton	80%
Skin	50%
Lymphadenopathy	33%
Otitis media	30-40%
Hepatosplenomegaly	20%
Pulmonary involvement	10-15%
Exopthalmos	
Pituitary dysfunction	
Systemic manifestations	
Cirrhosis	
CNS involvement	

Pulmonary histiocytosis

- 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 findings:

- Peribronchial inflammation with cyst formation leading on to fibrosis.

- Usually asymptomatic
- May present with milder symptoms to severe respiratory failure
- HRCT is pathognomic
- BAL
- Lung biopsy

- Treatment: LCH International Study Protocol is followed.
- Prognosis depends on the risk organ involved.
- Poor prognosis

Take home message

- 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.

Acknowledgements

- Dept. of Radiology
- Dept. of Hematology
- Dept. of Pulmonology
- Dept. of Paediatric surgery

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